Cardiac Catheterization for Congenital Heart Disease

From Fetal Life to Adulthood

Gianfranco Butera
Massimo Chessa
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Editors

Second Edition

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Foreword: "Nothing Endures But Change"

This concept was stated by Heraclitus of Ephesus in his book *On Nature* in the sixth century B.C. A long time has passed from the period of this pre-Socratic philosopher; however, the same concept applies today and truthfully to the field of interventional cardiology. The only way to be alive and up-to-date is to continuously improve and change. Therefore, it is with this thought we welcome the second edition of the *Cardiac Catheterization for Congenital Heart Disease*. The second edition follows the popular "how-to" format of the previous edition and is the result of a worldwide cooperation of many international experts, who put together their knowledge and expertise.

The first edition was published in 2014 and many new transcatheter therapies have been developed in the field of interventional cardiology for CHD, including the expanded use of balloon-expandable valves for dysfunctional RVOTs, conduits, and valve-in-valve therapies in all four cardiac valves. The initial use of self-expanding valves in the RVOT to treat severe pulmonary regurgitation has continued to be developed around the globe. The increased role of advanced imaging, such as 3D Rotational Angiography (3DRA), has allowed interventional cardiologists to use fluoroscopic overlay to not only improve the precise positioning of devices such as stents and valves but also to potentially decrease radiation exposure by reducing the need of additional angiograms.

The role of integrating advanced imaging modalities in the cath lab, such as real-time MR-guided cardiac catheterizations

and potential interventional procedures without the use of radiation, has been developed and promoted by centers around the world. In addition, 3D printing has become popular in both planning complex surgical and interventional procedures, and educating patients and families in their complex CHD. Finally, the editors have included a much-needed section on quality improvement of both results and care in our ongoing quest of delivering the highest form of interventional cardiology for our patients.

As Heraclitus stated "nothing endures but change," he also said "the eyes are more exact witnesses than the ears"…which is the way interventional cardiologists view improving our procedures to achieve our core value and mission of treating our patients.

Respectfully,

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Preface

"Our progress is not to assume that we have arrived but to continually aim for the goal."
Bernard of Chiaravalle, Benedictine monk

Six years have passed since this book was printed and distributed for the first time. It was in English, but the Chinese version arrived just few years later.

Many changes have occurred in the world in this period, including some unexpected events as the experience of a pandemic spread. We thought that uncontrolled infections were historical events that occurred during the "dark ages" centuries ago. Once more, life and reality are more creative than humans.

In the smaller world of congenital interventional cardiology, several less dramatic changes have occurred. Our goal, as Bernard of Chiaravalle opined, remains the same and we are continually striving to achieve it. We all aim at improvement and at providing new answers to unsolved questions and problems.

As a result, we welcome and commend to you the second edition of this book. A good proportion of the content is new since new approaches and technologies continue to evolve. These include the transcatheter treatment of sinus venosus ASD, of PDA in premature babies, the use of self-expanding valves in large RVOTs, the development of engineering methods including modeling and 3D printing, the great development of imaging techniques, and the integration of imaging modalities to make procedures safer, quicker, more reproducible, and most importantly more effective. To these topics we have also added new

chapters covering the approach to the mid-aortic syndrome, to PA-VSD-MAPCAs management, hybrid access, percutaneous solutions for large RVOTs with the current available technology, RVOT stenting in infants, and PFO closure. Furthermore, a chapter on tools and techniques usually used by "adult cardiology colleagues" was included in order to share knowledge about allied and potentially useful devices. Finally, as we are determined to strive for the best, a full and comprehensive chapter on quality evaluation and improvement has been included. Nowadays, excellence requires analysis of our own practice in order to continuously improve our results.

We are extremely proud that this book is the product of the experience of more than 50 world-class authors from all over the globe. It is thanks to them that this dream has come true … and for the second time!

A Roman philosopher, Seneca, said "Optimae ideae pertinent ad omnes" (the best ideas belong to everyone). We are grateful to each author for having shared their valuable experience and best ideas for the benefit of everyone and in particular for the benefit of our young colleagues and our patients.

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Part I

General

1 Patient Information and Informed Consent

Maarten Witsenburg

1.1 Introduction

Interventional (and diagnostic) catheterization is an important tool in congenital heart disease. It has evolved from atrial septostomy in the 1970s to a wide range of procedures including device closure of various defects and percutaneous valve implantation nowadays.

As any form of invasive study or treatment, it is not without risks and serious complications may occur. Therefore, it should only be performed after balancing the advantages and risks of the procedure [1]. The risk associated with the use of ionizing radiation for these procedures should also be kept in mind, especially because of the young age of many of these patients.

The patient (or his or her legal representative) has to agree on the suggested treatment, but can only do so after having been informed appropriately. The combination of the duty to inform and the agreement of the patient with the treatment plan is called informed consent.

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Informed consent is an essential step in any diagnostic or interventional cardiac catheterization in a patient with congenital cardiac disease.

1.2 Background

Healthcare ethics is based on the moral concepts of benevolence, autonomy, absence of malice, equity, and responsibility. Autonomy implies that the patient (and/or the legal representatives) can only consent after the provision of adequate information. The major elements in a valid consent process are sufficient understanding, sufficient information, and freeness from duress [2, 3].

In the ESC-EACTS myocardial re-vascularization guidelines, it is stated that information should be "objective and unbiased, patient oriented, evidence based, up-to-date, reliable, understandable, accessible, relevant, and consistent with legal requirements" [3].

1.3 Information and Consent in Clinical Practice

In a non-emergent setting, the indication for a diagnostic or interventional cardiac catheterization should be discussed within a multidisciplinary team including at least the (pediatric) cardiologist, interventional cardiologist, and cardiac surgeon. For non-complex cases a written and locally approved protocol can be an alternative for the discussion within the multidisciplinary team.

In such a heart team, the indication, risks and benefits, possible other treatment options, and timing of procedure are discussed. This team decision is written down in the patient record, as well as the team members who were involved in the discussion.

Once the decision is made, the patient (or legal representative) is informed. It is important to take enough time to discuss the

reason for treatment, its timing, risks, and possible treatment complications. Use of terminology that the patient understands is essential. An illustrated information sheet can be very helpful. One should realize that a lay person as a patient will always have a major lack of knowledge, even after an extensive discussion with the interventional cardiologist. The consent will therefore for a major part be based on the patient's trust in the treating physician. After the patient has consented, this is documented in the patient record. Depending on local rules and practice, the consent can be given orally or in writing.

In emergencies, time may be lacking to fulfill the steps mentioned above. A typical example is severely hypoxic neonate with d-transposition in need of urgent balloon atrial septostomy to improve atrial mixing. In such cases the information needs to be given after the procedure, including explanation of possible complications that may have occurred.

1.4 Conclusion

Recommendations for treatment in congenital heart disease will rarely have a higher than 1C level of evidence. As such expert opinion plays a major role. Even for procedures that have been used extensively for many years, the implications, including complications, have become clearer recently. The complex trajectory from indication for treatment to the diagnostic or interventional procedure itself are at risk for cognitive bias [4].

In addition the availability of an extending range of devices might sometimes result in using these for questionable indications.

The important point is that any interventional cardiologist should act in a responsible way before, during, and after the intervention. Whenever complications may have happened, he/she should be able to explain the problem, both to the patient and to colleagues, and how steps were taken to minimize any further harm.

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2 Anaesthesiological Management of the Paediatric Patient in the Catheterisation Laboratory

Giuseppe Isgrò and Marco Ranucci

2.1 Introduction

The widespread use of therapeutic cardiac catheterisation in the management of congenital heart disease requires the presence of a trained paediatric cardiac anaesthesiologist with the ability to provide both safe and consistent sedation or general anaesthesia to paediatric cardiac patients. Specific knowledge of the pathophysiology of congenital cardiac lesions and the clinical implications of diagnostic and therapeutic procedures are essential.

Sedation is often preferred to general anaesthesia, in particular for diagnostic procedures, because mechanical ventilation can cause haemodynamic disturbance and can alter the results of the study. General anaesthesia is applied mainly for interventional procedures (i.e. percutaneous valve implantation, atrial septal defect or ventricular septal defect closure, patent ductus arteriosus closure, or STENT implantation), during which it is essential to keep the patient deeply relaxed to permit the precise deployment of the device.

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Other factors influencing the decision to use sedation or general anaesthesia are patient related: age, clinical conditions and complexity of cardiac lesions.

Diagnostic and interventional procedures in the catheterisation laboratory carry risks for the patient such that continuous patient monitoring is essential.

The anaesthesiologist should contribute to the treatment of complications associated with cardiac catheterisation and, obviously, pre-empt and manage issues arising from sedation and anaesthesia. Finally good anaesthetic practice means that after the procedure the patient is delivered to a post-anaesthesia care unit or directly to the intensive care unit in the best condition possible.

2.2 Anaesthesia

2.2.1 Preoperative Consideration

Preoperative clinical evaluation is mandatory to assess the general condition of the patient and the type of cardiac disease and make plans for post-procedural care. Those patients affected by severe cyanosis should be hydrated prior to cardiac catheterisation, to minimise dehydration.

Fasting should be planned according to the age, clinical condition, and related laboratory investigations.

Routine preoperative tests (ECG, chest X-ray, lab investigations) are required and evaluated by the anaesthesiologist—in some cases, review of echocardiography. An assessment should include scrutiny of previous anaesthetic records and prior premedication.

Certain patients including chronically cyanotic patients are at risk of post-procedural bleeding, so that packed red cell units, fresh frozen plasma and concentrated platelet units are quickly available according to the procedure.

Strict attention to intercurrent illness is required, and if necessary, the catheterisation procedure should be postponed whilst this resolves.

2.2.2 Premedication

Drugs for premedication are administered to reduce anxiety and promote cooperation. Additional benefits include induction of anaesthesia without memory of this stressful time and reduced adrenergic stimulation that can be deleterious, particularly certain anomalies (i.e. tetralogy of Fallot, uncompensated ventricular septal defect with pulmonary hypertension and anomalous origin of left coronary artery arising from the pulmonary artery).

Children under 6 months of age or those that are very sick often can be managed without premedication as this can be deleterious under some circumstances.

Many drugs are available for premedication; the most commonly used are ketamine, midazolam, fentanyl and morphine. Dexmedetomidine, a new centrally acting alpha 2-adrenoceptor agonist, has been used in the setting of cardiac catheterisation laboratory safely with good results.

The choice of the drug alone or in combination must be decided by the anaesthesiologist after assessment of the patient and according to local experience and protocols.

2.2.3 Sedation and Anaesthesia

Sedation and general anaesthesia can be administered according to the preoperative condition, including the risk of developing post-procedural deleterious effects related to cardiac catheterisation and anaesthesia drugs use (i.e. pulmonary hypertension).

Pathophysiology of any cardiac lesion should be discussed beforehand with the paediatric cardiologist to reduce the risk of anaesthesia delivery, although modern anaesthesia drugs have reduced impact on cardiovascular system (Table 2.1). Sevoflurane, a volatile anaesthetic, has very little effects on systemic pressure and heart rate. Dexmedetomidine is thought to be protective for postoperative atrial fibrillation.

Table 2.1 Anaesthetic drugs

Drug	Induction	Maintenance
Ketamine	0.5–2 mg/kg	0.01–0.05 μg/kg/min
Midazolam	0.1–0.3 mg/kg	1–3 μg/kg/min
Propofol	1–2 mg/kg	3–5 mg/kg/h
Sevoflurane	3–5%	1–2%
Fentanyl	3–5 μg/kg	1–2 μg/kg/min
Morphine	0.1 mg/kg	1–2 μg/kg/min
Cisatracurium	0.1–0.2 mg/kg	1–2 μg/kg/min
Dexmedetomidine	1 μg/kg	0.2–1.4 μg/kg/h

Midazolam is safely used to maintain sedation, usually in combination with fentanyl or morphine.

The use of muscle relaxants that permit to keep the patient ventilated under general anaesthesia is nowadays safe, because the introduction of many newer agent with low rate of adverse effects; the combination of modern volatile anaesthetics and modern muscle relaxants have reduced to very rare event the incidence of malignant hyperthermia.

Cisatracurium, a non-depolarising muscle relaxant, a cis-isomer of atracurium, is indicated in paediatric anaesthesia because of the absence of histamine release; its half-life is 22–29 min and it is eliminated through the Hoffman metabolism, so it can be used safely in patients with poor renal function.

2.3 Monitoring and Anaesthetic Equipment in the Cardiac Catheterisation Laboratory

2.3.1 Electrocardiogram

Electrocardiogram is used for continuous monitoring of heart rate, rhythm and ST changes throughout the pre-, intra- and post-procedural phases.

2.3.2 Blood Pressure

Systemic blood pressure may be monitored noninvasively during the most common procedures by an automated oscillometric technique.

During risky procedures or in very sick patients, it may be necessary to monitor invasive blood pressure, by cannulation of an artery. Thereby arterial cannulas, transducers, and flushing devices must be present in the laboratory.

2.3.3 Pulse Oximetry

It provides a continuous and noninvasive monitor of oxygen saturation, which is mandatory during both sedation and general anaesthesia in paediatric cardiac patients, who are at risk for the development of hypoxia.

2.3.4 Capnometry

Is a continuous and noninvasive method of measurement of expired carbon dioxide and is very useful to monitor the adequacy of ventilation during general anaesthesia or to detect malfunction or failure of the anaesthesia machine. Moreover, it provides a useful information related to the quality of pulmonary perfusion and can reflect haemodynamic changes.

2.3.5 Temperature Monitoring

Temperature monitoring is very important especially in the newborns, who are at risk for hypothermia because of their relatively large surface area and the inefficiency of their thermoregulatory mechanisms. Cutaneous temperature may be monitored by adequate probes. Central temperature, if required, can be measured using a nasogastric probe.

In order to avoid hypothermia in children, it is important to warm the environment and the inhalatory gases by a humidifier. Warming of intravenous fluids may be needed. The use of heating blankets is also recommended especially in the newborn.

2.3.6 $ScvO_2$ (Continuous Mixed Venous Oxygen Saturation) Monitoring

Paediatric and adult patients with severe cardiac disease, who undergo catheter laboratory interventional procedures, can be monitored with respect to cardiac output. In congenital heart disease patients, it is usually either not possible or desirable to insert a pulmonary artery catheter designed for output measurement. Currently central venous catheters with oximetry are available to continuously monitor venous saturations. These catheters are usually inserted into the right jugular internal vein like a normal central venous line, with the same dimension and length (PediaSat and PreSep catheters—Edwards Lifesciences, Irvine, CO) (Fig. 2.1).

The continuous monitoring of venous saturation can help to identify sudden changes in haemodynamic status, rapidly changing when cardiac output decreases or increases.

This parameter is included also in the management of the early goal-directed therapy (EGDT) for critically ill patients.

2.3.7 NIRS (Near-Infrared Spectroscopy) Monitoring

Another tool of haemodynamic monitoring is near-infrared spectroscopy (NIRS) (Fig. 2.2). NIRS is used in many clinical situations to continuously monitor cerebral and splanchnic perfusion and has the potential to provide information on the adequacy of systemic oxygen delivery. Some authors have demonstrated a good correlation between NIRS and $ScvO_2$, but NIRS cannot precisely predict $ScvO_2$ value, though it can be used for trend monitoring.

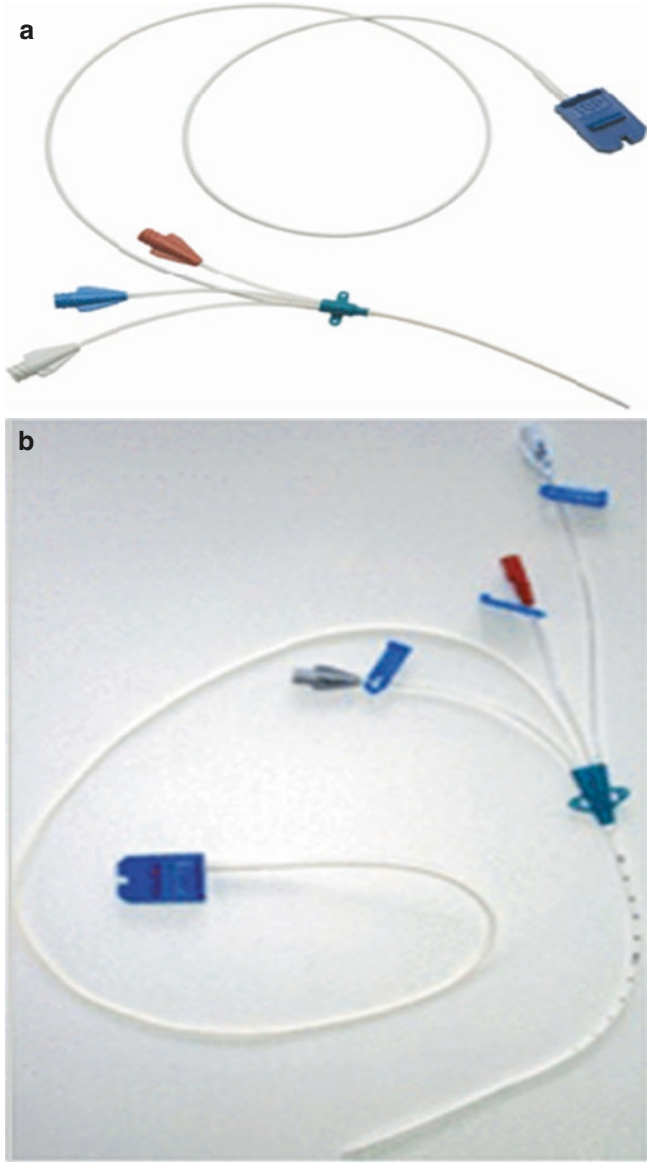

Fig. 2.1 (**a**, **b**) PediaSat catheter (paediatric) (**a**) and PreSep catheter (adult) (**b**). (Courtesy of Edwards Lifesciences)

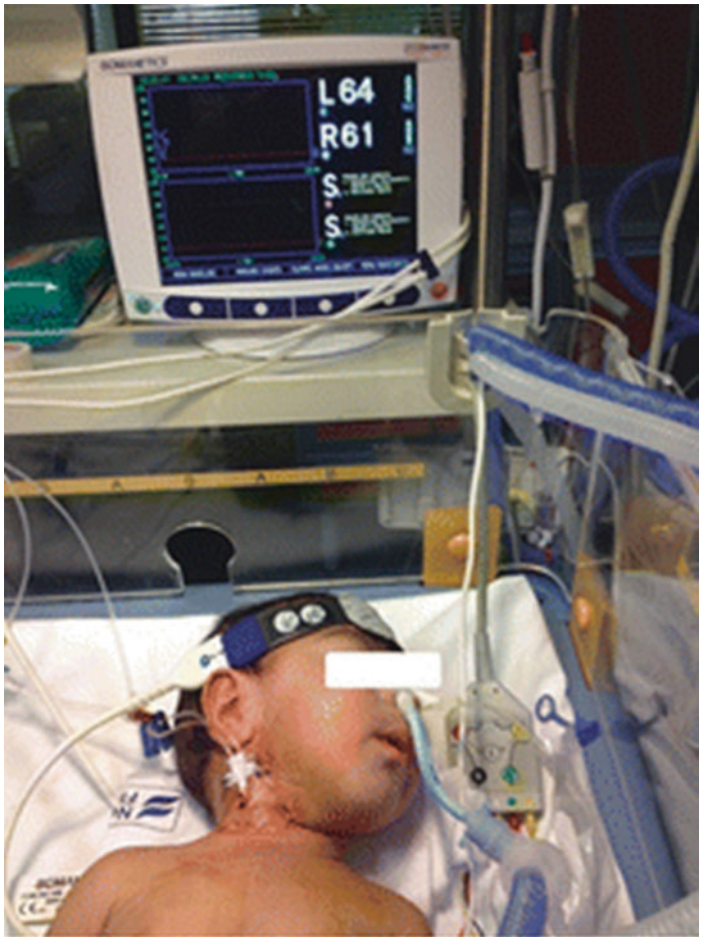

Fig. 2.2 NIRS monitoring

2.3.8 Anaesthetic Equipment

- Different sizes of cannulas for venous and arterial cannulation.
- Different sizes of central venous catheters.
- Different sizes of face masks.
- Different sizes of endotracheal tubes.
- Airway management equipment and difficult airway management equipment available.
- Suction apparatus and different sizes of suction catheters.
- Mechanical ventilator with inhalatory anaesthetic agents.
- Scavenging setup for waste inhalational agents.
- Sedative, analgesic and anaesthetic drugs.
- Resuscitation drugs.
- Intravenous infusion set—intravenous fluids (crystalloids and colloids).
- Defibrillator.
- Stethoscope.
- Self-inflating manual resuscitation bag.
- Foley catheters and nasogastric tubes.
- Blood gas analyser.

2.4 Ventilation Strategies During Cardiac Catheterisation Procedures

There is no preferred ventilation strategy in the cardiac catheterization laboratory, but spontaneous breathing, in particular during diagnostic procedures, is preferable to mechanical ventilation to maintain more natural intrathoracic physiology and consequentially to obtain the best acquisition of more accurate hemodynamic data.

General anaesthesia with positive pressure ventilation provides a secure airway and control of $PaCO_2$, but increased intrathoracic pressure may alter hemodynamic parameters.

However, general anaesthesia with positive pressure ventilation is not contraindicated, but requires a correct approach to the

patient and to the pathophysiology of the underline cardiac disease; in some particular case is better to avoid high PEEP (Positive End Expiration Pressure), and use correct I:E time (Inspitation-Expiration ratio) is necessary to have a less impact on venous return, mainly in patients with right heart failure, hypovolemia or Fontan physiology.

The effect of pulmonary pressure on pulmonary venous flow depends on the relative filling state of the pulmonary circulation.

During hypovolemic condition an increase in lung volume reduces venous return to the left ventricle, while in a fluid overloaded state, an increase in lung volume would shift blood, thus increasing pulmonary venous flow to the left ventricle.

Change in lung volume can alter the diastolic compliance of the left ventricle because of direct compression of the chamber and also due to elevated right ventricular pressure that results in a conformational change of the interventricular septum.

As well as good ventilator setting is fundamental, the optimal Endotracheal Tube placement is very important: displacement in right or left main bronchus or placement upon the Tracheal Carina, can alter the hemodynamic parameters during diagnostic procedure.

Spontaneous ventilation might maintain more natural intrathoracic physiology and consequentially can result in the acquisition of more accurate hemodynamic data.

However, oversedation during spontaneous ventilation can cause airway obstruction, hypoventilation and subsequent respiratory acidosis.

This increases PVR and might alter shunt physiology and alter hemodynamic measurements.

Basically patients having even minimal sedation should have their airway and $EtCO_2$ monitored throughout the procedure to avoid hypercapnia, to minimize adverse events related to sedation and ventilation.

2.5 Fluid Management

The intravenous fluid regimen during Cardiac Catheterisation is guided by the age of the patient, clinical condition (fluid overloaded or not), reason for admission in cath lab, presence of any deficit and presence or absence of shock and/or multi-organ failure in particular renal failure.

In congenital heart patients, intraoperative management of intravascular fluids is a priority.

Hypovolemia might be present at the beginning of the procedure, particularly in small infants and children, secondary to dehydration occurring during prolonged periods of preoperative fasting (NPO).

Hypovolemia is particularly important in very young, cyanotic, erythrocytotic or shunt-dependent patients.

In these circumstances it is preferable to administer intravenous (IV) isotonic fluids to maintain hydration during the fasting period prior to catheterization.

Careful attention to blood loss is particularly important in neonates who have a small blood volume and in cyanotic patients.

Blood transfusion is mandatory to mantain normal level of haematocrit that should be 40–45% in newborn.

Cyanotic patients that experience blood loss, may require red cell transfusions despite relative normal level of haematocrit.

In this case is highly important to asses DO_2 (Oxygen Delivery) and Lactates levels to address the best fluid therapy.

Volume overload can occur during longer procedures, particularly those involving multiple angiograms, and is less tolerated in patients with congestive heart failure or shunt lesions.

Fluids restriction and diuretics administration is required in this case.

2.6 Complications of Cardiac Catheterisation

Complications related to cardiac catheterisation include arrhythmias, acute valvular regurgitation, hypotension, desaturation, vessel/myocardial rupture, pericardial tamponade and cardiac arrest.

Many of these complications are self-limiting and do not require treatment, e.g. minor arrhythmias. In all cases, a cardiac surgeon should be available, and for particularly difficult risky cases on standby.

Other complications are not related with the procedure itself, but to the positioning of the patient on the table: ulnar nerve injury, brachial plexus injury, pressure lesions.

Staff members are responsible for the positioning of the patient.

Airway oedema with stridor (laryngeal spasm) is frequent after prolonged catheter laboratory procedures, and the anaesthesiologist must consider this complication after extubation and promptly intervene; the treatment includes the administration of intravenous steroids, diuretics and occasionally the need for noninvasive ventilation (CPAP—continuous positive airway pressure) for some hours after the procedure.

Vascular thrombosis can occur after catheterisation procedures often associated with established patient-related risk factors (i.e. newborn, small infant); heparin or fibrinolytic therapy may be required, and in very rare instances, vascular surgical input can be required.

Another important complication is acute renal failure (ARF) especially in those children that undergo same day to catheter laboratory procedures and surgical procedures. It is directly correlated to the amount of iodine-containing contrast administered. If possible, surgery should be delayed following angiography (Table 2.2).

Table 2.2 Suggested emergency drugs

Glucose 33%
Atropine
Epinephrine
Norepinephrine
Dopamine
Isoproterenol
Furosemide
Etacrinic acid
Diazepam
Calcium chloride or calcium gluconate
Sodium bicarbonate
Lidocaine
Corticosteroids
Aminophylline

Suggested Reading

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3 Antibiotics and Anticoagulation

Luciane Piazza

The use of antibiotics in the catheterization laboratory (Cath lab), to prevent infectious endocarditis has changed along the years. Despite the increased number of strategies and the advancements in diagnosis and antimicrobial therapy, surgical techniques and management of complications, infective endocarditis (IE) still carries high rates of mortality and morbidity. IE typically requires prolonged courses of antibiotics and often valve or conduit replacement. IE prophylaxis have been proposed for patients with cardiac disease potentially at risk of IE following bacteremia during invasive procedures.

It is important here to highlight two concepts:

(a) prophylaxis—is the administration of an antimicrobial in the periprocedural period in the absence of clinical infection to prevent an infectious complication.
(b) bacteremia—is the presence of the bacterial within the bloodstream without clinical signs or symptoms of infection.

In the Cath lab, the interventions to prevent infections consist on a primary action and an antibiotic prophylaxis.

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Primary Prevention: Consist of appropriate infection–control measures and dedicated techniques. The most important recommendations for sterile techniques are (a):

1. For physicians (Table 3.1).
2. Environmental catheterization of the laboratory (Table 3.2).
3. For patients (Table 3.3).

In order to protect the hemodynamic laboratory and the entire team of workers, it is necessary to accurately study the clinical case in advance by choosing the necessary materials, avoiding by search materials in other rooms or other storage areas during the procedure. All healthcare personnel should be aware that the door must always be closed and for this reason, to prevent contamination it is also necessary:

Table 3.1 Recommendation for physicians

Hand wash	Is the most important procedure to prevent infections. The recommendation is: hand scrub for 2–3 min containing an antiseptic agent. For the next application, it should be better avoid scrub and just wash with antiseptic solution to prevent skin irritation and dermal abrasion
Body protection	To care the patient is necessary wear caps, masks, gowns, and gloves. Sterile gowns and gloves must be worn in a sterile way Current issue: If you are not sure or if the patient's clinical history of infectious disease is unknown, it is recommended an appropriate mask to prevent droplets or by air infections, according to WHO

Table 3.2 Environmental catheterization of the laboratory

Clothes prevention	Accuracy of clothes is necessary, specially for shoes, in order to reduce contamination
Cleaning of the room	The laboratory must be cleaned in accordance with the requirements
Air vents	The efficiency of negative pressure chamber is mandatory in order to maintain the required conditions. It is extremely important to emphasize that is necessary to work with the hemodynamics room door closed

Table 3.3 Recommendation for patient preparation

aHair removal	Remove hair at the access site using clipper or depilatory on the day of the procedure
Skin cleaning	2% chlorhexidine with alcohol is the most effective antiseptic agent, but could be substituted with povidone-iodine. Take care in neonates and premature infants because of increased risk of skin irritation and absorption (b)
Drapes	Nonporous drapes are necessary to cover the area. It should be large enough and must contain a small adhesive part attached to the skin around the area involved in the intervention

- Avoid discussing the cases with the open door.
- Do not leave the hemodynamic room with the personal protective equipment.

3.1 Antibiotic Prophylaxis

Prophylactic intravenous antibiotic should be routinely administered to high-risk patients (Table 3.4) undergoing cardiac catheterization. The principles of antibiotics prophylaxis are based on:

- The choice of the antimicrobial agent.
- The timing of the first administrated dose.
- The duration of the prophylactic regimen.
- Although prophylaxis should be restricted to high-risk patients according to the guidelines, worldwide common practice in the cath lab is to extend preventive measures to all congenital heart disease patients.
- Usually this guideline (Table 3.5) is used to prevent the IE. Although most catheterizing physicians do not give antibiotic prophylaxis for standard diagnostic catheterization. The antibiotics are given in prolonged or complicated procedures. Other clinical conditions may be cause of bacteremia. For this reason the physician has to consider each situation in order to better identify the most adequate indication for prophylaxis. For example: preterms, neonates, and patients with immune compromise.

Table 3.4 Pathology with higher risk of IE

Prosthetic cardiac valve
Prosthetic material used for cardiac valve repair
Previous infective endocarditis
Congenital heart disease • Unrepaired cyanotic CHD including palliative shunt and conduits • Completely repaired congenital heart defects with prosthetic material or devices placed by surgery or by catheter intervention, during the first 6 months after the procedure • Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic devices
Cardiac transplantation recipients who developed cardiac valvulopathy

Table 3.5 Antibiotic prophylaxis—dose and regimen

Situation	Agent	Adults	Children
Standard	Ampicillin or Cefazolin or ceftriaxone	2 g 1 g	50 mg/kg—Single dose 30–60 min before procedure
Allergic	Clindamycina or Vancomicyn	600 mg 500 mg	20 mg/kg—Single dose 30–60 min before procedure

– Prophylactic treatment in patients that carries nasal *Staphylococcus aureus* is recommended prior the cardiac surgery. Therefore, the decolonization is the indicate pre-catheterization for patients with cardiac surgery program in the same hospitalization or where the patient remains intubated for subsequent cardiac surgery. This trick is important to prevent the surgical site infection.
– Nasal Swab is performed within a day of admission:

(A) Nasal swab positive for staphylococcus aureus methicillin-sensitive (MSSA):
  I. Patient >12 year: intranasal mupirocin prophylaxis: morning and night for 5 days.
  II. Patient <12 year: Mupirocin off label—used with informed consent.

(B) Nasal swab positive for staphylococcus aureus methicillin-resistant (MRSA):
Procedere come descritto sopra I. II.
III. Infusion of vancomycin 1 h before catheterization. Duration of infusion: 1 h.
IV. Be sure to use the devices according to the regular protocols of each hospital facility for contact insulation.

3.2 Anticoagulation in Catheterization Laboratory

The unfractionated heparin (UFH) is a commonly used antithrombotic drug in the cath lab for prevention of thromboembolic formation; it is also important in preventing peripheral arterial and venous thrombosis. In the most common access for cardiac catheterization, femoral artery, femoral vein, although jugular vein, brachial or radial arteries, UHF is used on some occasions. In all interventional procedures pharmacological thromboprophylaxis should only be considered in absence of contraindications like active or potential bleeding or severe thrombocytopenia.

The heparin has a short half-life (about 1.5 h) and easy reversibility, associated with many years of clinical experience. These characteristics have allowed the heparin as an anticoagulant of choice in the cath lab and in surgical operation. The major limitation associated with the UFH use is the unpredictable dose–response relationship. The dosage must be evaluated in order to reduce excessive bleeding (Table 3.6). It can be difficult to maintain the therapeutic level. Due to pharmacokinetics parameters, UHF often requires frequent monitoring and consequently requires a dose adjustment.

It is necessary to control the serum activity of heparin through the activated clotting time (ACT) 15 min after the bolus. Usually, the ACT is repeated 1 h after the first heparin bolus. It can be repeated in any doubt situation such as: bleeding or suspected thrombotic formation. Do not forget to repeat the ACT before removing the introducers.

Table 3.6 Type of procedure and heparin dosage

Type of procedure	Heparin dosage in bolus
Diagnostic venous catheterization	75 UI/kg
Diagnostic arterial catheterization	100 UI/kg
Venous and arterial diagnostic catheterization	100 UI/kg
Interventional catheterization	100 UI/kg

Table 3.7 Reversal of heparin therapy

Time since last Heparin dose, min	Protamine dose, mg/100 U Heparin
<30	1.0
30–60	0.5–0.75
60–120	0.375–0.5
>120	0.25–0.375

The protamine sulfate is used to reverse the effect of heparin when the quantity of heparin is too high in plasma, or when the effect is no longer required and in case of toxicity. (Table 3.7).

Maximum dose of 50 mg. Infusion rate a 10 mg/mL solution should not exceed 5 mg/min.

Hypersensitivity reaction to protamin sulfate may occur in patients with known hypersensibility reaction to first or those previously exposed to protamine therapy or protamin-containing insulin.

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4 Angiography: Basics and Contrast Media

Jose Luis Zunzunegui

The cardiac catheterization laboratory plays an important role in the management of children with congenital heart disease by not only enabling diagnosis but, in many cases, providing definitive therapy. This chapter focuses on the importance of adequate planning of the study, optimizing image formation, management of fluoroscopy and cine parameters, and basic knowledge regarding the use of contrast media that allow the cardiologist to lower radiation dose without sacrificing image quality.

4.1 Cardiac Catheterization Laboratory Equipment Overview and Basic Roentgenology

The X-ray tube is a glass tube containing a vacuum with a cathode (negative terminal) and anode (positive terminal). An electric current passes through a tungsten filament coil (cathode) and heats it, such electrons are "boiled off" the filament (thermionic emission). These electrons are accelerated toward the anode within the tube by application of a large electrical voltage, measured in kilovolts peak (kVp), across the cathode and the anode. This stream of electrons is the tube current, measured in milliamperes (mA). The kinetic energy of these high-velocity electrons, after striking the

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spinning tungsten disc (anode), is transformed mostly to heat energy and a few X-ray photons. The point at which the electrons impact on the tungsten target is called the focal spot of the X-ray tube. The energy carried by each X-ray photon depends on the applied voltage (kVp), while the rate of X-ray production depends on the tube current (mA). As the X-ray passes through the patient, it undergoes a change. Some of the X-rays are scattered in different directions from the original beam, while the others are absorbed by the tissues; this latter process is known as attenuation. The quantity of X-rays removed varies according to the mass of the patient with the remaining emerging as a beam on the other side. Scattered rays confer no imaging benefit and are a radiation to both the patient and catheterization laboratory personnel. The flat panel detector signal chain is described schematically in Fig. 4.1.

X-rays that reach the target are converted into electrons once again by interacting with the input phosphor and photocathode;

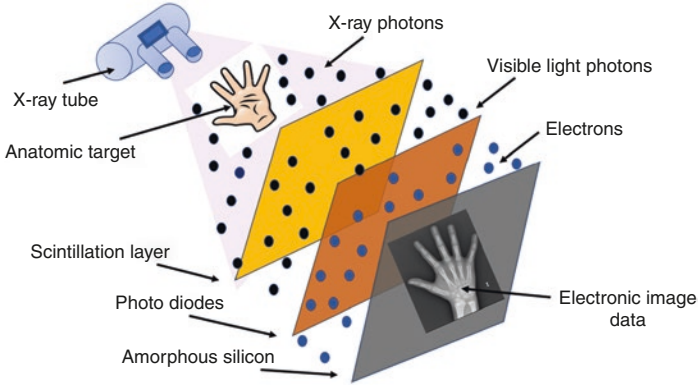

Fig. 4.1 Flat panel detector signal chain; with indirect digital X-ray imaging, an X-ray tube sends a beam of X-ray photons through a target. X-ray photons not absorbed by the target strike, a layer of scintillating material that converts them into visible light photons. These photons then strike an array of photodiodes which converts them into electrons that can activate the pixels in a layer of amorphous silicon. The activated pixels generate electronic data that a computer can convert into high-quality image of target, which is then displayed on a computer monitor

the electrons are then focused and accelerated onto the anode where they strike and emit visible light. The emitted light is then focused and transmitted to a television monitor for viewing and/or storage. An important feature of imaging chain is the *automatic exposure control* (AEC) that exists to ensure relatively constant image brightness. AEC is accomplished by a feedback mechanism from the digital video processor to the X-ray generator; if conditions change such that fewer X-rays exit the patient (e.g., table has been moved such that a more radiopaque part of the body is now being imaged), then feedback to the X-ray generator will increase the quantity or intensity of the X-ray in order to maintain equal image brightness.

The catheterization laboratory is capable of several imaging modalities. Fluoroscopy is used for real-time viewing and should provide sufficient image quality to view small guidewires. Fluoroscopy imaging should be set to the lowest possible radiation with usable image quality. Nowadays, variable-rate pulsed fluoroscopy is the standard; the X-ray beam is pulsed at 30 or 15 pulses per second or fewer (the lower the pulse frequency, the less the radiation dose, at the expense of a "jerkier motion"). The duration of each pulse is also known as the exposure time and is expressed in millisecond (ms), with typical setting for pediatric cardiac fluoroscopy ranging from 1 to 4 ms per pulse.

Images designed for permanent storage and review are usually obtained in acquisition (cine) mode, although higher quality modern fluoroscopic runs can usually be stored as a lower quality but acceptable alternative in many circumstances. Cine requires approximately 15 times more radiation per frame and should be used sparingly. Cine is always pulsed, and rates of 15, 30, and 60 frames per second are typically available in pediatric catheterization laboratories (whereas most adult studies are performed at 15 frames per second). Faster frame rates are necessary to view rapidly moving structures throughout the cardiac cycle (e.g., prosthetic valve leaflets), or in the setting of extremely high-flow rates through a vascular bed (e.g., arteriovenous fistula), and particularly if the patients is tachycardic. Any modification to the standard parameters of the X-ray beam

(mA, kV, ms) usually does not produce any significant benefit in image quality and tends to increase the dose of radiation [1, 2].

4.2 Tactics for Radiation Dose Reduction and Image Quality Improvement

4.2.1 Diagnostic Information Should be Obtained Primarily Noninvasively

Determination of important anatomic variants (e.g., systemic venous anomalies) will help in planning of the procedure (e.g., site of vascular access) and will serve to minimize the number of angiograms needed to clarify the anatomy. One should avoid obtaining angiograms that provide redundant information already known from noninvasive studies (echocardiography, MRI) "just because we are there."

4.2.2 Position Patients Properly (Isocentered and Straight on the Table)

Having patients in the isocenter keeps the heart at the center of the field whichever angulated views are used. Prolonged fluoroscopy during changes in angiographic projection is therefore avoided. Another benefit of having the patient positioned correctly straight on the table is that cardiovascular structures can be consistently related to skeletal and tracheobronchial landmarks (e.g., fossa ovalis, the ductus arteriosus, etc.) with minimal trial and error or wasted fluoroscopy.

4.2.3 Use the Lowest Acceptable Frame Rate During Pulsed Fluoroscopy and Cine Angiography

Always use pulsed fluoroscopy, never continuous fluoroscopy. Be prepared to change the frame rates frequently during a case

depending on the type of structure that is being imaged (e.g., fast-moving vs. slow-moving; venous or arterial).

4.2.4 Do Not Use Fluoroscopy to Make Changes to the Patient/Table Position or Collimator/Shields

Patients should be moved first to the approximate desired location, and then fluoroscopy should be used very brief to check the position, followed by further adjustment. This is especially important when patients need to be moved by an assistant during the case (e.g., to reposition the arms).

4.2.5 Remove Unnecessary Body Part (or Instruments) from the Field

A typical example of this is leaving the arms in the path of the beam. Leaving the arms in the field results not only in needless radiation exposure to the arms but also in an overall increase in radiation exposure to all the patient's tissues because the radiopaque arms drive the AEC to compensate with increase radiation output. The same can be said for the operator's hands and for any radiopaque instrument in the field.

4.2.6 Always Perform a Test Injection of a Small Amount of Contrast Material Using Fluoroscopy Prior to Acquiring an Angiogram

Fluoroscopy of the test injection can be useful to correct the angiographic projection prior to the actual angiogram, it can aid with determining the correct magnification mode (to prevent the need for panning if the magnification is too high), and it can be stored and reviewed to help make these determinations.

4.2.7 Use the Lowest Acceptable Magnification Mode

The replay zoom features might be helpful in making measurements, at no radiation cost to the patients. Electronic magnification should be used sparingly, because of the substantial increase in the radiation dose it requires. Remember that excessive magnification requires panning to search for the structures of interest, leading to a further increase in radiation dose.

4.2.8 Use Collimators and Partial-Thickness Shields

Collimators are extremely beneficial overall in reducing the volume of tissue exposed to the primary beam and in reducing scatter; reducing scatter is, in turn, beneficial for reducing exposure to laboratory personnel and improving image contrast. As a general rule, the collimators should be visible within the field, and studies should not be performed with the collimator wide open.

4.2.9 Center the Region of Interest Correctly in the Field

The center of the field has the least amount of image distortion; therefore, an angiography should not intentionally be performed at the periphery of the field. Furthermore, bringing the region of interest to the center of the field allows for tighter collimation and less exposure of unnecessary patient tissues to X-rays.

4.2.10 Keep the Image Intensifier as Close to the Patient as Possible (and the X-Ray Tube as Far Away as Possible)

The farther the intensifier is from the patient, the higher the input dose and the scatter to the laboratory personnel. A distant

intensifier also results in geometric magnification, which introduces geometric blur.

4.2.11 Use the Angiographic Projection That Reduces Operator Exposure Whenever Possible

For example, generally the right anterior oblique projection moves the X-ray tube away from the operator, while the left anterior oblique projection moves it closer.

4.2.12 Decrease Beam-On Time

Fluoroscopy must not be applied while discussing or contemplating the next maneuver. It is important to remember that if the eye is not on the screen, the foot should not be on the fluoroscopy pedal.

4.2.13 Remove Anti-Scatter Grids When Catheterizing Small Children

A significant reduction in radiation dose is possible without compromising image quality.

4.2.14 Use X-Ray Stand Position Memory

Useful projections can be stored in the systems memory, allowing rapid return without the need for fluoroscopy to verify position.

4.2.15 Use Biplane Fluoroscopy, Roadmap, and Overlay Features

These features allow vessels of interest to be found with minimal trial and error.

4.2.16 Catheter Selection

An end-hole catheter is useful for selective, relatively small-volume injections by hand, such as into coronary arteries, aortopulmonary collaterals, and other small- or medium-sized arteries. Contrast injection into the cardiac chambers, main pulmonary trunk, or aorta should be made through a multi-side-hole catheter. Multiple side holes facilitate high contrast flow rates, high velocity of injection, and minimal catheter whip.

4.2.17 Contrast Delivery

In general, for anatomic definition, contrast should be delivered through the catheter as rapidly as possible, generally in 1 or 2 s. As a general rule, the volume of contrast recommended in each injection could be 1–1.5 cc/kg in a cardiac chamber or the aorta and 0.5–1 cc/kg in pulmonary branches (maximum volume of 30–40 cc per injection). A high-flow rate is much important than volume for a good anatomic definition. Viscosity of contrast medium is inversely related to temperature; therefore, warming the contrast medium may facilitate high-flow injection through lower profile catheters. Most catheter laboratories keep contrast in a warming cabinet, and injectors usually have a device to keep warm the contrast throughout the procedure [1, 2].

4.3 Contrast Media

The remarkably high tolerance of modern contrast media has been achieved through successive developments in chemical pharmacological technology. Nonetheless risks associated with contrast media (CM) have not been completely eliminated, and adverse reactions of varying degree continue to occur. Consequently, it is imperative that anybody administering contrast agents is familiar with the characteristics, indications, and potential side effects of these agents.

All intravascular iodinated CM are based on a tri-iodinated benzene ring. High-osmolar contrast media (HOCM) are the oldest agents. They are relatively inexpensive, but their utility is limited. They are monomers (single benzene ring) that ionize in solution with a valence of −1. Their cation is either sodium or meglumine. A major advance was the development of nonionic compounds. They are monomers that dissolve in water but do not dissociate in solution. Hence, with fewer particles in solution, they are designated low-osmolar contrast media (LOCM). The most recent class of agents are dimers that consist of a molecule with two benzenes (again, each with 3 iodine atoms) that do not dissociate in water (nonionic). These compounds are called iso-osmolar contrast media (IOCM) (Fig. 4.1). The toxicity of CM decreases as osmolality approaches that of serum. HOCM have an osmolality of 1570 mosm/kg H_2O, while IOCM have an osmolality similar to serum at 290 mosm/kg H_2O.

Since the purpose of these agents is to deliver iodine in sufficient concentration for imaging, the ratio of iodine atoms to particles in solution becomes important. The ratios are as follows: HOCM 5, LOCM 3.0, and IOCM 6.0. Currently used iodinated agents are cleared almost completely by glomerular filtration. With reduced renal function, there is excretion primarily in the bile and through the bowel. Circulatory half-life is 1–2 h, assuming normal renal function. In modern clinical practice ionic CM are rarely used in catheterization laboratories. IOCM have the lowest osmolality and more iodine atoms per molecule, producing the best contrast image. However, these are very expensive so the nonionic monomers (LOCM) remain the most widely used even in pediatric patients (Fig. 4.2).

4.4 Contrast Reactions

4.4.1 Anaphylactoid Reactions

These are essentially anaphylactic reactions but are not initiated by an allergen-IgE complex. Indeed the pathway by which the mast cells become stimulated has not been clarified. The reaction

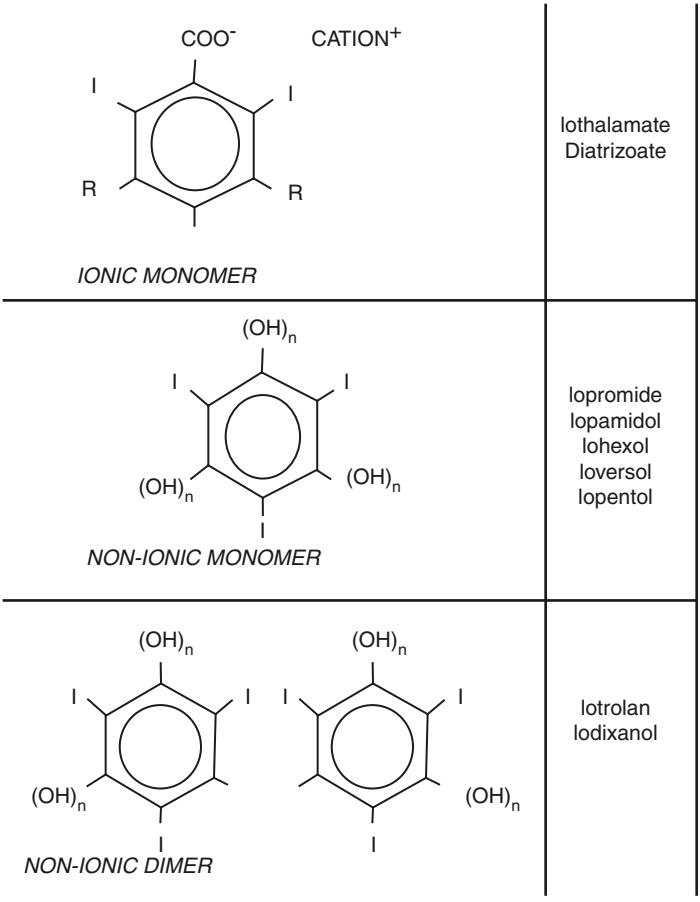

Fig. 4.2 Chemical structure of iodinated contrast agents and examples of contrast media

can occur even the first time contrast is administered, and the severity is not dose related. Treatment is similar to other conventional anaphylactic reactions.

Patients who are at increased risk for anaphylactoid reaction may benefit from premedication. Such patients include those with asthma, allergies, or a history of a prior moderate or severe

reaction to contrast. In this situation methylprednisolone and diphenhydramine are used.

4.4.2 Nonanaphylactoid Reactions

4.4.2.1 Chemotoxic, Organ Specific

Nephrotoxicity

Although institutional criteria vary, in general acute renal failure is defined when serum creatinine raises 25–50% or 0.5–1 mg/dL. Serum creatinine peaks in 3–5 days but may be elevated as early as the first day. In young children creatinine levels may not be sensitive enough to detect renal failure; in these patients cystatin C levels or glomerular filtration values may be a more appropriate test. Risk factors for renal insufficiency induced by contrast are age >65 years, diabetes, end-stage liver disease, and severe congestive heart failure. Clinical manifestations are highly variable and may range from completely absence of urine to oliguria. Most effects are temporary and reversible. In mild cases, serum creatinine returns to normal in 2 weeks. When severe, dialysis may be necessary. The major preventive action against nephrotoxicity is to ensure adequate hydration. One possible protocol would be 0.9% saline at 100 mL/h, beginning 6–12 h before and continuing 4–12 h after intravascular iodinated contrast medium administration. Pediatric infusion rates are variable and should be based on patient weight.

Cardiovascular Toxicity

Patients with underlying cardiac disease have an increase incidence and/or severity of cardiovascular side effects. Pulmonary angiography and intracardiac and coronary artery injections carry the highest degree of risk. Possible reactions include hypotension, tachycardia, and arrhythmias. More severe but uncommon reactions include congestive heart failure, pulmonary edema, and cardiac arrest.

Neurotoxicity

Iodinated contrast agents cause a change in the blood–brain barrier due to their hypertonicity. These risks are reduced when low- or iso-osmolar agents are used. Potential reactions include headache, confusion, seizures, altered consciousness, visual disturbances, and dizziness.

Thyroid Dysfunction

Iodine-based contrasts media (IBCM) exposure has been associated with increased risk of thyroid dysfunction, especially in children, most commonly hypothyroidism. A single intravenous IBCM bolus can contain approximately 150 times de recommended daily amount of iodine intake for children 1–8 years old. The thyroid gland responds to the excess iodine exposure, by temporally reducing thyroid hormone production over 24–48 h. At present, the incidence and duration of thyroid dysfunction in children from IBCM exposure, as well as clinical significance of this finding, are incompletely characterized. In all reported cases, the infants were either premature and had serious underlying conditions. Although iodine-induced dysfunction can occur in any individual, it is most commonly reported in those with predisposing conditions, such as underlying thyroid disease and chronic or end-stage renal disease.

4.4.2.2 Vasovagal Reactions

These are characterized by bradycardia and hypotension. When a vasovagal reaction occurs, the patient should be put into the Trendelenburg position and atropine and IV fluid (saline or lactated Ringer's) administered if clinically necessary [3].

For most patients intravenous contrast media are well tolerated and require no special attention for appropriate indications. However, a standardized list of pre-imaging screening questions can help to identify patients who require additional consideration to optimize contrast media administration to their specific needs (Table 4.1) [4].

Table 4.1 Screening question for patients prior to intravenous contrast administration and action ítems for positive screening results

1. Allergy to IV contrast media? If yes, what was the reaction? Did you receive any treatment for it? • If yes, ensure the appropriate precautions have been taken prior to administration of intravenous contrast. If an appropriate premedication regimen has not been administered, contact the referring clinician prior to contrast administration
2. Active asthma symptoms (wheezing, shortness of breath)? • If yes, the examination should be performed at a facility where a code team is immediately available unless the same class of intravenous contrast media has been previously administered without incident
3. Kidney transplant, single kidney, kidney cancer, dialysis, diabetes mellitus, high blood pressure requiring medication, surgery within the last 6 weeks, other kidney problems? • Dialysis – If anuric, proceed – If oliguric, discuss with referring clinician prior to IBCM • If yes to a different question, same action items as question 4
4. Acute illness (e.g., multiple episodes of vomiting/diarrhea or unable to eat or drink for 24 h or more within the last week)? • If yes, check recent serum creatinine and calculate eGFR (eGFR is not helpful in the context of acute kidney injury [AKI] or dialysis) • If AKI or CKDe, discuss with referring clinician or nephrologist prior to contrast administration
5. IV contrast exposure in the last 48 h? • If yes, there is a potentially increased risk of CIN when administering additional doses. If the patient has additional risk factors for CIN (see questions 3 and 4), discuss with referring clinician prior to repeat contrast administration, unless the study is emergent
6. Thyroid dysfunction? • Consider discussion of potential risk of superimposed iodine-induced thyroid dysfunction with referring clinician
7. Thyroid cancer or Graves disease? • Confirm that radioactive iodine nuclear medicine study or therapy is not planned for at least 2 months following IBCM administration

AKI acute kidney injury, *CIN* contrast induced nephropathy, *eGFR* estimated glomerular filtration rate, *ICBM* iodine-based contrast media

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5 Angiography: Radiation Exposure and Standard Projections

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5.1 Radiation Exposure Today

Conventional X-ray examination represents 93% of the total of examinations but contributes only to 5% of the collective dose; diagnostic catheterization (41%), interventional catheterization (43%), and CT (11%) are responsible for about 95% of total collective effective dose. Both patients and working staff are at a potential risk to radiation. Factors such as age, body size, distance between the hands and body and X-ray generator, configuration of the of the X-ray equipment, number of cases per day, and length of study contribute to a relatively higher level of exposure in pediatrics. The National Academies' Biological Effects of Ionizing Radiation 7th Report in 2006 (BEIR VII) states its research priority in infants exposed to diagnostic cardiac catheterization.

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5.2 Potential Hazards of Radiation Exposure [1]

5.2.1 Deterministic Risks

Radiation-induced skin reaction may not appear after 2–3 weeks after exposure; threshold for mild transient skin erythema is about 2 Gy, main erythema and epilation are expected with peak skin doses exceeding 6 Gy, 10–15 Gy may cause telangiectasis and chronic radioderma, and 16–18 Gy causes skin ulcerations. High doses of radiation can damage the conjunctiva, iris, sclera, and blood vessels of the retina, but the lens is the critical site for it may sustain irreversible damage from a relatively low dose of radiation and formation of cataracts in the posterior pole of the lens.

5.2.2 Stochastic Risks

Malignancy is a stochastic effect of radiation [2], meaning that there is no clear exposure threshold for development. Most radiation-induced damage is rapidly repaired, but occasional mis-repair of DNA breaks can result in point mutations, chromosomal translocations, and gene fusions linked to induction of cancer. Despite lack of a safe lower threshold, there is no question of the carcinogenic effects of organ doses in the excess of 100 mGy. Some organ tissues are at greater risk (such as the brain, skin, and thyroid) than others (gonads). The incremental fatal cancer risk is estimated at 4% per Gy unit. According to estimates of the BEIR VII report, the lifetime attributable risk values of cancer incidence from a single cardiac catheterization were 2.1 and 0.8% for female and male patients, respectively. In the typical patient dose range related to diagnostic and interventional use of X-rays (0–50 mSv), the associated cancer risk cannot be deduced from epidemiological data owing to a lack of statistical power so risk estimates for late effects have been based on a linear no-threshold model extrapolated from high-dose data as obtained in the lifespan study of atomic bomb survivors that support the concept that no radiation

doses, no matter how small, can be considered safe without a threshold safe dose. This model assumes that the DNA damage is proportional to the dose and that cellular responses operate equally efficient at low and high radiation doses. An attractive approach to study the deleterious effects of low-dose X-ray exposure is the use of biomarkers. DNA double-strand breaks are the most relevant type of lesions responsible for late effects of ionizing radiation and chromosomal aberrations, and micronuclei in peripheral blood lymphocytes are validated biomarkers of somatic chromosomal damage and intermediate end points in carcinogenesis.

5.3 Special Problems of Medical Radiation in Children [2]

For any given dose, children are three to six times more sensitive to the induction of cancer as they have more rapidly dividing cells and longer life expectancy than adults. Also, for a given procedure, dose is larger in a small infant than in an adult, and organs are closer resulting in more radiation dose. Children with congenital heart disease frequently undergo repeat imaging, with each examination adding to the cumulative lifetime risk.

Corresponding estimated lifetime attributable risk of cancer in the age 0–15 years is 1 in 804 (1 in 1717 for fatal cancer) for male subjects receiving 7.1 mSv and 1 in 331 (1 in 859 for fatal cancer) for female subjects receiving 9.4 mSv. However, risks are 1.9–2 times higher for child aged 1 year (1 in 382 for males and 1 in 156 for female patients both for fatal and nonfatal cancer) than for a 15-year-old. Cancers occur after a latency period of at least 5–10 years for most solid cancers and approximately 2 years for leukemia. Surprisingly, only two cohort studies, one from J. MacLaughlin and another from B. Modan, with controversial and confronting conclusions have assessed the association between the risk of cancer in children and radiation exposure during pediatric catheterization.

5.4 Terminology [1]

Total *air kerma* (K, Gy units) is the procedural cumulative X-ray energy delivered to air at the interventional reference point. It is used to monitor patient dose burden and is associated with threshold-dependent deterministic skin effects. New laboratories deliver radiation doses ranging from 23.4 nGy per pulse at a 25-cm flat-panel detector field to 56.6 nGy per pulse at a 16-cm field.

The International Commission on Radiological Protection (ICRP) recommends the use of *effective dose* (E) to evaluate the effects of partial exposure and relate this to the risk of equivalent whole-body exposure. The E characterizes stochastic cancer risk. It takes into account both the type of radiation and the nature of each organ being irradiated reflecting the different importance of tissue types to the danger to the whole organism. The unit for *effective dose is the Sievert* (*Sv*). One Sv carries a 4% chance of developing a fatal cancer in an average adult and a 0.8% chance of hereditary defect in future offspring. Modern cardiac interventional procedures, angiography and interventions, produce effective doses of 4–21 mSv and 9–29 mSv, respectively, and are therefore relatively high (1 mSv is the equivalent of approximately 10 chest X-rays). Published effective doses for pediatric catheterization range from 2.2 to 12 mSv, but there is a wide variation from one center to another in indications, child's age and weight, follow-up, etc.

Dose area product (DAP)—the standard unit is Gray square centimeter – is defined as the absorbed dose multiplied by the area irradiated, and it is the measure reflected in angiographic studies indicating the total X-ray energy delivered to the patient as a result of fluoroscopy and cine-film sequences (Fig. 5.1). It represents the radiation dose in the air at a given distance from

Fig. 5.1 (**a**, **b**) Dosimetric doses report in two regular angiographic studies. Total DPA dose related to cine acquisition with frame/s and projection. Radiation in the cath lab is generated using two different modes: fluoroscopy or "cine" angiography. Fluoroscopy involves 95% of the total X-ray operation time but only causes 40% of the total radiation dose; cine represents only 5% of the total X-ray tube operation time but 60% of the total radiation exposure

a

Protocolo de examen

```
Info pacient:
Nomb:                                        Sexo: F  ID:
-------------------------------------------------------------------------------
Pos. d. paciente:  HFS                                          04-Sep-13 10:54:17

Pos. d. paciente:  HFS                                          04-Sep-13 10:54:28

4    CARD          FIXED    LV 3040                  5s  30F/s  04-Sep-13 12:08:43
A  73kV   99mA    3.4ms  0.0CL large       22cm    75.7µGym²  9.4mGy   0LAO  25CRA 146F

4    CARD          FIXED    LV 3040                  5s  30F/s  04-Sep-13 12:08:43
B  73kV  293mA    3.5ms  0.0CL large       20cm    85.3µGym²  16.6mGy  90LAO   0CRA 146F

5    CARD          FIXED    LV 3040                  5s  30F/s  04-Sep-13 12:21:42
A  73kV  122mA    3.4ms  0.0CL large       22cm    90.4µGym²  11.3mGy  0LAO  25CRA 145F

5    CARD          FIXED    LV 3040                  5s  30F/s  04-Sep-13 12:21:42
B  73kV  406mA    3.5ms  0.0CL large       20cm   116.1µGym²  22.6mGy  90LAO   0CRA 145F

6    CARD          FIXED    LV 3040                  6s  30F/s  04-Sep-13 12:28:52
A  73kV  103mA    3.4ms  0.0CL large       22cm    89.7µGym²  11.2mGy  0LAO  25CRA 168F

6    CARD          FIXED    LV 3040                  6s  30F/s  04-Sep-13 12:28:53
B  73kV  336mA    3.5ms  0.0CL large       20cm   112.8µGym²  21.9mGy  90LAO   0CRA 168F


***Datos de expos. acumulada***                                 04-Sep-13 13:36:48
Pie.: Dr.                Expos.: 6      Fluoro:  21.2min Total: 1034.6µGym²  148.3mGy
A    Fluoro:   20.8min     451.8µGym²   53.1mGy          Total:  707.6µGym²   85.1mGy
B    Fluoro:    0.4min      12.8µGym²    2.2mGy          Total:  327.0µGym²   63.3mGy
```

b

Protocolo de examen

```
Info pacient:
Nomb:                                        Sexo: F  ID:
-------------------------------------------------------------------------------
Pos. d. paciente:  HFS                                          05-Sep-13 08:51:39

1    CARD          FIXED    Card <12kg               4s  30F/s  05-Sep-13 11:04:26
A  62kV  722mA    3.6ms  0.0CL large 0.6Cu 16cm     8.8µGym²   2.1mGy   0LAO  32CRA 109F

1    CARD          FIXED    Card <12kg               4s  30F/s  05-Sep-13 11:04:26
B  62kV  728mA    3.9ms  0.0CL large 0.6Cu 16cm     5.1µGym²   1.6mGy  90LAO   0CRA 109F

2    CARD          FIXED    Card <12kg              10s  30F/s  05-Sep-13 11:05:15
A  62kV  728mA    3.8ms  0.0CL large 0.6Cu 16cm    25.5µGym²   6.2mGy   0LAO  32CRA 290F

2    CARD          FIXED    Card <12kg              10s  30F/s  05-Sep-13 11:05:15
B  66kV  273mA    3.4ms  0.0CL large 0.3Cu 16cm    16.3µGym²   5.2mGy  90LAO   0CRA 290F

3    CARD          FIXED    Card <12kg               4s  30F/s  05-Sep-13 11:08:21
A  66kV  358mA    3.5ms  0.0CL large 0.3Cu 16cm    12.5µGym²   3.5mGy  43RAO  23CRA 112F

3    CARD          FIXED    Card <12kg               4s  30F/s  05-Sep-13 11:08:21
B  66kV  736mA    4.3ms  0.0CL large 0.3Cu 10cm    11.6µGym²   6.8mGy  48LAO  30CRA 112F

4    3D            DYNAAUT  SaDR-L                   5s  30F/s  05-Sep-13 11:15:55
A  67kV   99mA    3.4ms  0.0CL small 0.1Cu 48cm    76.6µGym²   2.8mGy  99RAO   0CRA 133F

5    CARD          FIXED    Card <12kg               3s  30F/s  05-Sep-13 11:20:26
A  62kV  553mA    3.5ms  0.0CL large 0.6Cu 22cm    12.9µGym²   1.4mGy   0LAO   0CRA  99F

5    CARD          FIXED    Card <12kg               3s  30F/s  05-Sep-13 11:20:26
B  66kV  344mA    3.5ms  0.0CL large 0.3Cu 20cm    10.3µGym²   2.1mGy  90LAO   0CRA  99F

6    CARD          FIXED    Card <12kg               4s  30F/s  05-Sep-13 11:21:17
A  62kV  589mA    3.5ms  0.0CL large 0.6Cu 22cm    10.8µGym²   1.7mGy   0LAO   0CRA 108F

6    CARD          FIXED    Card <12kg               4s  30F/s  05-Sep-13 11:21:17
B  66kV  375mA    3.5ms  0.0CL large 0.3Cu 20cm    12.3µGym²   2.5mGy  90LAO   0CRA 108F

7    CARD          FIXED    Card <12kg               4s  30F/s  05-Sep-13 11:44:42
A  66kV  347mA    3.5ms  0.0CL large 0.3Cu 16cm    13.5µGym²   3.9mGy  37RAO   1CRA 127F

7    CARD          FIXED    Card <12kg               4s  30F/s  05-Sep-13 11:44:42
B  66kV  525mA    3.5ms  0.0CL large 0.3Cu 20cm    29.0µGym²   4.4mGy  45LAO  32CRA 127F

8    CARD          FIXED    Card <12kg               8s  30F/s  05-Sep-13 12:11:44
A  62kV  727mA    3.8ms  0.0CL large 0.6Cu 16cm    16.2µGym²   4.8mGy   8RAO   1CAU 236F

8    CARD          FIXED    Card <12kg               8s  30F/s  05-Sep-13 12:11:44
B  62kV  727mA    3.8ms  0.0CL large 0.6Cu 20cm    21.3µGym²   3.5mGy  81LAO   6CRA 236F


***Datos de expos. acumulada***                                 06-Sep-13 07:42:13
Pie.: Dr.                Expos.: 15     Fluoro:  56.8min Total:  485.9µGym²  99.5mGy
A    Fluoro:   36.9min     120.1µGym²   31.6mGy          Total:  297.0µGym²  58.1mGy
B    Fluoro:   19.9min      82.9µGym²   15.4mGy          Total:  188.9µGym²  41.4mGy
```

the X-ray tube multiplied by the area of the beam at that distance. Coronary angiography and interventions produce DAPs in the range 20–106 Gy cm^2 and 44–143 Gy cm^2, respectively. In order to estimate the risk of radiation-induced sequelae, the dose area product (Gy cm^2) must be converted to the effective dose (mSv).

In computed tomography (CT), *dose length product* (DLP) is the standard dose measurement reported, expressed in mGy.cm. Important differences in doses are obtained with various CT protocols with saving algorithms, with/without cardiac gating, type of gating (prospective/retrospective), etc. Early studies with retrospective gating and no dose modulations delivered doses >15 mSv. Estimated lifetime risk of cancer is as high as 1:143 for a 20-year-old woman if a scan was performed retrospectively with no modulation (old explorations).

The scanner-derived DLP and the catheterization-derived DAP do not allow comparisons. Effective dose (mSv) allows cross-modality comparisons of radiation doses. In cardiac CT scan, the most common method of estimating the effective dose is the use of a conversion factor applied to the dose length product (DLP); conversion factors previously used in the literature varied between 0.014 and 0.019 mSv/mGy·cm and are based on tissue weightings published and updated by the ICRP. There are also PC-based programs that calculate effective doses with data from DAP, projection angle, kV, field size, duration of exposure, frames per second during acquisition, and patients' height and weight. Conversion factors for conventional invasive coronary angiography have also been used but are based on older ICRP tissue weightings; published conversion factors vary widely from 0.12 to 0.26 mSv/Gy cm^2 with 0.26 mSv/Gy cm^2 the most common frequently simplified to 0.20 mSv/Gy cm^2. An average cardiac computed tomography and angiographic examination are equivalent to almost 100 and 300 chest X-rays, respectively (a single chest X-ray = 0.02 mSv).

5.5 How to Manage Radiation Doses for Invasive Cardiac Procedures [1]

5.5.1 Preprocedure

Use dosimeters and proper shielding: shields, lead curtains, aprons, and glasses. Learn and know about radioprotection and know your own screen dose assessment. Study on your equipment how to store fluoro, adjust pulse, and frame rate. Plan your study.

5.5.2 Procedure

Keep in mind the current mandate and use radiation "as low as reasonably achievable" (ALARA concept). Limit fluoro and cine. Store fluoro when image quality is not required. Achieving an adequate image, as opposed to the highest quality image that is possible, is a basic principle. Image intensifier low-level modes should be used as often as possible: try to work, both in fluoroscopy and in acquisition modes, with the lowest frame rate. Distance between the X-ray tube and the patient should be maximized, keeping the intensifier or flat-panel detector as close to the patient as possible. Use the lowest degree of magnification required for accurate interpretation. Minimize radiographic beam time ("cine" acquisition creates 12–20-fold higher-dose intensities than fluoroscopy mode). Acquisitions represent 60–70% of the total DAP during a typical study. Collimation is an efficient radiation-reducing factor, and modern systems have virtual control of collimation. There are less irradiating angulations: 20° right anterior oblique gets the lowest patient DAP, cranial and caudal angulations rose the doses significantly and are maximum in left lateral angulations. The operator fatigue also raised radiation exposure to 28% due to more and longer radiographic runs after working for more than 6 h. Remember, an adequate use of

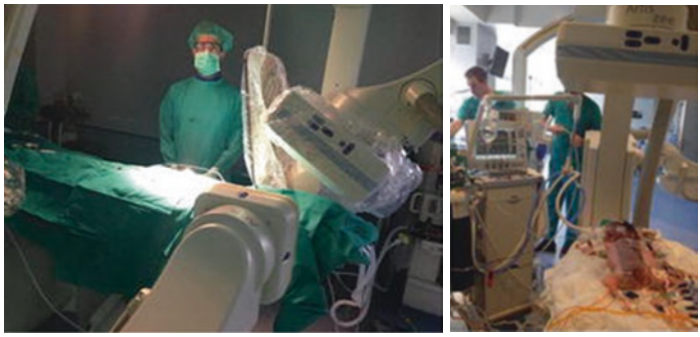

Fig. 5.2 Pediatric patients are often very sick and need ventilator, pumps, GE tubes, etc. Patient should be close to flat detector or intensifier; operator should use shields, curtains, glasses, collars, and apron. All these cautions make often impossible to achieve convenient angulations or biplane combinations to allow catheter, sheaths, or wire manipulations in small children

filters, especially for small (<15 kg) and the simpler rule than doubling the source to operator distance will decrease operator dose to approximately one quarter. Operator and personnel exposure are directly related to the dose area product: when operating in a biplane cine-acquisition mode, scattered radiation multiplies by a factor between 5 and 21. Pediatric cardiologists could easily achieve a lens opacification threshold so they should wear glasses, apron, and collars and should correctly use shields, curtains, etc. (Fig. 5.2).

5.5.3 Postprocedure

Document radiation dose in records. Follow high-dose patients for the next weeks and refer to appropriate consultant if skin effects appear.

5.6 Angiographic Projections

Structural heart procedures are focused on chambers and structures larger than the 3-mm field of coronary procedures and are at greater risk of the limitations and artifacts of the *Z*-axis. Angiography produces a two-dimensional projection, and so multiple orthogonal views to minimize the impact of the "*Z*-axis" on the image are needed to understand heart and vessels in three dimensions. In the past, X-ray systems were fixed, and oblique and angled projections were achieved by changing the position of the patient on the table. Fortunately today, modern C-arms can be turned to get such projections.

The main idea is to get axial, non-overlapped, or foreshortened profile of the various structures; many and different angulations will be needed with great variations for the same structure or disease in different patients. Although bidimensional vascular angiogram will continue to lay a central role, advanced digital imaging and detailed three-dimensional reconstructions will enable more accurate diagnosis, minimizing contrast dye burden and radiation to patient and operators.

Every case should be prepared in anticipation, paying special attention to previous angiographies, CT, and MRI because in most cases, the projection which will optimize the profile could be anticipated looking at 3D reconstructions coming from these studies. Imaging workstations include packages that attempt to facilitate multimodality (fluoroscopy, ultrasound, CT, and MRI) fusion, but there are still important limitations for their use in real time such as the time consumed and the requirement of an expertise not easy to be gained; for instance, special attention needs to be paid to measurements as there are important differences for the same lesion between different techniques. Do not forget that angiographic images have a better temporal resolution, whereas MRI and CT images are merged pictures of the diameters during the whole cardiac cycle. The future will be the integration of imaging modalities; the information gained by one technique enhances and is incorporated in an additive manner to the information acquired by other technique.

Angiographic computed tomography (ACT) provides cross-sectional CT images from a rotational angiography run using a C-arm-mounted flat-panel detector. The volume set obtained can be manipulated on a separate workstation in the interventional suite to generate a 3-dimensional angiographic picture combined with CT-quality soft tissue imaging that can be used in real time during the procedure. ACT is useful to define the optimal camera angles (Fig. 5.3) for a planned intervention under standard biplane or single-plane fluoroscopic guidance allowing to choose the best

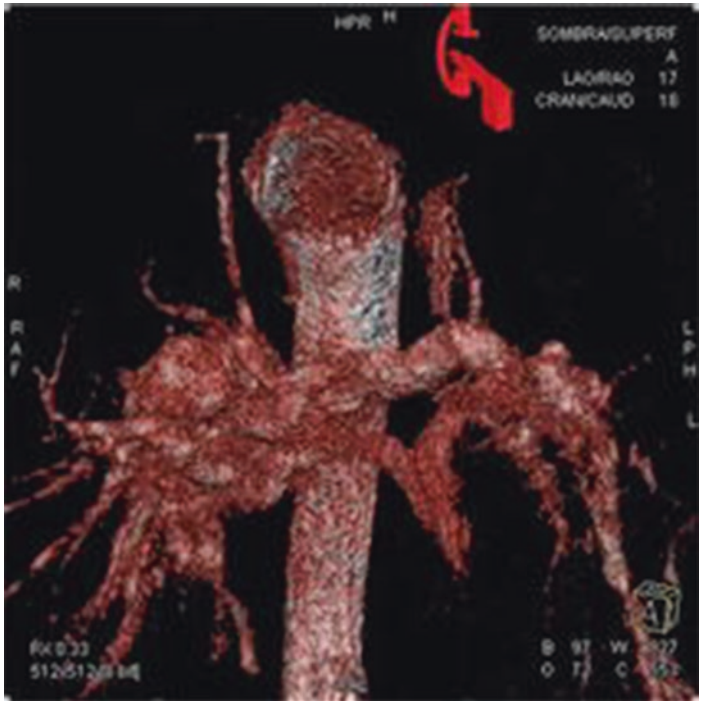

Fig. 5.3 3D reconstruction from an angiographic computed tomography (ACT) acquired and processed during the catheterization. Best C-arm angulation can be predicted upon figure movement. Aortogram from an infant with pulmonary atresia and ventricular septal defect with multiple aortopulmonary collaterals

oblique angulations. There is still limited experience with this modality that needs some extra training: images obtained are time-averaged over the duration of 5-s arm rotation over 210ª, there are dropouts of the signal in areas of very tight stenosis or adjacent to stented regions, images are hand manipulated by the degree of windowing during post-processing, etc. Anyway, in selected cases, ACT has advantages over conventional CT: images are easily obtained in the same procedure and can be used to guide catheter manipulations serving as a 3D overlay road mapping, total radiation seems to be similar to length cine acquisition, and contrast dose is less or equal to that used to CT. But we lack enough information on the added benefits of that technique. Do we need the additional radiation and contrast exposure to perform preprocedural CT scans and C-arm procedural scans on some patients?

Although there is no general agreement, biplane equipment both reduces total contrast dose (not an insignificant problem) and helps to figure out the area of interest but not always with a significant total X-ray dose reduction. Projections used for angiocardiography include frontal, lateral, right, and left oblique, with or without axial (craniocaudal or caudocranial) angulations. The choice of a set of projections will depend upon the information required, equipment capabilities, and the physical constraints to patient access. Standard biplane configurations include RAO/LAO and frontal or lateral projections, with additional cranial or caudal tilt, but possible combinations are endless with many local or personal variations.

A cookbook for every intervention in every case is not possible and is not possible in this short chamber. There are a number of "rules of thumb": the first step is to achieve the correct degree of steepness or shallowness. After that, the degree of cranial or caudal tilt should be chosen (Table 5.1).

Attention should be paid not only to get fine pictures but to get pictures that can be used. It is worthless to get an angulation such that the image generator position will preclude to work with catheters, sheaths, wires, etc. (Fig. 5.4).

Table 5.1 Recommended projections

Projection	Angles, plane A	Angles, plane B
Conventional RAO	40° RAO	
Frontal	O°	
Shallow LAO	1–30°	
Straight LAO	31–60°	
Steep LAO	61–89°	
Left lateral	90° left	
Cranially tilted RAO	30° RAO + 30° cranial	
Cranially tilted frontal (sitting up view)	30–45° cranial	
Cranially tilted shallow LAO	25° LAO + 30° cranial	
Cranially tilted mid LAO (long axis oblique)	60° LAO +30–30° cranial	
Cranially tilted steep LAO (hepatoclavicular view)	45–70° LAO +30° cranial	
Caudally tilted frontal	45° caudal	
AP and LAT	0°	Left lateral
Long-axis oblique	30° RAO	60° LAO +20–30° cranial
Hepatoclavicular view	45° LAO + 30° cranial	120° LAO + 15° cranial
Specific lesions	Angles, plane A	Angles, plane B
Pulmonary stenosis	0° + 30° cranial	Left lateral
RVOT-MPA (sitting up)	10° LAO + 40° cranial	Left lateral
Long axial for LPA biplane	30° RAO	60° LAO + 30° cranial
LPA long axis		60° + 20° cranial
ASD	30° LAO + 30° cranial	
PA bifurcation and branches	30° caudal + 10° RAO	20° caudal
Left ventricular outflow tract obstruction	RAO	Long-axis oblique
Aortic coarctation	0°/shallow RAO/ shallow LAO	Left LAT/long axis oblique

Table 5.1 (continued)

Projection	Angles, plane A	Angles, plane B
Ventricular septal defect perimembranous		Long-axis oblique
Ventricular septal defect inlet and muscular		Hepatoclavicular view
Ventricular septal defect outlet	RAO	
Patent ductus arteriosus	30° RAO	Left lateral/left lateral + caudal
Mustard superior baffle obstruction	30° LAO + 30° cranial	
Mustard inferior baffle obstruction	Frontal	
Surgical fistula between supraortic arch and branch pulmonary artery	Shallow RAO/ LAO	
Fontan operation, tunnel/conduit obstruction	0°	Left lateral
Fontan operation, fenestration	Shallow RAO/ LAO	

5.7 Specific Lesions (In Situs Solitus with Left Aortic Arch) [3] (Fig. 5.5)

5.7.1 Secundum Atrial Septal Defect and Fenestrated Fontan

Secundum atrial septal defects are best profiled in the 30° LAO + 30° cranial. If balloon sizing is performed, this projection will elongate the axis of the balloon allowing proper measurements.

5.7.2 Ventricular Septal Defect

The location of the defect should be well studied before catheterization so the best projection could be chosen. For the perimembranous defect, the mid-cranial LAO projection at about 50–60°

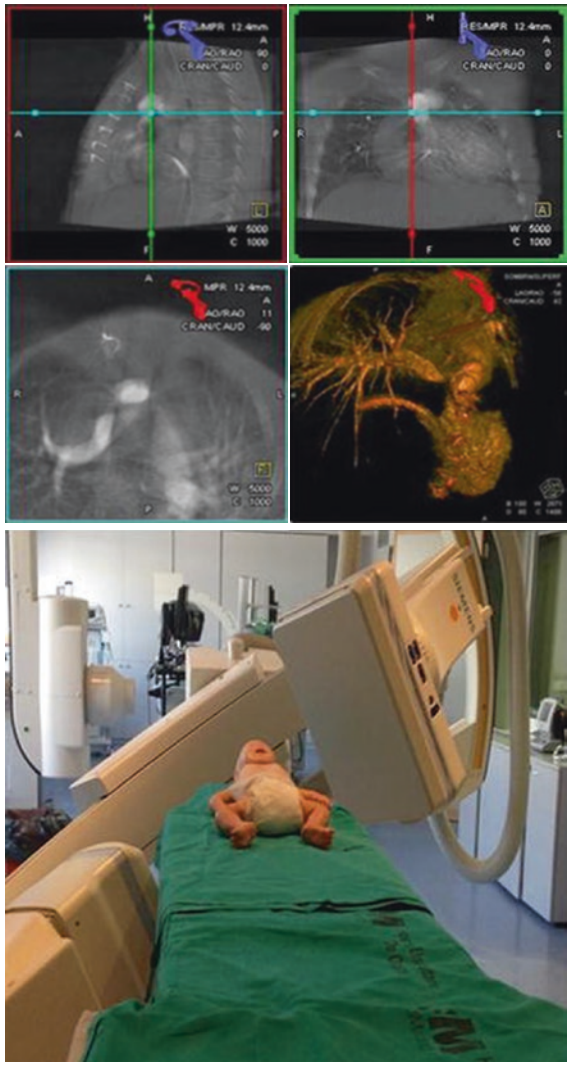

Fig. 5.4 In some cases despite the fact that tomographic views and 3D reconstructions can help to choose a theoretically adequate C-arm angulation, this cannot actually be achieved. In some occasions, it is mechanically unaffordable and in others there are important limitations when working which make it impractical

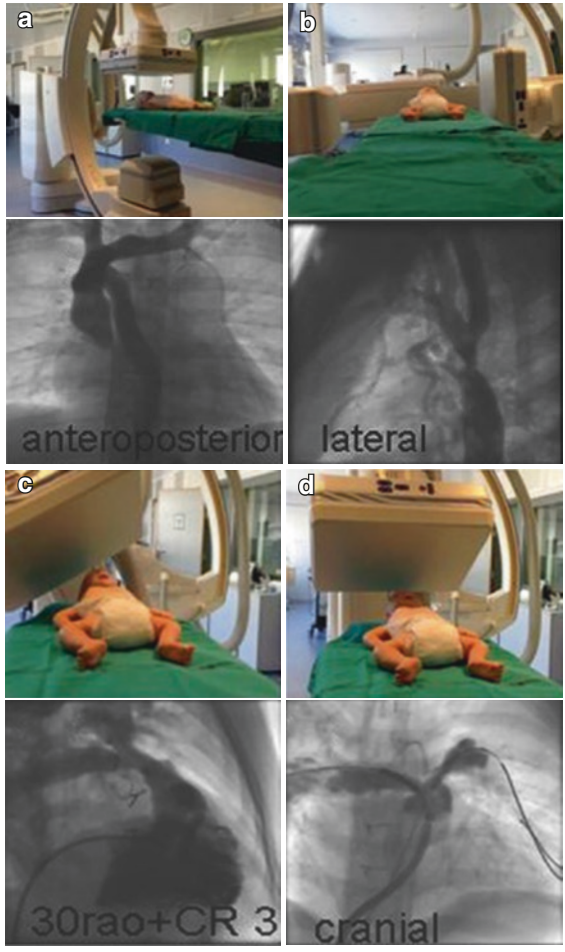

Fig. 5.5 (**a**) Mustard with severe superior vena cava stenosis, anteroposterior projection. (**b**) Native aortic coarctation in lateral projection. (**c**) Postoperative severe supravalvular stenosis in a shallow right oblique with 30° cranial projection, (**d**) severe bilateral stenosis at the origin of both pulmonary arteries post arterial with operation in a shallow cranial angulation. (**e**) Interatrial septum in a shallow right oblique with 30° cranial angulation, (**f**) aortic arch "opened" in a deep left lateral oblique 70° with a shallow 20° cranial angulation ("long axis"). (**g**) Multiple muscular ventricular septal defects 40° left oblique plus 40° cranial ("four-chamber view"), (**h**) postGlenn patient with no stenosis at the anastomosis studied in a shallow left oblique with a shallow caudal angulation

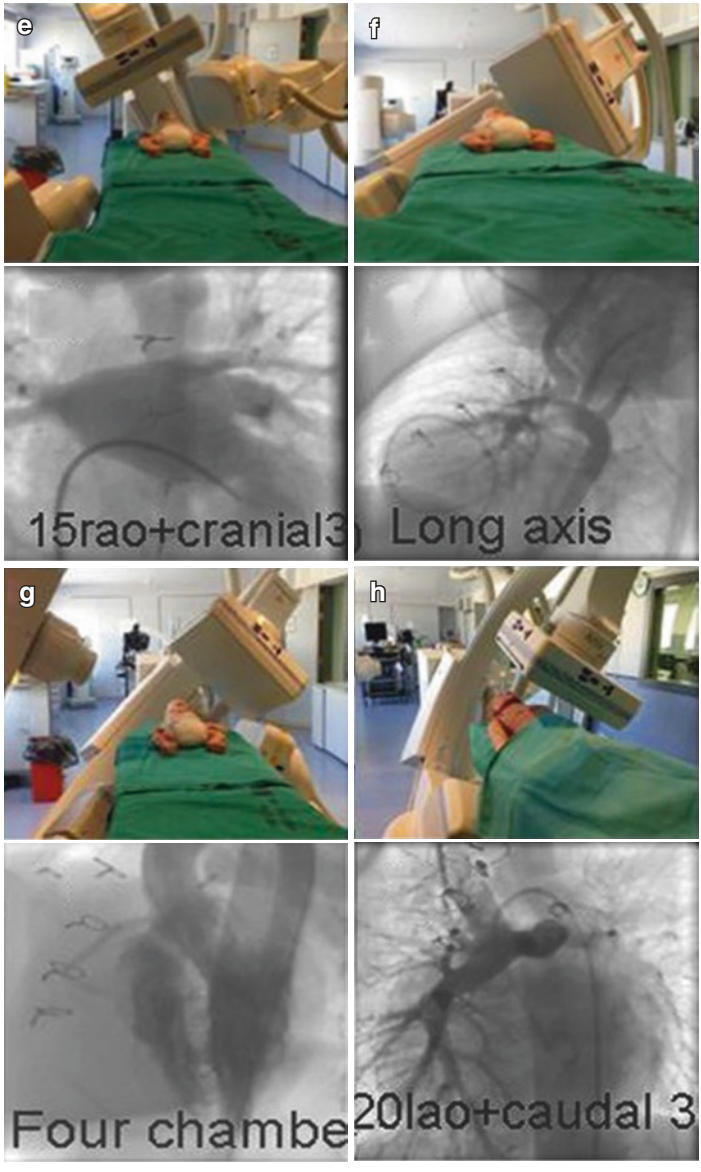

Fig. 5.5 (continued)

LAO and as much cranial tilt as the conditions allow are the best. Posterior defects are better outlined in a four-chamber view. Simultaneous orthogonal RAO in biplane system will help to profile the defect. RAO view will outline the high anterior and infundibular (outlet) defects. Patients with muscular VSD frequently have more than one defect, so that multiple projections are useful to evaluate the entire septum, probably beginning with a four-chamber view.

5.7.3 Patent Ductus Arteriosus

In most of the cases, closure can be straightly performed just with the lateral plane. If not well defined, a simultaneous or added shallow RAO will nicely demonstrate the ductus. In patients where ductal arch and aortic arch are overlapped some caudal tilt in the plane will help.

5.7.4 Surgical Fistula Between Supraortic Arch and Branch Pulmonary Artery

Usually, the vessel profile is obscured by the aortic arch with frontal projection, so depending on fistula, side shallow RAO or LAO is needed.

5.7.5 Aortic Valve

In the setting of normally related great arteries with ventricular arterial concordance diameter of the aortic valve, it is best performed using biplane in the long axis and RAO projections. Supravalvular aortic stenosis would need additional injections to profile coronary ostia.

5.7.6 Coarctation of the Aorta

It could be outlined in PA and LAT and shallow or steep LAO/RAO. If working in biplane, 30° LAO and left LAT + 15° caudal tilt minimize ductal bump or diverticulum overlapping. Be cautious with the transverse aortic arch that is best studied in a left posterior oblique, especially if a stent should be implanted near the head and neck vessels.

5.7.7 Mustard Baffle

If looking for superior baffle obstruction, 30° LAO + 30° cranial best outlines the lesion. Pay attention to ascending aorta as it is really close. If looking for inferior baffle lesions, a frontal projection adequately shows obstruction profile. Leaks are more difficult to categorize because of the many spatial possibilities so start with a frontal projection.

5.7.8 Bidirectional Cavopulmonary Connection

Caval to pulmonary connection is toward the anterior surface of the right pulmonary artery rather than on the upper surface, and so AP projection will overlap the anastomotic site; a 30° caudal + 10° LAO will open the area and allows an outline to the full extent of the right and left pulmonary arteries, and plane B in the left LAT with or without 10° caudal angulation will profile the anterior-posterior dimension.

Venous collaterals can be examined in AP + LAT.

5.7.9 Fontan Operation

In Fontan patients whether lateral tunnel or extracardiac connection, both superior and inferior caval veins and pulmonary circulations should be studied to determine if venous pathways are

patent and whether venous collaterals have developed. Fenestrations are best profiled with some degree of right or left anterior obliquity, and collaterals are best studied in AP and LAT projections.

5.7.10 Pulmonary Valve Stenosis, Tetralogy of Fallot, and Pulmonary Valve Atresia with Intact Ventricular Septum

In pulmonary stenosis and right ventricular outflow tract lesions, AP projection will foreshorten the structures. A 30° cranial with 15° LAO will open the infundibulum allowing visualization of the pulmonary valve and main and branch pulmonary arteries. The best definition to measure the valve is the left LAT projection with an additional 10–15° caudal angulation of the lateral detector to separate branch vessels (in that case, there will be foreshortening of the outflow tract).

5.7.11 Branch Pulmonary Artery Stenosis

These represent the most difficult angiographic studies. In each case, modifications for general rules must be made and there are many personal preferences.

A cranial tilt frontal projection with a left LAT or RAO/LAO projection is frequently the first injection and allows seeing proximal and hilar regions. Since there is frequent overlapping, these standard views can be modified by increasing or decreasing the degree of RAO or LAO and adding caudal or cranial tilt.

For the right pulmonary artery, a shallow RAO projection with a 10–15° cranial tilt separates the upper and middle lobe branches, and a left LAT with 15° caudal tilt will open up all the anterior vessels.

To study the posterior leftward direct left pulmonary artery, a 60° LAO with 20° cranial with a caudal tilt on the lateral detector is recommended.

If the main pulmonary artery is aneurysmatic and obscures the confluence, a steep 30° caudal in plane A plus 10–20° RAO will open the bifurcation.

5.7.12 Total Anomalous Pulmonary Venous Connection

Inject in the main pulmonary artery or separately in each branch if pulmonary venous hypertension is known or suspected, and film in PA and LAT projections.

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3. Benson L, Mikailian H. Angiography. In: Sievert H, Qureshi SA, Wilson N, Hijazi ZM, editors. Percutaneous interventions for congenital heart disease. London: Informa UK; 2007. p. 15–31.

Catheters and Wires

6

Adam Koleśnik
and Grażyna Brzezińska-Rajszys

6.1 Diagnostic Catheters

Diagnostic catheters are thin-walled tubes introduced into patient's vessels and the heart via the valved introducer sheaths. Structure of the catheter, its geometry, and other characteristics depend on the purpose it serves. There are many designs and technical solutions created by numerous manufacturers of catheterization equipment. Catheters are named according to their shapes, people who designed them, or the vessels they are supposed to enter. The basic principle of catheter selection, however, is that they must serve the purpose they are suitable for. Thus, in a

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pediatric cardiac catheterization laboratory, one often uses catheters designed for procedures other than those being performed. Nevertheless, there are some basic catheter categories that the operator has to be familiar with.

6.1.1 Anatomy of the Catheter

Although diagnostic catheters usually look like simple plastic tubes, their construction is quite complex. Materials used should be safe for the patient, assure maneuverability, respond to the torque applied, be kink-resistant, be resistant to the pressures generated during contrast injection, and assure good visibility on fluoroscopy.

Several properties are crucial when selecting a catheter. The outer diameter is traditionally given in French (F), representing outer circumference in millimeters (corresponding to about 0.3 mm of outer diameter), inner lumen diameter in decimal fraction of inch (e.g., 0.035″), length in centimeters, maximal pressure in pounds per square inch (psi), and maximal flow in milliliters per second (mL/s).

There are some discrepancies in describing proximal and distal direction of the catheter. For the purpose of this chapter, the tip of the catheter will be called its distal end and the Luer lock adapter its proximal end.

Catheter manipulation requires application of torque to its part outside the patient. This torque has to be transmitted to the tip. Besides, as mentioned before, the catheter has to be kink-resistant and provide some support while passing through the vessels and/or chambers. This is why shafts of the catheters are usually composed of a plastic material (nylon, polyethylene, polyurethane, PTFE) braided with thin metal meshwork. Depending on the manufacturer, the catheter size and the distal ends of catheters can be made of braided or unbraided material. The tip itself usually lacks reinforcement to assure its softness and minimize the risk of vascular wall injury. The distal tip of the catheter may have an additional radiopaque marker to improve its visualization. Some

of the catheters have a single end hole for injection of the contrast medium, for pressure measurements, and for advancing guidewire, while other catheters such as angiographic catheters have multiple side holes for even contrast distribution. It is recommended to avoid any pressure injections of contrast through a catheter with end hole only. Balloon-tip catheters have a CO_2 inflatable balloon at their tips. This balloon is supposed to allow free floating with the bloodstream and prevent tangling between the chordae tendineae in the cardiac chambers. Other catheters have hydrophilic coating that makes them slippery and facilitate their gliding through tortuous vessels. Sizing catheters have additional radiopaque markers embedded in their shafts at known distances, for precise calibration and measurements.

6.1.2 Types of the Catheters

6.1.2.1 Angiographic Catheters

The main purpose of the angiographic catheters is the appropriate visualization of anatomy by means of the injection of contrast medium into blood vessels or cardiac chambers. Multiple side holes at the end of the catheter help to distribute the contrast evenly and deliver it efficiently during ventriculography or angiography. End hole allows for over-the-wire insertion of the catheter. The angiographic catheters can withstand high pressure and flow of the contrast medium, without recoil of the catheter during the injection.

There are angiographic catheters of various curves available in the market. Special shapes have been designed for a variety of purposes, e.g., pulmonary angiography. Despite their different shapes and other features, the main principles remain the same.

Berman angiographic catheter is a balloon-tipped catheter without the end hole (Fig. 6.1a). Thus, it cannot be advanced over a guidewire. Since it has a straight tip, the curved wire can be placed inside the catheter to shape it and support it when entering the desired location. The CO_2 inflatable balloon helps to cross the valves with the blood flow. However, in the presence of interatrial

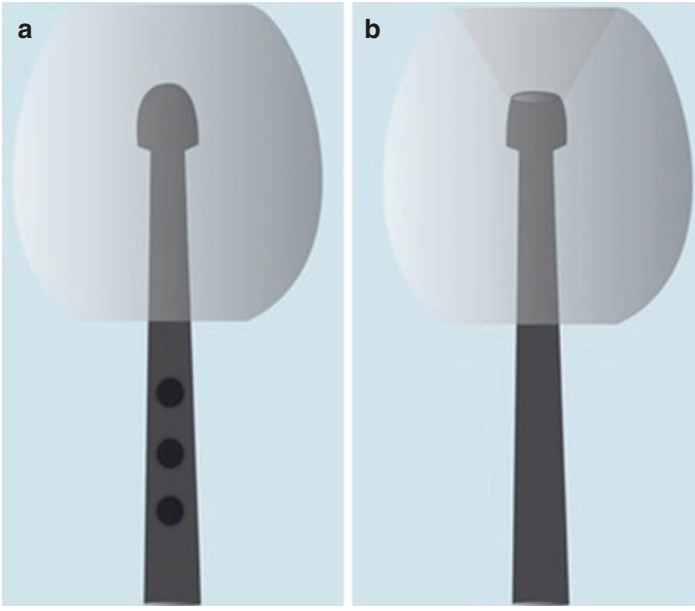

Fig. 6.1 Tips of floating catheters. (**a**) Berman angiography balloon catheter. (**b**) Pulmonary wedge balloon catheter

or interventricular communications, it can be used to catheterize left heart structures as well. Antegrade approach to the aorta is feasible also in patients with transposition of the great arteries, double outlet right ventricle, or functionally univentricular hearts or in the presence of large ventricular septal defects.

Moreover, the balloon catheter can be used to occlude the distal parts of the vessels and perform occlusion arteriography. Balloon occlusion descending aortography helps to force blood flow through aortopulmonary collateral arteries in the tetralogy of Fallot and other congenital cardiac malformations with pulmonary stenosis or atresia. In fenestrated Fontan patients, one can occlude the fenestration with the balloon tip in order to evaluate changes of blood pressure in the Fontan circulation. All these and many more applications make the Berman angiographic catheter an especially valuable item in the catheterization laboratory inventory.

6.1.2.2 Pulmonary Balloon Wedge Catheters

The pulmonary balloon wedge catheter (Swan-Ganz) is a single end-hole, balloon-tipped catheter, originally invented to measure right heart pressures. Its balloon tip makes it float to the distal pulmonary arteries (Fig. 6.1b). When it reaches the desired position, the inflated balloon occludes the antegrade flow in the vessel. Thus, the pressure in the pulmonary veins and the left atrium can be measured. When placed in the pulmonary vein, one can measure the pressure in the pulmonary arterial bed, based on exactly the same principle as the antegrade pulmonary wedge measurement. However, in the hands of interventional cardiologist, the pulmonary balloon wedge catheter becomes more widely used for advancing the guidewire for interventional procedures, selective pulmonary arteriography, simulation of vessel occlusion, and many others. With the balloon inflated at its tip, it should cross the tricuspid valve safely and minimize the risk of its injury during the following interventions, such as balloon valvuloplasty, pulmonary artery angioplasty, or stent placement. In case of extreme pulmonary artery hypoplasia, injection of the contrast medium through the catheter wedged in the peripheral pulmonary vein with consecutive flush with saline results in retrograde visualization of the pulmonary arterial vessels. Antegrade placement of the Swan-Ganz catheter in the Blalock–Taussig shunt is used, after inflation of the balloon, to simulate the shunt occlusion and monitor pressure changes in the pulmonary arteries. Undoubtedly, the pulmonary wedge catheter should always be available for use in the catheterization laboratory shelf.

6.1.2.3 Curved Catheters

A large variety of curved catheters are designed for selective catheterization of blood vessels. As mentioned before, their names often suggest their particular application. However, the interventionist searching for “right ventricular outflow tract catheter” or “right Blalock–Taussig catheter” would be unsuccessful in finding these. The operator should base selection of the most useful equipment on personal preferences, experience of other specialists, knowledge of catheter properties, and the patient’s anatomy. Most of the curved catheters have a single end hole. They can be

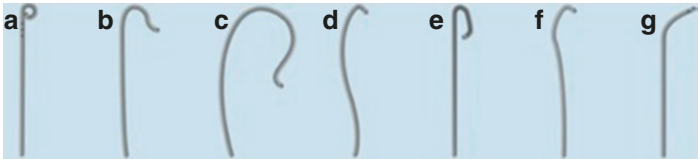

Fig. 6.2 Shapes of selected torque-controlled catheters (see text). (**a**) Pigtail catheter, (**b**) Amplatz left coronary catheter, (**c**) Amplatz right coronary catheter, (**d**) internal mammary catheter, (**e**) Judkins left coronary catheter catheter, (**f**) judkins right coronary catheter, (**g**) multipurpose catheter

used for selective angiography, pressure measurement, and guidewire placement.

Selected curved catheters are presented in Fig. 6.2. Some of these, described below, deserve some more attention.

Coronary catheters are designed to easily intubate the normal coronary arteries. Judkins and Amplatz catheters are the most popular (Fig. 6.2b–e). Among them, Judkins right coronary catheter (JR) is one of the most widely used in the cardiac catheterization laboratory. The distal part of the catheter is gently rotated to find support in the ascending aorta, and the tip bends at almost a right angle to reach the orifice of the right coronary artery.

In pediatric catheterization laboratory, this shape has proved to be useful in the selective catheterization of Blalock–Taussig shunts and collateral vessels, entering the right ventricle outflow tract, and many other procedures.

Other applications of Judkins right coronary catheter include crossing restrictive interatrial communications (especially in hypoplastic left heart syndrome patients), crossing interventricular septal defects, and many others.

Internal mammary catheters with their C-shaped tips can be used to enter vessels having origins at acute angles. Their applications include selective catheterization of Blalock–Taussig shunts and collateral vessels.

Multipurpose (MP) catheters have their distal ends curved at an obtuse angle (Fig. 6.2g). Usually there is at least one side hole near the tip. Such catheters, in accord with their name, can serve multiple purposes such as angiography, pressure measurement,

and selective catheterization. They can be used to cross a tight coarctation, enter the branches of the aortic arch, reach the left atrium from the femoral vein and the right atrium, and place the guidewire in the pulmonary vein before atrial septal defect device closure.

6.1.2.4 Special Catheter Types

The Multi-Track angiographic catheter (NuMED) has a short lumen for the guidewire at its tip. Thus, the tip can be introduced over the wire to a desired location, while the shaft remains free and multiple side holes remain open for use. This allows injection of contrast medium for angiography and measurement of pressures without losing the position of the guidewire.

Microcatheters are superthin catheters that can be introduced through standard lumen (0.035″–0.038″) single end-hole catheters for selective catheterization of small-sized vessels. Once the vessel is catheterized, one can deliver a microcoil through the guidewire lumen to occlude it. In pediatric and congenital cardiology practice, closure of small collateral vessels appears to be the major indication.

Guiding catheters are single end-hole angled catheters in shapes similar to diagnostic catheters, but with much larger lumen, which permits advancement of interventional equipment. They are widely used in coronary interventions to introduce rapid exchange balloon catheters with a side hole for the guidewire. The walls of the guiding catheters usually possess a three-laminar structure with metal braiding in the middle layer. The size of the guiding catheter, given in French, reflects its outer circumference (as opposed to the introducer sheaths sized by their lumen circumference). In congenital heart disease patients, application of guiding catheters is limited.

6.1.3 Selection of the Catheter and Catheter Manipulation

Every operator has own experience-based preferences. Nevertheless, some points need to be considered. First of all, the

goal of the procedure has to be specified. For example, in the right heart, where catheterization for idiopathic pulmonary hypertension angiography is not a standard component, the pulmonary wedge catheter is an obvious choice. Should right ventriculography or pulmonary angiography be planned, the Berman floating angiography catheter or multipurpose catheter is a reasonable choice. Also the size of the catheter should match the objectives, because it determines the maximum flow and contrast injection pressure. In case the diagnostic catheterization is followed by the intervention, one has to check if the inner lumen can accept the appropriate guidewire or the device. Length is another parameter worth considering. It can be really annoying when after long time of manipulation the catheter is too short to reach its destination.

With all kinds of catheters, it is important to remember the anatomic details, find support sites for the catheter tip and the shaft, and consider the use of a guidewire to position a catheter or to shape it. Wherever possible, biplane fluoroscopy should be used during the procedure. Manipulation principles are different for floating catheters and torque-controlled catheters. Some guidelines and tips and tricks are presented below.

6.1.3.1 Floating Catheters

Floating catheters (Berman, Swan-Ganz) are very soft, and their response to torque is limited. They should float freely in the direction of the blood flow. Nevertheless, especially in difficult anatomy or valvar insufficiency, it may be difficult to manipulate them. Additionally, floating catheters are packaged in a curved manner, and it may be impossible to straighten them completely. The problem can start just after crossing the introducer sheath. Without the balloon inflated, they can enter side branches. Inflation of the balloon makes the catheter float to the right atrium. Once the right atrium has been reached, it is possible to enter the superior vena cava. Sometimes it is possible to direct the tip posteriorly with a gentle torque and push the catheter up to the superior vena cava. In case of failure, insertion of a straight guidewire may solve the problem. Without a guidewire inside and without the balloon inflated, the catheter is likely to enter the left atrium

via an atrial septal defect or patent foramen ovale, if present. To enter the left pulmonary veins, the catheter should be directed posteriorly. Otherwise, it will enter the left atrial appendage. Inflation of the balloon of catheter placed in the left atrium, close to the interatrial septum followed by a clockwise torque, may help to cross the mitral valve and enter the left ventricle. If this maneuver does not work, one can use the stiff end of the guidewire bent in a U-shape to angle the distal end of the catheter. Guidewire will also help to transmit the torque. In the left ventricle, the catheter will float toward the apex. Again, a curved stiff end of the guidewire may help to bend the catheter (with the balloon inflated) toward the interventricular septum and push it up into the aorta. To avoid the tension onto the ventricular wall, the guidewire should be partly withdrawn from the catheter, to permit free floatation of the balloon.

In most of the cases, the catheter floats from the right atrium to the right ventricle. If this does not happen, an angled guidewire tip may be helpful to curve the catheter. Another solution is to find support for the catheter tip in the atrial wall, push it further to the atrium to make it bend, and then pull it back. The catheter should recoil and jump into the ventricle. The third method is to create a loop in the right atrium by pushing the catheter with some clockwise torque. Once the loop has been created, the catheter may enter the right ventricle and float to the outflow tract. The atrial loop can also be helpful to reach the right ventricular outflow tract, when the catheter keeps floating toward the ventricular apex. If it is stuck at the apex, a coiled guidewire can help to free it. Shaping the catheter with a guidewire is also useful to manipulate into the branch pulmonary arteries.

There are some issues to be kept in mind:

1. The balloon is able to accommodate more CO_2 than just one syringe; the more the balloon is inflated, the easier the floating is, but caution is needed as the balloon can rupture with too much CO_2.
2. The catheter can be straightened or bent using the guidewire; the guidewire helps to transmit the torque.

3. Creating a loop can help to manipulate with the catheter; it is better to straighten the catheter as soon as its final destination has been reached.
4. Especially in blood vessels, the balloon can obstruct them and alter the blood pressure; it should be deflated during measurements of pressure.
5. In selected applications, obstruction of the vessel with the balloon tip can help to make a selective contrast injection or perform an occlusion test; always remember where the hole(s) is(are).
6. When performing balloon occlusion angiography, the vessel occlusion time should be kept to a minimum; after inflation of the balloon, the catheter will float downstream—pull it back to the desired position, and deflate the balloon as soon as the angiography has been performed.

6.1.3.2 Torque-Controlled Catheters

Most of the catheters are torque-controlled. The torque is applied by the operator at the proximal end of the catheter.

Most of catheter tips lack braiding. That makes them soft and susceptible to kinking. While crossing the blood vessels, angled tips may tend to enter side branches/tributaries. If this problem occurs, they should be introduced over a guidewire. This is mandatory for pigtail catheters. Applications of selected catheters have been discussed already.

The importance of selection of an appropriate catheter shape is undisputed. Sometimes the shape has to be modified to reach a desired location. One can use a guidewire to make the angled catheter straight. On the other hand, the stiff end of a guidewire can be used to apply additional curvature. Entering the right ventricular outflow tract with the Judkins right coronary catheter is a good example. The catheter introduced to the right atrium will tend to move to the superior atrial wall, the right atrial appendage, the superior vena cava, or the left atrium through the atrial septal defect. One can, however, bend the catheter with an angled guidewire and then withdraw it to allow entry into the right ventricle. Torque applied to the catheter will direct the tip to the outflow tract. Should a new shape be permanent, one can reshape it by

placing the catheter in hot water or in steam with a stiff, preshaped guidewire inside. When the new shape is achieved, the catheter has to be cooled in saline.

Sometimes it is relatively easy to enter an origin of the blood vessel, but the guidewire makes the catheter recoil instead of entering the vessel. This may happen in major aortopulmonary collateral arteries and Blalock–Taussig shunts. If the catheter shaft is pushed excessively, an angle between the shaft and the tip inside the vessel may become too acute. Under such conditions, the guidewire tip is unable to straighten the catheter tip, it pushes the catheter further, and the tip recoils. Hence, it is better to straighten the angle by pulling the catheter down.

6.2 Guidewires

There are plenty of guidewire types and designs used in cardiac catheterization laboratory, produced by numerous manufacturers. Spring guidewires are composed of inner core made of stainless steel or nitinol, accompanied by a fine, steel safety wire and outer fine steel winding. Most of spring wires are coated with polytetrafluoroethylene and sometimes heparin to prevent clotting. In the distal part of the guidewire the core narrows or there is just a safety wire and outer winding. It makes the tip soft and limits a risk of injury to vascular or cardiac wall. The soft tip can adapt to a vessel shape, cross stenotic areas, and tortuosities. Wires with "floppy" tips can be especially useful in such setting. Tips of guidewires are straight or curved. J-tips are the most frequently found. The stiffer the wire is, the more support it provides to diagnostic or therapeutic catheters. Guidewires with a core wire extending from the proximal to distal end can transmit the torque 1:1 or near to it, which makes them more maneuverable. Tips commonly have platinum, gold, or tungsten elements to make them more radiopaque. Also guidewire shafts can be coated with, e.g., polyethylene/tungsten material, to enhance their visibility on fluoroscopy.

Guidewires with hydrophilic coating are slippery when wet. They glide through tortuous vessels easily. It can be difficult to

manipulate them, so a special plastic torque device is very useful. Hydrophilic wires have to be wet all the time, because they become sticky when dried.

Steerable guidewires have an additional filament attached to a proximal handle. An operator can change the shape of their tip by moving the handle. Thanks to the nitinol core, the tip returns to its initial shape. Such guidewires are used to navigate through tortuous vessels or cross the stents without passing between the struts.

Pressure wires are equipped with a pressure transducer at their tip. Initially, pressure wires were designed to measure pressure gradients across stenotic coronary arteries to assess fractional flow reserve. Gradually, other applications were developed, e.g., measurement of pressure gradients through stenotic valves or vessels such as banded pulmonary arteries in patients after hybrid procedures for hypoplastic left heart syndrome.

Size of a guidewire is given in fraction of inch. The length is measured in centimeters. Especially long (260–300 cm) exchange wires are used to exchange long catheters (Fig. 6.3). Some guidewires can be additionally extended using extension wires.

Guidewires are used to guide diagnostic catheters, therapeutic catheters, guiding catheters, and introducer sheaths through the heart or the blood vessels. Selection of the guidewire has to match the purpose of its usage. One has to consider:

1. Diameter of the guidewire: the operator should know what size the catheter is able to accommodate. Generally, back-bleed ports with flush port should be used to prevent bleeding and formation of thrombi. It becomes especially important in huge catheter lumen/guidewire disproportion, since the bleeding can be significant. In case a flush port is not available, one has to remember to rinse the wire with heparinized saline frequently. If the intervention is needed, the size of the wire has to be chosen according to the lumen of interventional equipment, e.g., a balloon catheter.
2. Length of the guidewire: when it is too short, it will not leave the catheter tip or may be unable to reach a desired position. It can also be impossible to exchange catheter over the wire. As mentioned before, some guidewires can be extended if needed.

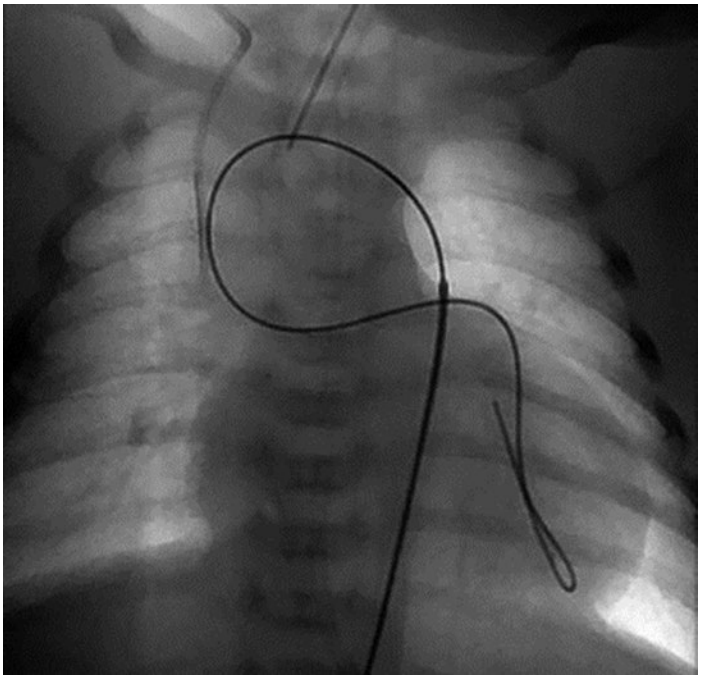

Fig. 6.3 Guidewire (Cook Inc., Bloomington, IL) position before stent placement to critical left pulmonary artery stenosis in a 3-week-old patient with complex congenital heart defect and duct-dependent pulmonary circulation after modified right Blalock-Taussig shunt. Guidewire introduced retrogradely from the aorta via the right Blalock-Taussig shunt, right and left pulmonary arteries to the distal branch of the left lower lobe pulmonary artery. Soft part of guidewire curled in the distal pulmonary artery branch for better support for the stent implantation

3. Hydrophilic coating: it helps to cross tortuous vessels and narrow stenoses. In spite of softness of hydrophilic catheter tip, it can easily perforate the heart or vessel wall, especially while exiting the catheter. That is why rather standard guidewires and not hydrophilic nor thin coronary wires are recommended to cross a critically stenotic aortic valve. The latter are more likely to perforate valvar leaflets.

4. Length of a soft tip: long, soft tips can enter the planned location but appear too extensive to support interventional equipment. On the other hand, guidewires with short tips are more traumatic.
5. Stiffness: it is safer and easier to advance catheters or sheaths using a stiff wire because of the better support. Softer wires can kink and lose their position or just make advancement of equipment impossible.
6. Shape of a tip: J-tipped guidewires are considered to advance more easily without entering side branches or tributaries. Nevertheless, the vessel diameter has to be large enough to accommodate the tip. Curved tips of some guidewires help to manipulate with the torque to reach a chosen location. Steel-core wire tips can be formed by an operator to get the best shape she/he needs.
7. Torque transmission: wires with a core continuous from proximal to distal end transmit the torque better than those lacking the core at their tips. The longer the floppy tip is, the more is the torque transmission limited.
8. Trackability and steerability: the shaft of the wire should be able to follow its tip through the tortuosities or narrowings in accord with operator's maneuvers.

As described in the section about catheter manipulation, stiff proximal end of a guidewire can be used to bend or shape catheter tips. One has to be cautious to not exit the catheter with a stiff end of the wire, as it can damage or perforate vascular structures or walls of cardiac chambers.

6.3 Introducer Sheaths

Introducer sheaths are used to assure safe vascular access, allow the insertion of catheters and interventional equipment, and help to guide the devices through tortuosities of the cardiovascular system. The sheaths are usually equipped with a back-bleed valve to prevent an excessive blood loss and a side port for flushing, pressure measurements, and, occasionally, contrast infusion.

The sheath is a thin-walled plastic tube composed of the material rigid enough to prevent kinking in the blood vessels. Their size reflects the inner diameter of the tube, i.e., the diameter of the dilator used to allow the smooth passage through the vascular wall. Thus, four French introducer sheaths can accommodate four French catheters. The outer dimension of the sheath is wider and depends on the thickness of the material the tube is made of. Dilators have a long, tapered tip sticking out of the sheath. The dilator and sheath locked together are introduced to a blood vessel over the wire. Size of the dilator inner lumen should be known to the operator, especially in case there is a need to exchange the sheath and use the one of the other size.

Back-bleed valves and side ports can be an integral part of the introducer sheath or be separate devices attached to the Luer lock at the end of the sheath. Usually, the back-bleed valve incorporated into the sheath is a latex diaphragm with a hole that permits insertion of the equipment. Resistance of such valve can significantly influence the effectiveness of manipulation and transmission of the torque applied to catheters and wires. In case of some introducer sheaths, the structure and rigidity of their valves make the effective.

Short introducer sheaths are used to maintain the vascular access and manipulate with the equipment. Their length should match the anatomy of the vessels—the sheath should not end opposite to a vascular wall, since it can produce complications such as vascular wall injury and bleeding.

Long sheaths are used to straighten blood vessels and create a smooth tunnel for diagnostic and, especially, interventional equipment, such as stents, occluders, vascular plugs, biopsy forceps, or transseptal needles. Tip of the sheath usually has a radiopaque marker. They can be straight or curved. The curvature should be chosen according to the delivery site and the route to be passed. For example, 45° curvature is suitable for ASD closure and 180° curvature is used for ADO I—type devices. Most of them can be recurved using hot steam or air to meet the needs of particular procedures. The curvature of steerable sheaths can be changed by the operator during the procedure.

Non-braided sheaths (e.g., Mullins sheaths) are used most frequently. Occasionally, however, it is necessary to use braided kink-resistant sheaths (e.g., Cook Flexor sheath) to cross tortuous vessels or acute bends in the cardiac chambers. Some sheaths have a hydrophilic coating to improve trackability.

The long sheath is delivers assembled with the dilator over the guidewire. The wire size should exactly match the diameter of dilator lumen. Most of the time, the sheath is introduced through the skin directly. Nevertheless, sometimes it is better to insert it through the short sheath. Keep in mind that the size of the sheath reflects its inner diameter! If you want to use sheath-in-sheath method the outer sheath should be at least two sizes bigger than the inner one.

Transseptal sheaths are curved similar to the transseptal needle and the lumen of the dilator follows the diameters of the needle. It affects the size of the guidewire—in standard Brockenbrough needle the dilator houses 0.032″ guidewire.

The use of specific sheath has to be carefully planned. Not only the kind of the device, shape of the sheath and its size have to be considered. Even the best plan can be ruined when the long sheath is too short for the length of the balloon catheter or pre-mounted stent system.

Suggested Reading

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Balloons

7

Caroline Ovaert and Duarte Martins

7.1 Introduction

A large variety of balloon catheters are currently available for catheterization of patients with congenital heart disease. Balloons are required for miscellaneous indications and types of procedures.

Direct *dilation* of stenosed valves and vessels, with or without balloon-expandable stent placement, remains the most important indication for using a balloon catheter. Industries have, over the last years, produced a wide panel of balloons, available in various lengths and diameters and with improving balloon characteristics that will be described in the next paragraphs. Some of those balloons have specifically been designed for use in children and adults with congenital heart lesions but several of them are primarily intended for treatment of other lesions. When performing

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complex heart catheterizations and balloon dilations in patients of different ages and weights, it is mandatory to have a broad spectrum of balloon catheters available in the catheterization laboratory, to be able to face the different clinical and technical situations.

The *atrial septostomy* or enlargement of a restrictive foramen ovale (Rashkind maneuver) is usually performed with specific balloon catheters designed for this purpose.

Balloons may be useful for the *analysis of dimensions* of heart defects such as atrial septal defects, before percutaneous closure. Sizing balloons have been specifically designed for this purpose.

Balloons may also be used for *test occlusion* of a defect (foramen ovale, atrial septal defect, ventricular septal defect) during hemodynamic assessment. They may be used to temporary *interrupt flow* in a collateral vessel in order to stabilize occlusion devices. Finally, balloons may be useful during *complication management.* No specific balloons are designed for those last purposes.

7.2 Direct Dilation of Valves and Vessels

7.2.1 Characteristics of Balloons

Dilation balloons are static balloons, which means that they expand to a fixed diameter when inflated to a certain maximum pressure (also called 'nominal' pressure). The d*iameter and nominal pressure* of a balloon are usually indicated by the manufacturer (Fig. 7.1a, b). When the pressure delivered to the balloon increases beyond the nominal pressure, wall tension increases proportionally and may cause balloon rupture [1]. The 'rated burst pressure' indicates this level of pressure where the balloon is likely to rupture. The maximum pressures tolerated will vary with the material used for the fabrication of the balloon but also with the diameter. Large-diameter balloons will have lower nominal and burst pressures than smaller balloons made of the same fabric. Indeed, wall tension is for a same pressure level, higher in larger-diameter balloons (Laplace's law: pressure = tension/radius) (Fig. 7.1c) [1]. This also means that, in large balloons, lower pressure is needed when compared to small balloons, to generate

a

Diameter (mm)	Length (cm)	Nominal* (ATM)	RBP† (atm)
12	2	6	18
	4	6	18
	6	6	18
14	2	6	18
	4	6	18
	6	6	18
16	2	6	18
	4	6	18
	6	6	16
18	2	6	16
	4	6	16
	6	6	16
20	2	6	16
	4	6	16
22	2	4	14
	4	4	14

Fig. 7.1 Balloon characteristics: diameter, pressure and compliance. (**a**) Each balloon will come with a pressure table, indicating nominal and rate burst pressure for each diameter. (**b**) Rated burst pressure is often labeled on the balloon catheter itself, for easy access. (**c**) Schematic representation of Laplace's Law solved for wall tension. (**d**) Superposition image of a noncompliant balloon at nominal and rated burst pressure. Note the absent increase in diameter and disappearance of the waist. (**e**) Superposition image of a compliant balloon at nominal and rated burst pressure. Note the overall increase in diameter while retaining a small waist

identical wall tension and, as a correlate, clinical efficacy. Four categories of balloons, according to pressure characteristics, are currently available in pediatric and congenital heart catheterization: low-pressure, medium-pressure, high-pressure, and ultrahigh-pressure balloons. They will be further described below.

Compliance of the balloon is an important characteristic [2, 3]. Completely noncompliant balloons will have a fixed diameter all along the balloon; at nominal pressure and even if pressure is increased above nominal pressure (Fig. 7.1d). In compliant balloons, the diameter may increase, especially in the areas of the balloon facing less resistance from the surrounding structures (Fig. 7.1e). This is important to know as this may be source of complications. Indeed, if a compliant balloon larger than the vessel is used to dilate a resistant stenosis, an increase in pressure may tear the 'normal' vessel adjacent to the stenosis.

The *morphology* of a balloon is characterized by the diameter, the length and the 'shoulders' of the balloon. The shoulders are the end-parts of the balloon, where the diameter reduces to the shaft's diameter (Fig. 7.2). Most balloons have radio-opaque markers to indicate where the working part of the balloon ends and where the shoulders start. Short shoulders are usually preferred in pediatric heart catheterizations as the shoulders tend to increase the length of the rigid part of the total balloon catheter. This can make manipulation in small heart and vessels more difficult and dangerous with worse hemodynamic tolerance [2, 3].

The *profile* of the balloon catheter is determinant especially in small children who will undergo multiple heart catheterizations and in whom vessel patency is crucial. The profile will depend on the balloon characteristics and the shaft of the catheter. High-pressure noncompliant balloons are often made of thick material that will require larger introducers. Larger balloons have a higher profile. The catheter shaft contains the balloon lumen and the wire lumen. If a balloon catheter accepts a 0.035″ wire, the profile will be increased as compared to a catheter accepting only a 0.014″ wire. A large enough balloon lumen is needed to be able to inflate and deflate rapidly the balloon but will increase the profile.

Most balloons for dilation in pediatric and adult congenital heart lesions are '*over-the*-wire' *balloons* meaning that the whole catheter will track over the wire. This is different from most coronary angioplasty balloons where the wire tracking is limited to the distal part of the catheter shaft. The MULTI-TRACK™ balloon dilatation catheter (NuMED Inc., NY, USA) designed for mitral

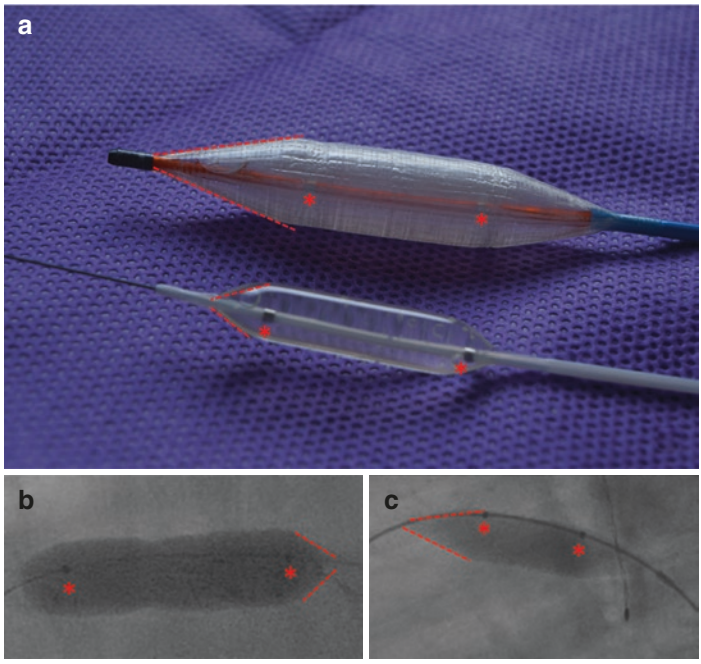

Fig. 7.2 Balloon characteristics: morphology. (**a**) Picture of a Tyshak II® balloon (lower balloon) and an Atlas® balloon (upper balloon), displaying balloon usable length between two radiopaque markers (marked *), and shoulders (dashed lines). Angiographic images of the same balloons (respectively **b** and **c**). Note the marked difference in shoulder size compared to balloon usable length

valve dilation has also a limited section (1 cm) at the distal tip of the catheter for wire tracking.

Other characteristics of balloon catheters include flexibility, kink resistance, pushability, stretchability, and indeflation time that all differ between balloons. The advantages and disadvantages of one balloon will have to be put in balance with advantages and disadvantages of another one as no ideal 'multi-use' balloon exists. It is important to have several balloons available in the catheterization laboratory but having the whole range of

commercially available balloons is impossible and unnecessary. Based on his own experience, the interventional cardiologist will have to select a few balloons with different and complementary characteristics to be able to cover all the clinical uses.

7.2.2 Low-Pressure, Medium-Pressure, High-Pressure, and Ultrahigh-Pressure Balloons

Balloons can be divided in categories according to the maximal pressure they can sustain: low-pressure, medium-pressure, high-pressure, and ultrahigh-pressure balloons. The limit between the medium- and high-pressure balloons is not well defined, and for this reason, they are here described in the same group. Table 7.1 summarizes some of the characteristics of currently available balloon.

Low-pressure balloons are characterized by their high compliance with low nominal and burst pressure rates (less than 10 atm). They come in various lengths and diameters and usually have small profiles and flexible shafts. They are very useful for balloon dilation of pulmonary and aortic valves in neonates, infants and children and may be used for dilation of aortic coarctation or vein stenosis in the young child. The low pressure they sustain and their high compliance make them unsuitable for dilation of 'pressure-resistant' stenosis in pulmonary arteries and for stent placement. When low-pressure dilation fails to open sustainably a stenosis, the analysis of how the balloon behaves during inflation and deflation with the low-pressure balloon remains very useful to understand the lesion. It will be possible to differentiate a long segment stenosis from a localized stenosis or a pressure-resistant lesion from a compliant lesion that recoils after balloon deflation. This will guide subsequent intervention. Low-pressure balloons are most of the time hand inflated by the operators. The use of an indeflator remains however recommended to avoid balloon rupture.

The second category includes the *medium- and high-pressure balloons* with burst pressure rates between 10 and 20 atm for most of them. The shaft is often stiffer and the profile higher than for

Table 7.1 Non-exhaustive list of commercially available and frequently used balloons in congenital heart catheterization

Pressure	Name	Company	Diameter (mm)	Introducer (F)	NP (atm)	RBP (atm)	Wire
Low-pressure							
– Compliant, low burst pressure (<10 atm) – Small profiles and flexible shafts – Useful for pulmonary or aortic valves, arterial or venous stenosis in the young child – Unsuitable for dilation of 'pressure-resistant' stenosis and for stent placement – Indeflator recommended to avoid balloon rupture	Tyshak Mini® Tyshak II®	Numed Numed	4–10 4–12	3–4 4–6	3–4.5 3–4.5	3.5–6 3.5–6	0.014″ 0.021–0.035″
Medium and high pressure							
– Burst pressure: 8–20 atm – Stiffer shaft and higher profile – Useful for pressure-resistant lesions (not responding to the 'low-pressure' balloons) especially in pulmonary arteries or calcified conduits. Indicated for stent insertion – Indeflator recommended for controlling the inflation pressure	Opta™ Pro Ultra-thin™ SDS Z-med II™ Z-med II-X™ Powerflex™ Mustang	Cordis Boston Sci Numed Numed Cordis Boston Sci	3–12 4–10 4–10 8–30 3–12 3–12	5–8 5–7 5–7 7–16 5–8 5–6	6 2–6	6–10 12 13–15 3–15 8–15 24	0.035″ 0.035″ 0.025–0.035″ 0.035″ 0.035″ 0.035″

(continued)

Table 7.1 (continued)

Pressure	Name	Company	Diameter (mm)	Introducer (F)	NP (atm)	RBP (atm)	Wire
	Advance® 35LP Mullins-X™ Cristal Balloon Sterling Balloon™	Cook Numed Balt Boston Sci	3–12 12–25 8–40 2–10	5–7 9–16 6–15 4–6	5–10 3–8 bar	8–15 9–14 10–14	0.035″ 0.035″ 0.035–0.038″ 0.018″
Ultrahigh pressure							
– Ultrahigh molecular weight polyethylene (UHMWPE) – Noncompliant, burst pressures, >20 atm – Useful for in-stent stenosis, stenosis adjacent to stents, pressure-resistant stenosis, to rupture stent cells (overlapping stents) – Special ‘high-pressure’ indeflator	Conquest® Atlas® Gold	Bard Bard	5–12 12–26	6–8 7–12	8 4–6	20–30 12–18	0.035″ 0.035″

the balloon catheters of the first category. They are very useful for pressure-resistant lesions (not responding to the 'low-pressure' balloons) especially in pulmonary arteries or calcified conduits and are indicated for stent insertion. They must be inflated with an indeflator to control the inflation pressure. A large panel of balloons is currently available in this category (Table 7.1).

The *ultrahigh-pressure balloons* are more recent in the pediatric cardiology field. They are completely noncompliant balloons, made of ultrahigh molecular weight polyethylene (UHMWPE). This very resistant fabric supports very high pressures, often up to 30 atm or more. They are useful for treating in-stent stenosis or stenosis adjacent to stents, often by rupturing the previously inserted restrictive stent. They may also be helpful to rupture stent cells when stents cover side branches or to dilate very resistant non-stent-related stenosis. Some of those balloons, originally for vascular use, have particularly long shoulders, and this has to be taken into account when choosing the appropriate balloon length, especially in small children. The ultrahigh-pressure balloons need to be inflated with special 'high-pressure' indeflators.

7.2.3 Cutting Balloons

The Boston Scientific Cutting Balloon® (Boston Scientific, MA, USA) is made of a noncompliant balloon with 4 sharp steel blades embedded longitudinally on its surface (Fig. 7.3a). Other bladed balloons are on the market but their experience in pediatric or congenital interventions is scarce. The rationale is to create 4 controlled tears in a thickened intima and media, without over-dilating the vessel, avoiding by there a noncontrolled deeper tear which puts the vessel at risk for aneurismal dilation or rupture (Fig. 7.3b). The bladed and cutting balloons were initially designed in the 1990s to dilate resistant coronary stenosis. Since the late 1990s, they are also used to dilate pressure-resistant stenosis in pulmonary arteries, in-stent stenosis and restrictive atrial septal defects (Fig. 7.3c–f) [2, 3].

The Boston Scientific Cutting Balloon® comes in diameters between 2 and 8 mm and has a burst pressure of 8 atm. The

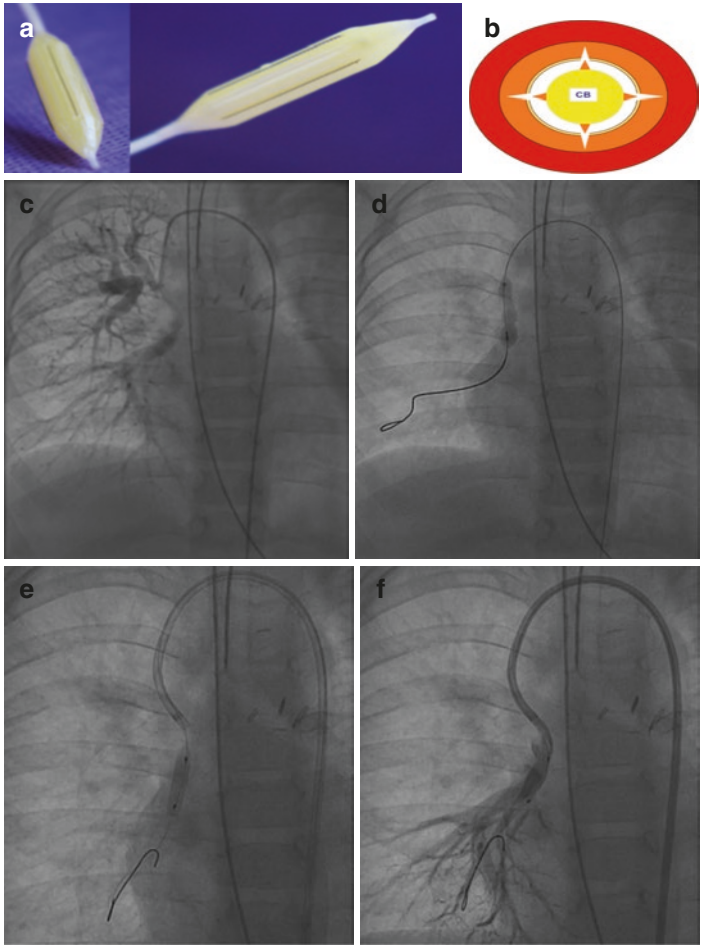

Fig. 7.3 Cutting Balloons: (**a**) Expanded cutting balloon, displaying blades. (**b**) Schematic section of a cutting balloon (CB) and vessel, showing mechanism of action. (**c**) Pulmonary angiogram from a patient with PA VSD with unifocalized MAPCA to the lower right lobe, displaying anastomotic stenosis. (**d**) A compliant balloon shows multiple resistant lesions. (**e**) Balloon dilation with a cutting balloon. (**f**) Final angiography showing marked enlargement of the lumen

maximal diameter limits its use to small vessels. The use of a long sheath to advance and retrieve the balloon is highly recommended in order to avoid damage to cardiac and vascular structures with the sharp blades. Inflation and deflation must be very slow, with the help of an in deflator, in order to allow proper opening and refolding of the blades. Balloon dilation with a cutting balloon may be followed by angioplasty with a standard compliant balloon or by stent insertion.

7.2.4 Coronary Balloons

Coronary balloons with our without premounted stents must be available when performing congenital heart catheterization. They can be used for various interventions such as pulmonary valve dilation after radiofrequency perforation of pulmonary atresia, branch pulmonary artery dilation, ductal stenting or sometimes coronary interventions.

Coronary balloons come in various diameters, usually ranging between 1.5 and 5 mm and various lengths (8 to 40 mm). They can be compliant or noncompliant, and are usually of the 'monorail' type (not 'over the wire'), with a low profile. Nominal and burst pressure with maximal diameters may vary but are always clearly mentioned.

7.2.5 Other Special Balloon Dilation Catheters

The NuMED *balloon in balloon (BIB®) catheter* (NuMED Inc., NY, USA) is a triaxial catheter. One lumen is for tracking over a guide wire, while the 2 others are to inflate 2 balloons (Fig. 7.4a). A small balloon (inner balloon) is inside a larger balloon (outer balloon) (Fig. 7.4b). The inner balloon inflates to half the diameter of the outer balloon and is 1 cm shorter. The rated burst pressure is different for each size. The double-balloon catheter allows incremental inflation which is very helpful for stent placement in

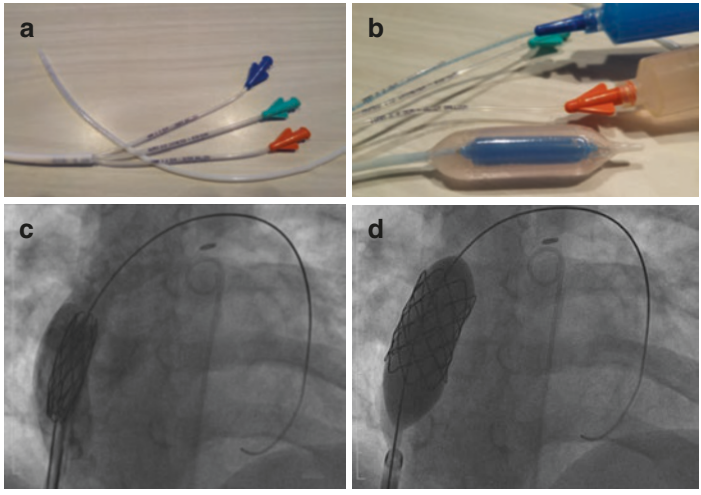

Fig. 7.4 Balloon-in-balloon (BIB): (**a**) Triaxial catheter: 1 lumen (green) is for tracking over a guide wire, 1 lumen (indigo) is for dilation of the small inner balloon, 1 lumen (orange) is for inflation of the larger outer balloon. (**b**) The inner balloon (inflated in indigo dye) is completely inside the outer balloon (inflated in orange dye), inflates to half of its diameter and is 1 cm shorter. (**c**) Stent inflated over the inner balloon alone, allowing repositioning. (**d**) Incremental inflation of the outer balloon, safely securing the stent against the vessel wall

large vessels. The inner balloon provides initial expansion of the stent and acts as a tool to hold the stent in place while the outer balloon is inflated. The outer balloon is then inflated securing the stent against the vessel wall (Fig. 7.4c, d).

The *Nucleus*™*, Nucleus-X* ™ *balloon catheter* (NuMED Inc., NY, USA) and the *Inoue-Balloon catheter* (Toray Industries, Inc., Houston, TX, USA) have a 'dumbbell' shape and are specifically designed for valve dilation. Inoue balloons are more intended for mitral valve dilation as the Nucleus balloon may be used for aortic or mitral valve dilation. The smaller central part has to be located at the level of the valve annulus. The larger external parts will help to stabilize the balloon (Fig. 7.5).

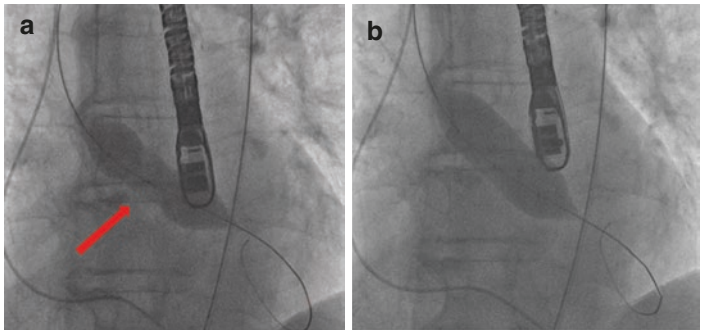

Fig. 7.5 *Dumbbell*-shaped balloon: images of a Nucleus™ balloon (Numed) used for aortic valve dilation. (**a**) The balloon is incompletely inflated and has the dumbbell morphology; the narrowest part is at the level of the aortic annulus (red arrow). (**b**) The balloon is completely inflated with loss of the dumbbell shape

7.3 Septostomy Balloons

Septostomy balloon catheters are specifically designed to cross the foramen ovale and to perform the Rashkind atrial septostomy maneuver (Fig. 7.6a, b). The balloons are round and noncompliant.

The Edwards 5F atrioseptostomy catheter (Edwards Lifesciences Corporation, CA, USA used to be largely used but has been recently made unavailable. The Edwards 5 F balloon catheter had a single lumen for inflation of the balloon. The catheter was rather stiff and has a curve at the end to facilitate crossing of the foramen ovale. The balloon took larger volumes of contrast (4 cc) than the other atrioseptostomy balloons which was useful when a large atrial septal defect was needed.

The NuMED Z-5™ atrioseptostomy catheter (NuMED Inc., NY, USA) is a good alternative dual-lumen balloon catheter. The catheter is soft and easily progresses over a wire, which is helpful when crossing of the restrictive foramen ovale is difficult. The balloon catheter exists in 2 sizes (1 cc balloon, 4 F catheter and

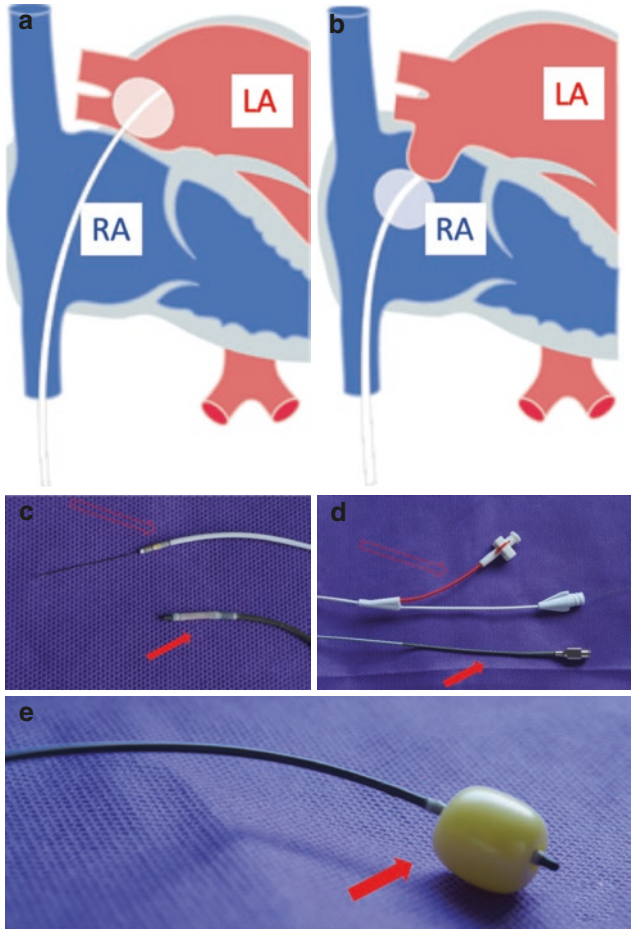

Fig. 7.6 Septostomy Balloons. (**a**, **b**) Specifically designed to cross the *foramen ovale*, to perform the depicted Rashkind atrial septostomy maneuver. (**c**) Distal tip of a Numed Z-5™ atrioseptostomy catheter (open red arrow) and a Medtronic Rashkind balloon septostomy catheter (full red arrow). Note the capacity of the former to travel over the wire, while the latter has a more stiff and curved tip to tackle de *foramen ovale* directly. (**d**) Proximal tip of a Numed Z-5™ atrioseptostomy catheter (open red arrow) and a Medtronic Rashkind balloon septostomy catheter (full red arrow). Note the double lumen of the former. (**e**) Fully inflated Medtronic Rashkind balloon septostomy catheter

2 cc balloon, 5 F catheter), the smallest being particularly useful in small preterm babies. This catheter is normally designed for septostomy under fluoroscopic guidance (with use of a wire). It can still be used for atrioseptostomy at the bedside, under echocardiographic guidance. The shaft is softer than the Edward's one but can be stiffened, if needed, by inserting a wire up to the end of the catheter. Figure 7.6c–e show the differences with the Edwards catheter.

The Medtronic 6F, 50 cm Rashkind Balloon Septostomy Catheter (Medtronic Inc., MA, USA) was another alternative but has been recently withdrawn. It has only one lumen for balloon dilation, like the Edwards balloon, and only takes 2 cc of liquid. The catheter shaft is also slightly softer than the Edwards catheter.

7.4 Sizing Balloons

Sizing balloons have been introduced for measurement of atrial septal defects during percutaneous atrial septal closure. Two types of sizing balloon exist based on the method used for sizing.

The first method is the 'pull-through' technique (Fig. 7.7a). The balloon used is a soft, compliant and spherical balloon (Equalizer™, Boston Scientific, MA, USA) that is inflated in the left atrium. While deflating slowly, the balloon is pulled across the septal defect. The diameter of the balloon when it crosses the

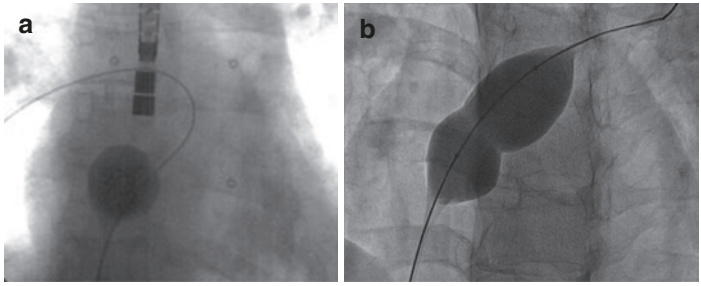

Fig. 7.7 Sizing Balloons. (**a**) **'**Pull-through' technique. (**b**) 'Static' technique

defect will be considered as the stretched 'atrial septal defect diameter'.

The other method is the 'static' method (Fig. 7.7b), during which a soft, compliant, low-pressure and elongated balloon (Amplatzer™ Sizing Balloon II (St. Jude Medical Inc., MN, USA), PTS® and PTS-X™ (NuMED Inc., NY, USA)), is inflated across the atrial septal defect until a waist is seen on the balloon. The diameter of the waist will inform on the atrial septal defect diameter.

7.5 How to Use Balloons

7.5.1 Preparation, Introduction and Inflation

There are different ways to prepare a balloon. A dilution of contrast with normal saline is needed. The ratio of 1 unit of contrast and 3–4 units of saline is often preferred but may vary according to the type of balloon used, the purpose and operator's habits. Two syringes (in case of hand inflation) or 1 syringe and a pressure indeflator have to be filled with this diluted contract and connected to a three-way stopcock. The size of the syringes depends on the size of the balloon used. The whole system (syringe, indeflator and stopcock) has to be thoroughly de-aired before connecting to the balloon. Once this is done, the system will be connected to the balloon, with the balloon in line with the pressure indeflator. De-airing of the balloon can be performed by gently inflating the balloon at no pressure and removing the air bubbles subsequently. However, this technique has the disadvantage of unfolding the balloon which will then lose its 'profile'. Refolding the balloon is sometimes possible but time-consuming. The usual way of de-airing is the 'negative prep' technique [3]. A strong negative pressure is applied to the balloon with the indeflator or with the syringe, and this negative pressure is maintained by blocking the indeflator or syringe. The three-way stopcock is then turned to connect the balloon with the other syringe which will then allow the balloon catheter to passively fill with the contrast.

This maneuver is repeated a few times to allow full replacement of air by contrast. At the end, negative pressure is again applied to the balloon while entering the balloon catheter into the sheath and lesion. The wire lumen of the balloon catheter needs to be flushed before introduction.

Almost all balloons are currently introduced into the vascular system through vascular sheaths. In the early years of balloon dilation, the profile and deflation characteristics of the balloon catheters were such that direct introduction over the wire, without a sheath, was preferred to the use of very large sheaths. Direct introduction of a balloon catheter in a vein carries however a high risk of vessel trauma and use of a sheath lessens this risk. With the current characteristics of balloons and sheaths, the use of a sheath has to be the standard and direct introduction should be avoided [3].

Easy advancement of the balloon catheter requires a good and stable wire position. The wire should be as stiff as possible and the diameter should match the diameter of the balloon wire lumen. Good positioning of the balloon often requires gentle pushing and pulling maneuvers with the wire and balloon. Once good positioning is obtained, the balloon will be inflated under fluoroscopic control. The use of a pressure indeflator is recommended for all dilations, but in some conditions and with some balloons, a gentle hand inflation may be authorized. When dilating a valve or main vessel, several short dilations with rapid inflation and deflation have to be performed to avoid significant hemodynamic interference. In a distal pulmonary vessel, a slower and more sustained dilation may be performed [2, 3].

7.5.2 Deflation and Withdrawal

Once the dilation is finished, negative pressure should be applied in order to empty the balloon completely before leaving the dilated lesion. It is important to check on fluoroscopy that the balloon is completely emptied. If during this negative pressure maneuver blood comes back, this means that the balloon

has ruptured (see below). Negative pressure is usually maintained while leaving the heart and vessels. However, occasionally, in stiff balloons, this can augment the stiffness of the balloon folds and increase trauma to the vessel and heart. Extra care has to be taken when pulling the balloon back to groin, and in case of resistance, one should avoid to pull harder but try with gentle re-advancements and rotations, and sometimes small re-inflations and deflations, to extract the balloon catheter. Withdrawal into the sheath is sometimes difficult. In those cases, gentle re-inflation followed by deflation with negative pressure may improve balloon folding and make withdrawal easier. If only the distal part of the balloon refuses to enter the sheath, the sheath might have to be taken out together with the balloon catheter, leaving the wire in place [3].

7.6 Double-Balloon Technique

Insertion of two balloons to dilate valves may be useful when the valve annulus is too large for the available balloons. This may be the case for pulmonary or mitral valve in adult patients (Fig. 7.8a, b). Using two balloons may also be useful in smaller patients, to reduce the sheath size. Insertion of two smaller balloons in two separate veins will require smaller sheaths than one large balloon through one access vein. Additional benefit may be the hemodynamic tolerance. Large balloons need a long time to inflate and deflate. Two smaller balloons will more rapidly inflate and deflate and in addition, with the double-balloon technique, even at full inflation, there will still be some residual flow between the balloons which may improve hemodynamic tolerance [3]. There are several formulas to calculate the effective diameter of the 2 balloons inflated together. According to the formula used and the combination of balloons, the calculated effective diameter will be approximately 15–30% smaller than the sum of the 2 diameters (Fig. 7.8c) [2].

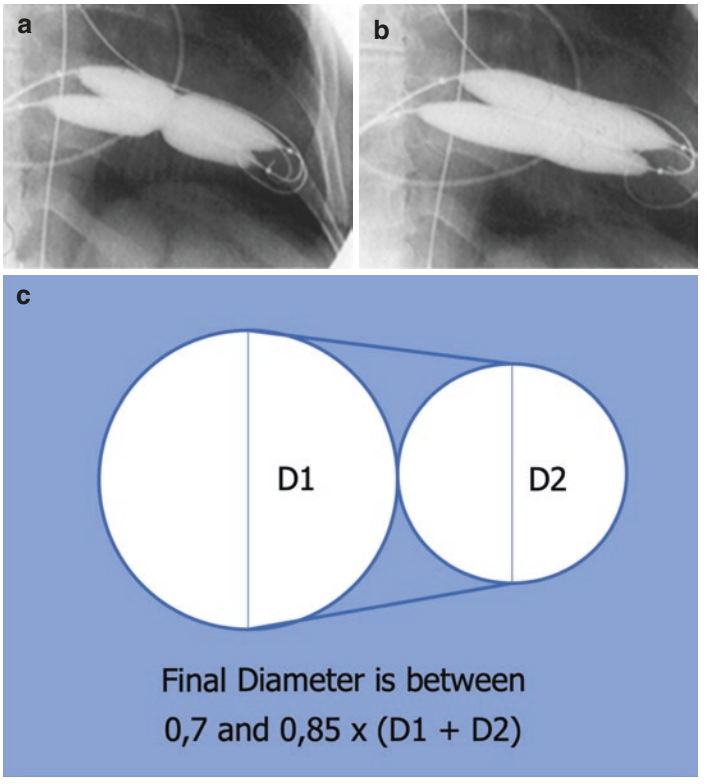

Fig. 7.8 Double-Balloon technique. (**a**) Double-balloon technique for mitral valve dilation. Here the balloons are not fully inflated and the waists are still visible. (**b**) Full inflation of the balloons, causing the waists to disappear. (**c**) Schematic representation of the obtained diameter

7.7 Complications

Balloon rupture is the most frequently encountered complication. Different types of balloon rupture have to be distinguished as they may have different consequences in particular for balloon retrieval.

A balloon may be punctured by an adjacent stent strut, especially when the stent is fractured. Calcified vessel walls may also

puncture a balloon. The puncture may be recognized on fluoroscopy when contrast extravasates out of the balloon in a localized area. It is however often noticed while applying the negative pressure to deflate the balloon: blood flows back together with the contrast. The contrast leakage prevents appropriate pressure increase inside the balloon which may limit vessel or stent expansion and complicate balloon retrieval. Connecting the balloon to the dye injector and injecting dye under high pressure may be a way to improve balloon and stent expansion which will then allow safe retrieval of the balloon.

The balloon can also tear over a long distance. This is usually the result of balloon inflation with an excessive pressure or in case of dilation of a very calcified lesion (such as a conduit). On fluoroscopy this is noticed as sudden disappearance of the contrast. Most often, the tear is longitudinal and there will be no problems to retrieve the balloon entirely (Fig. 7.9a). Less frequently balloons can tear circumferentially which is a more serious complication as the distal part may fold over the tip of the balloon catheter (Fig. 7.9b–d). Excessive traction on the balloon catheter has to be avoided as the distal balloon end may come loose and will then be free-floating in the lumen. It is crucial to maintain stable wire position as the distal balloon end remains over the wire. The use of a snare will usually be needed to stabilize and retrieve the balloon. Figure 7.10 details a useful technique to retrieve circumferentially burst balloons.

Inability to deflate a balloon may happen and is usually the sign of a localized rupture or puncture of the balloon at the proximal end, with the proximal deflated end of the balloon obstructing the communication between the balloon and the balloon catheter lumen. In these cases, when hemodynamic instability occurs, it may be preferable to burst the balloon.

Fracture of the shaft of the balloon is very rare and unlikely with single-used balloons but may happen with reused and re-sterilized balloons.

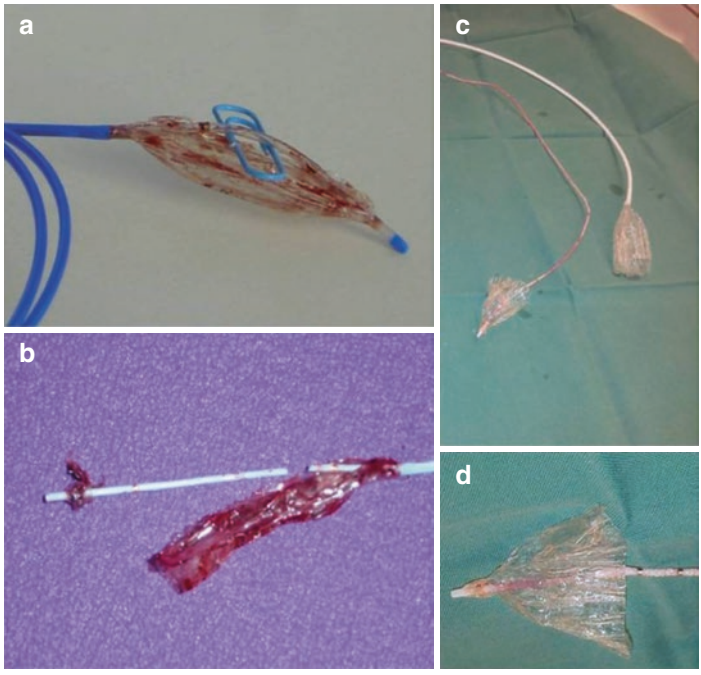

Fig. 7.9 Balloon Rupture: Types of tears. (**a**) Longitudinal tear. The blue paper clip shows the balloon shaft exposed by the longitudinal tear (courtesy Dr. Joseph De Giovanni, Birmingham, UK). (**b**) Circumferential tear. (courtesy Dr. Joseph De Giovanni, Birmingham, UK). (**c**, **d**) Circumferential balloon rupture associated with a shaft rupture. The distal part remained on the internal shaft (**d**), the proximal part on the external shaft (**c**). (Courtesy Prof R. Berger, Groningen, Netherlands)

7.8 Basic Requirements

When starting a pediatric and congenital catheterization activity, the large number of different procedures that the interventionalist may encounter, as well as the broad range of patient's ages and sizes makes it difficult to figure out what material is mandatory on the shelf. Table 7.2 summarizes what to our opinion is the 'minimum balloon set-up' to be able to start and cover most frequent pediatric and adult congenital interventions.

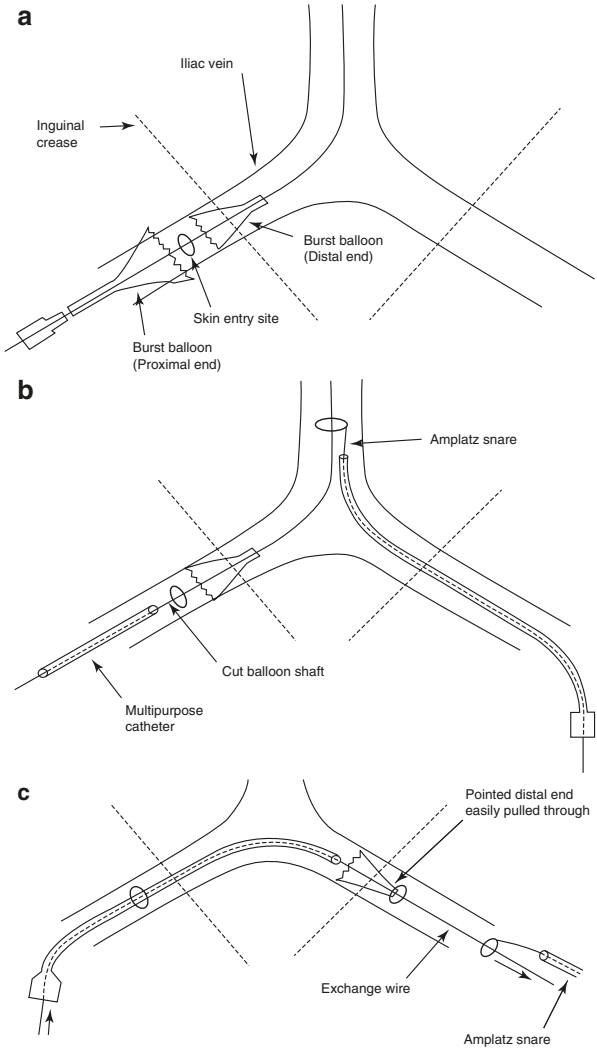

Fig. 7.10 Technique for retrieval of circumferentially burst balloons. (**a**) Wire position has to be conserved. (**b**) The wire is snared distally, using the contra-lateral vein. The proximal part of the balloon will be taken out of the body and the balloon shaft will be cut. A multipurpose catheter will be advanced on the wire to push the distal end of the catheter and balloon toward the contra-lateral vein. (**c**) The distal end of the balloon will easily be pulled through the contra-lateral sheath. (Courtesy Dr. Jo De Giovanni, Birmingham, UK)

Table 7.2 Basic Setup

Indication	Stock	Additional
Balloon Atrial septostomy	Septostomy balloon—keep at least 2 in stock	Stock up if need anticipated
Valvuloplasty	Low-pressure balloon (such as Tyshak II)—keep one of each diameter 5–9/20 mm for neonatal lesions	May order a range of 2 or 3 balloons for older patients Coronary balloons may be useful after valve perforation in PA IVS
Vessel treatment	Low-pressure balloons kept for valvuloplasty may be useful for vessel treatment in infants and small children Keep a small range of medium-pressure balloons (such as Powerflex Pro) for lesions in older children (6, 8, 10, and 12 mm, for example) High-pressure balloons (such as Atlas Gold) are useful for resistant lesions in adolescents and adults. Keep a small range of diameters (12–24 mm), usually the most useful length is 4 cm	Longer low-pressure balloons (i.e., 30 mm) are usually more stable for vessel treatment. Shorter high-pressure balloons (2 cm) may be more effective in resistant lesions. Coronary noncompliant balloons perform well for very small lesions Consider a cutting balloon for resistant pulmonary peripheral lesions
Stent delivery	*Match to locally available stents* Max LD 16, 26 and 26 mm—BIB 25, 30 and 40 mm. CPstent 28, 34 and 39 mm—BIB 30, 35 and 40 mm Consider diameters 12, 14, 15, 16, 18, 20 mm (large vessel treatment) 22, 24 mm (landing zone for melody valve)	Consider keeping short high-pressure balloon (i.e., Atlas Gold 2 cm) for resistant stenosis or stent flaring
Sizing	ASD closure: keep 25, 30, and 40 mm/30 mm sizing balloons for sizing of large defects or those with floppy or irregular rims	RVOT sizing: keep a 25/40 mm low-pressure balloon (such as Tyshak II) for testing dilated RVOT for valve implantation

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Stents

8

Sebastian Góreczny and Eric Rosenthal

8.1 Introduction

Stent implantation in congenital heart disease became available in the late 1980s with a rapid uptake in the 1990s. While standard balloon dilatation was a successful approach to the treatment of stenotic lesions, limitations were apparent. Fibrotic stenotic lesions allowed controlled dissection with remoulding of the vessel wall during the healing phase but more elastic lesions, long-segment stenoses, hypoplastic vessels, stenoses related to kinking or tension on a vessel rarely responded well often with immediate vessel recoil. Balloon oversizing in this setting could lead to vessel tearing with dissection flaps, vessel rupture with haemodynamic collapse and late aneurysm formation.

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Stent implantation prevented the immediate elastic recoil, allowed the vessel to be dilated only to the required diameter and sealed small intimal flaps to the vessel wall. Stenosis relief was superior both acutely and in the long term with a lower risk of acute vessel complications. By not overdilating the vessel, stent implantation could be used early after cardiac surgery.

Issues unique to the paediatric population include small patients' size limiting vessel access and difficulty in advancing the rigid stent through a tortuous vascular route. After somatic growth, stent redilation is needed until the patient is adult size. Stents unable to be dilated to adult size result in a fixed stenosis after growth.

8.2 Indications

Initially limited to patients big enough to accommodate an appropriate sheath and stent that would not need redilatation, the encouraging early results and improvements in stent, balloon and delivery sheath design widened the indications (Table 8.1) [1–3]. Stents are implanted as a bridge in neonates and infants with elective surgical removal during the next stage of treatment. Indeed, stents that are eventually dilatable to adult size can now be introduced through 6 F sheaths. A hybrid surgical approach further expanded the benefits with cooperation between surgeons and interventionalists. Covered stents can seal native aneurysms and fistulae and those resulting from surgery, balloon dilation or bare-metal stents (Fig. 8.1). Coronary artery interventions are increasing.

8.3 Stent Features

Given the diversity of lesions and patient size range, a single type of stent does not suit all situations. Stent implantation is an art of choosing the best device for a specific patient and condition. It is better to be experienced with a limited range of stents (Table 8.2) rather than trying to master all.

Table 8.1 Indications for stent implantation in congenital heart defects

1. Branch and peripheral pulmonary artery stenosis
(a) Postsurgical
(b) Native
2. Pulmonary vein stenosis
(a) Postsurgical repair of TAPVD
(b) Native
3. Aorta and branches
(a) Native coarctation
(b) Re-coarctation
(i) Surgery
(ii) Balloon dilation
(c) Aneurysms
(d) Abdominal coarctation
4. Right ventricle outflow tract
(a) Pulmonary atresia after perforation
(b) Tetralogy of Fallot
(c) Conduits
(i) Standalone
(ii) Preparation for percutaneous valve implantation
5. Arterial duct in duct-dependent
(a) Pulmonary circulation
(b) Systemic circulation
6. Surgically created shunts
(a) Blalock–Taussig
(b) Central
(c) Sano
7. Systemic vein stenosis
(a) Postsurgical (Fontan, Senning, Mustard)
(b) Pacemaker electrodes, central lines
8. Major aorto-pulmonary collateral arteries (MAPCAs)
9. Intracardiac communications
(a) Atrial septum
(b) Atrial fenestration
10. Sealing of fistula or communication with covered stent
(a) Patent arterial duct
(b) AV fistula
(c) Fontan fenestration
11. Coronary artery stenosis
(a) Kawasaki
(b) Post arterial switch
(c) Left internal mammary bypass

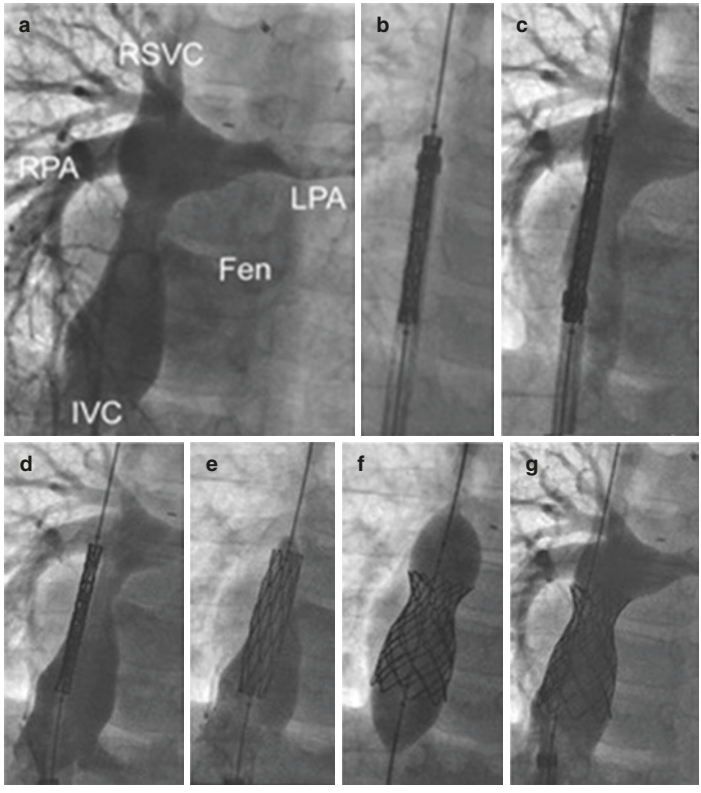

Fig. 8.1 Covered stent (Premounted Cheatham Platinum on a balloon-in-balloon) implantation into a lateral tunnel of an 8-year-old boy with exercise-induced cyanosis and protein-losing enteropathy. Angiogram in (**a**) shows a tunnel stenosis before the branch pulmonary arteries and a patent fenestration. After passing a long sheath over a stiff guide wire placed in the SVC, the stent is advanced (**b**). Angiography via the long sheath is used to position the stent (**c**, **d**). The inner balloon is inflated with angiography confirming position in the stenosis and continued flow across the fenestration (**e**) followed by the outer balloon (**f**). Final angiogram shows the fenestration to be occluded (**g**) and the stenosis dilated. Note the position and shortening of the stent between (**d**) and (**f**) so that the final position does not obstruct the right pulmonary artery. *RSVC* right superior vena cava, *RPA* right pulmonary artery, *LPA* left pulmonary artery, *FEN* fenestration, *IVC* inferior vena cava

Table 8.2 Commonly used stents in congenital heart disease

Stent	Material	Cell design	Nominal diameter (potential)	Length (mm)	Sheath size (F)	Guide wire (″)	Mounting	Common usage
Sinus-Superflex-DS[a]	N	O	4–9	12–24	4	0.018	Self-expandable	Neonate and infant lesions
Zilver 518[b]	N	C	4–10	20–80	5	0.018	Self-expandable	Neonate and infant lesions
Protégé RX[c]	N	O	6–10	20–60	6	0.014	Self-expandable	Neonate and infant lesions
Palmaz Blue[d]	Cc	C	4–7 (12)	12–24	4–5	0.014	Aviator plus	Neonate and infant lesions
					5	0.018	Slalom	
Palmaz genesis medium[d]	Ss	C	4–8 (12)	12–24	5–6	0.018	Slalom	Neonate and infant lesions
					6	0.035	Opta Pro	
							Unmounted	
Formula 535[b]	Ss	O	4–10 (20)	12–60	5–6	0.035	High-pressure balloon catheter	PAs, atrial septum, RVOT, coarctation
Palmaz genesis large[d]	Ss	C	5–10 (12)	19–79	5–6	0.018	Slalom	PAs, atrial septum, RVOT
					6–7	0.035	Opta Pro	
							Unmounted	
Visi-Pro[c]	Ss	O	5–10 (14)	12–57	6–7	0.035	High-pressure balloon catheter	PAs, atrial septum, RVOT

(continued)

Table 8.2 (continued)

Stent	Material	Cell design	Nominal diameter (potential)	Length (mm)	Sheath size (F)	Guide wire (″)	Mounting	Common usage
Valeo Lifestent[e]	Ss	O	6–10 (20)	17–56	6–7	0.035	High-pressure balloon catheter	PAs, atrial septum, RVOT, coarctation
Genesis XD[c]	Ss	C	10–12 (18)	19–59	8	Depending on the balloon catheter	Unmounted	Pulmonary arteries
Double Strut LD[d]	Ss	O	9–12 (18)	16–76	8	Depending on the balloon catheter	Unmounted	Pulmonary arteries
Mega LD[4]	Ss	O	9–12 (18)	16–36	9	Depending on the balloon catheter	Unmounted	Pulmonary arteries
Max LD[d]	Ss	O	12 (26)	16–36	11	Depending on the balloon catheter	Unmounted	Coarctation, pulmonary arteries, veins
Advanta V12[f]	Ss[j]	O	12–16 (22)	29–61	9–11	0.035	High-pressure balloon catheter	Coarctation, pulmonary arteries, veins
Covered Cheatham Platinum[g]	Pi[j]	C	12–24 (26) (up to 34 mm for 10 Zig)	16–60	12–14	0.035	Unmounted or premounted (NuDEL) on BIB catheter	Coarctation, pulmonary arteries, veins, RVOT

Cheatham Platinum 8 Zig[g]	Pi	C	12–24 (26) (up to 34 mm for 10 Zig)	16–60	10–12	0.035	Unmounted or premounted on BIB catheter	Coarctation, pulmonary arteries, veins, RVOT
BeGraft[h]	Cc[j]	O	12–24 (30)	19–59	9–14	0.35	High-pressure balloon catheter	Coarctation
Andrastent XL[i]	Cc	H	15–25	13–57	8–9	Depending on the balloon catheter	Unmounted	Coarctation, pulmonary arteries, veins, RVOT
Andrastent XXL[i]	Cc	H	20–32	17–57	10–11	Depending on the balloon catheter	Unmounted	Coarctation, pulmonary arteries, veins, RVOT

C closed, *O* open, *N* nitinol, *H* hybrid, *Ss* stainless steel, *Cc* chromium cobalt, *Pi* Platinum-iridium
Potential diameter from reported experience—not confirmed by the manufacturer
[a]Optimed
[b]Cook Medical
[c]Medtronic
[d]Cordis
[e]Bard
[f]Atrium
[g]Numed
[h]Bentley InnoMed
[i]Andramed
[j]Covered with an expanded polytetrafluoroethylene (e-PTFE)

Attributes of an ideal stent include:

- Safe delivery to the target lesion.
  - Low profile allowing use of a small sheath and crossing of tight stenoses.
  - Flexibility and easy trackability through tortuous pathways.
  - Premounting to ease introduction and passage through the sheath and vascular system.
  - Highly radio-opaque for precise positioning.
- Performance at the site of implantation.
  - Expansion without shortening.
  - High radial force.
  - Conformation to vessel curvature.
  - Smooth edges that do not damage the balloon or vessel wall.
  - Side-branch flow that is not compromised.
  - Minimal neointimal proliferation and non-thrombogenic.
  - Capacity to redilate to adult size.
  - Retrievability if malpositioned.
- Additional features.
  - Covering for aneurysms and fistulae (not compatible with side-branch patency—unless perforated and stented through side branch struts!)
  - MRI compatibility for follow-up.
  - Drug delivery to prevent restenosis.

Types of stent (Fig. 8.2):

- *Closed-cell design*: The original traditional closed-cell design consists of regular cells that do not have direct communication with each other. With expansion the cell changes configuration but all have the same shape—becoming shorter but wider with a high radial force at all diameters. They are inflexible and straighten a vessel rather than conform to its shape.
- *Open-cell design*: A lack of a bridging connection between some adjacent cells allows them to merge into larger areas during stent expansion. This gives greater access to side branches allowing balloon dilation through the cells to improve flow

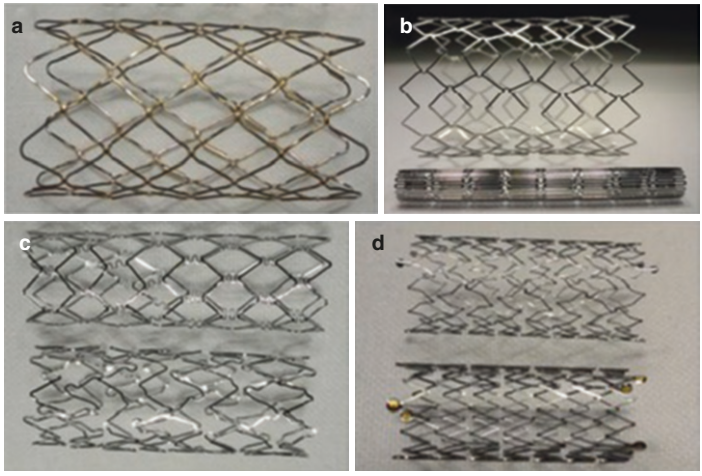

Fig. 8.2 Large balloon-expandable stent in (**a**) expanded closed-cell (Cheatham Platinum) stent and (**b**) expanded and unexpanded hybrid cell (Andramed) stent. Medium-sized balloon-expandable stents in (**c**) closed cell (Palmaz) above and open cell (Valeo) below. Medium-sized self-expanding stents in (**d**) Zilver upper and Sinus-Superflex lower

(Fig. 8.3). They are more flexible, can pass around tighter curves and conform to the vessel shape. They do not crimp as well onto a balloon but the irregular outer surface anchors it to the target lesion reducing the risk of stent migration. They shorten less especially when expanded sequentially but lack radial strength at large diameters. Restenosis may occur due to neointimal hyperplasia through the larger open cells.

- *Hybrid design*: Some stents are designed with an association of open and closed elements in order to keep together radial force, flexibility and anchoring properties.
- *Premouted stents*: Open or closed cells are available in a range of diameters and lengths and can be manufactured in custom lengths and larger diameters. They are quick to prepare and can be advanced safely without a long sheath as they adhere firmly to the balloon catheter.
- *Self-expanding stents:* These are not used as often in congenital heart disease as they have a much lower radial force than

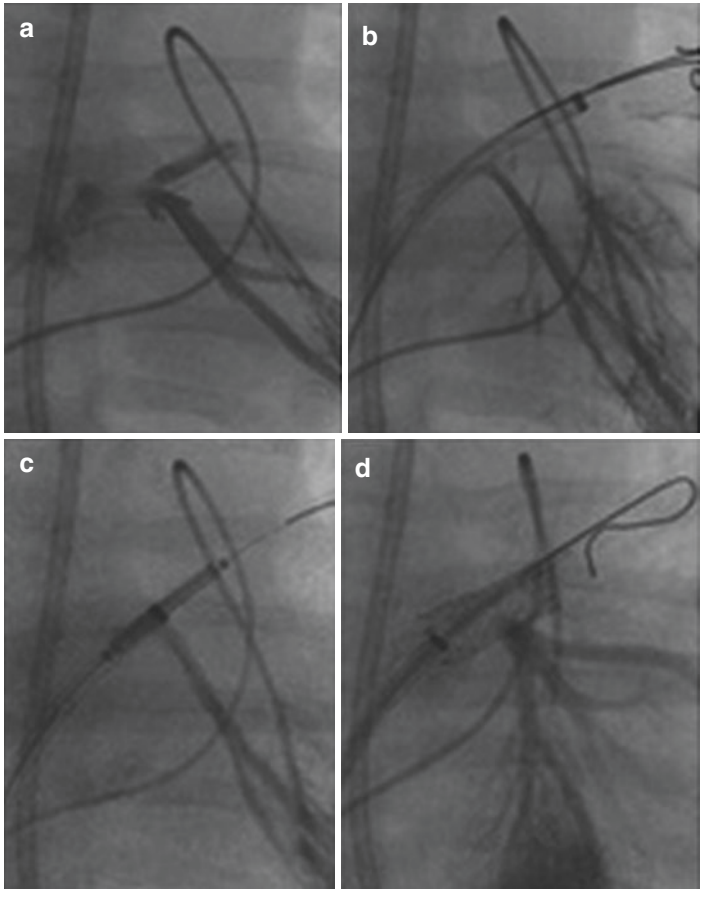

Fig. 8.3 Premounted Visi-Pro stent implantation into a native common left pulmonary vein stenosis after two previous cutting balloon dilations in a 15-month-old boy. Tight stenosis in (**a**) shown by pulmonary artery wedge injection. Long sheath advanced over two guide wires into upper lobe branch and stent uncovered guided by pulmonary artery wedge injection (**b**, **c**). After stent implantation the inferior pulmonary vein is jailed (**d**) and the origin easily dilated with a coronary balloon (**e**) due to the open-cell design with opening of the ostium (*circle* in **f**). CT angiogram 18 months later confirms patency of upper and lower veins into the stent (**g**)

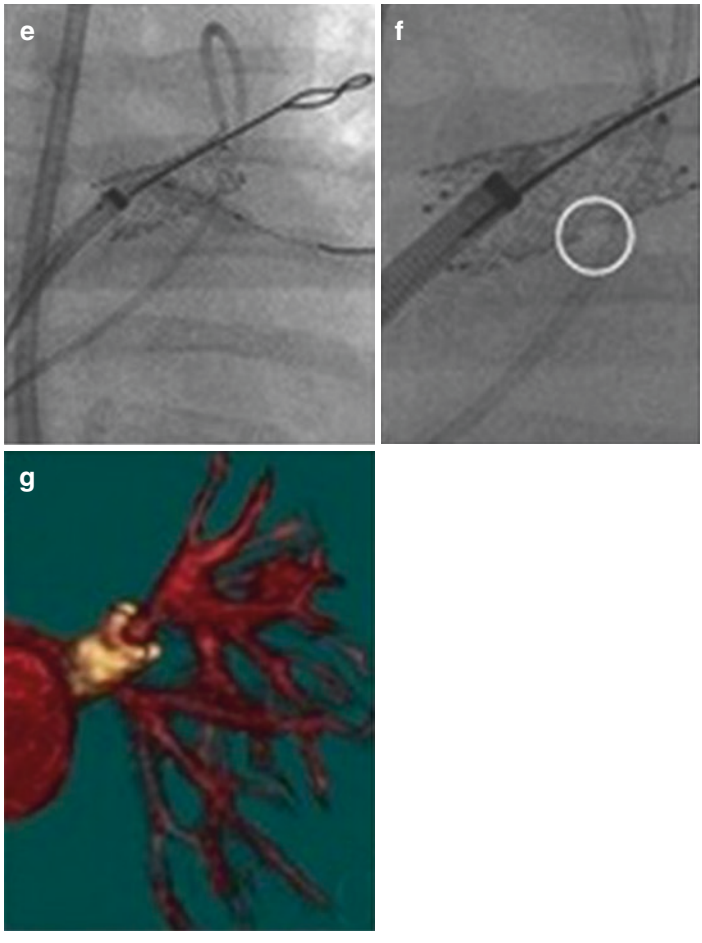

Fig. 8.3 (continued)

balloon-expandable stents but conform well to the vessel shape.

- *Covered stents*: Increasing role in native coarctation and pulmonary conduits allowing full dilation with a reduced risk of vessel damage compared to bare stents. More recently for closure of sinus venosus ASDs.

- Coronary stents: A huge range is available for use in coronary arteries as well as other lesions in neonates and infants.
- *Growth and biodegradable stents*: Metals or polymers that are absorbed by the body or stents with weakened joints that allow easy balloon disruption and a new larger stent to be implanted have been tested but are yet to reach commercial release.
- *Stent grafts*: These are used for aneurysms and dissections of the aorta and beyond the scope of this review.

8.4 Stent Implantation

The basic principles of stent implantation are common to most lesions (Table. 8.1). *Meticulous attention to detail and a structured approach are critical to success without complications*:

- Pre-procedure imaging: Echocardiography, MRI and CT scanning allow the lesion (length, diameter, side branches, adjacent vessel diameters, extrinsic structures (bronchus, coronary artery), aneurysms) and access vessels to be evaluated which when put into the clinical context ensure that:
  - Appropriate stents, sheaths, guide wires, etc. are available.
  - Vascular access is tailored to the lesion (jugular, brachial, carotid, transhepatic, trans-septal, double access, hybrid).
  - Angiographic planes are chosen to reduce contrast and radiation during the procedure.
  - Special measures arranged (transoesophageal echocardiography for atrial septal stenting; radiofrequency perforation for aortic atresia; bronchoscopy, coronary angiography, coils and plugs for hepatic access; surgical standby or ECMO for high-risk patients or lesions).
- Procedure

  Most stenting procedures are performed under general anaesthesia with strict aseptic technique.
- Access

  This depends on the lesion, patient's size and the available vessels. Usually a direct course is preferred if possible. In very

small children or when access is limited or the course is tortuous, a carotid or iliac cutdown or hybrid approach may be needed. For large-bore arterial access, a vascular preclosure suture may be appropriate.

- Angiography

  Good quality images profiling the stenosis (ideally two orthogonal planes) with measurements of the lesion and adjacent vasculature are essential for the final choice of stent size and length and serve as a reference for stent placement. Alternatively, three-dimensional rotational angiography (3DRA) or fusion of computed tomography (Fig. 8.4) or magnetic resonance imaging may be used for guidance of stent implantation [4]. This will be described in details in Chap. 61.
- Predilation

  Balloon dilatation of tight stenoses/subatretic is occasionally needed to introduce the sheath and balloon/stent assembly. Predilation to the planned stent diameter is generally avoided except in special situations. If balloon inflation abolished the stenosis, in distensible lesions, the stent might be insecure after placement and be displaced on balloon withdrawal. In potentially non-compliant lesions (branch pulmonary artery stenosis), predilation testing is important as the stent may obstruct or fracture (Figs. 8.5) if the lesion cannot be dilated; initial high-pressure or cutting balloon dilation may allow subsequent stenting. Balloon inflation can mimic the effects of the stent on adjacent structures (coronary arteries during RVOT stenting, left main bronchus after Norwood surgery (Fig. 8.6)).
- Stent choice

  Many factors influence the stent choice for a particular patient and lesion—not least an operator's experience and preferences (Table 8.2). *One important determinant is the current and final size of the target vessel.*
- Guide wire and sheath placement

  Different catheters and guide wires are used to cross the lesion to as distal and stable a position as possible—*the time spent at this stage is essential to ensure a smooth procedure.* The guide wire to carry the stent balloon assembly is then passed into position. The thickness is dictated by the lumen of

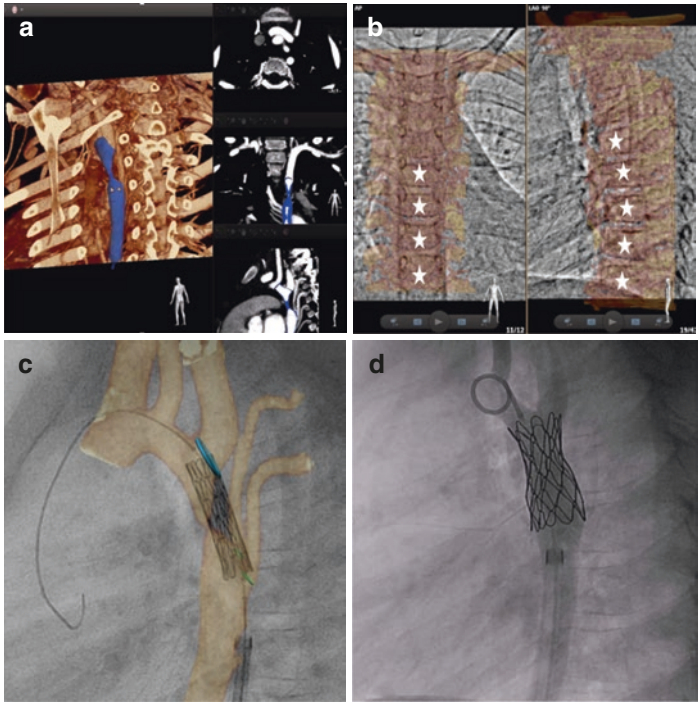

Fig. 8.4 Coarctation of the aorta (CoA) stenting with fusion of computed tomography. A raw computed tomography dataset was manually segmented to expose the narrowing and highlight the nearby vessels **(a)**. Stored fluoroscopy in anterior-posterior and left lateral projections with vertebral bodies (white stars) of the mid and lower thoracic spine served as a reference for matching the 3D reconstruction with the fluoroscopy **(b)**. A Cheatham Platinum stent was positioned with guidance of the 3D overlay **(c)**. A three-dimensional rotational angiography was preformed to evaluate the final outcome **(d)**. Images with permission from Góreczny S et al. 3D image fusion for live guidance of stent implantation in aortic coarctation–magnetic resonance imaging and computed tomography image overlay enhances interventional technique. Postepy Kardiol Interwencyjnej. 2017;13:269–272

the balloon catheter that the stent is mounted on and is usually as stiff as possible.

In most instances a long sheath is advanced across the lesion (Figs. 8.1 and 8.3). It facilitates safe stent placement without displacing the stent when negotiating a tortuous course, tight

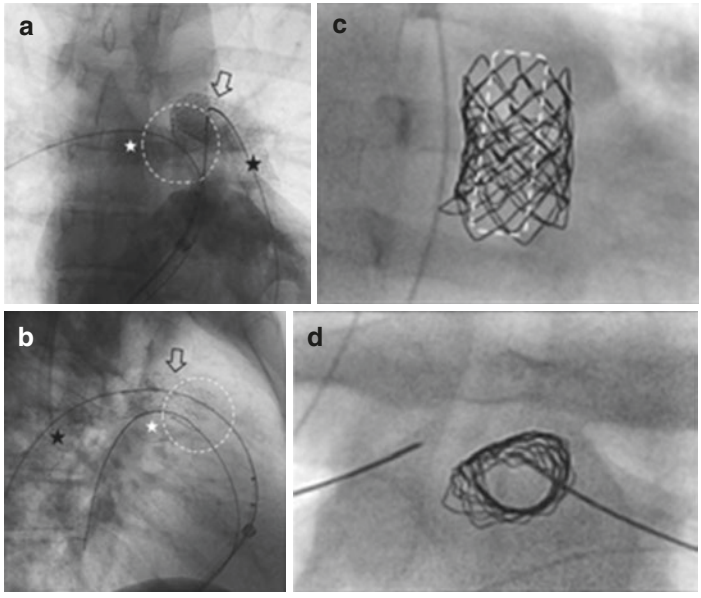

Fig. 8.5 Examples of complications after stent implantation. Proximal LPA & RPA stenoses were previously treated with two stents Isthums and Valeo, respectively (**a**, **b**). The stent (black star) in the LPA embolized distally several months after the procedure. During implantation of a new stent (LD Max, arrow) into the LPA, the previously implanted stent (white star) in the RPA was compressed (white dashed circle). A guidewire placed electively across the RPA stent allowed subsequent redilation of the proximal end. A frame in antero-posterior projection from routine fluoroscopy of Melody valve (after presenting with a Cheatham Platinum stent) shows several insignificant fractures (white dashed rectangle) without fragmentation (**c**). An image in caudal projection reveals compression of the right anterior wall of the valve and stent (**d**). A Cheatham Platinum stent fracture (white dashed circle) late after recoarctation stenting in a patient after surgical treatment of interrupted aortic arch (**e**, **f**). Due to stable position of the fractured struts and close proximity of head vessels originating from the arch through a single trunk, the stent was redilated without implantation of another stent. Left pulmonary artery Palmaz Genesis stent facture in patient after Glenn shunt (**g**). Fractured parts of the stent were reconnected with implantation of a Cheatham Platinum stent (**h**). *LPA* left pulmonary artery, *RPA* right pulmonary artery

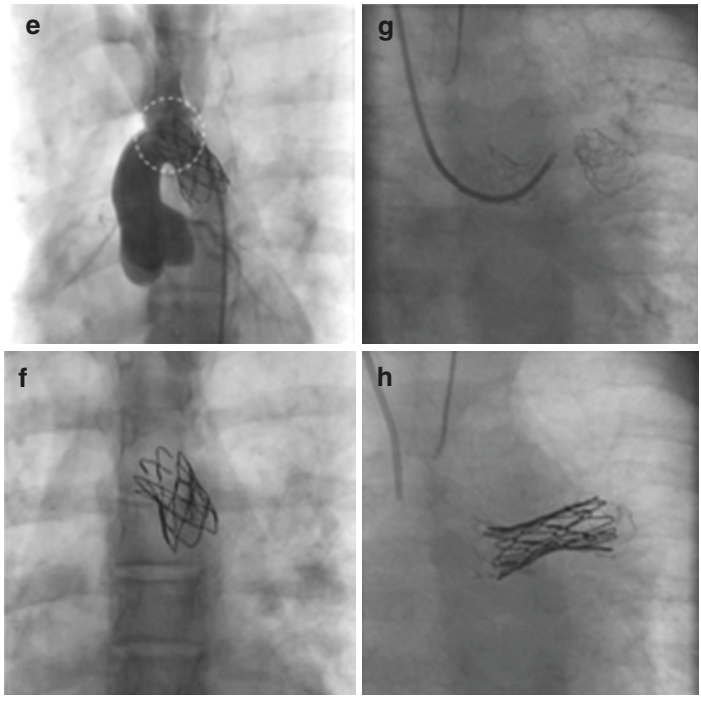

Fig. 8.5 (continued)

bends and stenoses. It allows angiography for proper stent positioning and pressure monitoring. After stent implantation it allows safe balloon withdrawal and placement of a larger balloon if needed and gives control in the event of complications. In small patients, advancing a long sheath over a stiff wire can cause significant tricuspid regurgitation and hypotension. Positioning the tip of the sheath in the right atrium and using the balloon/stent assembly as the sheath "dilator" during advancement can shorten the period of haemodynamic compromise and avoid sheath kinking on dilator removal. Hydrophilic and kink-resistant sheaths also facilitate the procedure. The alternative is to use a short sheath with a premounted stent (a less rigid system)—stent positioning relying

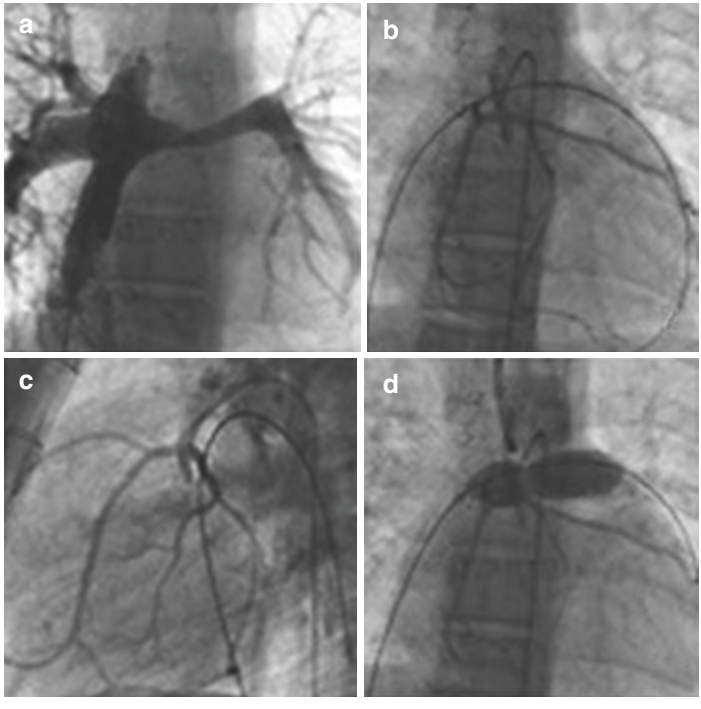

Fig. 8.6 Stent implantation into left pulmonary artery after stage III Norwood procedure in a 17-year-old boy (**a**). MRI scanning raised concerns of proximity of both the left main bronchus and native aorta to the stenosed segment confirmed on angiography in AP and lateral projections with the guide wire in position (**b**, **c**). Trial balloon inflation to the stent diameter was performed with simultaneous native aorta angiography and bronchoscopy without compromising either structure (**d**, **e**). A Cheatham Platinum stent was implanted (**f**, **g**)

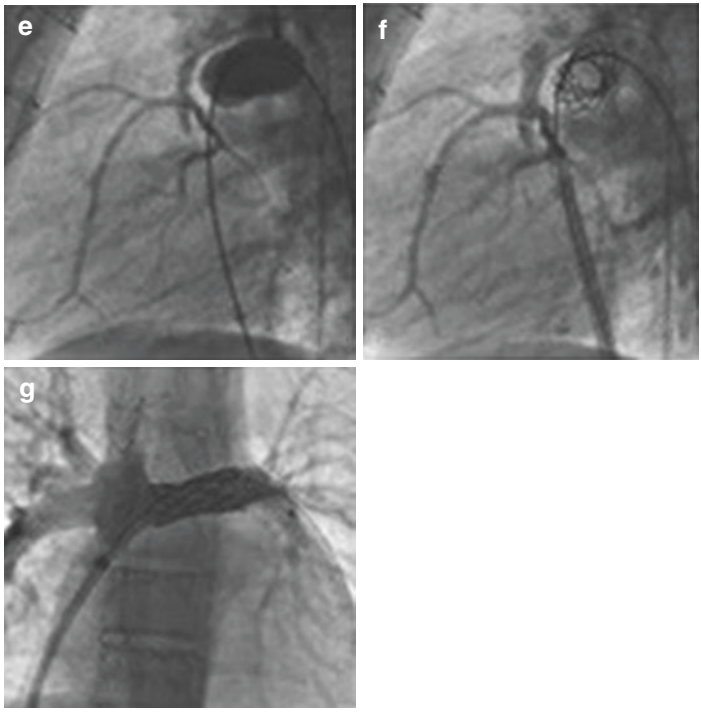

Fig. 8.6 (continued)

on previously acquired landmarks or a separate angiographic catheter.

- Mounting

  Unmounted stents are centred and manually crimped onto the balloon. A stiff guide wire in the balloon prevents compromise of the lumen. Gradually increasing manual force is used symmetrically around the circumference and along the length of the stent. Poor stent adherence can be overcome by application of contrast to the balloon to act as temporary "glue"; umbilical tape wrapped tightly around the stent enhances the crimping; partial balloon inflation allows the stent to grip better. *It is important to match the length of the stent and the balloon.* Too short a balloon results in the ends not inflating—a

risk for stent displacement when the balloon is withdrawn. If too long a balloon is used, the distal end may "milk" back from a small distal vessel causing deployment too proximally. The balloon should therefore ideally be only a few mm longer than the stent itself.

- Stent introduction

  Premounted stents pass easily through the valve of the sheaths. With hand-mounted and covered stents, the stent or covering may be displaced off the balloon or stent if passed directly through the valve. A plastic or metal introducer provided in the stent packet or a short section of another sheath protects the stent during introduction through the valve. Before deploying the stent, it is important to confirm that the stent has not slipped off the balloon—else withdrawal and remounting may be necessary.
- Stent positioning

  At the target site, the long sheath is withdrawn leaving the stent in place. Multiple contrast injections through the sheath (or additional angiographic catheter) are used to fine-tune the position (Fig. 8.1). Still frames with landmarks (bones, trachea, temperature probe), image overlay and roadmapping can be used to help in the final stent positioning. Reliance on these alone may be compromised by distortion of the anatomy by the stiff guide wire/stent balloon assembly. The whole balloon as well as the stent must be uncovered or the proximal part of the balloon may not inflate.
- Stent deployment

  The balloon is inflated with an indeflator up to the recommended pressure to avoid balloon rupture. The primary operator controls the stent balloon assembly and guide wire to reposition the stent if it moves, e.g. if the balloon only inflates proximally pushing the stent distally. The rate of inflation varies—some operators prefer a slow inflation; others a rapid inflation (that gives less scope for repositioning). A balloon-in-balloon results in less stent shortening and an opportunity to reposition the partially expanded stent before full inflation (Fig. 8.1). Rapid ventricular pacing-induced hypotension helps

to maintain the position of coarctation and transverse arch stents.

After deployment the balloon is deflated and angiography used to confirm the stent position. It is important to fully deflate the balloon as withdrawal of a partially inflated balloon may displace the stent. A long sheath can be advanced over the deflated balloon and into the stent to reduce the risk of displacement and allow repeat angiography and placement of a larger/higher pressure balloon if necessary.

8.5 Complications

The larger sheaths and stiffer guide wires used may increase the frequency and severity of complications associated with cardiac catheterisation though they are in general low. *Acute stent-related complications can largely be prevented by meticulous attention to detail.* When they occur, however, it is vital to maintain guide wire position for remedial action with the stent and vessel still accessible.

- Stent malposition or migration

  Minor malposition is dealt with by recapturing the stent with the same / larger balloon and "repositioning" it. If this is not possible, then an overlapping stent is placed to complete treatment of the lesion. If the stent is free floating, recapturing and repositioning may be possible if the stent is still on the guide wire—an alternative is to deploy it in a "safe" position that does not compromise other vessels or will not become stenotic with growth (IVC or descending aorta). If the stent cannot be repositioned with other vascular tools (snares, bioptomes, tip deflectors) and its position causes haemodynamic compromise or is free floating, then surgery is required. Withdrawal of a partially deployed stent to the access site may allow a minor surgical cutdown to remove it.
- Stent embolization

  In rare occasion a stent may embolize after initially successful implantation. For free floating stent a similar approach

as described for stent malposition and migration might be used. If the stent is stable in a distal branch another stent might be implanted proximally to cover the stenosis (Fig. 8.5). If blood flow to the side branche(s) is compromised balloon dilation of struts might be necessary.

- Balloon rupture

  Balloon rupture before the stent is fully expanded is dealt with by rapid contrast injections either by hand or a power injector. If this fails it may be possible to withdraw the balloon from the stent (stabilising the stent with the long sheath or snaring the stent from another access may help) and replace with a new one.
- Side-branch compromise

  Uncovered stents only rarely obstruct a side branch to a hemodynamically significant degree, though late endothelialisation may further compromise flow. Compression of a side branch that exits acutely close to the stenosis may also occur. Open-cell stents can be opened into the side branch to improve flow (Fig. 8.3), but if compression is a concern, then a second guide wire +/− balloon into the side branch can help preserve it during stent deployment. Covered stents over a major side branch need perforating if there is insufficient collateral flow.
- Vessel dissection and rupture

  Minor dissections tend to heal. Excessive dilation of very tight stenoses, dilation much above the diameter of the normal adjacent vessel and sharp edges of some stents can lead to acute dissection and even rupture. Management involves balloon tamponade followed by covered stent implantation across the area. If this is not possible, emergency surgery may be needed. While it is tempting to implant oversized stents to eliminate or reduce the need for further redilatation, significant overdilation that is tolerated initially may lead to aneurysm formation.
- Stent fracture

  Stent fracture may occur immediately after implantation but more frequently is detected weeks later. Fracture of a single or a few struts usually has no clinical significance, whereas complete fracture with stent separation leads to recurrence of the stenosis. Very tight stenoses, sharply angled lesions, muscular

structures and external compression (e.g. the sternum) all increase the risk. Balloon inflation to redilate the stent is rarely successful and risks the free-standing wires puncturing the balloon. Usually a second stronger stent (+/− covered) is implanted to bridge the gap and relieve the stenosis (Fig. 8.5).

- Restenosis

  Restenosis caused by the patient's growth relative to the fixed diameter of the stent is managed by stent redilation. Neointimal proliferation to a degree that compromises the lumen occurs infrequently and unpredictably. It too responds to further dilation of the stent +/− addition of a further stent inside the first (bare or covered). Dilatation beyond the manufacturer's maximum has been described with many stents although each has a limit and may cause a fixed stenosis after growth. Surgical removal or incision and patching the lesion may be needed unless an ultra-high-pressure balloon is able to disrupt the stent—simultaneously deploying a new larger (covered) stent inside the first.

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9 Transcatheter Valve Devices in Congenital Heart Disease

Doff B. McElhinney

Transcatheter valve implantation has revolutionized therapy for cardiac valve disease, particularly dysfunction of the aortic and pulmonary valves, and for failure of surgical bioprostheses in all valve positions. There is ongoing development of new devices and transcatheter therapies for atrioventricular valve dysfunction and to meet currently unserved indications for aortic and pulmonary valve disease. In patients with congenital heart disease, transcatheter valve devices have been used primarily for pulmonary outflow obstruction or regurgitation, secondarily for replacement of dysfunctional tricuspid valve prostheses, and occasionally for left-sided or native tricuspid valve disease. The purpose of this chapter is to provide an overview of transcatheter valve devices used to treat patients with congenital heart disease, as well as con-

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genital heart disease-focused valve devices undergoing clinical trials.

9.1 Balloon-Expandable Valves

The first approved devices for transcatheter pulmonary valve replacement are balloon-expandable prostheses consisting of a valve mounted within a balloon-expandable stent frame. The primary application of balloon-expandable valves is treatment of stenotic and/or regurgitant surgical conduits or bioprosthetic valves [1–7], although the availability of larger balloon-expandable valves has led to more frequent placement within patched or native right ventricular outflow tract anatomies [8–10].

9.1.1 Medtronic Melody Valve

The Melody valve (Medtronic Inc., Minneapolis, MN) comprises a valved segment of bovine jugular vein mounted within a platinum-iridium balloon-expandable stent, and was developed specifically for treatment of dysfunctional right ventricle-to-pulmonary artery conduits (transcatheter pulmonary valve replacement) (Fig. 9.1). The entire length of the stent is covered by the walls of the jugular vein segment, which is sewn to the stent along both ends and at each node. The Melody valve is produced in 2 sizes, which utilize the same stent frame but contain different sized jugular vein valves. The original valve (PB1810) is prepared at 18 mm inner diameter and recommended for expansion on 18, 20, or 22 mm balloons, and the PB1610 has a working range that is 2 mm smaller. However, the Melody valve has been shown to maintain competence at much smaller diameters, and up to expanded diameters of 24–25 mm. The platinum-iridium stent frame is essentially identical to the CP stent (NuMed Inc). There is extensive clinical literature on outcomes after Melody valve implant [1–5, 8].

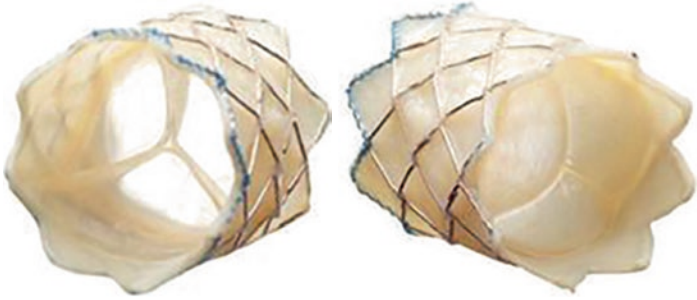

Fig. 9.1 Photographs of the Melody valve viewed from the outflow (left) and inflow (right) perspectives

9.1.2 Edwards Sapien XT and Sapien 3 Valves

The Sapien XT and Sapien 3 transcatheter heart valves (Edwards Lifesciences, Irvine, CA) are second- and third-generation iterations of the original Cribier valve and the first-generation Sapien valve, which were developed primarily for transcatheter aortic valve replacement (Fig. 9.2). Both of these valves comprise a trileaflet bovine pericardial valve that is mounted within a balloon-expandable cobalt-chromium stent frame. The stent is only partially covered, by a polyethylene terephthalate (PET) fabric skirt, which is sewn to the frame and to which the leaflets are sewn separately. The major structural differences between the Sapien XT and Sapien 3 are a cuff at the base of the device to help reduce paravalvar regurgitation in the aortic position, and a greater overall device height. The geometry of the stent frame is also different, but the material (cobalt-chromium) and structural characteristics are similar. The clinical literature on transcatheter pulmonary valve replacement with the Sapien valve is limited but growing [6, 7, 9, 10].

Fig. 9.2 Photographs of the Sapien XT (top) and Sapien 3 (bottom) valves, with leaflets in the open position

9.2 Self-Expanding Valve Replacement Devices

9.2.1 Medtronic Harmony Valve

The Harmony valve (Medtronic Inc., Minneapolis, MN), which is currently available in 2 different sizes, comprises a self-expanding nitinol stent frame, with 6 independent zigs, and an internal fabric covering that is sutured to the frame along the length of each zig. The ends of the device are flared to secure it within the right ventricular outflow tract, and the smaller central portion houses a trileaflet porcine pericardial tissue valve. The image below depicts the smaller of 2 Harmony valve sizes, which has a 22 mm valve, measures 34 mm fully expanded at the outflow end, 42 mm at the inflow end, and in its unconstrained form is approximately 55 mm

long. The Harmony valve received premarket approval from the U.S. Food and Drug administration in March 2021, and although there is limited clinical experience, the preliminary results are generally encouraging [11, 12] (Fig. 9.3).

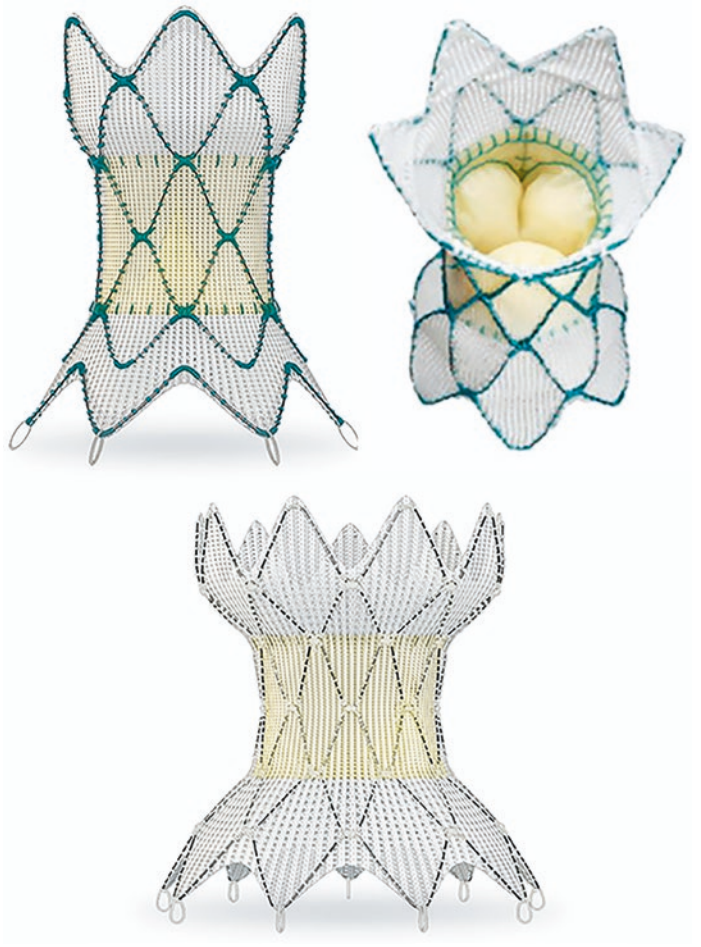

Fig. 9.3 Photographs of the Harmony valve: (Top) the 22 mm size device, viewed from the side (left) and from the inflow (right) aspects; (Bottom) the 25 mm device, viewed from the side

9.2.2 Edwards Alterra Adaptive Prestent

The Alterra adaptive prestent (Edwards Lifesciences, Irvine, CA) is unique among the devices described in this chapter, insofar as it comprises a docking station only, which is designed as a landing zone for the previously discussed Sapien 3 valve. The concept of the "adaptive present" derives from the prototypical infundibular reducer devices described by Boudjemline et al. in 2004 [13]. The Alterra prestent is composed of a 48-mm-long laser-cut nitinol frame covered internally with polymer fabric that is sutured to the frame. The central portion of the frame is designed as the landing zone for the 29 mm Sapien 3 valve, and the outflow and inflow segments are symmetrically flared to a larger diameter (47 mm). The distal cell is uncovered, and the tips of the distal-most and proximal-most rows of the device bend outward to protrude into the wall of the RVOT and PA. Early results in the 15 patients were encouraging [14] (Fig. 9.4).

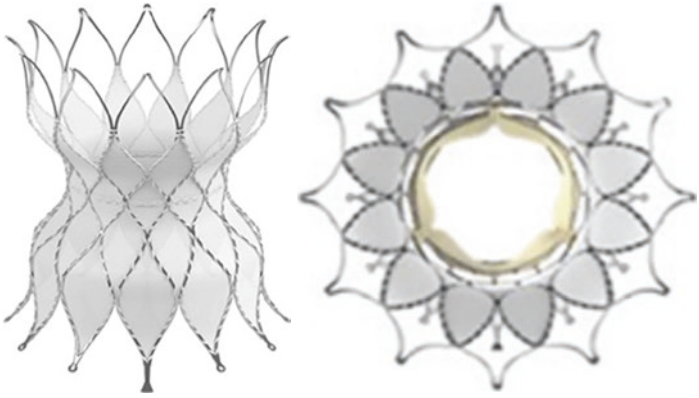

Fig. 9.4 Photographs of the Alterra prestent from the side (left), and from the outflow perspective with a Sapien valve implanted (right)

9.2.3 Venus P Valve

The Venus P Valve (Venus Medtech, Shanghai, China) comprises a laser-cut self-expanding nitinol frame that is covered internally with porcine pericardial tissue and houses a trileaflet porcine pericardial valve. The valve-containing central portion is either 20 or 30 mm in length, and ranges in diameter from 20 to 32 mm in 2 mm increments. The frame is secured within the right ventricular outflow tract by flared proximal and distal segments, which 7 mm and 10 mm wider than the central portion, respectively, and each 10–14 mm in length. Radiopaque markers indicate the distal extent of covering and the level of the valve. The device is manufactured in a variety of diameters) and lengths (20 and 30 mm straight sections lengths. Several multicenter studies on the Venus valve have been published [15–18] (Fig. 9.5).

Fig. 9.5 Photograph of the Venus P valve from the side perspective

9.2.4 Taewong Pulsta Valve

The Pulsta Valve (TaeWoong Medical Co, Gyeonggi-do, South Korea) is comprised of a treated porcine pericardial valve that is mounted within a self-expanding frame of knitted double-stranded nitinol wire that is partially covered internally by porcine pericardium. The valve ranges in diameter from 18 to 28 mm and is hand-sewn to the central portion of the nitinol frame, which has uncovered sections in the inflow and outflow that flare to diameters 4 mm larger than the valve. The length varies in accordance with diameter, from 28 to 38 mm. The first human implant of the Pulsta valve was reported in 2017 [19], and procedural and 6-month follow-up results were reported for a 10-patient feasibility trial in 2018 [20] (Fig. 9.6).

9.2.5 Med-Zenith PT-Valve

The Med-Zenith PT-Valve (Beijing Med-Zenith, Beijing, China) is comprised of a porcine pericardial valve that is mounted within a self-expanding nitinol frame that is covered internally by porcine pericardium (Fig. 9.7). The frame has a cylindrical central segment that supports the valve, with symmetric outward flaring of the inflow and outflow portions. The valve diameter is manufactured in three sizes (20, 23, and 26 mm) and the size of the frame ranges in largest diameter from 28–44 mm and in length from 38–54 mm. The initial human experience with the PT-Valve was reported in 2019, with successful implantation in 4 patients with severe PR [21].

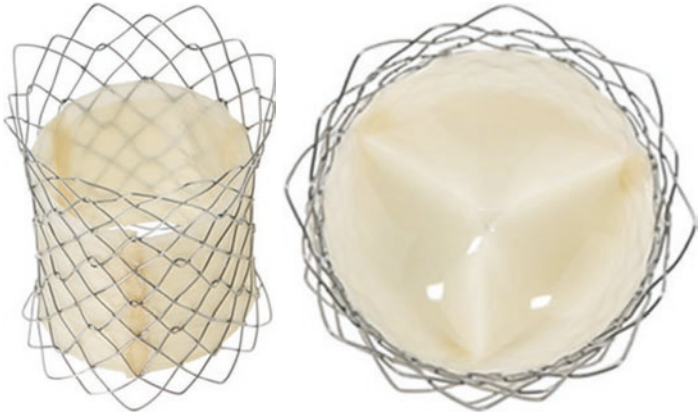

Fig. 9.6 Photographs of the Pulsta valve viewed from the side (left) and showing the valve leaflets from the outflow end (right)

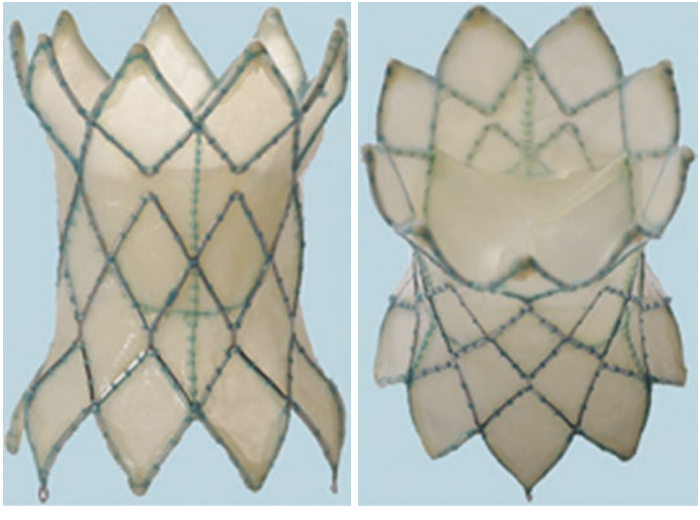

Fig. 9.7 Photographs depicting the PT-valve from the side (left) and showing the valve from the outflow end (right)

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10 "Adult" Tools Relevant for Congenital Heart Disease

Aphrodite Tzifa and Charalambos Kavvouras

10.1 Equipment

10.1.1 Cardiac Catheters

Appropriate catheter selection is an important decision, since the best choice of guide catheters makes the procedure simpler without exposing the patient to any unnecessary risks or unnecessary screening time. The operator should have good knowledge of the different types of catheters available and should know which one to use and when.

Catheter lengths used are 80 to 110 cm depending on the anatomic characteristics and the access site. Catheter selection depends on the approach (radial or femoral), the height of the patient, and the aortic diameter and curvature. Guide catheters are available in diameter sizes from 5 to 8 F with each F being equivalent to 0.33 mm with inner lumen diameters (ILD) between 0.056 and 0.091 in.

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An operator has a big variety of catheters to choose from, such as the Multipurpose, NIH, Judkins coronary catheters, the Amplatz, Pigtail, Cobra catheters, etc. During the early years of interventional cardiology, catheters were large, stiffer and difficult to handle with higher rates of catheter-associated complications. As technology evolved catheters became thinner with lower profile and today small sized guide catheters can be used, even for the most complex interventions in pediatrics, without risk. The shape of the catheter chosen depends on the target lesion for which intervention is being planned, taking into consideration origin characteristics, such as acute takeoff and its distal course.

Right heart catheterization is often performed with a wedge (arrow) catheter. In order to cross the tricuspid valve, it is suggested that the balloon tip is inflated to avoid going through chordae of the tricuspid valve subvalvar apparatus, which may result in chordal rupture when the catheter is exchanged for a long sheath over a wire. In patients with severe pulmonary hypertension due to severe dilation of the right heart chambers or due to severe tricuspid valve regurgitation, advancing the Swan–Ganz catheter to the pulmonary artery and PA wedge position may be challenging, hence a Berman Catheter can be used instead. The Berman catheter does not have an end-hole and cannot be used over a guide wire, however a wire can be placed inside the catheter to shape it and support it when entering the desired location. Besides the above-mentioned catheters, special shape catheters used in adult interventional radiology or cardiology, such as the RIM catheter, the LIMA catheter, etc. are also occasionally used in pediatrics, in order to reach vessels with acute angulations, origin and course.

Guide catheters of 5–9 Fr, routinely used for coronary interventions in adults, can be utilized in the congenital lab to deliver ductal devices and vascular plugs for embolization of anomalous vessels (arterial ducts, aortopulmonary collaterals, coronary artery fistulas, etc.) and for closure of paravalvular leaks. Guide catheters can also be used for delivery of small diameter stents or coronary stents for branch pulmonary artery stenting, arterial duct stent implantation, RVOT stenting, etc.

10.1.2 Coronary Wires

Coronary wires of 0.014″ are frequently used in congenital interventions across all age groups but are particularly helpful in facilitating neonatal and infantile procedures, as indicated below.

Indications for coronary guidewire use:

(a) Crossing the aortic or the pulmonary valve for balloon dilation in neonatal and infantile aortic and pulmonary stenosis.
(b) Crossing and stenting of a patent ductus arteriosus (PDA).
(c) Palliative stenting of aortic coarctation or transverse arch in neonates and infants, when surgery is contraindicated.
(d) Stenting of severely stenosed branch pulmonary arteries or recanalization of occluded pulmonary arteries.
(e) Perforation of an atretic pulmonary valve: When an RF perforation system is not available, the stiff end of a coronary guidewire has been employed to perforate the valve with care. In these cases, the back end of a BMW universal guide wire (Abbot) or one of the stiffer coronary wires, used for total chronic coronary occlusions, such as the Conquest Pro wire (Asahi Intecc Co. Ltd. 3–100 Akatsuki-cho, Seto, Aichi 489–0071, Japan) have been tried successfully.
(f) Recanalization of severely stenosed or chronically occluded small vessels, such as femoral vessels.

10.1.2.1 Types of Coronary Guidewires

The most commonly used coronary guidewires in pediatrics are the Luge guidewire (Abbott), the Choice PT Extra Support (Boston Scientific, USA), the BMW universal guidewire (Abbott) and the Conquest Pro wire (Asahi Intecc Co. Ltd., Japan). It is also worth mentioning the Safesept wire, commonly used for transseptal punctures in adults. Its use can be expanded to Pediatrics, in order to perforate the septum in small children, but it has also been used to perforate atretic pulmonary valves when an RF perforation system is not available. Once it has gone through the os created, its distal tip curls up to form a "j," thereby preventing cardiac or vascular trauma.

10.1.3 Coronary Balloons

Coronary balloons have been routinely used over the past few decades for congenital cardiac interventions. They are available in diameters of 2–5 mm. *Over-the-wire balloons* are more preferable, as they have better torquability and pushability. However, *monorail balloons* are also used, when the over-the-wire options are not available.

Of late, *drug-eluting balloons* have also been added in the Pediatric Cardiologists' armamentum for lesions that commonly develop intimal proliferation, such as the pulmonary veins, in cases of idiopathic pulmonary venous stenosis or in patients with Williams syndrome, whose vessels present significant intimal thickening and proliferation.

Indications for use of coronary balloons in congenital heart disease:

- Fetal cardiac interventions: balloon dilation of fetal valves or atrial septum.
- Pre-dilation of nearly atretic vessels, such as branch pulmonary arteries or nearly atretic isthmus, prior to further, larger size ballooning and stenting (Fig. 10.1).
- Pre-dilation of atretic or nearly atretic valves. When the passage through the valve is a pinhole, pre-dilation using a coronary balloon (diameter 2.5–3 mm; length 15–20 mm) may be needed.
- Balloon dilation of stenosed femoral vessels.
- Balloon dilation of Blalock–Taussig shunts.
- Balloon dilation of PDA.
- Balloon dilation of neonatal SVC/carotids/MAPCAs.
- Dilation of stent struts to create an orifice for a branch pulmonary artery.

10.1.3.1 Types of Coronary Balloons

Coronary balloons include standard balloons and non-compliant balloons (NC) that can reach higher atmospheres without bursting. Different types of coronary balloons most frequently used in

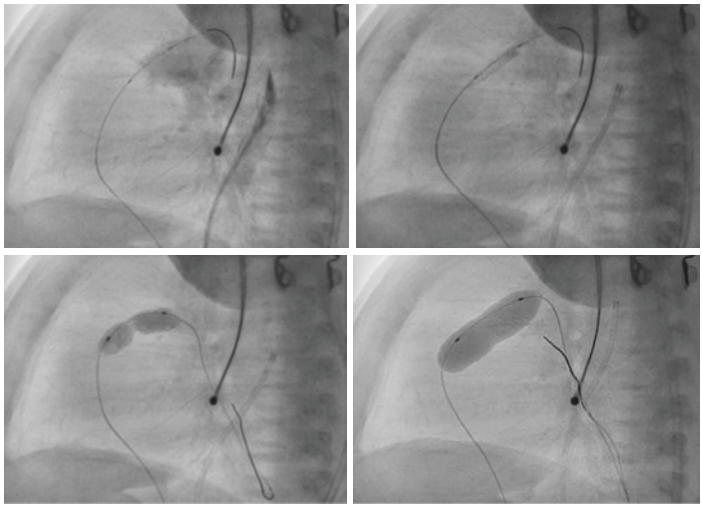
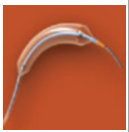


Fig. 10.1 PA/IVS neonate: Pulmonary valve perforation and initial dilation with coronary balloon 2.5 mm up to final size 8 mm

congenital heart disease include, but are not limited, to the following. Their characteristics are given in Table 10.1.

- **Apex balloon** (Boston Scientific). They are available in a wide array of diameters from 1.5 mm to 5.0 mm and lengths ranging from 8 mm to 40 mm. The OptiLEAP balloon material provides sizing flexibility. All sizes are available in Monorail and Over-the-Wire Catheter platforms. Boston Scientific state that the APEX Balloon Catheter is 35% more pushable than the MAVERICK™ Balloon Catheter.
- **Maverick balloons** (Boston Scientific). The Maverick Over-The-Wire Balloon Catheter has excellent crossability, flexible TrakTip Design for navigating through tight lesions and tortuous anatomy. The **Maverick XL** Monorail Balloon Catheter is available in wider (up to 6.0 mm diameters). In specific, the Maverick XL balloon diameters include: 4.0, 4.5, 5.0, 5.5, and 6.0 mm.

Table 10.1 Types of coronary balloons

							
Balloon name	Apex balloon	Maverick balloons	NC Quantum Apex	NC Trek	NC Euphora	NC Sprinter	Cutting balloons
Manufacturer	Boston Scientific	Boston Scientific	Boston Scientific	Abbott	Medtronic	Medtronic	Boston Scientific, MA, USA
Balloon material/design	OptiLEAP/ RX and OTW	DynaLEAP/RX	N/A/RX and OTW	CrossFlex/ Multilayer	Proprietary nylon blend/ Light	Soft Fulcrum Plus/RX	Noncompliant Nylon/RX
Guide catheter compatibility	5F	5F (Maverick XL requires 6Fr)	6 F	5Fr	5 F	5 F	6F
Balloon diameter (mm)	1.5–5.0	1.0–5.0 (Maverick XL goes up to 6 mm)	2–5.0	2–5.0	2–5.0	2–5.0	2–8.0
Balloon length (mm)	8.0–40	9.0–30	6.0–30	6.0–25	6.0–27	6.0–27	6.0–15
Nominal implantation pressure	6 atm	6 atm	12 atm	12 atm	12 atm	10 atm	6 atm

- **NC Quantum** Apex catheter (Boston Scientific). The company state that this is the best balloon in this category with 27% better distal end flex, 26% improved tip flexibility, 23% less tip catch and 23% improvement in recross, excellent visibility via platinum iridium marker bands. Rated burst pressure is 20 ATM (18 ATM RBP for 4.5 mm and 5.0 mm diameters). They are available in Monorail™ or Over-the-Wire Designs.
- **NC Trek** (Abbott). Available from 2 mm to 5.0 mm balloon sizes. Indicated for balloon dilatation of the stenotic portion of a coronary-artery or bypass graft stenosis or coronary-artery occlusion and also for balloon dilatation of a stent after implantation. In Pediatrics it can be used to dilate tight small lesions, shunts and also implanted coronary stents in non-coronary lesions. Abbott have tested post-deployment stent expansion with the MULTI-LINK VISION and MULTI-LINK ULTRA stents.
- **NC Euphora** (Medtronic) noncompliant balloons. They are available in up to 5 mm in diameter. They have been bench tested vs. NC Trek and NC Quantum Apex balloons. They have high RBP up to 20 atm, **excellent recross** in difficult cases and can be re-inflated up to 20 times to RBP without bursting. They are very useful in dilating postoperative scar lesions in the pulmonary arteries.
- **NC Sprinter** (Medtronic). Available in sizes 2.00–5.00 mm and lengths: 6, 9, 12, 15, 21, 27 mm. Nominal pressure is 10 Atm and rated burst pressure 18 Atm.
- **Cutting balloons** (Boston Scientific Cutting Balloon®, Boston Scientific, MA, USA). They are used to dilate pressure-resistant stenosis in pulmonary arteries and veins, by creating four controlled tears in thickened intima and media, as well as relieve in-stent stenosis when severe intimal proliferation has developed. They come in diameters of 2–8 mm and have a burst pressure of 8 Atm. Care should be taken to deliver and withdraw them in a long sheath, Slow inflation allows for the wings to expand evenly exposing the blades. Deflation should

also be slow in order to refold the wings adequately and cover the blades before withdrawal into the sheath. This will ensure avoidance of peel off and detachment of a blade.

10.1.4 Coronary Stents

Coronary stents have been used in pediatric interventional cardiology for the past few decades. Initially they were utilized to perform ductal stenting. They now have a wide range of applications, mostly in the neonatal and pediatric population. Published studies have shown excellent medium and long-term results in maintaining vascular patency and deferring surgical procedures in high-risk patients.

Indications for implantation of coronary stents in congenital heart disease:

- Hybrid procedures—PDA stent implantation (Fig. 10.2).
- Ductal stenting for duct-dependent pulmonary or systemic circulation (Fig. 10.3).
- Stent implantation of Blalock–Taussig shunts, either for shunt recanalization (Fig. 10.4) or shunt size upgrade.
- Stent implantation of stenosed MAPCAs.
- Stent implantation for stenosed pulmonary veins.
- RVOT stent (in pulmonary atresia after valve perforation or in severely cyanotic Tetralogy of Fallot as an alternative to Blalock–Taussig shunt).
- Palliative procedures of stent implantation in the neonatal arch (Fig. 10.5) or branch pulmonary arteries in inoperable or high-risk cases (Fig. 10.6).
- Stenting of the atrial septum. Coronary stents may be used to ensure a patent interatrial communication in patients with HLHS, either during fetal or neonatal life. In addition, in neonates placed on ECMO, atrial septal stenting with coronary stents up to 5 mm may be performed, although larger diameter stents are most commonly used to stent the atrial septum postnatally.

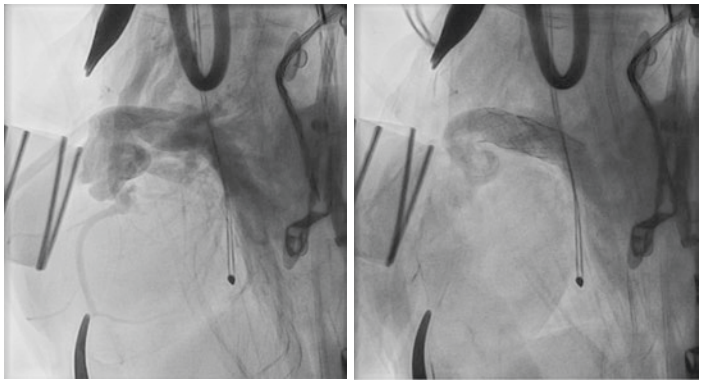


Fig. 10.2 Hybrid procedure for HLHS. PDA stent implantation

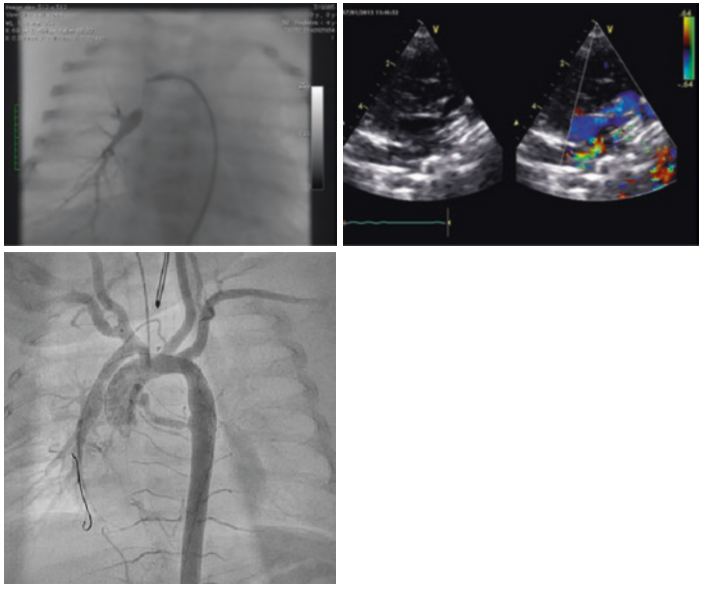


Fig. 10.3 10-day-old symptomatic neonate with non-confluent branch pulmonary arteries and RPA from a restricted right sided duct: Ductal opening with implantation of 2 overlapping coronary stents of 3.5 mm in diameter

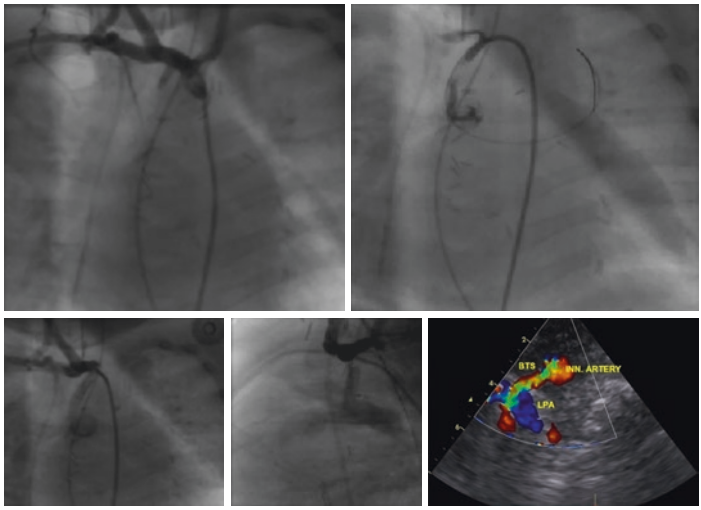


Fig. 10.4 6-month-old with blocked BT shunt: Emergency recanalization with 2 overlapping coronary stents of 4 mm in diameter

10.1.4.1 Types of Coronary Stents Most Frequently Used in Congenital Heart Disease

Coronary stents come in diameters of 2.25–5 mm and varying lengths. The most commonly used stents in pediatric cardiac interventions are given below and also in Table 10.2:

- VeriFLEX TM bare metal stent (ex-Liberte Bare metal stent, Boston Scientific). It is a stainless steel stent premounted on an Over-The-Wire or Monorail Balloon Catheter. It has two radiopaque markers which aid in the accurate placement of the stent and is available in diameters of 2.75 to 5.0 mm. It has been very frequently used in PDA stenting as a single stent or >1 overlapping stents.
- Rebel bare metal stent (Boston Scientific). It is a platinum chromium stent, available in diameters of 2.25 to 4.5 mm.
- Integrity stent (Medtronic Co). It is a cobalt-chromium stent, available in diameters of 2.25 to 4.0 mm and varying lengths.

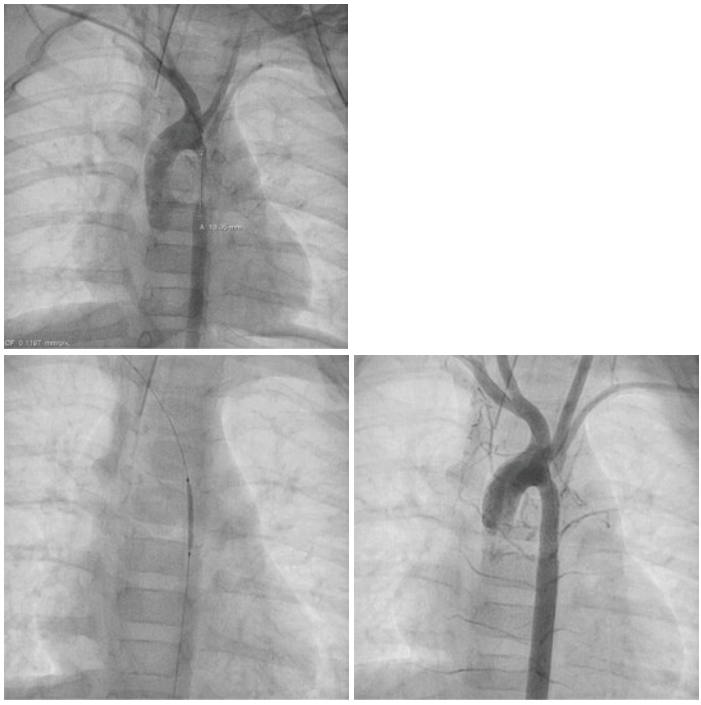


Fig. 10.5 18-month-old Williams patient with multiple recurrencies of aortic recoarctation: Implantation of a drug-eluting Resolute onyx stent of 5 mm diameter

- Vision (Abbott). Cobalt-chromium stent, available in diameters of 2.75 to 5.0 mm and varying lengths.
- PTFE covered stents of small diameters: The most frequently used stent in this group is the GRAFTMASTER® (previously JoGraft; Jomed, and now GRAFTMASTER; Abbott Vascular, Santa Clara, CA, USA), which is constructed using a sandwich technique, whereby a layer of ePTFE is placed between two 316 L stainless steel stents. A similar technology is applied in the Direct-Stent® (InSitu Technologies Inc., St. Paul, MN, USA), while the BeGraft coronary stent graft system (Bentley Innomed GmbH, Hechingen, Germany) combines a single

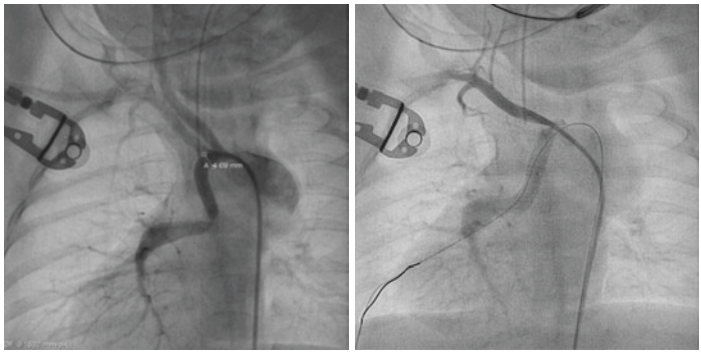


Fig. 10.6 1-year-old with severe failure to thrive, disconnected branch pulmonary arteries with RPA from Rt sided duct with distal severe stenosis and desaturation of 50%: Implantation of 2 overlapping coronary stents of 4.5 mm in diameter within the duct and origin of right pulmonary artery

layer cobalt-chromium stent platform with an ePTFE membrane clamped at the stent ends.

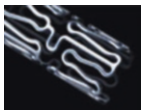
- Drug-eluting stents (i.e., Resolute Onyx stent, Medtronic) for use in the pulmonary veins for idiopathic pulmonary vein stenosis and also aortas of small syndromic patients, such as in Williams syndrome.

In addition, it should be mentioned that growth and biodegradable stents (metals or polymers that are absorbed by the body or stents with weakened joints that allow easy balloon disruption and a new larger stent to be implanted) have been tested but are yet to reach commercial release.

10.1.5 Delivery Sheaths

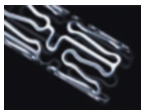
- The **Agilis NxT introducer** (Abbott) is designed to replace a fixed curve introducer. It helps reaching difficult areas of the heart, by providing a stable platform for improved catheter manipulation and contact. The introducer's steerable sheath tip can be adjusted to aid various anatomic approaches with just

Table 10.2 Types of coronary stents

						
Stent name	VeriFLEX TM (ex-Liberte bare metal stent) Monorail or over the wire	Rebel Bare metal stent	Integrity Bare Metal stent	Vision	Covered BeGraft stent	Covered Graftmaster (ex-Jomed stent)
Manufacturer	Boston Scientific	Boston Scientific	Medtronic Co	Abbott	Bentley Innomed	Abbott Vascular
Stent material/ design	Stainless steel	Platinum Chromium/ RX	Cobalt - chromium (F-562) single layer	Cobalt - chromium (L-605)/RX and Otw	Cobalt- chromium (L-605) single layer	Stainless steel (316 L)/ sandwich design
Guide catheter compatibility	5 Fr	5 Fr	5 Fr	5 Fr	5 Fr	6 Fr (≤4.0 mm) 7 Fr (4.5 and 4.8 mm)

(continued)

Table 10.2 (continued)

						
Stent name	VeriFLEX TM (ex-Liberte bare metal stent) Monorail or over the wire	Rebel Bare metal stent	Integrity Bare Metal stent	Vision	Covered BeGraft stent	Covered Graftmaster (ex-Jomed stent)
Stent diameter (mm)	2.75–5 mm	2.25–4.50 mm	2.25–4.0 mm	2.75–5.0 mm	2.5–10 mm	2.8–4.8 mm
Stent length (mm)	8.0–32	8.0–32	9, 12, 15, 18, 22, 26, 30	8, 12, 15, 18, 23, 28	8.0–24	16–26
Nominal implantation pressure	9 atm	9 atm	9 atm	9 atm	11 atm (2.5–4.0 mm) 10 atm (4.5–5.0 mm)	15 atm

one introducer. The steerable sheath is offering outstanding agility, stability and versatility. The atraumatic tip reduces the likelihood for injury during transseptal punctures. Its inner lumen is 8.5 Fr, hence it can accommodate telescopic passage of another long sheath of 6 Fr and below, or passage of a wide variety of catheters. The Agilis sheath is commonly used for closure of mitral valve paravalvular leaks (particularly helpful in septal/ medial location of PVLs) or other mitral valve interventions, such as balloon dilation of stenotic rheumatic mitral valves, as well as in electrophysiology procedures.

- The **GORE Dry Seal sheath (Gore) combines enhanced flexibility, kink resistance, and a hydrophilic coating, facilitating access to challenging anatomies and branch vessels, such as through the common iliac arteries, and into tortuous anatomies, such as the RVOT and the branch pulmonary arteries**. It is available in 12 to 26 Fr diameters, in 1Fr increments, and has a working length of 28 cm. The Gore Dry Seal sheath adapts to a large number of different devices and is pressurized to create a seal, hence no manipulation is required to maintain hemostasis, decreasing procedural blood loss and reducing the potential need for transfusion. It is frequently used for delivery of transcatheter valves into the right heart, as well as complex branch pulmonary artery stenting procedures.
- The **Medtronic Flexcath Advance steerable sheath** is a unidirectional, deflectable sheath used to help position mostly ablation catheters. It offers 135 degrees of deflection and it may facilitate better access to inferior pulmonary veins. It is fitted with a valve to allow for introduction, withdrawal, and exchanging of catheters and wires.
- The **DIREX™ Steerable Sheath** (Boston Scientific) is a low profile, bidirectional steerable sheath that provides agility and stability through excellent catheter access and support. Bidirectional steering allows dual deflection for maximum maneuverability. The steerable sheath facilitates access to hard-to-reach sites. Its braided shaft provides exceptional torqueability, pushability, and kink resistance. It is compatible with guidewires up to 0.038″/0.97 mm. It facilitates passage of a wide variety of catheters.

10.2 Transcatheter Imaging Tools

10.2.1 Fractional Flow Reserve

Fractional flow reserve (FFR) utilizes a specialized guide wire to measure blood pressure within a coronary artery. Large randomized studies have shown that FFR is superior to angiographic assessment for the detection of hemodynamically important coronary-artery stenoses and that use of FFR to guide coronary revascularization improves clinical outcomes. The instantaneous wave-free ratio (iFR) is a recently developed physiological index used to assess the severity of stenosis. The iFR is calculated by measuring the resting pressure gradient across a coronary lesion during diastole when microvascular resistance is low and stable. The benefit of iFR includes the ability to obtain an instantaneous lesion assessment without the need to administer a hyperemic agent, such as adenosine.

According to the American College of Cardiology (ACC) guidelines on coronary revascularization, FFR is reasonable for the assessment of angiographic intermediate coronary lesions (50–70% diameter stenosis) and can be useful for guiding revascularization decisions in patients with CAD (Class IIa, Level A). A recent consensus document on the use of FFR has suggested to expand its use in coronary stenoses up to 90%, given that up to 20% of lesions between 70 and 90% stenosis are not hemodynamically significant if FFR demonstrates the absence of hemodynamic relevance, the risk of future events associated with a stenosis is low, and cannot be further reduced by PCI with modern stents.

This *micromanometer pressure wire* can also be used as an alternative device for rapid and safe determinations of pulmonary artery pressures in children e.g., with aortopulmonary artery shunts, thus avoiding the passage of a catheter through a small shunt. In addition, FFR is occasionally a useful tool for assessment of the functional severity of coronary fistulas.

10.2.2 Intracardiac Echocardiography

Intracardiac echocardiography (ICE) allows for real-time assessment of cardiac anatomy during interventional procedures and guides catheter manipulation in relation to the different anatomic structures. In contrast to transesophageal echocardiography (TEE), ICE can be performed by the primary operator under conscious sedation, without the need for endotracheal intubation. It also reduces fluoroscopy exposure to both the patient and the operator by shortening the procedure time and facilitates early recognition of complications such as thrombus formation or pericardial effusion.

In the current era, two different ICE technologies exist, namely the radial or rotational ICE which uses a single piezoelectric crystal mounted at the tip of a 6- to 10-French catheter. A rotating transducer provides cross-sectional images in a 360-degree radial plane perpendicular to the long axis of the catheter. *Rotational ICE* operates at imaging frequencies of 9 to 12 MHz, which is useful for near-field imaging of up to 6 to 8 cm but limited for far-field imaging. The *phased-array ICE* consists of a 64-element transducer mounted on the distal end of an 8- or 10-French steerable catheter that can be deflected in 4 directions (anterior, posterior, right, and left). This catheter produces a wedge-shaped image that is displayed on a conventional ultrasound workstation. Phased-array ICE has several advantages over the mechanical rotational systems, including greater depth of penetration (up to 15 cm), greater maneuverability, and the ability to acquire Doppler and color flow imaging. Because of these advantages, phased-array ICE is preferred in the majority of interventional cardiology procedures, and the remainder of this article will focus exclusively on this ICE modality.

ICE provides comprehensive assessment of the atrial septal defects and the surrounding tissue rims. It can therefore be utilized for various interventional procedures such as ASD or PFO closure, VSD device closure, mitral valvuloplasty, and many electrophysiological procedures, such as transseptal puncture.

Insertion and withdrawal of the phased-array ICE probe will result in imaging more superiorly and inferiorly. Axial rotation allows for sweeping of the image through multiple planes. Three-dimensional ICE has recently become commercially available but limited data exist regarding the role of 3D ICE in percutaneous transcatheter procedures at present. The use of 3D ICE offers the potential to provide greater anatomic information during structural interventions but requires additional investigation to fully define its role.

Further Reading

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11 Hemodynamic Assessment: Pressures, Flow, Resistances and Vasoreactive Testing

Juan Pablo Sandoval Jones
and Lee N. Benson

11.1 Hemodynamic Assessment

Hemodynamics, a word derived from the Greek meaning blood power, is the study of the physical properties of the circulation of blood, including cardiac function and peripheral vascular physiology and the physical laws that control blood flow. These elements can be monitored in the catheterization laboratory and provide insights into cardiac performance in healthy individuals and insight to cardiac performance in patients with congenital or acquired heart disease. Invasive measurements during cardiac catheterization can document systemic and pulmonary arterial pressures, vascular resistances, and cardiac output. Such information can be used to guide clinical decision making and define the treatment strategies for patient care.

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G. Butera et al. (eds.), *Cardiac Catheterization for Congenital Heart Disease*, https://doi.org/10.1007/978-3-030-69856-0_11

11.2 Pressure Evaluation and Waveforms (Table 11.1)

The graphical representations of pressure waveforms play an important role in understanding the hemodynamic assessment. Accurate acquisition and interpretation allows diagnosis of cardiac function (or dysfunction) based on the interrelated events within the cardiac cycle, while inaccurate measurements can lead to misinterpretation and potential harm.

Pressure changes within the cardiac chambers and vessels are generally recorded through membrane transducers transforming the pressure signal to an electrical signal, which is filtered, amplified, and displayed as a change in pressure over time. The transducer may be located at the tip of a catheter, as a high-fidelity pressure sensor that provides high-fidelity representations of the pressure changes. A major disadvantage of such systems is cost and difficulty in multiple reuses, limiting their application to research. More commonly the pressure transducer is outside of the body, and the pressure waveform is transmitted from the intravascular catheter to the transducer through a column of fluid.

Table 11.1 Tips for accurate pressure evaluation

Step 1
Check setup (transducer leveling, zeroing, and calibration)
Select appropriate fluid-filled catheter (short, large bore, stiff, with side holes, or end hole)
Assure there are tight connections between catheter(s) and transducer(s), avoid fluid leakage
Remove all air bubbles in the circuit
Step 2
Perform assessment of wave configuration with close correlation to the ECG
Check for artifacts that can distort waveform tracing (e.g., over- or under-damping)
Check an arterial blood gas to confirm normocapnia and rule out respiratory pathology
Measure at end expiration
Obtain pressure waveform data before injecting contrast

Common to all catheter-based hemodynamic acquisitions and critical for data accuracy is the transducer pressure setup, which requires special attention to check that transducers are leveled, zeroed, and calibrated. For transducers that are fixed to the table, the mounting should be at the phlebostatic axis which is located at the fourth intercostal space and 1/2 the anterior-posterior diameter of the chest, the midaxillary line, which approximates the location of the right atrium. The next step is zeroing the transducer which refers to the establishment of a reference point for subsequent pressure measurements. The stopcock to the transducer membrane is opened to air (atmospheric pressure) and electronically the signal set as zero by the hemodynamic system. Calibration of the system is performed by the physiological recorder to set the appropriate scale factor. Once this is complete, the transducer is ready to display accurate pressure measurements. If the transducer is placed above the true zero position, pressure measurements will be lower than the actual pressure and the opposite is true if placed below the true zero position. As such, it is important to pay close attention to this portion of the procedure, as small pressure changes may lead to errors in diagnosis and perhaps inappropriate therapies. Some transducer configurations are not fixed to the table and incorporated into a manifold which connects the catheter to the transducer and fluid flushes. In this case, the manifold can be placed anywhere on the table, but the transducer zeroed to the midaxillary line with a fluid-filled tube. When obtaining measurements in this configuration, the manifold must be placed at the same location as when zeroed. The pressure signal may suffer distortions as pressure wave oscillates through the fluid-filled system. In this regard, the fidelity of the pressure signal depends on the physical characteristics of the measuring system, i.e., the length of the tubing to transducer, the compliance characteristics of the catheter (tubing) wall, the size of the catheter lumen, and the viscosity of the fluid within the catheter. If the oscillations are muffled and far apart, the system is referred to as over-damped. This will underestimate systolic pressures and overestimate diastolic pressures. An example may be seen when catheters are kinked or in the case of a loose connection between within the lines. An under-damped system occurs when the

oscillations are too pronounced, the pressures are magnified, and systolic pressures are overestimated and diastolic pressures underestimated. As such these effects can be modified to some extent by changing the viscosity of the fluid, for example, mixing contrast or blood within the catheter. Air bubbles may also be inadvertently introduced anywhere from the tip of the catheter to the transducer membrane. Any amount of air results in damping, by lowering the natural frequency of the measuring system and allows high-frequency components of the signal to oscillate, producing pressure wave form overshoots (commonly seen at the systolic and diastolic inflection of a ventricular trace). Flushing the measurement system restores its natural frequency response and the fidelity of the signal.

11.3 The Pressures

11.3.1 The Right Atrium

Normal right atrial (RA) pressure is 2–8 mmHg. It is determined by atrial and ventricular compliance, atrioventricular valve function, and central venous pressure (CVP). The latter can be influenced by several dynamic factors such as cardiac output, respiratory activity, skeletal muscle contraction, sympathetic venous tone, and hydrostatic forces, components which can modify the pressure in the RA.

When the tip of the catheter is placed on the right atrium, the resulting waveform has two major positive waves (*a* and *v*) and two negative descents (*x* and *y*) (Fig. 11.1). The *a* wave is generated from the pressure rise following atrial contraction and occurs following the P wave on the ECG by approximately 80 ms and normally is the dominant wave. As such, an *a* wave is absent in patients with atrial fibrillation. Increased *a* waves can occur in restricted atrial emptying into the ventricle such as atrioventricular valve stenosis or noncompliant ventricular chambers. Following atrial contraction, pressure decline is represented by the *x* descent, due to atrial relaxation and the downward motion of

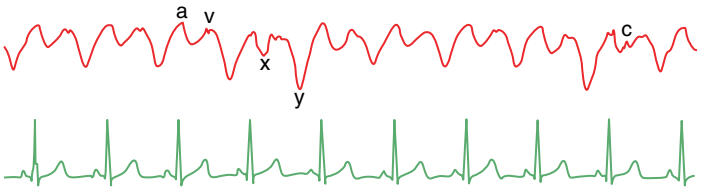

Fig. 11.1 *Upper panel*: venous pressure tracing from the right atrium denoting timing of the various wave components. The *a* wave is generated from the pressure rise following atrial contraction and occurs following the P wave on the ECG. The positive deflection following the nadir of the *x* descent is the *v* wave and is generated by passive venous filling of the atrium while the tricuspid valve is closed. *Lower panel*: the ECG

the atrioventricular junction during the early phase of ventricular systole. Occasionally, a third wave (*c* wave) may be observed as a small positive deflection during early ventricular systole when the atrioventricular valve is closed and bulges into the right atrium. If present, the *c* wave interrupts the *x* descent and follows the *a* wave by the same time as the PR interval on the ECG. Accordingly, patients with first-degree AV block may have increased *c* waves. As atrial relaxation continues, the *x* descent is now termed *x*′ and is present as pressure declines. The positive deflection following the nadir of the *x* descent is termed the *v* wave and is generated by passive venous filling of the atrium while the tricuspid valve is closed, the peak of the wave occurring at the end of ventricular systole, which corresponds to the end of the T wave in the ECG. Finally, the *y* descent reflects the fall in RA pressure when the atrioventricular valve opens and rapid emptying into the ventricle occurs. During inspiration the chest expands and pressure inside the chest becomes negative. The negative pressure is transmitted to the RA, and right atrial pressure falls during inspiration. In patients with congestive heart failure, poorly compliant pericardium or myocardium or cardiac tamponade right atrial pressure fails to fall or rise during inspiration. This abnormal finding often given the name of Kussmaul's sign and is clinically characterized as a paradoxical rise in the jugular venous pulse during inspiration and associated with an exaggerated *y* descent.

11.3.2 The Right Ventricle

Normal right ventricular systolic pressure is 20–30 mmHg and 2–8 mmHg at the end of diastole. It is important to remember that in normal physiologic circumstances, the right ventricle (RV) pumps the same stroke volume as the left ventricle (LV) but with considerable less amount of stroke work (~25%) because of the lower resistance of the pulmonary circulation. Once ventricular contraction begins, pressure increases in the RV forcing the closure of the atrioventricular valve, and in this early and extremely brief period, the pulmonary valve is closed allowing pressure to increase with no corresponding volume change (isovolumetric contraction). When the pressure exceeds that in the pulmonary artery (PA), the pulmonary valve opens with blood exiting the chamber. This is characterized as a rapid but sloped upstroke in the RV waveform (different from that of the LV) and occurs in most instances immediately after the onset of the QRS complex on the ECG. When repolarization occurs triggering the beginning of ventricular relaxation, pressure begins to fall, and when ventricular pressure falls below PA pressure, the pulmonary valve closes. Similarly, during this brief period, both the pulmonary valve and atrioventricular valve are closed, and pressure falls without change in volume (isovolumetric relaxation) until the diastolic pressure is lower than that of the RA, rendering the atrioventricular valve to open

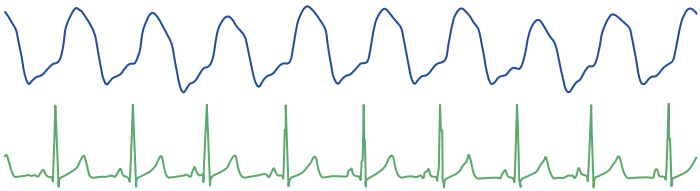

Fig. 11.2 *Upper panel*: typical appearance of the pressure wave generated by the right ventricle. Note the contour contrasted to that of the right ventricle in the presence of valvar stenosis as shown in Fig. 11.3. *Lower panel*: the ECG

(Fig. 11.2). End diastole is generally measured at the nadir of the RV pressure trace before the subsequent ejection. During early and late ventricular filling, pressure in the RV increases slowly, and when atrial contraction occurs, an *a* wave may appear on the ventricular waveform at end diastole. Under normal circumstances, the RV absorbs atrial contraction without any significant rise in pressure. In situations where the RV is noncompliant, the pressure may be transmitted through the chamber to open prematurely the pulmonary valve. An *a* wave may also be present under other pathological situations such as volume or pressure overload (i.e., pulmonary hypertension). The contour of the RV pressure trace can also indicate the form of pathology present. For example, in pulmonary valve stenosis with an intact ventricular septum, the systolic trace is triangular in shape, while in the setting of a ventricular septal defect (such as Fallot's tetralogy), the RV trace has the same contours as that of the LV, with a systolic plateau.

11.3.3 The Pulmonary Artery

Normal PA systolic pressure is 17–32 mmHg, and diastolic pressure 4–13 mmHg with mean pulmonary pressure ranging from 12 to 16 mmHg, reflecting the large cross-sectional area of the pulmonary circulation and low vascular resistance. Normally there is no systolic pressure gradient between the PA and RV unless there is valve, subvalve, or supravalve outflow obstruction (Fig. 11.3). An anacrotic notch is rarely seen in the upstroke of the pulmonary artery trace, while a dicrotic notch can be seen as pulmonary artery pressure drops, and the pulmonary valve closes and continues to fall until it reaches diastolic pressure. Elevated pulmonary pressures can be present in high-flow states with normal pulmonary vascular resistance (i.e., hypervolemia, large patent ductus arteriosus, or a ventricular septal defect) or high-resistance states (i.e., pulmonary vascular disease) or downstream obstruction (i.e., pulmonary vein or mitral stenosis).

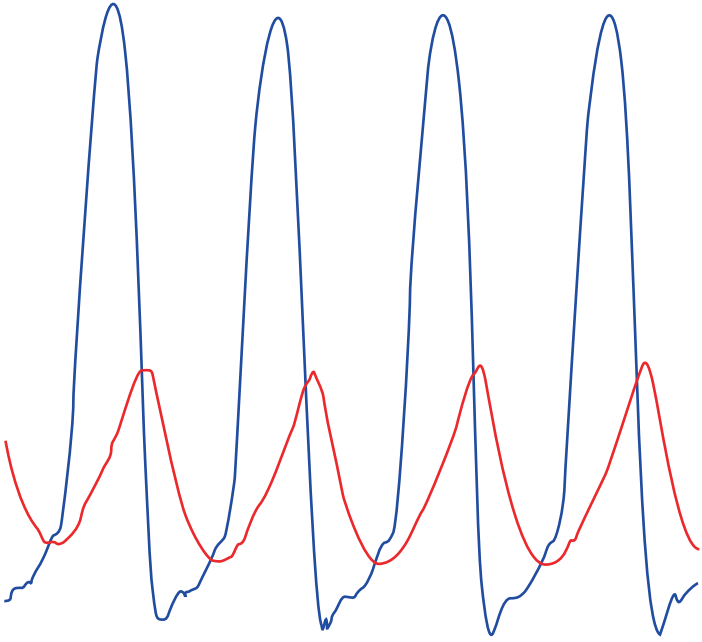

Fig. 11.3 A right ventricular trace in the presence of pulmonary valve stenosis. Note the triangular appearance contrasted to that in Fig. 11.2. Pressure transfer to the pulmonary artery is delayed and lower than that of the right ventricle due to the restricted outlet

11.3.4 Pulmonary Capillary Wedge Pressure

Pulmonary capillary wedge pressure (PCWP) is obtained during a right heart catheterization and is used as an estimate of left atrial pressure, and in the absence of pulmonary venous obstruction or mitral valve disease is a reflection of diastolic LV pressure (LVEDP). Normal mean PCWP values are between 2 and 12 mmHg. To measure the PCWP, a balloon-tipped end-hole catheter is maneuvered into a distal pulmonary artery and, with balloon inflation, occludes flow. This creates a static column of blood between the wedged balloon and the left atrium which reflects the pressure waveform of the left atrium. PCWP wave morphology resembles the left atrial wave-

form with positive *a* and *v* waves and negative *x* and *y* descents. The pressure tracing appears slightly damped and delayed (40–140 ms) relative to the left atrial waveform as the pressure wave must travel through the pulmonary capillaries. When assessing the PCWP, the operator should be careful and avoid balloon overinflation. This can occur with over-wedging which presents a false PCWP without recognizable *a* and *v* waves or can lead to pulmonary artery rupture which can be life-threatening. The PCWP measurement varies with the respiratory cycle where intrathoracic pressures vary with respiration and are transmitted to the pulmonary vasculature. Accordingly, the end-expiratory wedge pressure is often used when measuring the PCWP.

11.3.5 The Left Atrium

The left atrial (LA) pressure waveform has essentially the same shape as described for the right atrial pressure waveform, but the pressure is slightly higher with a dominant *v* wave. Normal mean pressures range between 5 and 12 mmHg. While LA pressure can typically be assessed indirectly by measuring the PCWP, direct measurement can be achieved in patients with an open communication at the atrial level (i.e., patent oval foramen or atrial septal defect) or by a transseptal atrial approach with a conventional transseptal needle or one having a radiofrequency tip. Such procedures can be guided by intracardiac echocardiography. An elevated *a* waves may be present in mitral stenosis or poor LV compliance. Great *v* waves may be seen in mitral regurgitation. The waveform will take the contour of an RV waveform in the presence of an atrial defect, with normalization of mean pressures.

11.3.6 The Left Ventricle

In adults, normal LV pressure is between 90–140 mmHg during systole and 5–12 mmHg at end diastole. In children, systolic and diastolic LV pressure varies with age. The same physiological events detailed for the RV apply to the left ventricle. However, the

waveform is morphologically different from the RV with a squared off configuration, due in part to the higher vascular resistance and time course of pressure decay (Fig. 11.4). When the mitral valve closes, there is a rapid upstroke during early isovolumetric contraction. Once pressure exceeds the pressure in the aorta, the aortic valve opens and blood is ejected into the systemic circulation, often with a prominent anacrotic notch. Ventricular pressure continues to rise during the ejection phase of systole and equals aortic pressure. As pressure in the LV begins to fall during relaxation, the aortic valve closes once pressure in the aorta exceeds LV pressure, marked by a dicrotic notch, which denotes the end of systole. As the pressure drops during isovolumetric relaxation, to below that of the left atrium, the mitral valve opens and diastole filling begins. Similar to what has been described for the RV, early and late filling phases of the LV show a slow gradual increase in pressure with atrial contraction contributing the remaining 10–25% of ventricular filling, where LVEDP is reached just before the abrupt rise in systolic pressure. End-diastolic pressure is measured coincident with the R wave of the ECG or post a wave and should be labeled as a post "a" wave measurement of LVEDP, distinct from the nadir of LV pressure at the end of isovolumetric relaxation (early filling). If an increased *a* wave (atrial contraction) is present before the LVEDP, poor compliance of the LV may be suspected. Additionally an elevated LVDEP is considered a marker of reduced diastolic performance and heart failure.

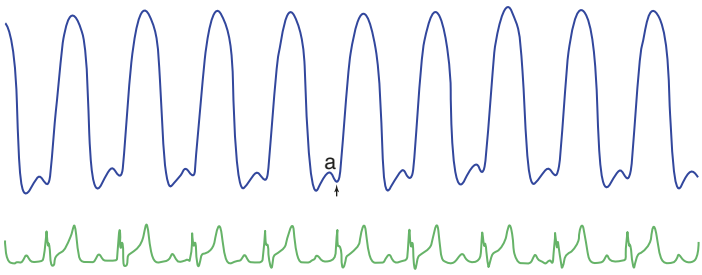

Fig. 11.4 *Upper panel*: typical left ventricular trace, note the trajectory compared to that of the right ventricle in Fig. 11.3. The *arrow* defines the post A wave end-diastolic pressure. *Lower panel*: the ECG

11.3.7 The Aorta

Normal aortic systolic pressure is 90–140 mmHg and diastolic pressure 60–90 mmHg in adults. Normal systolic and diastolic values vary in children according to age and height percentile. The waveform has a rapid upstroke and an anacrotic notch (a presystolic rise in pressure just before the aortic valve opens). Peak aortic pressure and a clear dicrotic notch due to closure of the aortic valve occur as pressure decays (Fig. 11.5). Under normal conditions, peak LV and aortic pressure are equal, and if a gradient of pressure is found between both structures, obstruction to the left ventricular outflow should be suspected (i.e., subvalvar, valvar, or supravalvular). In older patients, increased systolic pressure results from stiffness in the aorta and large arteries, whereas increased stroke volume plays a more important role in children and adolescents. A decreased diastolic pressure can result from a reduction in aortic blood volume at the onset of diastole and is generally age related in adults due to decrease in aortic elasticity. In children, a wide pulse pressure (systolic minus the diastolic pressure) may be seen in patients with patent arterial duct, aortic regurgitation, or surgical shunts (e.g., Blalock-Taussig shunt). Finally, a low or narrow pulse pressure is generally due to diminished left ventricular stroke volume and can be seen in the setting of aortic stenosis, congestive heart failure, or cardiac tamponade.

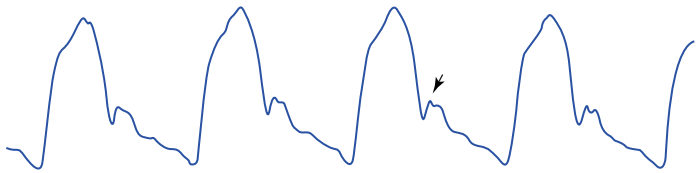

Fig. 11.5 A normal aortic pressure trace obtained from the ascending aorta. The waveform has a rapid upstroke and a clear dicrotic notch due to closure of the aortic valve as pressure decays (*arrow*)

11.4 Assessment of Cardiac Output

Measurement of cardiac output can be performed in the catheterization laboratory and offers insights to the patient's hemodynamic status. Cardiac output refers to the volume of blood pumped by the heart in 1 min, expressed in liters per minute, and often normalized for patient size by dividing by the body surface area (BSA) to obtain the so-called cardiac index (CI) measured in liters per minute per square meter ($L/min/m^2$). The normal value is 5–8 L/min in adults at rest or a cardiac index of >2.4 $L/min/m^2$. Different techniques are available for calculating cardiac output, and it is important to bear in mind the strengths and weaknesses intrinsic to each.

11.4.1 The Fick Method

In 1870, Adolph Eugen Fick developed a method to measure cardiac output. Fick's principle is fairly simple, observing that flow is proportional to the difference in concentration of some indicator (in this case, oxygen) in the blood as it enters and leaves an organ (in this case, the lungs) during steady state. The cardiac output can be determined from the difference in oxygen concentration in blood before it enters and after it leaves the lungs and from the rate at which the oxygen is consumed and is widely used for cardiac flow assessment in the catheterization laboratory. It assumes there is no intracardiac shunting and that pulmonary blood flow equals systemic blood flow. Measurement of blood arterial and venous oxygen content is obtained from sampling of blood from the pulmonary artery (low oxygen content) and from the pulmonary vein (high oxygen content). Sampling of peripheral arterial blood is often used as a surrogate for pulmonary venous blood. Determination of the oxygen consumption is more complex, but directly measured values preferred to assumed oxygen consumption from existing tables.

The Fick equation relates cardiac output to oxygen consumption and blood oxygen content and can be expressed as follows:

$$Q\left(\mathrm{l/min}\right) = \mathrm{VO_2}\left(\mathrm{ml O_2/min}\right) / \mathrm{arterial O_2 content} - \mathrm{venous O_2 content}\left(\mathrm{ml O_2/l}\right)$$

where Q is cardiac output expressed in liters per minute (l/min) and VO_2 is the oxygen consumption in ml O_2/min. The denominator of the Fick equation is the arteriovenous oxygen content difference (AV O_2) and is expressed as ml O_2/l of blood. This difference is calculated from the arterial (C_aO_2) and venous oxygen (C_vO_2) contents. The calculation of arterial and venous oxygen content (ml O_2/l) is straightforward, by calculating the oxygen capacity. A major calculation error is incorrect units, so one should take care to assure that the units (ml/l or ml/dl) are consistent throughout the equation. The oxygen capacity is the maximum amount of oxygen that sample (either arterial or venous) can bind. At normal oxygen tensions (i.e., room air), almost all oxygen in the blood is bound to the iron in the hemoglobin molecule and very little dissolved in the plasma. When breathing enriched oxygen, this fraction becomes more significant and must be taken into account; otherwise, the flow calculation will be in error (see below). As each gram of hemoglobin can carry 1.39 ml of O_2 (alternative values of 1.34 and 1.36 have also been used), the maximal amount of oxygen that can be carried (either venous or arterial) in 1 liter is

$$\mathrm{Oxygen\ capacity}\left(\mathrm{ml O_2/l}\right) = \mathrm{Hgb}\left(\mathrm{g/l}\right) \times 1.39\left(\mathrm{ml O_2/g Hgb}\right)$$

The oxygen content of the blood is the amount of oxygen in that specific sample (either arterial or venous) and can be estimated by the following formula:

$$\mathrm{C_aO_2}\ \left(\mathrm{ml O_2/l}\right) = \mathrm{Oxygen\ capacity}\ \left(\mathrm{ml O_2/l}\right) \times \mathrm{arterial\ oxygen\ saturation}\left(\%\right)$$

$$C_vO_2\left(mlO_2/l\right) = \text{Oxygen capacity}\left(mlO_2/l\right) \times \text{venous oxygen saturation}\left(\%\right)$$

For example, if the Hgb was 140 g/l, venous saturation 70%, the oxygen capacity would be 140 g/l × 1.39 = 194.6 ml O_2/l. The oxygen content would be 0.70 × 194.6 = 136.22 ml O_2/l.

If the patient is breathing enriched oxygen (F_IO_2 > 30%), the amount of dissolved oxygen becomes significant and must be accounted for in the flow equation. At body temperature, there is 0.000032 ml of O_2 per 1 ml of plasma at a partial pressure of oxygen of 1 mmHg. Thus, the solubility coefficient of oxygen in plasma is 0.00003 O_2 ml/ml plasma/mmHg O_2 tension. Therefore, the amount of dissolved oxygen in 1 l of plasma is 0.032 per P_xO_2 of the sample and has to be added to the equation above as

$$C_aO_2 = \text{oxygen capacity} \times \text{arterial oxygen saturation}\left(\%\right) + 0.032 \times P_aO_2\left(\text{mmHg}\right)$$

$$C_vO_2 = \text{oxygen capacity} \times \text{venous oxygen saturation}\left(\%\right) + 0.032 \times P_vO_2\left(\text{mmHg}\right)$$

Note carefully the units of the samples; if Hgb is measured as g/dl (not g/l), then it is multiplied by 10 to convert the units (deciliters to liters).

Ideally, the blood samples should be obtained simultaneously; the arterial blood obtained from the pulmonary vein and/or left atrium, although this is not always technically feasible. In this case, either the aortic, femoral, or radial artery can be used to determine arterial oxygen saturation. Similarly, a mixed venous saturation to calculate C_vO_2 should be obtained from the pulmonary artery in the absence of an intracardiac shunt. In the ideal setting, oxygen consumption should be measured rather than assumed [1]. It requires in most instances the use of a tight fitting hood that extracts all exhaled gas and passes it through a mixing system before measuring the concentration of oxygen. The difference between inhaled (room air oxygen concentration) and

exhaled oxygen concentration, with the known rate of flow maintained by the sampling pump, allows estimation of oxygen consumption [2]. This method is uncomplicated, but requires experienced personnel familiar with the methodology. More sophisticated equipment to allow measurement in an intubated patient is also available (spectrophotometer) but requires dedicated personnel for calibration and safe operation, and not readily available in most catheterization laboratories. Most frequently, oxygen consumption is *assumed* based on the patient's age, gender, and body surface area. The table provided by LaFarge and Miettinen [3] has been widely used to estimate oxygen consumption, albeit studies have shown those estimated values do not correlate well with measured data, particularly in small children [4, 5] (see appendix). Potential sources of errors that will interfere with accurate cardiac output when using the Fick method should be noted. The method assumes that the patient is in a steady state, stable and without alternating hemodynamics. If the blood samples are contaminated by an air bubble and not immediately analyzed, this may introduce error in the measurement. Finally when VO_2 is assumed, that estimation was obtained from healthy individuals, and extrapolating these values to pathologic conditions is uncertain, but it is widely used, and *probably* adequate for most situations, and do the calculations at the upper and lower ranges.

In pediatric practice, absolute flows are of less value than indexed flows. For example, if the VO_2 was 240 ml/min, and the BSA was 2 m^2, the indexed value would be 120 ml/min/m^2. As such, the VO_2 in infant <3 months is ~130 ml/min/m^2, 2–5 years ~150–200 ml/min/m^2, adolescents ~120–180 ml/min/m^2, adult females ~100 ml/min/m^2, and adult males ~110–120 ml/min/m^2.

11.4.2 Indicator-Dilution Method

Dye dilution measurement of cardiac output (which is seldom used today) involves the injection of a known amount of an indicator (an inert, non-metabolized, slow excreted dye) into the

circulation, which is then diluted in the blood. The blood is then sampled at a distant location from the injection site and the concentration of the indicator measured continuously, using a cuvette, during its first pass through the circulation, producing a dye concentration time curve. Flow is calculated as the amount of dye injected divided by the mean concentration of dye and the time over which it was sampled. The Stewart-Hamilton formula describes this relationship:

$$Q = \left(I \times 60\right) / \left(C_{\mathrm{m}} \times t\right)$$

where Q is the cardiac output; I the amount of dye injected (mg), 60 s/min (as a conversion from seconds top minutes); C_{m} the mean indicator concentration (mg/l); and t the total curve duration (sec). The method is accurate but requires complex equipment to perform, and other simpler methods are presently available for the clinical setting. Additionally, the calculation of flow from the time-concentration curve is only accurate in the absence of shunting, where the curve will become contaminated by early recirculation of dye. This can, however, be used to calculate shunt flow ratios from the contour of the curve, but requires several assumptions, the details of which are beyond the scope of this section.

11.4.3 Thermodilution Method

Thermodilution is a widely used method to measure cardiac output and is relatively simple to perform. It is a variation of the indicator-dilution technique using blood temperature as the indicator. A special balloon-tipped floatation catheter is placed in the pulmonary artery which has a thermistor mounted on its distal end and a proximal port opening into the right atrium. A small amount (5–10 cc) of a saline or dextrose solution, usually room temperature, is injected rapidly into the right atrium. A chamber (in this case the RV) is required to be present between the injection site and sampling site to allow complete mixing of the injectate. The

solution mixes with blood in the circulation, and the change in blood temperature is recorded by the thermistor. This change in temperature over time is inversely proportional to the blood flow. The procedure should be repeated two or three times and averaged values reported. There should be less than a 10% variance between samples. As in the dye method, thermodilution is inaccurate in the presence of intracardiac shunts, and results may be unreliable in low cardiac output states, severe tricuspid regurgitation, rhythm disturbances, and significant respiratory variations.

11.4.4 Angiographic Method

This method, once helpful in cardiac output assessment, is seldom used to in contemporary catheterization today. It estimates cardiac output from the stroke volume (SV) obtained from a left ventriculogram. The stroke volume is measured as the end-diastolic volume minus the end-systolic volume, and the output is calculated by multiplying by the heart rate (HR): $Q = \mathrm{SV} \times \mathrm{HR}$. The main limitation of this method is the requirement to correct for magnification, the projection used, and some assumptions regarding anatomy to apply the appropriate offset equations. In patients with rhythm disturbances and/or valvular insufficiency, and the complex anatomy common in congenital lesions, the method can be very inaccurate.

11.5 Assessment of Flows and the Q_p:Q_s Ratio

Flow calculations are based on Fick's principle and can be applied to both pulmonary (Q_p) and systemic blood flows (Q_s). Q_p can be estimated by the following equation:

$$Q_p = \mathrm{VO}_2 \Big/ \begin{pmatrix} \text{pulmonary venous } \mathrm{O}_2 \text{ content} \\ -\text{pulmonary arterial } \mathrm{O}_2 \text{ content} \end{pmatrix}$$

Similarly Q_s is estimated as

$$Q_s = \mathrm{VO_2} / \begin{pmatrix} \text{systemic arterial } \mathrm{O_2} \text{ content} \\ -\text{mixed venous } \mathrm{O_2} \text{ content} \end{pmatrix}$$

Finally, effective pulmonary blood flow (Q_{ep}) is the amount of deoxygenated blood that is pumped to the lungs.

$$Q_p = \mathrm{VO_2} / \begin{pmatrix} \text{pulmonary venous } \mathrm{O_2} \text{ content} \\ -\text{mixed venous } \mathrm{O_2} \text{ content} \end{pmatrix}$$

In a biventricular heart with no shunting, it is equivalent to Q_p. However, in complex cyanotic disease, oxygenated blood may be pumped to the lungs and is *ineffective* pulmonary blood flow. As such, the total pulmonary blood flow will be greater than normal and explains how a child can have an increased total blood flow and be cyanotic due to low effective pulmonary blood flows.

A fair assumption is to consider pulmonary venous O_2 content (PVO_2) as 95 or 98% if obtaining a sample from the pulmonary vein or the left atrium is not possible. A mixed venous oxygen saturation is needed to calculate C_vO_2 and best obtained from the most distal right heart chamber or site where there is no left-to-right shunt. A common practice is to obtain mixed venous saturation in the superior vena cava (SVC) and the inferior vena cava (IVC) applying the following formula:

$$\begin{aligned} \text{Mixed venous saturation} &= \left(3 \times \text{SVC sat} + \text{IVC sat}\right) / 4 \text{ or} \\ &= \text{SVC sat} - \left(\text{SVC sat} - \text{IVC sat}\right) / 4 \end{aligned}$$

The fact that mixed venous saturation more closely approximates the SVC has encouraged some clinicians to disregard contribution from the IVC completely as it is prone to sampling errors (i.e., the renal venous blood has a higher oxygen saturation than does hepatic venous blood). The same applies when samples are drawn from the RA where saturation could be low if collected near the coronary sinus. In the absence of a shunt, pulmonary and systemic flows are equal ($Q_p = Q_s$); however, the concept of a

single cardiac output calculation is invalid when shunting is present [6]. Calculation of the pulmonary to systemic flow ratio (Q_p:Q_s) can estimate the magnitude of shunts using the following equation:

$$Q_p : Q_s = \left(\text{Ao sat} - \text{MV sat}\right) / \left(\text{PV sat} - \text{PA sat}\right)$$

where Ao is the aortic saturation, MV the mixed venous saturation, and PV and PA saturations of the pulmonary vein and artery, respectively.

A Q_p:Q_s between 1 and < 1.5 is considered a small left-to-right shunt and of relatively small clinical consequence. A Q_p:Q_s > 1.8:1 indicates a large left-to-right shunt, while a Q_p:Q_s < 1 indicates a net right-to-left shunt. The same limitations of the Fick method apply to Q_p:Q_s calculations. The child must be in a steady state, the samples must be representative of the chamber of vessel, and the sample cannot be contaminated by the distal chamber (i.e., atrioventricular valve regurgitation). Small shunts are poorly detected, and in high-flow situations, the mixed venous sample may be high reducing the detection sensitivity. When taking the sample, do not let it equilibrate with room air; remember, oximeters are not accurate for saturation measurements if the Hgb is >200 g/l, where you will have to then do a blood gas. Sampling sites for the SVC are generally above the azygous vein, in the mid-lateral wall for an RA sample, and above the diaphragm for the IVC away from renal vein flow, which can contaminate the sample.

In addition to the above flow calculations, assessment of the efficiency of the heart to deliver oxygen to the tissues can be determined. Global oxygen delivery (DO_2), also known as systemic oxygen transport (SOT), is the amount of oxygen delivered to the whole body from the lungs. It is the product of total blood flow or cardiac output (Q_s) and the oxygen content of arterial blood (C_aO_2) and is expressed in ml/min: $DO_2 = Q_s \times C_aO_2$. The oxygen extraction ratio (O_2ER) is the ratio of VO_2 to DO_2 and represents the fraction of oxygen delivered to the microcirculation that is taken up by the tissues, $O_2ER = VO_2/DO_2$. The normal

O_2ER is 0.2–0.3, indicating that only 20–30% of the delivered oxygen is utilized. This spare capacity enables the body to cope with a fall in DO_2 without early compromise in aerobic respiration.

11.6 Resistance

Resistance in the vascular circuit is the difference in pressure between the two ends of the circuit divided by the flow. In the body, circulation is influenced by the resistance imposed to the heart by the vascular bed. For the right heart, the pulmonary vascular bed will determine pulmonary vascular resistance and can be calculated by the following equation:

$$\mathrm{PVR} = \left(\mathrm{mPAP} - \mathrm{mLAP}\right) / \mathrm{Qp}$$

where PVR is pulmonary vascular resistance, mPAP is mean pulmonary artery pressure, mLA_p is mean left atrial pressure (alternatively, pulmonary vein or PCWP may be used), and Q_p is pulmonary blood flow. Similarly, systemic vascular resistance can be calculated as follows:

$$\mathrm{SVR} = \left(\mathrm{mAoP} - \mathrm{mRAP}\right) / \mathrm{Qs}$$

where SVR is systemic vascular resistance, mAo_p is mean arterial pressure, mRA_p is mean right atrial pressure, and Q_s is systemic blood flow. Resistance units are commonly expressed as mmHg/l/min referred as Wood units, the term most commonly used by pediatric cardiologists. Resistance units can also be expressed as dyne·sec·cm^{-5}. To convert Wood units to dyne.sec.cm, multiply by 80. When Q_p and Q_s are indexed for body surface area, resistance should also be indexed and expressed as Wood units · m^2. Note should be taken that to correct for body BSA, the PVR is multiplied by the BSA. For example, if the BSA is 0.5 m^2, Q_p = 2 l/min, mPAP = 20 mmHg, mLAP = 8 mmHg, the PVR = (20–8)/2 = 6 Wood units, and PVRI = ((20–8)/2) × 0.5 = 3 Wood units · m^2.

11.7 Pulmonary Vascular Reactivity Testing

The assessment of pulmonary vascular reactivity plays an important role in the management and assessment of pulmonary hypertension. Following baseline hemodynamic assessment, the patient is exposed to 100% oxygen (for a minimum of 10 min) and repeat saturations and pressure measurements. When enriched oxygen is administered (F_IO_2 > 30%), the dissolved oxygen must be accounted for in the calculation. Failure to do so will underestimate of PVR. Situations that can increase PVR include hypoxia, hypercapnia, erythrocytosis, increased sympathetic tone pulmonary emboli, precapillary pulmonary edema, lung compression (pleural effusion), mechanical ventilation, and positive intrathoracic pressure. Common errors in PVR assessment include hypoventilation and acidosis producing pulmonary vasoconstriction, failure to calculate dissolved O_2, and underestimate AV O_2 difference which overestimates pulmonary blood flow and underestimates PVR, and in the setting of a septal defect assuming that no fall in PAP means no fall in PVR.

For example, a child with a VSD, Hgb = 100 g/l and VO_2 = 150 ml/min/m^2. *In room air*, Ao sat = 95%, PA sat = 80%, mLA_p = 6 mmHg, and mPAP = 60 mmHg.

The Q_p:Q_s will be 95–72.5/95–80 = 1.5:1.

The PVRI in room air will be calculated first by calculating the oxygen capacity as

$$\text{Oxygen capacity} = \text{Hgb}\ (\text{g/l}) \times 1.39 \times 100 = 139\ \text{ml}O_2\ /\text{l}$$

Oxygen content for the PA and PV can then be calculated as

$$\text{PA} = 139 \times 80\% = 111.2\ \text{ml}\ O_2\ /\text{l}$$

$$\text{PV} = 139 \times 95\% = 132.05\ \text{ml}\ O_2\ /\text{l}$$

And the pulmonary AV O_2 difference:

$$\text{PV} - \text{PA} = 132.05 - 111.20 = 20.85\ \text{ml}O_2\ /\text{l}$$

The pulmonary blood flow would be VO_2/pulmonary AV O_2 difference = 150 ml/min/m^2/20.85 ml O_2/l = 7.19 l/min/m^2. The PVRI (because the VO_2 was already indexed) would be 60–8/7.19 = 7.23 Wood units · m^2.

Now the child is given 100% oxygen to breath, and the values measured are mPAP = 60 mmHg, mLA_p = 8 mmHg, PA sat is 95%, and P*a*O_2 95 mmHg, and Ao sat 100% with a P*a*O_2 of 600 mmHg. Now if you do not take into account the dissolved oxygen, the A − VO_2 difference would be 6.95 ml O_2/l, pulmonary blood flow 21.53 l/min/m^2, and PVRI 2.4 Wood units · m^2. When the dissolved oxygen is taken into account (as it should be), the A − VO_2 difference is 22.1 ml O_2/l, and the pulmonary blood flow is 6.78 l/min/m^2. The PVRI then is 7.66 Wood units · m^2, showing no vasoreactivity.

In addition to an oxygen challenge, studies can include administration of nitric oxide (usually 40 ppm) and oral sildenafil. In those circumstances, a washout period between drugs should be allowed and measurements at return to baseline before the next drug challenge.

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12 Congenital Heart Disease: An Integrated Care Approach

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12.1 Introduction

There are important indications that children with congenital heart disease could encounter additional challenges when it comes to behavioral, emotional, and neuropsychological spheres, in comparison to their healthy peers [1–3] and that these challenges are also present during adulthood [4].

In this chapter, the psychological functioning of patients with congenital heart disease from childhood to adulthood and their families will be explored with specific reference to patients'

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empowerment. Practical advice will be provided when it comes to communicating with these patients and their families.

12.2 The Importance of Communication in the Relationship Between the Cardiologist and the Patients in the Prenatal and Neonatal Phase

Numerous studies have shown that most patients, and in the case of children their parents, want to be informed about their health conditions (type and diagnosis of the disease) and life expectancy (prognosis, treatments and their side effects) [5].

The diagnosis communication in the prenatal phase represents a fundamental moment and has been shown to have a positive impact on postnatal outcome with respect to morbidity, mortality, or quality of outcome [6].

In this phase of life, the communication of a prenatal diagnosis of CHD is of paramount importance. Often this diagnosis could lead parents to make the decision to terminate pregnancy. In this phase the opportunity to provide information to families before the baby's birth is invaluable, it is then relevant to establish a collaborative communication between the health care team and the family in order to offer anticipatory guidance, education, information on financial resources, and facilitate links to other parents [7].

The factors influencing the decision to terminate pregnancy have been identified both from cardiologists and parents. From the cardiologists' perspective, these factors are quality of life of the child, potential for neurodevelopmental delay, survival into adulthood, and severity of the CHD. For the parents the most important factors are quality of life of the child, moral/religious beliefs, and survival into birth and adulthood [8]. This shows that there are also different factors influencing the decision process in parents and physicians when deciding to terminate a pregnancy [9].

When discussing about this issue, the use of illustrations as a complement to oral information, is recommended, as it increases comprehension and satisfaction with obtained information. In

the current technologically developed environment, the overwhelming amount of information on the internet calls for compilation of easily accessible and reliable information sources via the internet [10].

Studies show that parents often experience anxiety. For this reason, it is advisable to provide families with supporting teaching materials about postoperative care. Health care professionals are able to provide a lot of information to empower those who are living on a daily basis with children with a heart defect such as other parents who underwent the same problem. In fact, very often the family is paired with one or two families with children sharing the same perspective [7].

Parents faced with their child's CHD diagnosis often have little time to make decisions. It is critical, therefore, that parents receive and understand information upon which to make informed choices regarding treatment options, where to have treatment, and whether or not to continue a pregnancy in the case of a prenatal diagnosis [11].

12.3 Congenital Heart Disease: Impact on Quality of Life and Psychosocial Aspects for Children and Adolescents

Despite the advances in cardiac treatment and an early correct diagnosis that could increase the survival of children with congenital heart disease, this condition influences the quality of life of children, adolescents and their parents. Being aware of the possible impact on quality of life of these patients and their families could help healthcare professionals, nurses in particular, providing suited care to the needs of these families, establishing priorities in their interventions, sensing predictors of a poor quality of life, promoting adherence to treatment and boosting compliance with treatment, and fostering greater satisfaction for these children, adolescents, and their parents [12].

In a study on this population, the Pediatric Quality of Life Inventory revealed that the perception of overall quality of life in adolescents with CHD was pretty low, especially when related to

physical, psychosocial, and school functioning, with the greatest difference in the domain of school functioning. Psychosocial factors included feeling angry and being worried about the future, and these were unrelated to the severity of the cardiac defect [13].

Structured psychosocial screening and mental health assessment using validated tools is recommended as part of standard care to facilitate early detection of risk and resilience factors and proactively engage with psychosocial services [14].

There is a growing interest in characterizing the neurodevelopmental outcomes of school-age survivors of cardiac surgery. In this cohort of children with complex congenital heart disease, a significant proportion of the children were at risk for inattention and hyperactivity, and nearly half were using remedial school services [15].

Children with coronary heart disease have greater difficulties when it comes to behavioral, emotional and neuropsychological aspects, compared to their healthy peers [1–3]. In some cases, this can generate parental overprotection, although this behavior has not been reported to be indicative of everyone or even most parents [16–18].

Even if the intelligence (I.Q.) is not severely impaired, these patients are at increased risk of speech and language impairments (including pragmatic skills), visual–spatial skills, executive functions, attention, and motor skills. Most of these patients achieve good psychosocial outcomes, although, according to parents and teachers' ratings, a substantial percentage of them is exposed to an increased risk [1].

Several factors play a role in adjustment to congenital heart disease, either improving the perception of quality of life or worsening it. Some buffer variables on congenital heart disease, as for example social support, play a role in increasing the perception of quality of life of patients during their lifetime. Social support has an impact on increasing resilience, promoting adjustment to illness [19].

Parents of children with chronic medical needs report increased parenting challenges, poor sleep, and maladjustment. The impact of parenting stress on both sleep and adjustment has yet to be evaluated for parents of infants and young children with congenital heart disease [20].

12.4 Psychological Functioning in Adults with Congenital Heart Disease

Behavioral cardiology and psycho-cardiology has come a long way in identifying the bio-behavioral pathways leading from chronic psychological stressors and perceived psychological distress, respectively, to hard cardiovascular disease end points [21] and we are now exploring also these mechanisms in congenital heart disease.

The continuous improvement in survival rates of patients with congenital heart disease (CHD) has led to a growing number of adults with CHD, especially those with complex heart defects [22]. These patients have been reported to be at an increased risk of psychological distress, neurocognitive deficits and social challenges [4].

In qualitative studies conducted on adults, it has been reported that having this cardiac condition can lead to feeling different, followed by attempts and difficulties to feel normal and to be perceived as normal by others [23–26].

Adults with congenital heart disease have made lifelong psychosocial adaptations on the basis of their congenital defects, with respect to physical as well as mental aspects and consider their situation as normal [27]. Patients over 30 years of age show a more negative perception of the disease compared to the other age groups [28].

The main psychological concerns reported by patients with ACHD in a recent study were generalized anxiety, health/heart related anxiety, depressed mood and difficulty in coping with a general medical condition [29].

When it comes to peers, adults with congenital heart disease very often experience difficulty in relationship and employment. This mainly occurs in adolescent and young adulthood due to a delay in the maturation progress and the inability to cope with different tasks at the same time [30].

Often there is a poor and incomplete understanding, of the patients and their families by the health care professionals, about the difficulties that they are experiencing due to the cardiac disease [31].

Most of the studies available reported that the psychological functioning in this population is more related to medical variables such as diagnosis and physical status. The major variables indicated as being predictors of psychological distress are [32]:

- Loneliness.
- Fear of negative evaluation.
- Imposed limits.
- Low capacity for physical exercise.
- Perceived health status.

For this reason, being part of an associations of reason can be very useful, since it offers an opportunity for patients and families to have a point of reference and also to participate in aggregative activities [33].

12.5 How to Help Families and Patients: Clinical Psychology Service, Patient Associations and Peer-to-Peer Programs

It has been reported by the patients with CHD and their families that there is a need for psychosocial care. These families and patients have to deal with many issues which might have an impact on how developmental tasks are dealt with.

There is a need for screening children with CHD about developmental, neuropsychological, and emotional problems. This was also has been recommended by the American Heart Association [3]. Preventive assessment enables the possibility of avoiding psychological problems later on in life.

Patients with congenital heart disease often cause feelings of alienation, isolation, marginalization, dehumanization and disempowerment. All this can have an impact on the body image, self-esteem and can lead to other psychological problems. Families and patients know the disease very well based on their experience and the desire to monitor their health, hence generating a good sense of empowerment [34].

Based on this, it is advisable multidisciplinary approach in the medical contexts, such as pediatric cardiology and adult congenital cardiology units, so as to involve the family. There is an emerging need of including psychologists, social workers and clinical nurse specialists to care for patients with congenital heart disease (CHD) and their families, who have specific training on these issues by the health care specialists [35].

According to the World Health Organization (WHO), peer-to-peer support represents an occasion to bolster patient resources, promote empowerment and to decrease the sense of isolation. Mutual aid is seen as a strategy to promote health in terms of strengthening social relations [36].

When it comes to peer-to-peer support, specifically for CHD, the American Heart Association has developed a very well structured program aimed at providing peer support, supervision and indications on when a referral to a psychologist is necessary [37]. Peer-to-peer support is very much indicated when it comes to loneliness and social problems issues [38].

In a recent perspective article, an integrated model for psychological care was presented and it shows how psychological care can make a difference in congenital heart health [39]. Clinical psychology service and support is beneficial for the patients with a high level of psychological stress, while peer-to-peer support is more appropriate when it comes to loneliness and social nature problems.

All CHD patients who are hospitalized prior to an intervention (both for cardiac surgery and for cardiac catheterization) it is recommended to perform a psychosocial evaluation through a structured psychological and psychometric assessment. This includes acquiring data about patients' family and working situation, previous psychological and psychiatric visits, lifestyle, and psychological functioning (anxiety and depression). The presence of the psychologist at the multidisciplinary medical meetings is also recommended [38].

Both clinical psychological services and peer-to-peer support have proven to be very effective in the case of dealing with a chronic disease condition, such as in the case of patients with ACHD. The use of an integrated approach has the advantage to

provide peer-to-peer support and social assistance. At the same time it allows to refer to a mental health professional in case of increased psychological distress [38].

12.6 Conclusions

In conclusion, it is important that cardiologists and health care professionals invest time and resources in understanding patients and parents' level of information needed in order to be able to understand their psychological needs and provide an adequate support.

Providing the desired level of information is key to the patients and parents' empowerment in order to fully involve them in the care process. An integrated approach is desirable to cope with the disease in a multidisciplinary way in order to offer to the patients and their families a humanized multilevel care.

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Part II

Vascular Access

13 The Usual Vascular Access

Daniel Tanase and Jochen Weil

The most frequent access for routine cardiac catheterisation is through the femoral artery and vein. Almost every position within the heard can be reached over these two vessels. The only exception is the pulmonary position in children with partial bidirectional cavo-pulmonary anastomosis. Therefore, access through one of the major veins of the upper part of body is needed, either the jugular or the subclavian veins.

The routine technique to introduce a sheath is the Seldinger technique, named after a Swedish radiologist Dr. Sven-Ivar Seldinger (1921–1998).

Accurate vessel puncture requires either orientation on the surface anatomy or direct visualization of the target vessel by ultrasound. Routine use of real-time ultrasound guidance reduces the number of attempts, time to access, and risk of vascular complications [1].

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Several transducers can be used for the ultrasound-guided puncture. The most appropriate is the one with the highest possible frequency and a small contact face (a hockey stick or a 12 MHz for newborns and small children, and a 6 MHz or linear transducer for adults).

Local anaesthesia effectively diminishes pain and reduces complications related to patient movement. Gradually injection of small volumes is preferable than an excess of volume that cause more pain and distorts the anatomy.

To minimize vascular access complications consider using a micropuncture kit (i.e. Cook Medical, Bloomington, IN) that allows access with a small 21-gauge needle, especially in small patients.

In general, forcing the guidewire almost never results in success [2].

13.1 Femoral Venous and Arterial Access

13.1.1 Positioning and Anatomic Landmark Guidance

Especially in small children a rolled towel under the buttock may facilitate access of the femoral vessels. The legs should be fixed in a slightly outward rotated position.

The landmark to be sought is the inguinal ligament, which runs between the superior iliac spine and the pubic tubercle (Fig. 13.1). This is not to be confused with the inguinal skin crease. The femoral artery crosses the inguinal ligament at approximately its midpoint and should be palpated there. The femoral vein runs closely medial.

Usually the vein is punctured first. As vessel diameter increases with age the point for cutaneous puncture for the vein is located more medial in the adult compared to the newborn.

The needle should be introduced at the level of the inguinal skin crease, or just below.

The angle between needle and skin also varies with age. It can be flat in the newborn (10°–20°) and ascends steeply in the adult

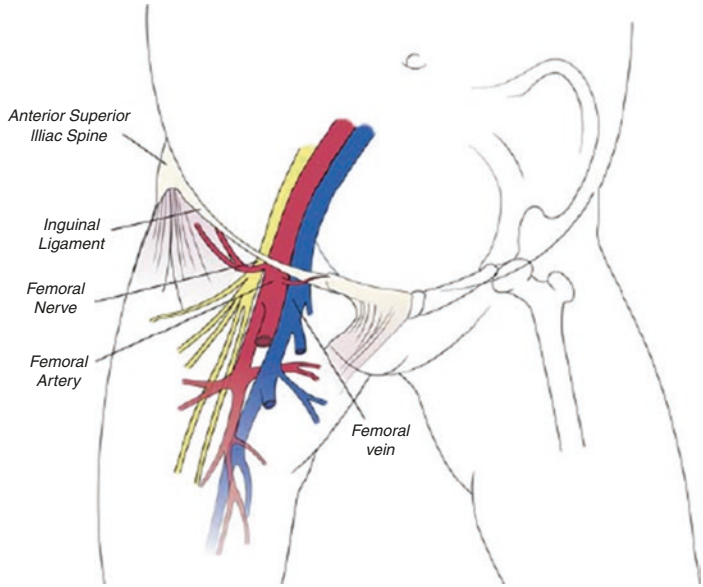

Fig. 13.1 Landmarks of femoral artery and vein access. (From Bergersen [3], with permission)

(45°–60°). In general, the more subcutaneous tissue is present the steeper the angle should be.

For femoral artery puncture, the needle should be directed towards the umbilicus. The direction of the vessel can also be checked by simultaneous palpation of the pulse with two or three fingers. The vein runs parallel to the artery.

For venous puncture it is helpful to advance the needle with a syringe attached under continuous suction. Once blood can be sucked easily, the syringe should be removed, while the needle is stabilized. Some operators prefer to puncture the artery without a syringe attached. Free pulsatile flow might be easily identifiable.

The guidewire should be introduced gently and should pass with minimal resistance. Once the guidewire is positioned, the needle should be removed and the sheath-dilator assembly advanced with a rotatory motion.

If the guidewire cannot be advanced gently but a backflow of blood still visible it is probable that the vessel was not hit appropriately. Than a slightly change of the direction or the angle of the needle might facilitate the advancement of the guidewire. If this is still not possible needle and guidewire should be removed and a new attempt performed.

13.1.2 Ultrasound-Guided Puncture

Real-time ultrasound guidance can be performed either in a long-axis or a short-axis approach. Especially in small children visualization of the vessels in the long axis might be advantageous because needle and vessel can be visualized at the same time. In adults, when vessel access should be performed steeper, the short-axis approach is an alternative.

For long-axis approach, the entire course of vessels should be visualized (Fig. 13.2). This gives information about the direction and the depth how far the needle should be advanced to penetrate the vessel wall.

The vein is compressible and located medial to the artery. But colour flow or Doppler imaging can distinguish the vessels, too.

Maximizing the transducer frequency, the sector field focus and adjusting the gain setting, able to detect low velocity flow, should optimize the image quality.

The transducer and the cord should be inserted into a sterile sheath (Fig. 13.3).

Once the entire course of the target vessel is visualized position of the transducer should not be changed.

The needle should be introduced closely to the contact face of the transducer and advanced in the same direction until it penetrates the wall of the vessel.

If the vessel and needle cannot be visualized at the same time, it is mostly because the tip of the needle is outside the ultrasound beam of the array. Changing the position of the transducer to pick

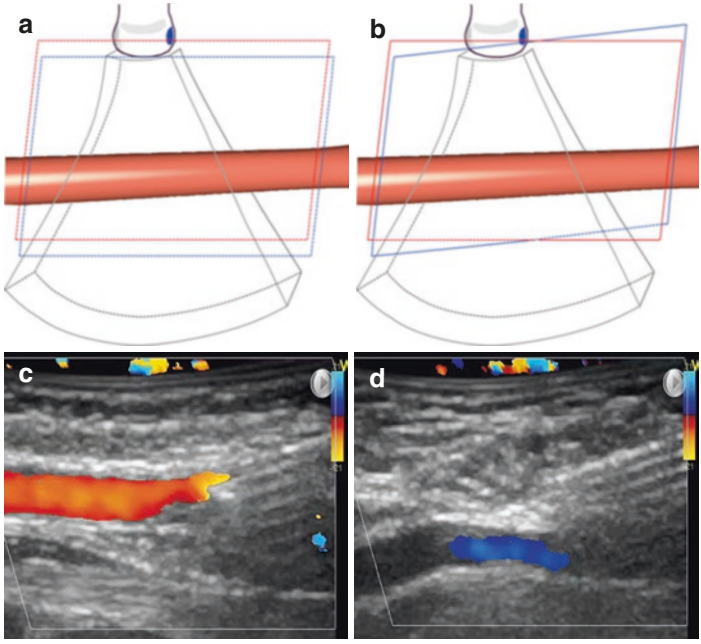

Fig. 13.2 Ultrasound-guided puncture. The transducer should be aligned to the course of the vessel—red plane in **a** and **b**. If the needle is advanced in the same plane—blue plane in **a**–the tip and distal parts of the needle become visible (**c**). Notice aliasing in the colour flow image in **c**, as the tip of the needle penetrates the vessel wall. In **b** the plane in which the needle is introduced—blue plane—is not aligned to the red plane of the transducer and the vessel. The vessel cannot be hit appropriately. In the corresponding ultrasound image (**d**) just mid parts of the needle are visible. The tip of the needle is near the vessel—as the vessel course is distorted—but not visible

up the needle is mostly not helpful. It is better to adjust the needle to the long axis of the transducer.

Introduction of the guidewire and of the sheath-dilator assembly are performed in the same manner as for anatomic landmark guidance.

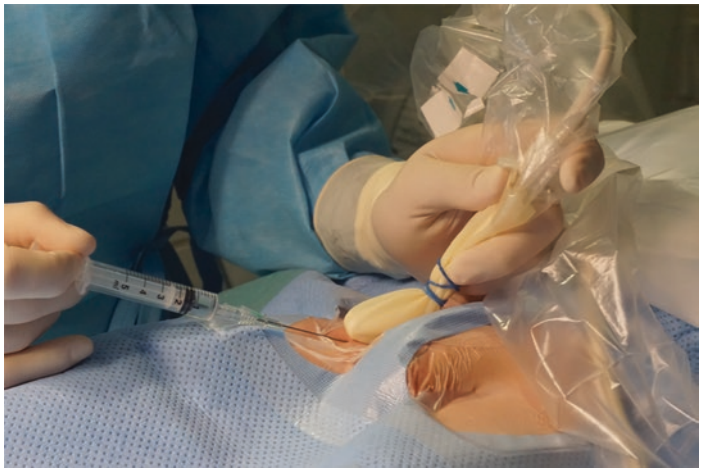

Fig. 13.3 The ultrasound transducer is placed inside a sterile sheath

13.2 Internal Jugular Vein Access

13.2.1 Positioning and Anatomic Landmark Guidance

The patient should be positioned supine with a rolled towel under the shoulders and the head turned to the contralateral side of the puncture. A slight Trendelenburg position of approximately 10°–15° would facilitate vessel access because it increases diameter of the vein. In contrary an overextension or overrotation of the head would inadvertently compress the jugular vein.

The essential surface anatomy is a triangle comprised of the sternal and clavicular heads of the sternocleidomastoid muscle medially and laterally and the medial third of the clavicle inferiorly, named the Sedillot's triangle. The internal jugular vein lies underneath this triangle and is located laterally to the carotid artery (Fig. 13.4).

The right internal jugular vein is preferred over the left one since:

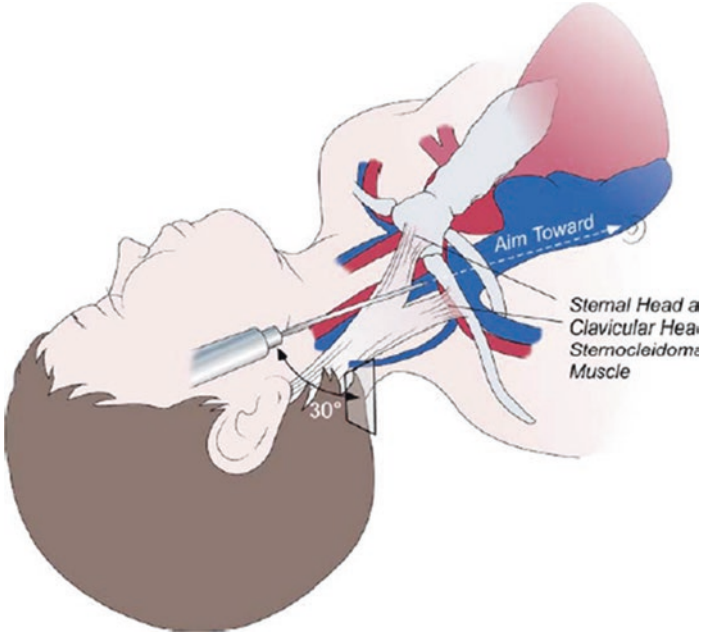

Fig. 13.4 Landmarks of the internal jugular vein access using the anterior approach. (from Bergersen [3], with permission)

- The apex of the lung is lower on the right side.
- The path to the atrium is more direct.
- There is less risk of damaging the thoracic duct.

There are three different approaches to the internal jugular vein.

For the anterior approach the needle is introduced at the medial margin of the sternocleidomastoid muscle approximately at the level of the thyroid cartilage and directed towards the ipsilateral nipple (Fig. 13.5).

For the middle route the needle enters the apex of the triangle formed by the both heads of the sternocleidomastoid muscle and the clavicle and also directed towards the ipsilateral nipple.

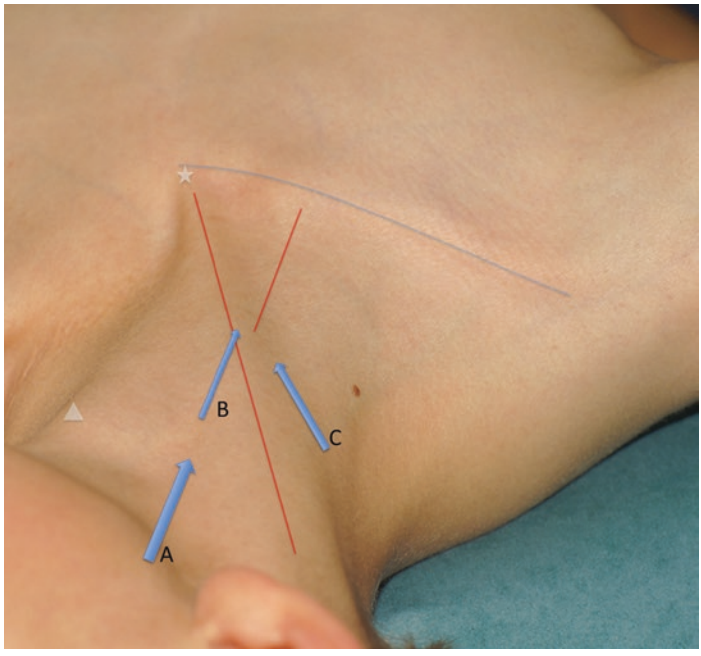

Fig. 13.5 Three approaches to the internal jugular vein. The blue line represents the course of the clavicle. The red lines show both heads of the sternocleidomastoid muscle. Star—suprasternal notch. Triangle—thyroid cartilage. (**A**) anterior approach; (**B**) middle approach; (**C**) posterior approach

In the posterior route the needle should be introduced along the lateral margin of the sternocleidomastoid muscle cephalad to the apex of the Sedillot's triangle. In this case it should be directed towards the suprasternal notch.

13.2.2 Ultrasound-Guided Puncture

For ultrasound-guided puncture of the internal jugular vein (IJV) the anterior approach is the most appropriate. Mostly the IJV lays antero-lateral to the carotid artery but anatomic variations with a complete anterior or lateral position are possible. A gentle probe

pressure will compress the vein and will help to avoid arterial puncture according to the close relationship of both vessels. But both vessels can be differentiated sufficiently either by pulsation of the artery or by colour flow Doppler.

The short axis will visualize both vessels at the same time, helping to distinguish them, while the long axis will help to rule out the direction in which the needle has to be advanced.

Puncturing technique is the same as for femoral vessel cannulation.

13.3 Subclavian Vein Access

Because of attachments of the subclavian vein to surrounding tissues the vein remains patent even in hypovolemic patients. A slight Trendelenburg position of the patient will not increase diameter of the vessel but will avoid complications of air embolism. The head should be kept neutral, as an excessive rotation to the contralateral side will increase the angle between the internal jugular and the subclavian vein therefore facilitating advance of the guidewire into the IJV than into the superior vena cava.

The subclavian vein lays between the first rib and the clavicular bone and is bordered posterior by the subclavian artery and brachial plexus (Fig. 13.6).

The anatomic landmark is the junction of the medial one third and lateral two thirds of clavicular bone. From medial this corresponds to the first slope of the bone when the anterior convexity goes on to the anterior concavity. The cutaneous puncture should be inferiorly and laterally to this point. The needle should be advanced parallel to the floor targeting the suprasternal notch.

Puncturing too close to the clavicle is disadvantageous because the needle has then to be directed posteriorly to negotiate with the clavicle. This increases the risk for pneumothorax.

The vein can be located by ultrasound, but because of the anatomic position between the clavicle and first rib real-time ultrasound localization during puncture is difficult. Thus, ultrasound localization without real-time ultrasound guidance does not offer advantages.

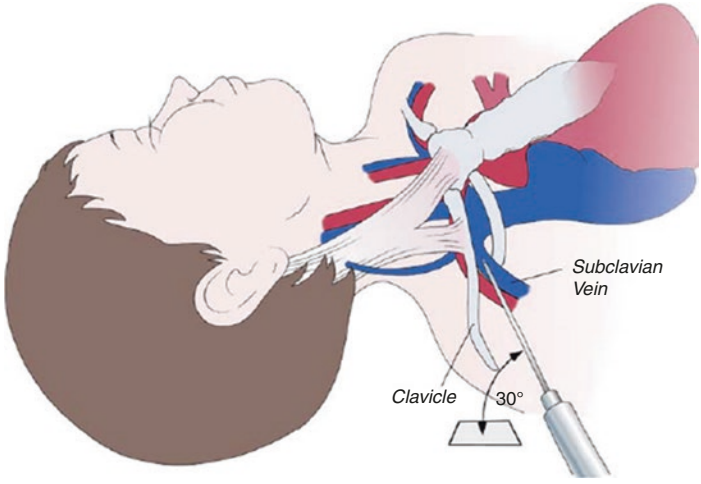

Fig. 13.6 Landmarks of the subclavian vein access. (from Bergersen [3], with permission)

13.4 Umbilical Venous Access

Catheterization of the umbilical vessels (two arteries and one vein) is well described in neonatal text books and is a standard procedure in the neonatal care.

The catheterization of the vessels is performed with a 3.5 or 5 F umbilical catheter. The 3.5 F catheter takes a 0.021″ and the 5 F catheter a 0.025″ guidewire.

If these catheters are in place, they can be cut with a scalpel short above the umbilicus and the sheath needed can be inserted over a guidewire.

13.4.1 Problems

Umbilical vein

- The guidewire and catheter are difficult to manoeuvre into the ductus venosus due to entering the portal vein.

Withdraw the catheter into the umbilical vein and give a small injection of contrast to delineate the course of the ductus venosus into the IVC.

Umbilical artery

- It might be difficult to advance the guidewire into the aorta due to the tight curves of the umbilical artery entering the iliac arty. A normal straight guide may be more suitable than a torque guidewire.

13.5 Radial Artery Access

13.5.1 Positioning and Landmarks

– The arm is abducted 90° and positioned on an arm support.
– Slight elevation of the wrist by a cotton swab and fixation of the hand.
– The landmarks are the distal ends of the radial and ulnar bone and the radial pulse.

13.5.2 Technique

Needle entry superficially with a 30° angle. No syringe attached. Advance until jerks of pulsating blood will flow.

13.6 Axillary Artery Access

In some patients access of the axillary artery provides an optimal approach for stenting of the patent duct in newborns with duct-dependent pulmonary circulation.

13.6.1 Positioning and Landmarks

- The arm is positioned over the head.
- Position a rolled towel under the shoulder—this should elevate the axillary region.

13.6.2 Technique

Palpation of the axillar artery is mostly possible. The clavicular bone indicates the further curse of the axillar artery.

An ultrasound-guided puncture is the most appropriate and helps to minimalize complications. The artery can be identified by a slightly pulsation. Advance the needle under real-time ultrasound control. Once free pulsatile flow is visible, advance the guidewire and insert the sheath using the Seldinger technique. A 4F sheath is usually well tolerated by a newborn.

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14 Unusual Access

Stephan Schubert and Felix Berger

14.1 Introduction

Vascular malformation, stenosis, or thrombosis of commonly used central arteries or veins (e.g., femoral or jugular, subclavian) gives rise to the need for alternative vascular access routes. Vascular access can be extremely difficult, especially in patients who have undergone multiple catheterization procedures at a young age or following placement of central lines in the aforementioned vessels. Preventing vessel stenosis, especially in very young patients, is a major challenge. The guidelines on cardiac catheterization recommend using the smallest possible sheath, especially for arterial access. Furthermore, the use of compression dressings after removal of the sheath and hemostasis may lead to vessel thrombosis or impairment of blood flow. Therefore, they should be avoided especially in small children. To prevent

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trauma to vessels caused by multiple puncture attempts in patients with complicated or unusual vascular access, the use of ultrasound-guided puncture may be preferential.

14.2 Ultrasound-Guided Puncture

Ultrasound-guided punctures increase safety and efficacy in patients with difficult vascular status, when access to small vessels is required, or for catheterization in preterm, hypotrophic, or newborn infants. It is also useful in patients with pulsatile or non-pulsatile assist devices, given that vascular access in patients who are fully anticoagulated or with reduced or absent pulse can be very difficult or cause serious bleeding [1]. In such cases, the use of a high-frequency ultrasound probe (10–12 Hz) with the option of color Doppler imaging is mandatory. Sterility can be ensured by using a single-use sterile probe tip. A 20G Abbocath radiopaque IV cannula (Hospira Inc., Lake Forest, USA) or 20G/50 mm length one-piece angiographic needle (Cordis, Johnson & Johnson, USA) and a short 0.018 in. wire may be used for vascular access in small children and in patients with a body weight of up to 20 kg. An example of an unusual access route (axillary artery) is illustrated in Fig. 14.1.

Ultrasound-guided puncture may also be used for uncommon vascular approaches such as transhepatic access.

14.3 Transhepatic Vascular Access

In patients with caval thrombosis or vascular malformation (absent SVC, IVC, or azygos continuation), those central vessels (caval veins or atrium) may not be accessible via conventional approaches. If the femoral or jugular access routes are obstructed or missing, a transhepatic approach may allow diagnostic or therapeutic catheterization or placement of a central venous line [2]. Transhepatic puncture is performed using the ultrasound-guided

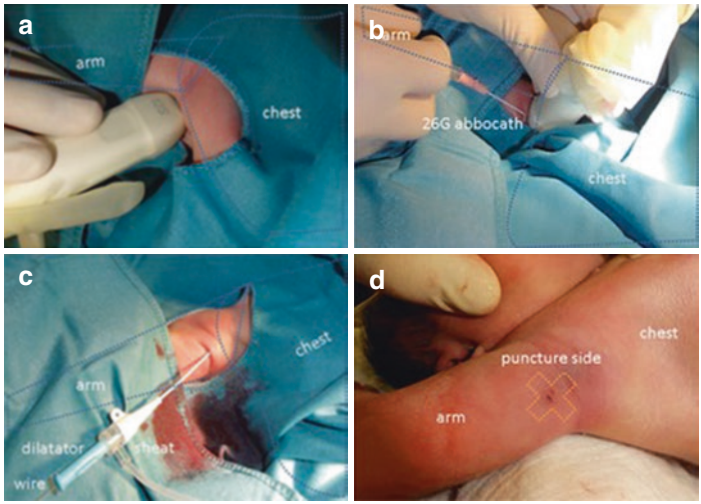

Fig. 14.1 Ultrasound-guided puncture: Right axillary arterial access with ultrasound-guided puncture for ductal stenting of a complex ductus-dependent defect in a 3-day-old infant with a BW of 2.9 kg. A 4.5–11.5 MHz ultrasound probe (GE Healthcare, Wauwatosa, USA) was used to visualize the axillary artery, and percutaneous puncture was performed with a 20G Abbocath. After vessel puncture and wire placement, a 3 French sheath (Balt, Montmorency, France) was introduced percutaneously and stent implantation (Coroflex blue) was performed

approach after sonographic visualization of the liver structure and especially liver veins [1]. We perform percutaneous puncture from the lateral or medial position after visualization of the concomitant structures and vessels. After passing the liver parenchyma, the liver veins should be reachable with the proximal part of the needle and aspiration should be attempted. If a withdrawal of blood is possible, correct position may be checked again by ultrasound and the introduction wire may be advanced into the atrium of the heart. Thereafter, the sheath or central catheter may be placed with or without pre-dilatation. The presence of hematoma can be excluded sonographically and liver enzymes checked, if a central line remains in place (Fig. 14.2).

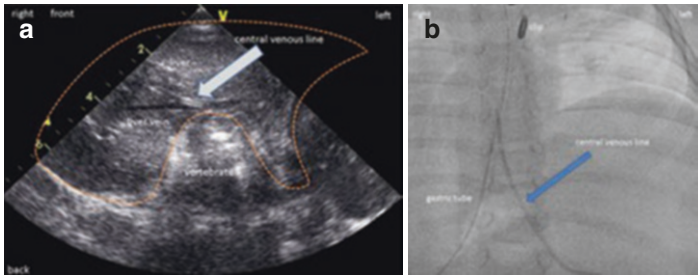

Fig. 14.2 Transhepatic central venous line placement: ultrasound-guided puncture by a left paravertebral and median percutaneous approach due to inverse abdominal situs with dextrocardia in a patient with caval thrombosis. (**a**) Sonographic picture after transhepatic placement of the central venous line. Visualization of liver veins and liver tissue is essential for percutaneous approach. (**b**) Fluoroscopic picture of central venous line with a transhepatic approach

14.4 Paravertebral Access

The vertebral–azygous–hemiazygous pathway may exhibit significantly enlarged collateral vessels in patients after corrective surgery of congenital heart disease, especially in those with modified Glenn or TCPC/Fontan operations or with obstructions or thrombosis of the superior caval vein. In these patients, the collateral pathways may connect to the pulmonary veins via the bronchial vascular system, potentially giving rise to a significant right-to-left shunt and, in some cases, cyanosis. With connection to the paravertebral veins, the vessels are often located extremely posterior, making retrograde access from the systemic or pulmonary veins difficult or impossible. In some cases, the proximal region may already have been occluded, but collateral flow still exists. MRI or computed tomographic visualization of the azygous or hemiazygous veins and their collateral vessels with CT-guided access through the paravertebral veins makes it technically possible to reach the vessel and applies a transcatheter occlusive strategy (see Fig. 14.4). Despite the direct proximity to the pleura,

access to the paravertebral veins may only be possible with a high degree of precision under real-time CT-based navigation. Thus, this approach can be recommended whenever access to the paravertebral veins is deemed mandatory.

Recently we reported a case of paravertebral access with the use of a CT scan [3]. Vascular access was performed and identified via a 4 mm paravertebral vein at the level of the 4th–5th thoracic vertebra as a small feeding vessel. Under real-time CT guidance, this vessel was punctured using a 4 Fr. Micropuncture introducer set (COOK Medical Inc., Bloomington, IN) and a 4 Fr. sheath (COOK Medical Inc.) were placed using the Seldinger technique (Figs. 14.3 and 14.4).

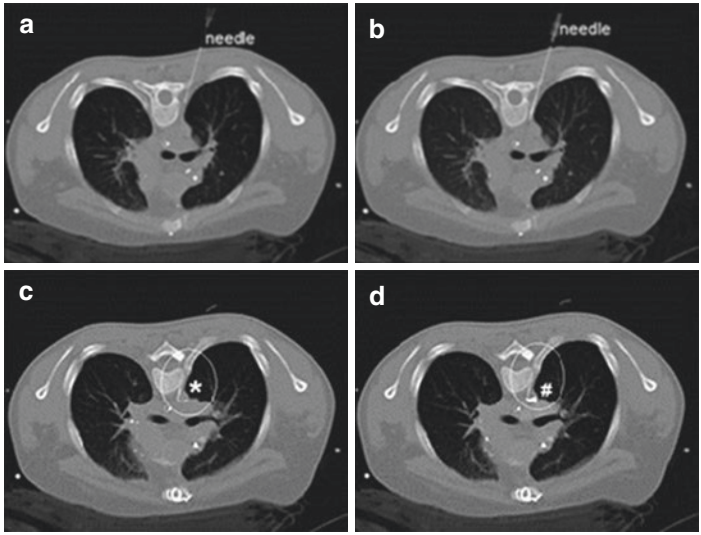

Fig. 14.3 (**a**) CT scan of the initial paravertebral approach (patient in prone position), with the needle passing through the paravertebral space. (**b**) Puncture of the paravertebral vein with a needle on the right side. (**c**) Introduction of a guide wire into the vertebral vein. (**d**) Insertion of a 4 Fr. introducer sheath over the wire into the paravertebral vein and application of contrast medium (#) to verify correct position of the sheath (From Schubert et al. [3])

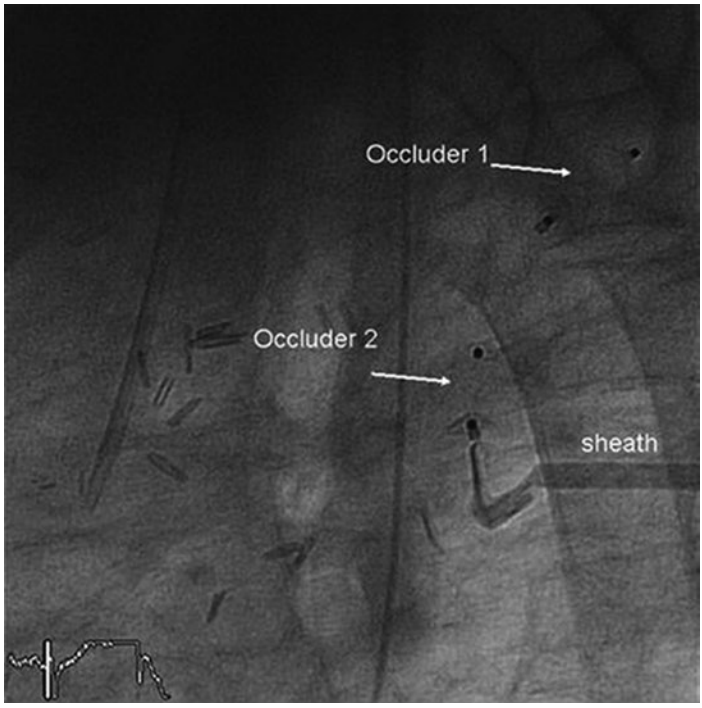

Fig. 14.4 Angiogram in lateral projection without contrast medium: two Amplatzer vascular plugs in situ. One occluder was placed in the inflow to the collateral; one was placed in the collateral itself. Access of the sheath through the vertebral vein is documented via paravertebral access (From Schubert et al. [3])

14.5 Iliac Venous Access

In patients with femoral vein stenosis or thrombosis, iliac access may offer an alternative approach. Again this may be achieved by ultrasound-guided puncture of the vessel. The transcutaneous puncture will be below the inguinal ligament, but the vessel entry of the needle will be positioned as high as possible with ultrasound-guided movement of the needle. On removal of the sheath, vascular pressure has to be applied and hemostasis achieved by adequate lower abdominal or inguinal compression. Ultrasonographic con-

trol of hemostasis is required. In patients with no suitable femoral or jugular venous access, iliac venous access may be a feasible alternative to the transhepatic access route. If femoral stenosis is apparent, treatment can and should be performed during or within sheath removal (see Fig. 14.5a–c).

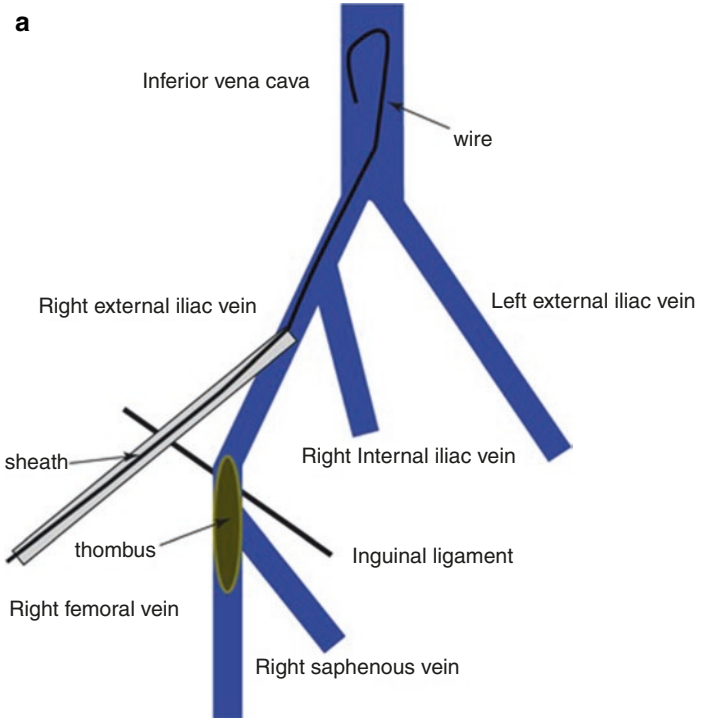

Fig. 14.5 (**a**) Iliac and femoral vessel area around the inguinal ligament. Venous puncture and sheath implantation can be performed via the right iliac vein under sonographic control. (**b**) Example of an angiographic and fluoroscopic picture in a 12-year-old girl with multiple vascular punctures and sheath/central line implantation and status post-stenting of the right iliac vein due to stenosis. (**c**) Balloon dilatation of iliac and femoral stenosis within sheath removal from the iliac vein. Complete treatment of the vessel stenosis after sheath removal was documented, saving this vessel as an access route for further catheterizations

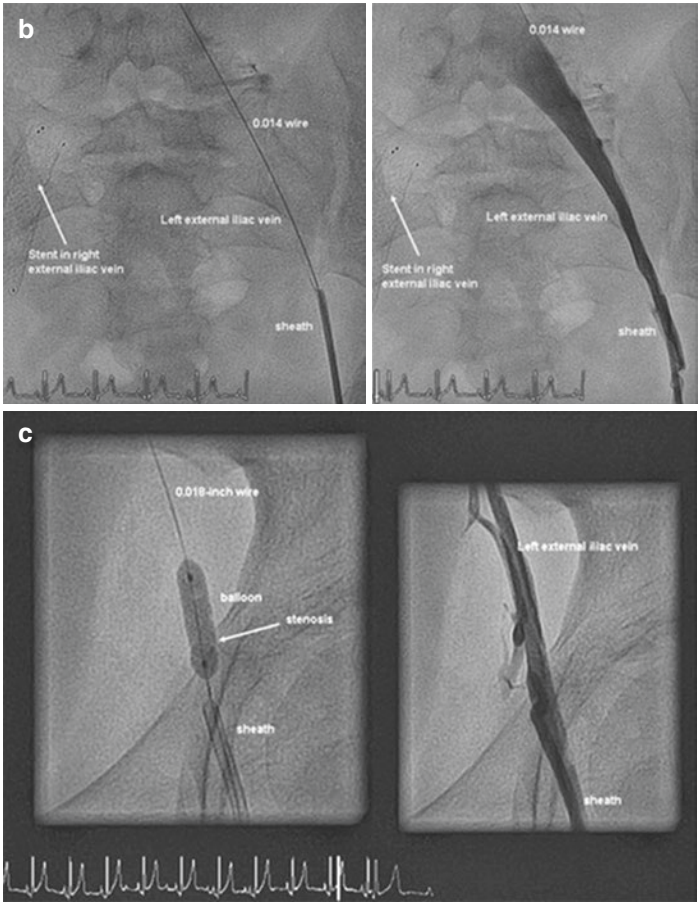

Fig. 14.5 (continued)

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Hemostasis

15

Zakhia Saliba and Ramy C. Charbel

15.1 Hemostasis

Hemostasis is an essential element in every vascular catheterization. Its main purpose is to stop both internal and external bleeding. A successful hemostasis should not compromise vascular patency once the material has been removed.

Factors that may affect hemostasis can either be patient related (blood thinners, blood disorders, history of cardiac surgery or prior catheterization…) or procedure related (several attempts to obtain vascular access, poorly anesthetized and not well immobilized patients, ratio of sheath size to vessel size, length and complexity of the procedure…).

The increasing trend in outpatient catheterization has made reduction of post procedural bleeding a priority.

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Femoral vessels are the most frequently used vessels in congenital catheterization at all ages, due to their larger caliber and their easier accessibility to the essential cardiac and vascular structures. They are usually engaged using the Seldinger technique. Vascular access technique and site dictate the type of maneuvers needed to achieve hemostasis.

There are no randomized controlled trials comparing commonly used techniques in achieving post catheterization hemostasis.

15.1.1 Manual Compression (MC)

MC remains the "gold standard" in achieving hemostasis of a femoral arteriotomy or venotomy. This is due to its effectiveness, its tolerability, its good safety profile, and its short learning curve. It is also a cheap maneuver that does not leave a foreign body in or around the vessel, thus reducing distant complications.

At the end of the procedure, the sheath is usually removed immediately.

When removing the sheaths, the operator should ensure arterial hemostasis first, and then remove the venous sheath. This approach is said to decrease the risk of arteriovenous fistula formation and it provides a useful means of treating a vagal reaction should the peripheral IV inadvertently be lost.

Before sheath withdrawal, it is essential to ensure the patency of the peripheral IV line that would be used shall the need for IV medication exist.

When pulling the sheath, the operator first prepares the area by clearing away equipment, syringes, and tubing. The sheath is then aspirated and flushed with saline to clear any thrombi.

To remove the sheath, the operator places left-hand fingers over the femoral artery (or below the vein) an inch more cranial (or caudal) than the skin incision. The skin is supported to minimize traction and vessel damage [1].

The operator applies gentle pressure using a sterile gauze and removes the sheath, taking care not to crush the sheath and "strip" clots into the artery.

It is crucial to monitor vital parameters and keep track of arterial pulsation with a pulse oximeter placed on the distal limb.

The distal limb coloration should be continuously checked during MC to ensure adequate perfusion.

If the pedal pulse is absent during compression, the pressure over the artery should be decreased periodically to allow distal circulation.

Keep in mind that arterial circulation can be compromised even after simple venous access.

Maintain adequate compression (enough to counter vascular pressure without compromising distal pulsation) for 10 min. Apply more gentle pressure for 2–5 min then apply a pressure dressing while keeping light compression.

Patients receiving antiplatelet treatment or in whom a larger sheath is used may need prolonged compression time.

To avoid rebleeding, maintain gentle MC until the patient is awake and their agitation has stopped.

If bleeding persists, maintain MC for 15 more minutes. If hemostasis is not achieved within 30 min and ACT >200 s, consider administering Protamine Sulfate for Heparin neutralization.

The Protamine dose to neutralize 100 units of heparin according to the elapsed time since the heparin was given:

- <30 min: 1 mg.
- 30–120 min: 0.5–0.75 mg.
- >120 min: 0.25–0.375 mg.

In general, 1 mg of Protamine neutralizes 100 units of heparin (not to exceed 50 mg/dose).

Vessel dissection and surgical restoration, or simple ligation is the last resort (rarely used in pediatric setting) whenever hemostasis is not achieved with conventional methods.

After compression, observe the site for 5 min to see if there is good hemostasis. Recompress if there is a hematoma or signs of bleeding. Recheck again in 5–10 min and consider application of a suitable pressure dressing.

An elastic dressing is applied on the puncture site with special care not to make it too tight. It should be a clear, plastic, waterproof, sterile dressing permitting visualization of the entry and surrounding tissues. It should be left for 12–24 h, with regular inspection every 15 min for 1 h, every 30 min for 2 h, then every 2 h until hospital discharge on the following day.

Patients should be kept in bed rest for 6–8 h after hemostasis whenever this is possible.

15.1.2 Vascular Closure Devices (VCD)

The use of hemostasis devices (VCD) has become more and more spread in many hospitals, particularly in adults (10–15% of all catheter-based procedures performed in adults utilize a VCD for femoral access site hemostasis). Those devices are conceived in the aim of improving patients' comfort and reducing hemostasis time. Most of them have a safety profile comparable to MC.

However, the use of these devices is still not very common in the pediatric population.

All VCDs reduce the time to obtain hemostasis. Each device has its own unique insertion technique, technical limitation and complications.

Based on their mechanism of action, VCD are divided into two categories: passive and active.

15.1.2.1 Passive Vascular Closure Devices

Hemostasis Pads

Coated with procoagulant substances to boost coagulation and hemostasis, these pads can be used in conjunction with MC. Compared to MC alone, hemostasis pads do not shorten the time to ambulation but they improve patient and physician's comfort.

Compression Devices: FemoStop (St. Jude Medical, USA) and ClampEase (Pressure Products Inc., USA)
Those devices are intended to replace humans with mechanical compression, either by using an inflatable bubble (FemoStop) or a clamp (ClampEase).

15.1.2.2 Active Vascular Closure Devices

Cardiva Catalyst (Cardiva Medical Inc., USA)
Used with MC, the Cardiva Catalyst is suggested for diagnostic or interventional procedures with sheath sizes up to 7 Fr.

Insert the apparatus through the existing sheath. Once the tip is within the arterial lumen, deploy a 6.5-mm umbrella-shaped disk coated with protamine sulfate. Remove the sheath and gently pull the disk against the arterial wall, where it is held in place by a tension clip. After 15 min (120 min for interventional cases), withdraw the device and lightly compress for 5 min.

The device is compatible with most patients and has been successfully used in pediatric patients.

Collagen Plug Device: Angio-Seal (St. Jude Medical, USA)
A three-component device, the Angio-Seal, includes a small and flat anchor, a collagen plug, and a suture.

Angio-Seal uses purified bovine collagen as a sealing substrate. The resorbable collagen plug induces platelet activation and aggregation, releases coagulation factors, and eventually results in the formation of fibrin and in thrombus generation. Angio-Seal achieves hemostasis by anchoring a collagen plug to the anterior vessel wall through a sheath delivery system.

Exchange the existing arterial sheath for a specially designed 6 Fr or 8 Fr sheath with an arteriotomy locator. Once proper positioning within the arterial lumen is secured, firmly hold the sheath in place and remove the guide wire and arteriotomy locator. Insert the device into the sheath until it snaps in place. Deploy the anchor and pull back against the arterial wall. This positions the collagen

plug just outside the arterial wall. Cut the suture below the skin level, leaving behind the anchor; collagen plug and suture will all dissolve within 2–3 months.

Collagen Plug Device: Mynx (Access Closure, USA)

Mynx uses a polyethylene glycol sealant that deploys outside the artery, while a balloon occludes the arteriotomy site within the artery.

Insert the device through the existing procedural sheath, and inflate a small balloon within the artery and pullback to the arterial wall. Place the sealant outside the arterial wall where it expands to attain hemostasis, and then deflate and remove the balloon through the tract.

Polyglycolic Acid (PGA) Plug Device: ExoSeal (Cordis Corporation, USA)

ExoSeal delivers an absorbable synthetic plug to the space adjacent to the arteriotomy using visual guidance for 6 Fr arteriotomy closure.

FISH (Morris Innovative, USA)

FISH uses an absorbable extracellular matrix "patch" made from porcine intestinal submucosa. It is indicated for procedures using 5–8 Fr sheaths.

Insert the "patch" through the arteriotomy to straddle the arterial wall. Release the "patch" from the device and pull a compression suture to hold the patch firmly in place.

Clip Device: StarClose (Abbott Vascular, USA)

StarClose uses a nitinol clip implant to reach hemostasis.

Insert the device into the arterial lumen, deploy the "wings," and pull against the arterial wall when the device is removed. Deploy the clip just outside the arterial wall to grip the edges of the arteriotomy and draw them together. The StarClose device is designed for invasive procedures using 5–8 Fr arteriotomies.

Suture Devices: Perclose (Abbott Vascular, USA)
Perclose offers suture-mediated VCD and has been successfully used in pediatric cases.

The hemostasis procedure requires several steps: positioning the device, needle deployment, suture capture, and needle removal. Vessel closure starts by replacing the vascular introducing sheath with the Perclose device. Guidewire access is unchanged until hemostasis is achieved. Three necessary components are included in each system: a closer, a clincher, and a knot pusher.

When the needle exit ports of the device lie just within the arterial lumen (as indicated by pulsatile exit of blood through "marker" lumens located adjacent to the needle exit point), the needles are deployed so that they exit the device within the arterial lumen, pass through the vessel wall, and are collected by a barrel located on the shaft of the device just outside the artery.

The barrel then conducts the needles and sutures to the surface through the sheath tract so that the two ends of each suture can be retrieved, tied together in a slipknot, and pulled down to the arterial surface to create a "surgical" closure of the arteriotomy. The delivery sheath and the guidewire are then removed as tension is maintained on the knot to achieve hemostasis [2].

15.1.3 Hemostasis in Non-femoral Access Sites

15.1.3.1 Jugular Access

The internal jugular vein is commonly used in congenital heart procedures, including catheterization in children with cavopulmonary anastomosis.

Simple MC for 5–10 min is usually sufficient for hemostasis after sheath withdrawal.

Some studies have showed the safety and efficacy of the Cardiva Boomerang Catalyst closure device in achieving hemostasis after jugular access.

Always beware of compromising the carotid artery vascularization when applying manual compression on the internal jugular.

15.1.3.2 Umbilical Access

In newborns 3 to 5 days old, umbilical arteries and vein are usually available for vascular access.

The umbilical vein can accommodate relatively large catheters and facilitate access to the heart. Less frequently, an umbilical artery can be cannulated and used for aortic and left ventricular catheterization.

Hemostasis is achieved after catheters and guide wires are withdrawn at the end of the procedure by simple light compression of the umbilicus for 5 to 15 min.

Shall the bleeding persist, the umbilical vein can be sutured directly or occluded with skin sutures.

A subsequent sterile non-compressive dressing should cover the umbilicus for 24 h.

15.1.3.3 The Transhepatic Access

Transhepatic access is an alternative for diagnostic and interventional procedures in patients with obliteration of the typical access sites.

The transhepatic vessels can accommodate relatively large sheaths.

This method provides relatively direct access to the atrial septum, left atrium and pulmonary veins.

It is possible in patients as small as 3.1 kg, but is mostly used in patients weighing more than 10 kg.

If the ACT exceeds 200 seconds, Protamine is administered (1 mg per 200 U heparin, maximum dose 25 mg, administered over a 5-min period) to lower the ACT below 200 s before the transhepatic sheath is withdrawn.

After removal of the sheath, the tract is obliterated with Gelfoam plugs to reduce the risk of significant hemorrhage.

The Gelfoam plugs are formed from small Gelfoam strips rolled to fit into the delivery sheath. The plug is advanced to the end of the sheath, which is then withdrawn while deploying the

Gelfoam plug. The intraparenchymal position of the sheath tip is documented by contrast injection.

Alternatively, an MReye Embolization Coil (Cook Inc., USA) or an Amplatzer Vascular Plug (AGA Medical Co., USA) can be used for this purpose. The coil or device is placed in the parenchymal tract between the hepatic vein and the liver capsule to minimize the risk of bleeding (Fig. 15.1). The sheath is then completely withdrawn and the puncture site covered with sterile dressing.

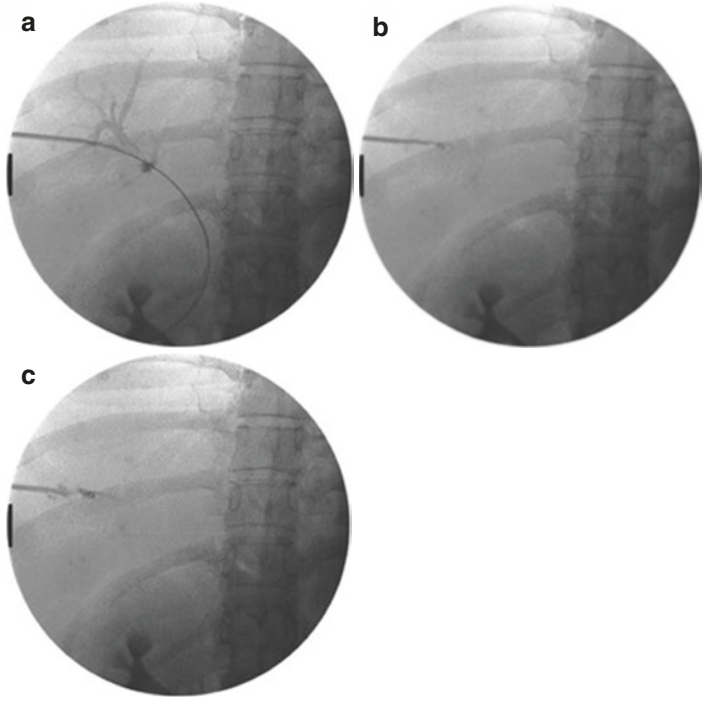

Fig. 15.1 Hemostasis after transhepatic catheterization: (**a**) A small volume (1–2 mL) of the contrast is injected to visualize the hepatic veins. (**b**) An MReye Embolization Coil (0.035 in. × 4 cm × 3 mm) is advanced to the end of the sheath and deployed between the hepatic vein and the liver capsule. (**c**) The intraparenchymal position of the sheath tip is documented by contrast injection

After catheterization, patients are monitored, their vital signs are checked, and the percutaneous transhepatic puncture site is inspected every 15 min for 1 h, every 30 min for 2 h, and then every 2 h until hospital discharge on the following day [3].

15.1.3.4 Surgical Vascular Access

Surgical access is usually reserved to cases when the Seldinger method fails or when larger introducer sheaths are used.

Sheaths are inserted through the artery or vein and secured with 2 purse-string sutures using Prolene 6/0.

After sheath's removal, the ends of the suture are drawn tight and the wound is closed like a purse.

Once the hemostasis is achieved, the subcutaneous tissues are closed with Vicryl 4/0 and the skin is sutured with Dermalon 4/0 (skin sutures are removed 2 weeks later).

15.1.3.5 Hybrid Procedures

Cardiac structures are directly punctured and sheaths are secured with two purse-string sutures using Prolene 5/0 or 6/0. Hemostasis is achieved the same way as in the surgical vascular access.

15.1.3.6 Fetal Interventions

In fetal interventions, the cardiac chambers are entered by a direct wall puncture through the maternal abdominal wall. At the end of the procedure hemostasis is usually achieved by a simple retrieval of needle, wire, and balloon. A sterile non-compressive dressing is applied to the maternal abdominal wall at the access site. No other hemostatic measures are needed.

Acknowledgments This is an update of the chapter written by Zakhia Saliba, Sami G. Slaba, and Elie B. Sawan, published in the first edition of this book.

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16 Access Complications and Management

Zakhia Saliba and Ramy C. Charbel

16.1 Access Complications and Management

Vascular access (VA) may lead to serious complications and high morbidity. Iatrogenic vascular injuries in children are affected by some anatomical and physiological characteristics: Children have thicker subcutaneous tissue resulting in less effective compression and they can be less compliant with post procedure ambulation recommendations. On the other hand, adults may have calcified arteries that could break under needle puncture, advanced stages of atherosclerosis, or arterial occlusive disease that would make the repair difficult.

Arterial vessels in young patients are more fragile and prone to vasospasm or sub adventitial injury if dilated extensively. Moreover, multiple access sites may be required in structural

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heart disease catheterization, while repeated cardiac interventions and multiple cardiac surgeries may result in incremental loss of sites. Thus, efforts should be made for preservation. It is essential to be familiar with the predisposing factors affecting the outcome of the procedure to avoid complications. These factors may be divided in 3 points:

1. Operator-related factors: Personal experience and ability to use ultrasound to guide access.
2. Patient-related factors: Age, anticoagulant medication, bleeding disorders, polyglobulia, connective tissue disorders, multiple cardiac surgeries or repeated catheterization using the same VA site.
3. Technical factors: the use of larger sheath in a smaller patient, complex and long exposure, unplanned access, simultaneous venous and arterial ipsilateral femoral access, insufficiently immobilized patient and the use of Vascular Closure Devices (VCD) for hemostasis.

Patients with one or more of the preceded conditions are to be carefully considered for preventive hemostasis after cardiac catheterization.

16.1.1 Preventive Measures

In order to decrease the incidence of complications' occurrence, some measures may be advocated:

- Discontinue anticoagulant/antiaggregant medication a few days prior to the procedure.
- Use deep sedation for complete VAS immobilization.
- Apply accurate disinfection for an aseptic environment.
- Avoid artery and vein entry in the ipsilateral femoral access.
- Encourage the use of ultrasound guiding for percutaneous access.
- Introduce the guide wire without resistance and check its intravascular position at fluoroscopy.

  - If the guide wire position is not secure, exchange the needle for a venous cannula and inject contrast to delineate the vascular anatomy (Fig. 16.1).
- Use stepwise predilatation if introducing larger dilator/sheath assemblies.
- Monitor adequate heparinization with ACT control during a long-lasting procedure.

An adequate hemostasis (refer Chap. 13) and meticulous patient follow-up in the few hours following catheterization, as well as a couple of weeks later, are crucial.

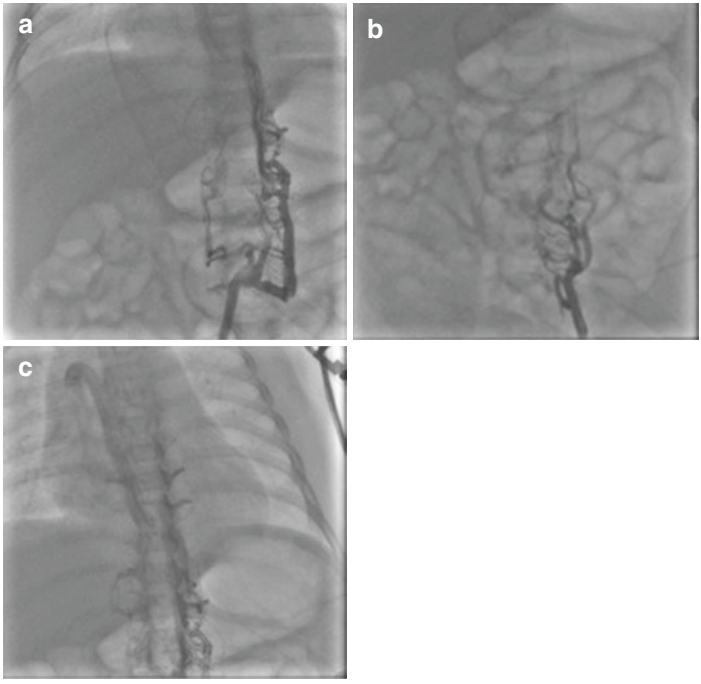

Fig. 16.1 Bilateral occlusion of the femoral veins in a 12-month-old child: dye injection in both veins (**a**, **b**) shows the contrast drainage to the right heart via a paravertebral collateral network to the right azygous vein and the right superior vena cava (**c**)

16.1.2 Complications Linked to the VAS and Techniques

16.1.2.1 Femoral Access

Percutaneous entry through the femoral artery and vein for cardiac catheterization is preferred because of the larger diameter of those vessels and the better accessibility of cardiovascular structures. To facilitate vessel entry and effective compression, the puncture should be above the femoral bifurcation but below the inguinal ligament.

A low stick, below the femoral bifurcation, may predispose to pseudoaneurysm, hematoma, arteriovenous fistula, dissection, and lymphocele, whereas a high stick may puncture the inferior epigastric artery or a posterior wall and cause retroperitoneal hemorrhage. Also, arterial or venous occlusion, femoral neuropathy, and pulmonary embolus may occur.

Hematoma

Arterial bleeding is a relatively common VAS complication where blood collects in the soft tissue. It is caused by blood loss at the VAS or by the perforation of an artery or vein and may occur if the arterial puncture is below the femoral bifurcation, making the femoral head unavailable to assist with compression.

Clinically, the skin surrounding the puncture site, where visible swelling is noticeable, is hardened. Hematomas vary in size and are often associated with pain in the groin area, which can occur at rest or with leg movement. Depending on severity, they can result in a decrease in hemoglobin and blood pressure and an increase in heart rate.

Managing a hematoma requires additional compression and immobilization of the leg, marking the area to evaluate for any change in size, providing hydration, monitoring serial complete blood cell counts, maintaining/prolonging bed rest, and stopping anticoagulant and antiplatelet medication if necessary, as well as blood transfusions if indicated.

If it is severe, it may require surgical evacuation but this is a rare occurrence in children. Many hematomas resolve within a few weeks as the blood dissipates and is absorbed into the tissue.

Acute Arterial Occlusion

Pulse loss after cardiac catheterization has been reported to occur in infants weighing less than 14 kg despite prophylactic use of heparin. The cause is usually vasospasm, especially in smaller patients. The classical 5 Ps (pain, paralysis, paresthesias, pulselessness, and pallor) are indicative of impaired circulation.

Recognizing limb ischemia in infants may be delayed because of their inability to communicate and the presence of more subtle signs such as decreased skin temperature and range of motion and skin discoloration.

As return of palpable pulses can be a false indicator of vessel patency, a rapid check of vasospasm (versus VAS thrombosis) should be made by ultrasound.

Due to the children's ability to develop a rich collateral network (Fig. 16.1), claudication or limb length discrepancy is less likely to occur.

Patients with markedly diminished pedal pulses at the end of catheterization should receive a heparin infusion at a rate of 12–17 units/kg/h to eliminate the risk of arterial spasm. This should begin with a bolus of 100 units/kg if the patients did not receive heparin during catheterization or of 50 units/kg if more than 2 h have elapsed after heparinization during the procedure. If the pedal pulse remains non-palpable or greatly diminished 4 h later, thrombolysis is considered and the patient should be transferred to the ICU. Thrombolysis instituted with tissue-type plasminogen activator using a bolus of 0.1 mg/kg followed by an infusion of 0.5 mg/kg/h for 2 h. Heparin infusion is then reinstituted at the same rate for 4 h.

If pulses become palpable during this period, heparin is continued for 6 h. Otherwise, a second course of thrombolysis is administered, again with a bolus of 0.1 mg/kg followed by an infusion of 0.5 mg/kg/h for 2 h with another subsequent heparin infusion

of 12–17 units/kg/h for 6 h. Patients have to be closely observed for complications, particularly for bleeding at the VAS. It is important to expose the pressure dressing so that bleeding can be instantly recognized and treated with manual compression (MC). The patient is kept NPO and remains in the supine position with the leg kept straight. Vital signs are monitored closely. This can lead to a patency rate of the target vessel of 95% [1].

Chronic Arterial Occlusion

Permanent arterial occlusion has been reported to range from 5.5 to 20% of cases.

If the chronic femoral artery occlusion causes no symptoms, the child should be monitored on a regular basis, but if there are symptoms, operative intervention is warranted.

In older children and adults, percutaneous recanalization of the occluded vessel may be attempted, first by passing a wire through the obliterated segment. The wire is then supported by a stiff catheter or dilator. Some use a straightened transseptal needle to complete the passage. During this maneuver, repeated contrast injections through the dilator or supporting catheter ensure needle or wire is following the proper "intravascular" course. After the wire passes into the proximal vessel, balloon angioplasty is performed with a balloon diameter slightly larger than that of the adjacent normal vessel.

Wire recanalization and balloon angioplasty are applicable to femoral and iliac veins and arteries, inferior vena cava and superior vena cava, as well as innominate vein. The lumen achieved is adequate for catheterization but may not remain patent long term. Stent implantation should therefore be considered to achieve a more uniform lumen large enough to ensure long-term patency.

Retroperitoneal Hemorrhage

Bleeding occurs behind the serous membrane lining the walls of the abdomen/pelvis and may occur if the arterial wall puncture is made above the inguinal ligament, resulting in the perforation of a suprainguinal artery or the penetration of the posterior wall.

It can be fatal if not recognized early, but can be diagnosed by computed tomography.

Patients usually display moderate to severe back pain, ipsilateral flank pain, vague abdominal or back pain, and abdominal distention (often not associated with obvious swelling hypotension and tachycardia). Ecchymosis and decrease in hemoglobin and hematocrit are late signs.

Retroperitoneal hemorrhage is managed by providing hydration, performing serial blood cell counts, maintaining bed rest, interrupting anticoagulant and antiplatelet medications if necessary, and performing blood transfusion if indicated. In rare instances, it may require surgical evacuation.

Pseudoaneurysm

A communicating tract between the tissue and, usually, one of the weaker walls of the femoral artery causes blood to escape from the artery into the surrounding tissue.

Possible triggers include difficulty with arterial cannulation, inadequate compression after sheath removal, or impaired hemostasis. It may occur if the arterial puncture is below the femoral bifurcation so the femoral head is not available to assist with compression.

Posttraumatic pseudoaneurysms in children are rare, and thus there is little information regarding treatment. In adults, they are more frequent with use of thrombolytics, antiplatelet agents, and anticoagulants.

Clinical signs include swelling at insertion site, large and painful hematomas, ecchymosis, pulsatile mass, or bruit and/or thrill in the groin. Pseudoaneurysms can rupture, causing abrupt swelling and severe pain.

One should suspect nerve compression when pain is out of proportion compared to the hematoma size. Nerve compression can result in limb weakness that takes several weeks to resolve.

Small femoral pseudoaneurysms commonly close spontaneously after cessation of anticoagulant therapy and require prolonged bed rest and monitoring, whereas large ones should be treated by ultrasound-guided compression or surgical repair.

Arteriovenous Fistula

An arteriovenous fistula is an abnormal connection between the arterial and venous system that bypass the normal anatomic capillary beds. Acquired AVF of the lower extremity is by far the most commonly occurring AVF due to the frequency of femoral percutaneous arterial and venous access.

The main causes include multiple access attempts, punctures above or below proper site level, and impaired clotting.

Long-standing AVFs can lead to limb edema, high-output cardiac failure, or aneurysmal degeneration of the artery. AVF can be asymptomatic, and result in bruit and/or thrill at VAS, and in swollen and tender extremities. In adults with pre-existing peripheral artery disease (PAD), it can lead to the onset or worsening of lower extremity ischemic symptoms.

In most cases, the communication between the artery and vein will spontaneously seal. Whereas, some require ultrasound-guided compression or percutaneous device closure. Surgical repair is rarely needed especially in children.

16.1.2.2 The Internal Jugular Vein

It is frequently used for access, particularly in patients with interrupted inferior vena cava or after Glenn operation.

Complications include puncture of the carotid artery, Horner's syndrome, air embolism, mediastinal hematoma, hemothorax, and carotid jugular fistula. Multiple attempts to obtain the jugular vein raise the incidence of complications, and patients should be closely monitored. Ultrasound-guided jugular vein catheterization is obviously superior to the landmark-based technique in terms of decreased incidence of complications.

When inadvertent carotid artery cannulation or hemothorax/pneumothorax have occurred, the appropriate management is first to stop the catheterization and pull out the catheter carefully. Patient monitoring should be performed in the ICU. The second step is local compression, which can stop bleeding, and then monitoring the vital signs of the patient. If they are stable, further checks such as physical examination, neurological examination,

Doppler ultrasound examination, plain chest radiograph, magnetic resonance angiography, and carotid angiogram could be considered. Treatment depends on clinical tolerance [2].

16.1.2.3 Transhepatic Access

Potential complications of transhepatic access include pneumothorax, hemorrhage, hemoperitoneum, hemobilia, cholangitis, liver abscess, sepsis, and hepatic vein thrombosis. In addition, intraperitoneal hemorrhage requiring laparotomy may occur.

The use of ultrasound to provide visualization of the liver and hepatic veins during transhepatic access is a major modification that may further decrease the likelihood of puncturing other structures during access, and thus decrease the risk of complications. To reduce the risk of significant bleeding, obliteration of the parenchymal sheath is advocated for hemostasis. This can be achieved with either coil embolization or Gelfoam plugs. When it occurs, parenchymal hemorrhage should be treated conservatively. If bad tolerance happens, exploratory laparotomy may be needed.

16.1.2.4 Umbilical Access

In newborns that are less than 4 days old, the umbilical vein is available for a rapid vascular access, but this can lead to several types of access complications including blood loss, vascular perforation, and thrombosis.

Umbilical catheter manipulation may predispose the newborn to necrotizing enterocolitis. To reduce the risk of its occurrence, 48 h of post-procedural fasting is recommended.

16.1.2.5 Vascular Closure Devices (VCD)

Many VCD are used in adults, but in children there is no consensus on the validity of any of the devices. Whereas VCD have reduced time to hemostasis and facilitated patient mobilization, their safety remains controversial.

The complications related to VCD use are generally the same as those observed in patients who are managed by traditional MC,

with the exception of device embolization. Moreover, severe periarterial infection and endarteritis requiring major surgery have been reported following the use of VCD. Their use is therefore only recommended in selected patients [3].

Acknowledgments This is an update of the chapter written by Zakhia Saliba, Georges S. Tabet, and Chady C. Kallassy, published in the first edition of this book.

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Transseptal Access

17

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17.1 Introduction

Transseptal (TS) puncture of the left atrium was first described in 1959 as a new, alternative method in accessing the left heart for the assessment of patients with acquired or congenital heart disease [1]. With the increasing volume of structural heart procedures and a growing number of congenital heart patients reaching adulthood, the operator needs skills to safely perform a TS puncture.

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17.2 Procedures Requiring TS

Transseptal is a cornerstone in procedures like percutaneous balloon mitral valvuloplasty, transcatheter mitral valve repair (MitraClip device Abbott Vascular), transcatheter mitral valve replacement and mitral valve-in-valve procedure, mitral paravalvular leak repair, LAA appendage closure, and left-sided ablation for atrial fibrillation (Fig. 17.1).

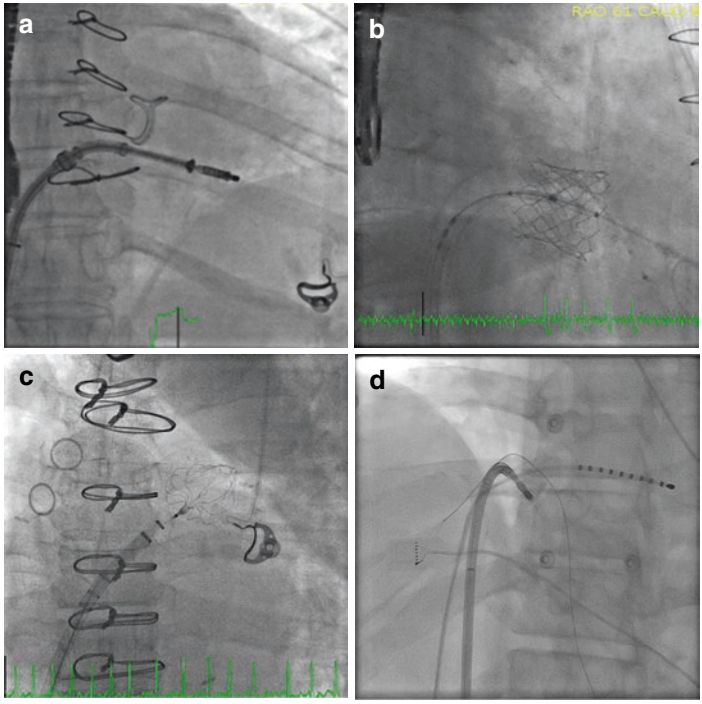


Fig. 17.1 Procedures requiring transseptal access. (**a**) Mitraclip (MitraClip device, Abbott Vascular), (**b**) Transcatheter mitral valve replacement (Edwards Sapien 3 valve, Edwards Lifesciences). (**c**) Left atrial appendage exclusion (Watchman device, Boston Scientific). (**d**) Left atrial fibrillation ablation

Although the fundamentals of transseptal puncture remain the same for all the procedures, each one of them requires a site-specific puncture for successful completion. In addition to structural heart interventions, transseptal puncture is also used for providing hemodynamic support using Tandem heart and for performing hemodynamic assessment in mitral and aortic valvular disease.

17.3 Understanding Transseptal Anatomy

A thorough knowledge of both atria, the *interatrial septum*, and adjacent cardiac structures is crucial for TS heart catheterization [2]. A majority of the atrial septation is formed by infolding of the right and left atrial walls (interatrial groove), with puncture outside the *fossa ovalis* valve (FO) and adjacent margins of its muscular rims (limbus), leading to perforation. The interatrial septum (IAS) is bounded posteriorly by a fold of the pericardium between the left and right atria, superiorly by the superior vena cava (SVC), anterosuperiorly by the noncoronary sinus of the aortic valve, antero-inferiorly by the coronary sinus, and inferiorly by the IVC. The supero-posterior rim is often referred to as the septum secundum. The aortic mound is located anterior and superior to the FO, overlying the aorta; posterior to the aortic mound is the transverse sinus or retroaortic space. More caudally, the pyramidal space constitutes the posterior septum where the right-sided pulmonary veins and their pericardial reflections forming the oblique sinus are located.

17.4 Transseptal Technique

The technique of *TS puncture* has a multitude of steps with variations made based on operator preference and patient-related factors (Fig. 17.2). First, a 0.032 or 0.035 in. J-tipped 135 cm guidewire is advanced to the SVC and an 8 F 62-cm-long sheath

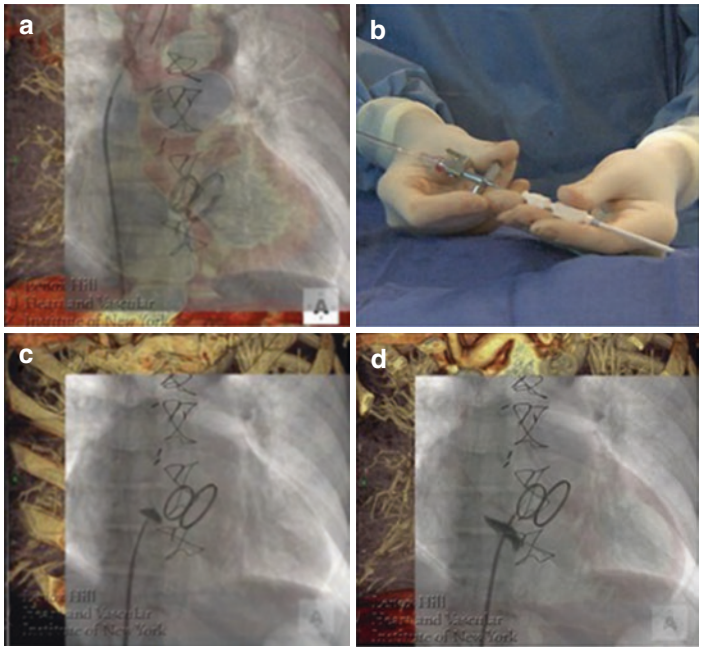


Fig. 17.2 Technique of transseptal puncture. (**a**) An 8 F 62-cm-long sheath is advanced over a J-tipped guidewire to the superior vena cava. The guidewire is removed and a 71 cm transseptal (TS) needle introduced under continuous flush. (**b**) With the needle approximately two fingerbreadths (1–2 cm) away from the sheath hub, the entire TS system is positioned at the 4–5 o'clock location and withdrawn caudally until it encounters two leftward jumps: SVC/right atrial junction and muscular interatrial septum. (**c**) TS tip subsequently engages the fossa ovalis (FO), confirmed by contrast injection. (**d**) Needle is briskly advanced puncturing the septum. (**e**) Once needle position is confirmed within the left atrium (LA), the entire system is advanced 1 cm. The dilator is disconnected from the sheath and the needle/dilator is turned toward the 12–1 o'clock location. (**f**) The sheath is advanced over the dilator into the LA and finally the dilator/needle removed. Passive back bleeding of the sheath de-airs the system and the patient is afterward anticoagulated

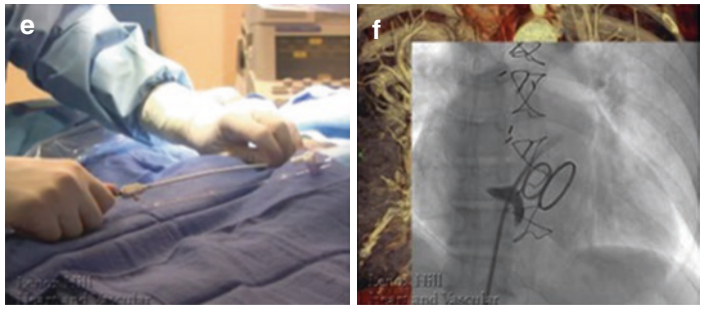


Fig. 17.2 (continued)

(SL or Mullins) placed. The guidewire is removed, leaving the sheath with its dilator locked in place. The dilator is bled back and flushed with a syringe to avoid thrombus or air introduction into the right atrium. Next, a 71 cm TS needle (standard Brockenbrough, BK, with 19° angle), selected to fit the length of the sheath, is attached to a manifold that allows for pressure monitoring, flush/discard, and contrast injection.

The needle is introduced while continuously flushing and is gently advanced through the sheath allowing it to rotate freely within the dilator. If resistance is met, especially at the location of the inferior vena cava/pelvic brim, the needle stylet should be reinserted to prevent piercing through the dilator/sheath. The needle tip must be kept within the lumen of the dilator, maintained approximately two fingerbreadths (1–2 cm) away from the sheath hub. Once the transeptal needle is juxtaposed to the tip of the dilator, the entire system is positioned at the 3–6 o'clock locations—both the sheath with sideport and needle indicator arrow pointed in the same direction. Typically the 4–5 o'clock location (45° from the horizontal plane) is most desired with 3 o'clock being directed toward the patient's left side (Fig. 17.3). The IAS is a posterior structure typically located at the 4–5 o'clock location; the aorta or retroaortic/transverse sinus is located at the 1–3 o'clock locations and should be avoided.

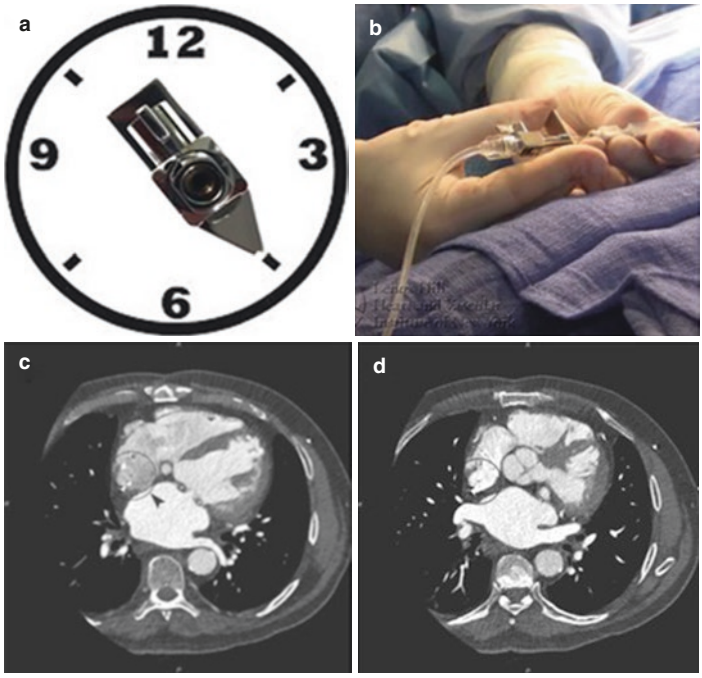


Fig. 17.3 Transseptal needle position. (**a**) The transseptal (TS) needle indicator arrow dictates the location of the needle tip. With the patient on a horizontal plane (3/9 o'clock), the sheath with sideport and indicator arrow are pointed in the same direction to the 4–5 o'clock location (45° from the horizontal plane). The interatrial septum with the fossa ovalis (FO) is typically a posterior structure located at this position. (**b**) Holding of the needle and the TS sheath/dilator requires maintaining the same distance from the distal dilator tip (1–2 cm) and concordant movements of the system. (**c**, **d**) A clockface superimposed on an axial slice of a cardiac CT reveals the location of the FO at 4–5 o'clock and the aortic valve more anterior at the 1–3 o'clock

Thereafter, the SVC/RA pressure tracing is recorded and the system withdrawn all together toward the IVC, without changing the relative distance between the sheath and needle. Upon descent from the SVC to FO, the system usually encounters two leftward jumps: first at the SVC/RA junction and second from the muscular IAS (in the region of the aortic mound) into the FO. The TS tip

should subsequently engage the FO, with the apparatus advanced slightly to firmly contact and tent the septum. A loss of RA pressure is typically noted and 3–5 cc of contrast gently injected to stain the IAS. Once the position is confirmed, the sheath/dilator is firmly anchored and the TS needle is briskly advanced, puncturing the septum. The transducer should reveal LA pressure, and additional contrast can be injected and oxygen saturation performed to confirm LA positioning. If LA pressure tracing is not noted, change the scale to assess for aortic pressure. Staining of the pericardium or aorta verifies inadvertent pericardial or aortic puncture. Until confirmation is made, the sheath/dilator should not be advanced.

The entire system is then advanced about 1 cm across the IAS, allowing the dilator to cross the septum. The dilator is disconnected from the sheath, and the needle/dilator is turned in a counterclockwise rotation, toward 12–1 o'clock, bringing the entire system anteriorly toward the center of the LA away from the posterior wall. The dilator/needle is fixed and the sheath advanced into the LA. Successively, the sheath is fixed and the dilator/needle removed slowly to avoid introducing air into the sheath. In addition, aspiration of the TS sheath can introduce air through the valve and is not recommended. Passive back bleeding with the sheath port positioned below cardiac level will allow for appropriate de-airing of the system. The sheath can then be flushed and the patient appropriately anticoagulated to achieve therapeutic ACTs between 250 and 300 ms, further adjusted according to the desired intervention.

17.4.1 Site-Specific TSP

17.4.1.1 Mitral Valve Interventions

Optimal transeptal puncture is crucial in mitral valve interventions [3]. The site of transseptal puncture for MitraClip depends on the origin of mitral regurgitation jet. For lateral and central jets, a superior and posterior puncture is selected; however, for medial jet an inferior and posterior puncture is favorable, avoiding excessive flexion of MitraClip system. A mid-fossa puncture is adequate for

PBMV and TMVR. Lateral paravalvular leaks (6–12 o'clock) might be easily reached with a central or anterior puncture. Medial paravalvular leaks (12–6 o'clock) are better approached from the transapical access, but if the transseptal approach is selected more posterior puncture is preferred (Fig. 17.3).

17.4.1.2 LAA Occlusion Procedure

A posterior TSP is required to achieve sufficient depth and maintain sheath coaxial to the long axis of LAA. There are small differences unique to the delivery of different LAA occlusion devices, depending on the delivery sheaths.

17.4.1.3 EP Ablations

With EP ablations, a relatively anterior puncture provides easier access to pulmonary veins which are located posteriorly in the left atrium (Fig. 17.4).

17.5 Knowledge of Imaging

17.5.1 Fluoroscopy

Traditionally, TS technique has been performed using *fluoroscopy*. The anteroposterior (AP) projection allows for identification of appropriate placement within the mid right atrium and against inadvertent placement into the right ventricle or LA through a patent foramen ovale (PFO). The placement of a pigtail catheter into the noncoronary cusp aids at the delineation of the posterior border of the aortic wall, as well as the aortic valve/root. It also provides active arterial blood pressure monitoring during the procedure.

In addition to the AP view, other views should be utilized including right anterior oblique (RAO) at 40–50° and left anterior oblique (LAO) at 30–55° [4] (Fig. 17.5). The location of the TS system within the anterior/posterior axis is evaluated in the RAO view and within the superior/inferior axis in the LAO view. In the RAO projection, the IAS is en face with posterior, superior, and inferior borders identified. The intended site of puncture is half-

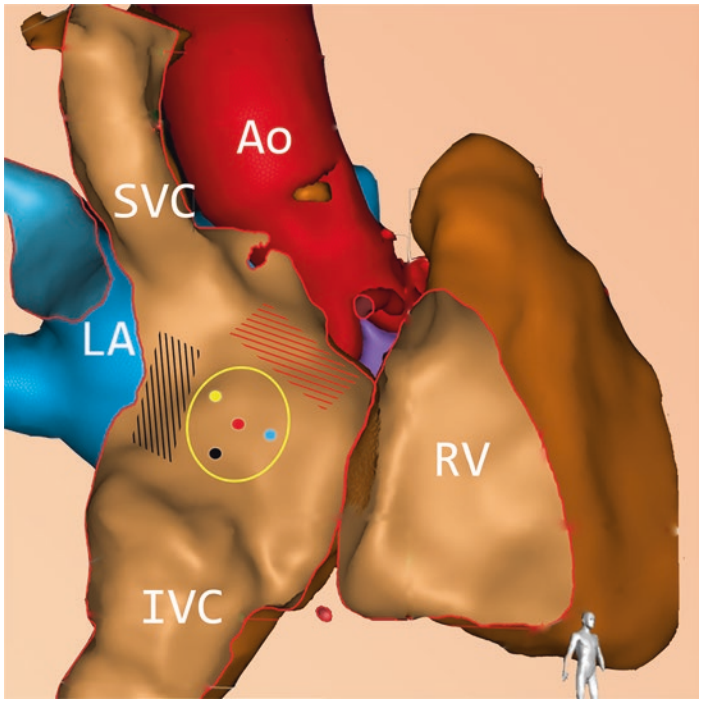


Fig. 17.4 Site-specific transseptal puncture. Yellow dot—MitraClip, medial paravalvular leaks, and mitral valve interventions; blue dot—EP pulmonary vein ablation; black dot—left atrial appendage closure and lateral paravalvular leaks; red dot—hemodynamic assessment. The risk of injury: red-striped rectangle—aortic root area, black-striped area—posterior perforation at the infold of the atria

way between the posterior boundary of the atria and a line drawn extending from the posterior aortic wall, approximately 1–3 cm below the noncoronary cusp. The angle at which the septum is punctured can be visualized with the needle directed away from the field of view. In the LAO projection, a line can be drawn extending from the posterior aspect of the pigtail catheter to the spine at a 45° angle. The intended puncture site is located approximately halfway between these two landmarks along this line with the needle directed to the right in a posterior direction.

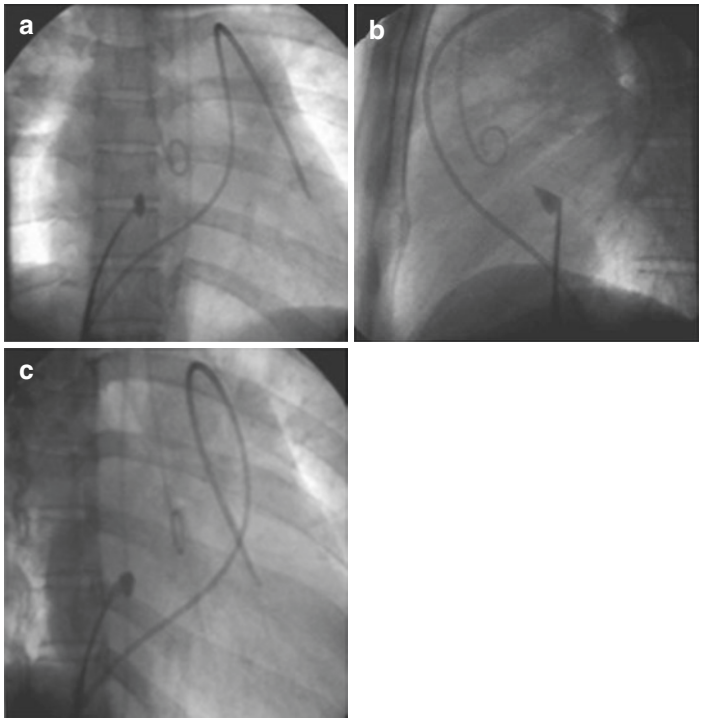


Fig. 17.5 Fluoroscopy for transseptal puncture. (**a**) In the anteroposterior (AP) view, the pigtail can be noted placed into the noncoronary aortic valve cusp, identifying the posterior border of the aortic wall and aortic root. A balloon-tipped catheter is also visualized within the right ventricular outflow tract into the left branch pulmonary artery. (**b**) In the left anterior oblique (LAO) view at 30–55°, a line can be drawn from the posterior aspect of the pigtail catheter to the spine at a 45° angle. Intended TS puncture is halfway along this line with the needle directed to the right (posterior). (**b**) In the right anterior oblique (RAO) view at 40–50°, the intended TS puncture is halfway between the posterior boundary of the atria and a line drawn extending down from the posterior aortic wall, approximately 1–3 cm below the noncoronary cusp

17.5.2 Echocardiography

Nonetheless, the distortion of the IAS decreases the efficacy of conventional fluoroscopy in identifying the anatomic landmarks. Echocardiography can offer high-resolution images of important cardiac structures, with transthoracic, transesophageal (TEE) and intracardiac (ICE) echocardiography. *3D TEE* enables even more comprehensive imaging of the heart, using either volumetric or multiplane 2D imaging [5, 6]. 3D imaging of the IAS closely parallels true anatomic inspection and is implicitly understood with the image easily rotated from the RA to LA perspective and intracardiac catheters and devices well visualized. Alternatively, ICE provides 2D and now *3D ICE* imaging with a clear definition of all intracardiac catheters, the IAS, septal tenting prior to puncture, and bubble visualization in the LA confirming needle position [7] (Fig. 17.6).

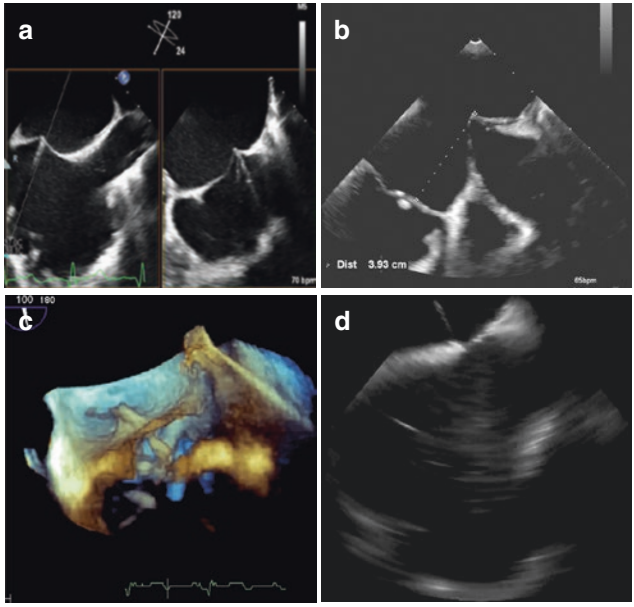


Fig. 17.6 Echocardiography for transseptal puncture. (**a**) With X-plane bicaval view a precise location for the transseptal puncture can be selected. (**b**) Measurement of height to the mitral valve annulus. (**c**) Assessment of the needle location in 3D. (**d**) Intracardiac echocardiography for transseptal puncture

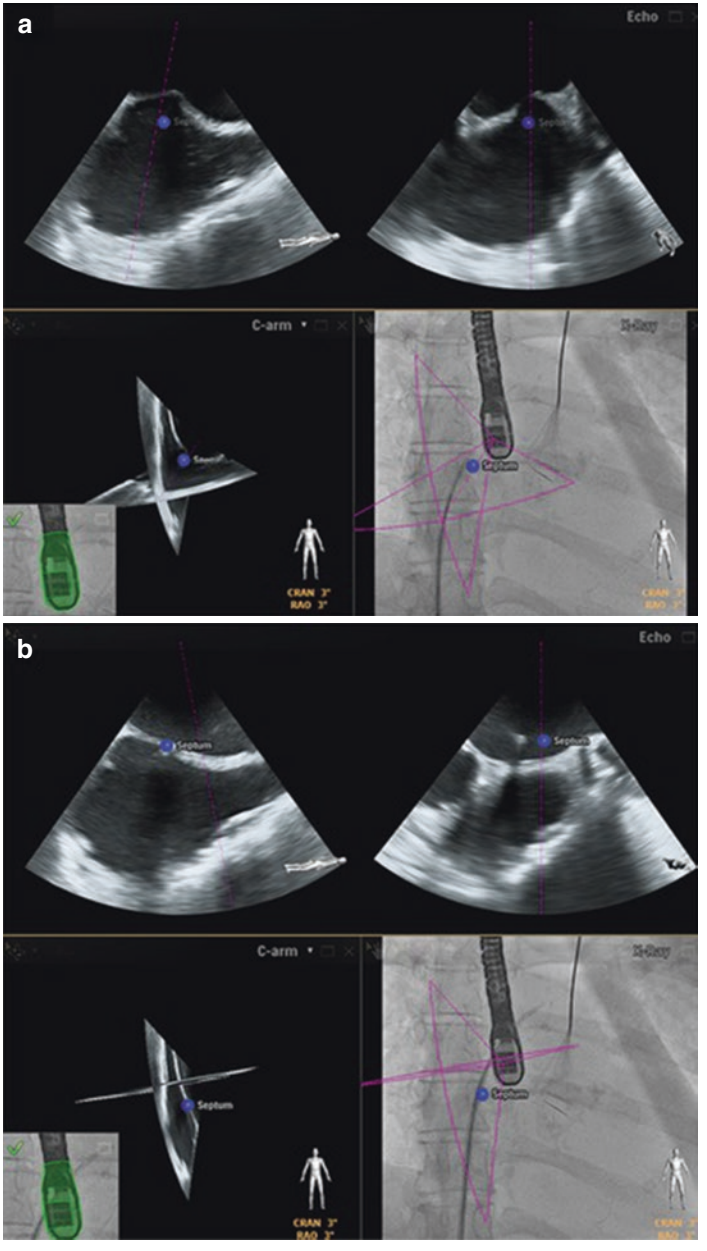
17.6 Advanced Imaging

With recent technological improvements, the integration of TEE with fluoroscopy in the catheterization laboratory, also known as *echo-fluoro imaging*, provides an alternative to traditional image-guided TS catheterization. The basis of fusion imaging relies on the utilization of live echo data and merging it with live fluoroscopy. Echo-fluoro software (Philips Healthcare, Best, Netherlands) automatically registers the 3D TEE field of view, in reference to the probe face plate, with fluoroscopy. After successful registration, a *TS landmark* can be placed on 2D x-plane and adjusted with confirmation in a 3D view. These landmarks are subsequently overlayed onto fluoroscopy to guide TS puncture (Fig. 17.7).

17.7 High-Risk Transseptal Anatomy

High-risk TS anatomy can vary and includes abnormal rotation of the cardiac axis; distortion of the IAS due to intra- or extracardiac causes; formation of an IAS aneurysm, abnormal fibrosis, hypertrophy, and/or calcification; and the presence of previously placed IAS closure devices. Recognizing these features is essential and modifying the approach to TS puncture necessary. Abnormal rotation of the cardiac axis can occur in the setting of significant ventricular hypertrophy or hypertrophic cardiomyopathy. Intracardiac causes of IAS distortion include LA and RA dilatation as well as many congenital heart defects. A dilated LA has bulging of the IAS toward the RA, making the FO convex. With TS needle

Fig. 17.7 Echo-fluoro imaging. (**a**) Echo-fluoro imaging provides an alternative to traditional image guidance with live echo data merged with fluoroscopy. Echo-fluoro software (Philips Healthcare, Best, Netherlands) automatically registers the 3D TEE field of view, in reference to the probe face plate, with fluoroscopy. After successful registration, a transseptal (TS) landmark (*blue dot*) can be placed on 2D x-plane (*upper frames*) and adjusted with confirmation in a 3D view. These landmarks are subsequently overlayed onto fluoroscopy to guide TS puncture (*right lower panel*). (**b**) Successful site-specific TS puncture performed with sheath/dilator advanced into the left atrium at the site of intended position


a
Echo
C-arm
CRAN 3°
RAO 3°
b
Echo
Septum
C-arm
Septum
CRAN 3°
RAO 3°

descent, the system is directed either too anterior or too posterior. On the other hand, a dilated RA has bulging of the IAS toward the LA. The FO is concave making it a challenge for the TS system to reach. For extracardiac distortion, severe scoliosis can alter the axis of the IAS and a dilated ascending aorta can cause bulging in the anterosuperior aspect of the IAS (Fig. 17.8a, c, d).

Surgically repaired IAS or presence of baffles/conduits can further alter anatomic landmarks with puncture and sheath advancement more difficult in the presence of *endothelialized patch material*. Multiple materials have been utilized including pericardium, Teflon (DuPont, Wilmington DE), Dacron (DuPont), and Gore-Tex (Gore, Flagstaff AZ); however, all can be crossed with minimal risk for residual shunt [8]. Increased thickness or calcification of the FO and local scarring at a prior puncture site can make repeat TS catheterization more challenging with lower success rates than first time punctures [9, 10] (Fig. 17.8b). Lastly, previously placed atrial septal defect (ASD) and patent foramen ovale (PFO) closure devices can equally alter anatomic landmarks with device overlapping varying parts of the septum and most endothelialized making sheath advancement more difficult.

17.8 Alternative Approaches and Advanced Techniques

Difficulties at engaging the FO may be related to many of the anatomical variations described [11]. Bending the standard BK needle to increase (dilated RA) or decrease (dilated LA) curvature and/or bending the patient with the right shoulder down may aid in engagement of the IAS. Alternative equipment such as Brockenbrough needles with additional length of 89 cm or accentuated curve (BK1, 53° angle) may be necessary to reach or engage the FO.

Additional efforts can be made to provide further evidence that the TS needle after puncture is located within the LA and reduce the risk of *LA free wall perforation* [12]. While stabilizing the TS system, the manifold can be removed from the TS needle and an 0.014″ coronary wire inserted and advanced either within the

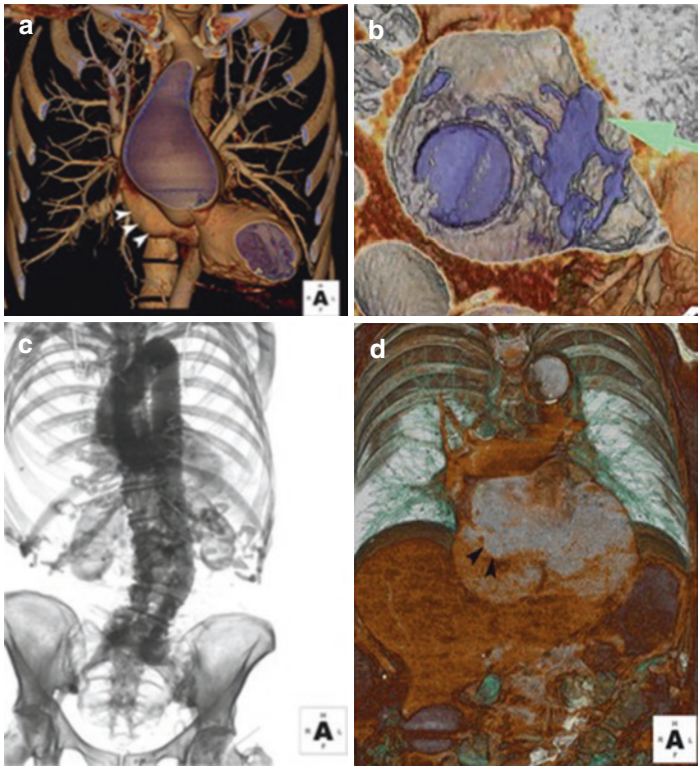


Fig. 17.8 High-risk transseptal anatomy identified by CTA. (**a**) Extracardiac distortion by an ascending aortic aneurysm can cause bulging of the anterosuperior aspect of the interatrial septum (IAS) (*white arrowheads*). (**b**) Extensive calcification (*green arrow*) of the IAS can be visualized in a postsurgical patient with limited location for transseptal puncture. (**c**, **d**) Patient with significant scoliosis leading to a more horizontal orientation of the IAS (*black arrowheads*)

body of the LA or into the left upper pulmonary vein. In addition to verifying location, the wire also minimizes the risk of perforation when the entire system is advanced across the septum.

Radiofrequency (RF) energy can provide an alternative method for TS access over conventional mechanical energy [13, 14] . This can be achieved by using a dedicated RF TS system (Baylis

Fig. 17.9 Radiofrequency NRG Transseptal Needle (Baylis Medical, Montreal, Canada)

Medical, Montreal, Canada) (Fig. 17.9) or direct application of RF to the end of a standard TS needle. The addition of RF cautery decreases the need for significant force applied to the septum for puncture, potentially improving the accuracy and risk of sheath/dilator inappropriate movement.

Another technique for transseptal puncture is using a *needle wire* system (Safesept, Pressure Products Inc., San Pedro,CA, USA). The needle wire system is a 120-cm nitinol guidewire with 0.014-inch diameter. It has a "J tip" which remains in straight inside the Brockenbrough needle but immediately prolapses upon entry into the left atrium allowing for safe crossing of septum [15] (Fig. 17.10).

Finally, increasing numbers of patients presenting for TS cardiac catheterization have undergone previous percutaneous ASD or PFO closure [16–18]. Areas of native septum not covered by the closure device and suitable for TS puncture can be considered. If not available, direct puncture through the device can be performed. Needle puncture of the device can be achieved via standard technique or with the use of RF energy. Once the needle has crossed the device, confirmed by imaging, the dilator is advanced into the LA. The TS needle is subsequently removed and a stiff guidewire then placed into the LA or left pulmonary vein. The tract is further enlarged using either a dilator or small septostomy balloon prior to advancement of the required TS sheath. Caution is necessary in cases where *ASD rims* were less

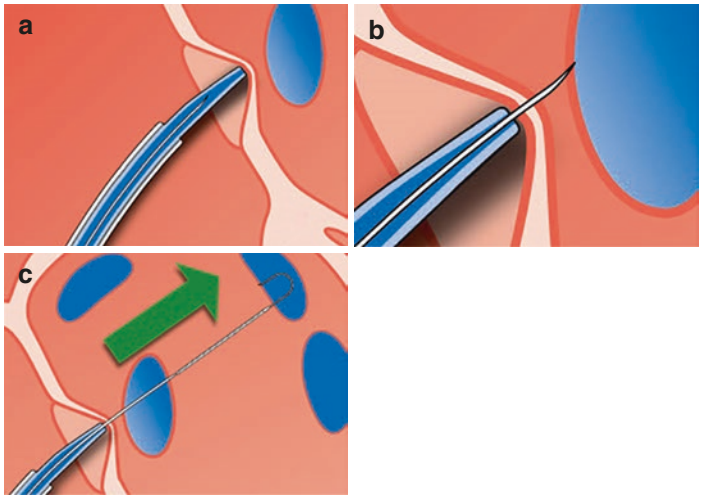


Fig. 17.10 SafeSept Transseptal Guidewire (Pressure Products, Inc., San Pedro,CA, USA). (**a**) tenting the interatrial septum. (**b**) transseptal puncture. (**c**) unsupported by the needle and dilator, the tip of the wire assumes a 'J' shape, rendering it incapable of further tissue penetration

than 5 mm or where inadequately supported large devices or very small devices are present. Traditionally, 6 months should have elapsed prior to attempting this technique to allow for endothelialization and securing of the device. At earlier time points, consideration for device retrieval can be considered.

17.9 Knowing the Contraindications

Despite the many potential indications, it is equally important to understand the contraindications to this procedure. The presence of *atrial thrombus or mass*, either in the right or left atrium, is an absolute contraindication. Organized thrombus specifically localized within the left atrial appendage is a relative contraindication and should only be performed by experienced operators with the ability to utilize advanced imaging modalities to reliably avoid the LAA with catheters and wires. The presence of smoke is not a

contraindication, and coagulopathy with an INR of >2.5 and/or a platelet count of <50,000 cell/dL is not recommended without reversal. In addition, a disruption of the normal inferior vena caval (IVC) flow excludes a traditional transfemoral venous approach to TS puncture.

17.10 Complications

TS puncture, on the whole, is a reasonably safe procedure with complication rates around 1%. Complications that can occur include the following: cardiac perforation, causing hemopericardium ± *pericardial tamponade* due to perforation of the RA or LA walls, LAA, or coronary sinus; *aortic wall perforation*; IVC perforation and retroperitoneal hematoma; cardiac arrhythmias such as atrial tachyarrhythmias and heart block; *systemic embolization* either from air, thrombus, cholesterol, or calcium; and death. Contemporary experience has revealed rates of tamponade ranging from 1 to 3%, systemic embolization less than 1%, and mortality of 0.1% [19–21]. The highest risk typically occurs either during the puncture or during advancement of the sheath into the LA. The factors that influence complication rates include the type of procedure, whether it is diagnostic or interventional, levels of anticoagulation, sheath size, left atrial pressure, presence and compliance of the pericardium, the use of imaging for TS guidance, and, most importantly, operator learning curve.

17.11 Conclusion

Recent expansion of left-sided diagnostic and interventional procedures has led to a resurgence of TS cardiac catheterizations. Understanding the indications/contraindications, IAS anatomy, the technical aspects of TS puncture and its associated complications are paramount. Alternative techniques for the difficult, high-risk patient should be recognized and employed. In addition, the use of multimodality imaging is essential for accurate TS localization and procedural safety. More advanced imaging such as

echo-fluoro imaging may play a greater role as site-specific TS puncture is required. Overall, TS access is a valuable procedure that can be successfully performed with minimal risk to the patient.

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Hybrid Access

18

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Luciana Da Fonseca Da Silva,
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Over the last three decades, the collaboration between pediatric interventional cardiology and congenital heart surgery has expanded tremendously, with two subspecialties joining for a common aim: to improve patient outcomes. Despite major technological advances in each of the two fields, both share a number of limitations, whether related to the intrinsic invasiveness of cardiac surgery and cardiopulmonary bypass (CPB), or due to limitations related to vascular access for cardiac catheter interventions. Hybrid procedures allow to bring both fields together and overcome the adversity of the invasive nature of CPB as well as the access limitations inherent to patient size or anatomy, which often preclude specific interventional strategies.

Having the attribute to be potentially safer, faster, and less invasive compared with traditional open heart surgery, the hybrid approach concept has brought to a commonplace the skills of the surgeon, who provides an optimal access to the heart and the skills of the interventional cardiologist who performs the interventional procedure, whether device deployment, balloon angioplasty or other. Among the most common hybrid access techniques performed are intraoperative stent implantations as well as perven-

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tricular muscular ventricular septal defect (VSD) closure [1–3]. The objective of this chapter is to describe various hybrid access approaches, as well as the surgical techniques and indications of the most common access sites for hybrid procedures in the treatment of congenital heart diseases.

18.1 Hybrid Carotid Access

The carotid artery approach is an advantageous technique since it provides a more direct route for the aortic valve—or at times the ductus arteriosus or Blalock–Taussig shunt—than trans umbilical or femoral access [4–6]. It also allows utilization of larger sheaths given that the carotid artery is larger in diameter than the femoral artery, particularly in small infants. After the description by Azzolina et al. [7] and given the risks reported with femoral route of acute limb ischemia, claudication, and growth restriction, the trans carotid cutdown approach is advocated for small patients whose peripheral access options are suboptimal, or when the anatomical features are such that the carotid retrograde approach would reach the lesion optimally. It may also be preferred for patients for whom a fragile and labile hemodynamic condition dictates expeditious procedures, such as critical neonatal aortic valve stenosis [5].

In this setting, a carotid artery cutdown is performed by the cardiovascular surgeon. The proximal right common carotid artery is exposed via a small incision in the neck. The carotid artery is carefully dissected to avoid injury to the vagus nerve (Fig. 18.1). The right common carotid artery is isolated, and the vessel is mobilized and secured using nontraumatic vascular clamps. A transversal arteriotomy is performed with insertion of an adequate size sheath directly into the carotid artery. The patient receives heparin (100 U/kg body weight, intravenously). A catheter is then passed through the sheath and into the ascending aorta or the site of interest. The procedure is subsequently completed by the interventional cardiologist.

This access facilitates balloon aortic valvotomy in neonates, particularly with prematurity (Fig. 18.2), or dilation of aortic

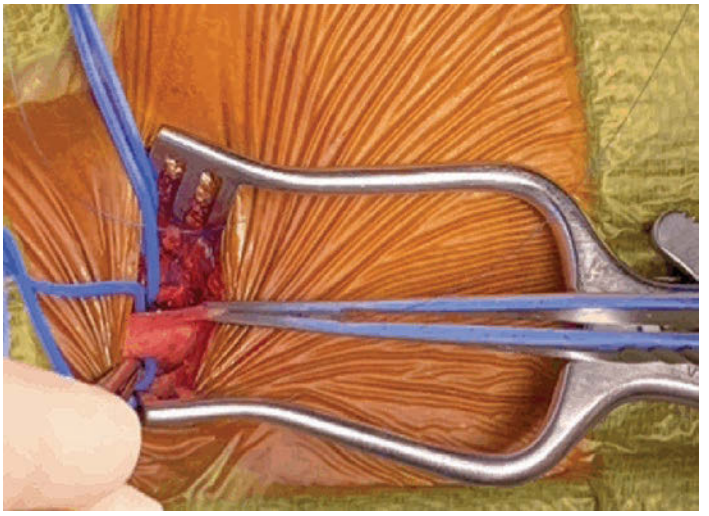


Fig. 18.1 Left carotid artery dissection. Blue vascular loops are placed around the artery

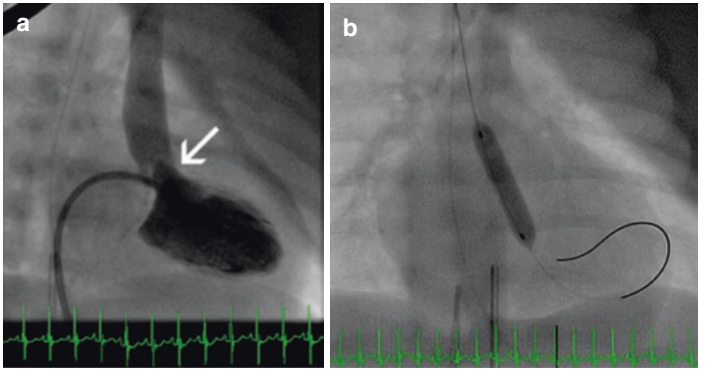
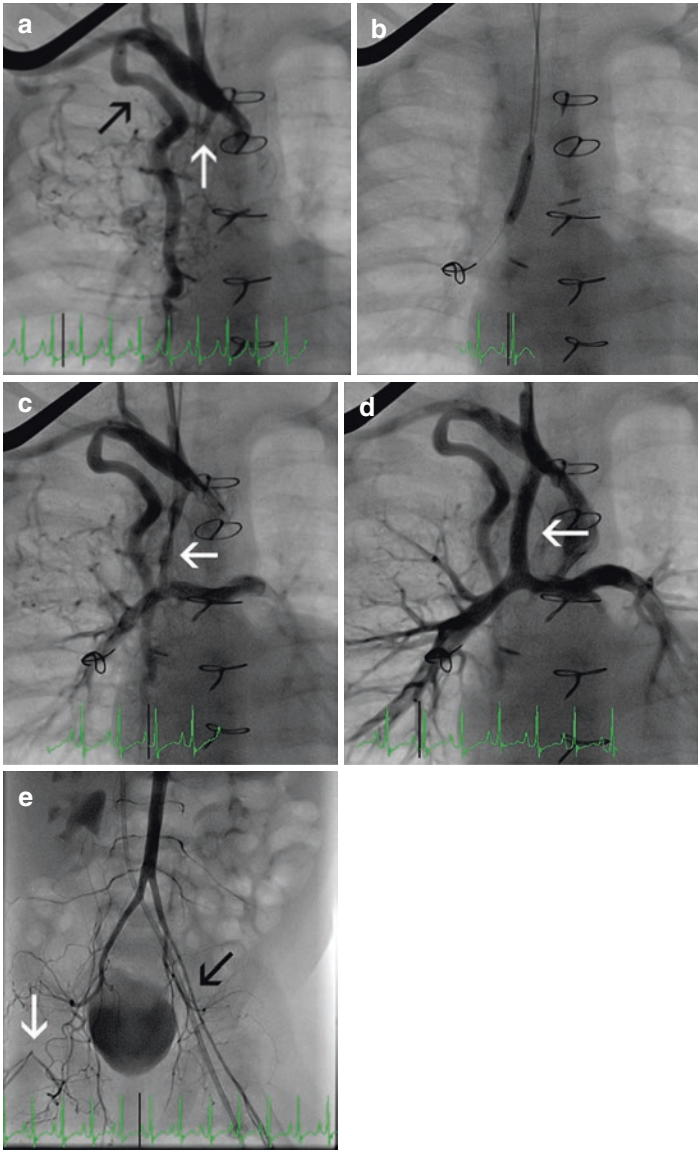


Fig. 18.2 Transcarotid aortic balloon valvotomy. (**a**) Left ventriculogram performed transvenously, typically via umbilical venous access, optimal in a premature infant, demonstrates doming thickened aortic valve. (**b**) Balloon valvotomy is performed via hybrid right carotid access

coarctation and stent implantation in children, using standard techniques [6, 8]. In addition, it can be a direct approach to a ductus arteriosus for ductal stenting or catheter intervention within a Blalock–Taussig shunt (Fig. 18.3). After the procedure, the sheath is removed, and the artery is reconstructed with interrupted sutures.

Although percutaneous carotid access has been reported as a safe alternative [9], most centers still continue to prefer the carotid hybrid approach. In adults, the carotid cutdown access can be used for transcatheter aortic valve implantation (TAVI). In this situation, the left common carotid artery is preferentially selected, since it provides superior coaxial alignment between the aortic root and the transcatheter heart valves (THVs) during deployment and affords simpler operating room configuration [10]. The proximal left common carotid artery is exposed via a small incision 2 cm above the left clavicle intravenous heparin is administered. Vascular clamps are used to achieve proximal and distal control of the carotid artery, and 5 F vascular access sheath is inserted. The stenotic aortic valve is then crossed in the usual fashion, using a straight-tip guide wire. A pre-shaped Amplatz Super stiff guide wire is then positioned in the apex of the left ventricle, and a 14 F sheath is inserted for the purposes of performing balloon aortic valvuloplasty. Thereafter, sequential dilation of the carotid artery is performed in selected cases with 16 and 18 F dilators, and an

Fig. 18.3 Transcarotid BT shunt recanalization. (**a**) Innominate artery angiogram performed via transcarotid access demonstrates complete occlusion of the BTS (white arrow), with prominent aortopulmonary collateral flow via large right internal mammary artery (black arrow). (**b**) After crossing shunt with coronary wire balloon angioplasty is performed. (**c**) Post angioplasty there remains significant shunt stenosis. (**d**) Stent implantation is performed demonstrating widely patent BTS (white arrow) post intervention. (**e**) Angiogram performed in the abdominal aorta demonstrates occlusion of the right common femoral artery (white arrow) and severe hypoplasia of the left common femoral artery (black arrow) precluding this access option, explaining the need for transcarotid intervention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18 F vascular access sheath is then carefully advanced into the ascending aorta. Standard TAVI implantation techniques are done. After valve deployment, the 18 F sheath is carefully retracted, and vascular clamps are applied to allow the artery repair using 6-0 polypropylene suture. A control angiogram is performed to assess artery patency.

18.2 Hybrid Axillary Access

Considering the same drawbacks from other retrograde or antegrade approaches, the axillary route has proven also to be a safe and effective way to access the heart for transcatheter interventions. Dua et al. described this approach as a good palliative initial procedure for severe congenital AS in infants <5 kg [11]. Moreover, this approach has been used for ductal stenting in critical right ventricular outflow tract lesions in the neonatal period and balloon angioplasty of aortic coarctation in critically ill newborns [12].

For lesions in the left side, the axillary artery is accessed by a "cut-down" technique. A small infraclavicular incision in the deltopectoral groove is made. The pectoralis muscle is separated along the muscle fibers and dissection of the axillary artery is carried out, without injuring the brachial plexus. Control of the vessel is established by encircling two rubber vessel loops proximally and distally around it. A 22-G catheter is used to puncture the artery through which a wire is advanced into the artery and exchanged for either a 3.5 or 4 F sheath. An angled 0.018″ wire is advanced across the aortic valve via a 3.5 or 4 F multipurpose catheter and a guidewire loop is formed in the left ventricular cavity.

After the aortic dilation, the catheter is then withdrawn and the sheath is removed. The arteriotomy is closed with 6-0 polypropylene sutures, followed by skin closure with interrupted sutures. To assess patency of the axillary artery, the right brachial pulse is palpated, and color and perfusion of the arm and hand are recorded post procedure.

18.3 Perventricular Access

Getting access to the heart for endovascular interventions is typically accomplished via percutaneous transvenous or retrograde arterial procedures due to lower procedural risk, but since the report by Levy and Lillehei [13], direct cardiac catheterization became an alternative for a wider spectrum of pathologies. Moreover, direct access to the heart has the advantages for relatively easier, shorter, and with a better coaxial alignment with intracardiac defects.

With the reports by Hozler et al. about the increased risk for procedure and device-related complications with percutaneous transcatheter VSD closure in patients <10 kg [14], the perventricular technique described by Amin, with it variants, became the preferred hybrid approach to address VSD device closure in selected patients [15–17]. Although most of the experience has been on muscular VSD device closure [18], there have been also reports on perimembranous and doubly committed VSD closure [19, 20]. Often times the indications for considering this technique include:

- Complex VSD posing difficulties for surgical patch repair without ventriculotomy
- Small patients (<5 kg) with inadequate peripheral vascular access
- Concomitant surgical procedure without CBP
- In the context of complex surgical repairs with the intend to limit the CBP time

Likewise, the experience garnered with percutaneous transcatheter pulmonary valve implantation along with the development of larger devices has prompted the use of the **perventricular hybrid approach for pulmonary valve implantation** with both Melody and Sapien prosthesis [21, 22]. In addition to less surgical trauma, and radiation exposure, this approach has no limitations regarding age and body weight, with the advantage that the entire

procedure can be done without the support of CPB and less blood transfusion requirements. Among the common indications are:

- Inadequate peripheral vascular access, either die thrombosis or small size
- RVOT complex anatomy limiting catheter course
- Ventricular arrythmias susceptibility precluding the use of long sheaths
- Patients at high risk for CPB

Although perventricular pulmonary valve implantation was initially used as a bailout strategy for failed percutaneous pulmonary valve implantation in patients with Tetralogy of Fallot (TOF) in whom a conduit repair had been used for reconstruction of the right ventricular outflow tract (RVOT) [23], it is now also considered as an alternative to surgical pulmonary valve replacement in patients with history of transannular patch repairs [24] where percutaneous implantation may be difficult. Often the hybrid approach in dysfunctional RVOTs requires downsizing of the main pulmonary artery and pre-stenting to create a proper landing zone and to minimize embolization risk.

These types of approaches can be accessed either through a full sternotomy, anterior thoracotomy, subxiphoid incision or transverse sternal split incision [25]. The location of the intracardiac defect and the anatomical characteristics will dictate which one would be the best access approach. In the following section, we will describe the subxiphoid incision and anterior thoracotomy technique for perventricular VSD closure, and full sternotomy for pulmonary valve replacement with main pulmonary artery banding.

18.3.1 Subxiphoid Incision Technique

A small longitudinal midline skin incision at the base of the xiphoid is performed. The inferior part of sternum is opened. The pericardium is accessed, and traction sutures are placed to the

edges of the pericardium to the thoracic wall to optimize exposure. Heparin is given intravenously (100 U/kg), and the activated clotting time is monitored to be longer than 250 s. Transesophageal echocardiography (TEE) allows to determine exact size, location, and distance between the puncture site and the ventricular septal defect. Additionally, the right ventricular free wall is palpated to locate the area of maximal thrill to aid in identifying the best puncture site. Then, a double 4-0 polypropylene purse-string is placed with the final goal to deliver the device orthogonally to the defect. The free wall is punctured within the suture using a trocar, after which the needle is removed, and a guidewire is inserted. The guidewire is advanced through the RV cavity, under TEE guidance and slowly passed through the VSD, and then directed into the left ventricular cavity. The dilator and delivery sheaths are introduced over the guidewire into the RV cavity. The dilator is removed, and the delivery sheath is advanced over the guidewire into the LV. The guidewire and the dilator are removed, and the sheath is allowed to bleed to remove any air entrapped inside it. The VSD device once loaded on delivery cable is inserted into the loading sheath, introduced into the delivery sheath, and advanced through the VSD, avoiding any injury to the LV free wall or mitral valve apparatus. Under TEE guidance, the left disc is deployed, and the sheath and the device are pulled back slowly until the left disc touches the ventricular septum. The waist and the right disc of the occluding device are released while the delivery cable is maintained under slight traction. The position of the device, any residual defect, and the motion of the aortic valve are assessed before complete deployment by TEE. The device is then fully released. Following device deployment, the delivery system is pulled back, and the purse-string is tightened. Any source of bleeding is controlled, a drain is placed in the pericardial space, and the wound is closed in the standard fashion. This approach has advantage to avoid sternotomy, allowing the incision to be small and located in an area of the chest which is less likely to be exposed, and therefore more esthetically pleasing.

18.3.2 Anterior Thoracotomy

A left parasternal ultraminimal intercostal incision made in the left sternal margin at the third intercostal space (<1 cm) and a pericardium hanging technique without sternal incision has also been described to occlude double committed VSDs [26]. The position of the purse-string suture on the right ventricular surface is determined using tweezers under direct visualization and TEE guidance. When the tweezers' head vertically points toward the VSD, a purse-string suture is placed at the position of the tweezers' head.

18.3.3 Full Median Sternotomy

In patients with prior history of cardiac surgery (such as those after TOF repair presenting for hybrid pulmonary valve implantation), careful entry is performed through a redo median sternotomy, and the RVOT and pulmonary trunk are fully dissected. Echocardiographic evaluations either by TEE or epicardial probe are performed in order to size the MPA and determine the puncture area. Location can also be planned based on prior imaging such as by computed tomography or cardiac magnetic resonance. Double purse-string sutures are placed approximately 2.5 cm proximal to the pulmonary valvar plane in the RV free wall and heparin (400 IU per kg of body weight) is given. Depending on the device used, downsizing of the MPA through a band placement or plication is required to achieve a diameter of 22–23 mm for the largest diameter Melody valve (outer diameter of 24 mm) or 27–28 mm for the largest diameter Sapien valve. Different materials have been used for banding including umbilical tapes, PTFE grafts, and felt strips. A radiopaque marker has to be included when placing the band, as well as anchoring to the pulmonary artery to prevent migration. For plication, a pledgeted longitudinal running suture provides a more elongated reduction of the RVOT, or horizontal mattress suture provides focal narrowing if calcification of a pre-existing RVOT patch is present [27].

Although the SAPIEN S3 and XT valves can be implanted without pre-stenting [28], stent implantation is often preferred in native RVOTs to offer an optimal landing zone. Pre-stenting is necessary for Melody valve implants to minimize the long-term risks of valve stent fracture. Percutaneous stent implantation can be performed after compliance testing and using for guidance the radiopaque marker positioned by the surgeon at the time of plication or banding of the main pulmonary artery. Subsequently, an 18-G needle is used to puncture the RV free wall where the purse-string suture was placed, and a 7 F sheath is introduced into the RV. After a guidewire is placed in the distal left pulmonary artery through the stent previously placed, the RV puncture is dilated to introduce the valve delivery system. After deployment, the purse-strings are tied down, and the valve is assessed by angiography and echocardiography. After the procedure is completed, the sternotomy is closed with a standard technique, with pericardial drains left in place.

Other groups have described the use of a porcine pulmonic valve mounted inside a self-expanding stent, adapting a delivery system used for transventricular aortic valve implantation. This technique avoids the use of pre-stenting and allows for a large valve implant, delivered via the main pulmonary artery, through hybrid access. Three sutures with pledges are placed at the proximal and distal sites of the valve externally to ensure fixation [29, 30].

18.4 Transapical Left Ventricular Access

The transapical hybrid approach is a well-established technique in the adult population for transcatheter aortic valve implantation [31] or for the treatment of prosthetic paravalvular leaks [32]. This approach provides access to both valves: retrograde to the mitral and anterograde to the aortic valve.

In patients with congenital heart defects, it has been used to address critical aortic valve stenosis in low-weight patients [33], as a rescue therapy where a retrograde approach was not possible [34], Fontan circuit patients without a secure retrograde access [35], for cardiac arrythmia ablations with mitral prosthetic valves

[36], or closure of paravalvular leaks [37]. This type of hybrid access can be performed through either a midline sternotomy or an anterior left thoracotomy.

18.4.1 Left Anterior Thoracotomy

Under general anesthesia, a parasternal transverse skin incision is made at the fourth or fifth intercostal space. Dissection of the intercostal muscles is done with electrocautery at the superior border of the inferior rib to avoid injuring the intercostal neurovascular bundle. Care must be taken in the medial side of the incision to avoid injuring the left internal mammary artery as well. The use of one lung ventilation is described to decrease the chance of entering the left pleural space. The pericardium is longitudinally opened followed by placement of traction sutures to the thoracic wall to optimize exposure. The left ventricular apex is identified, and after removal of some pericardial fat (in older patients), once the left anterior descending coronary artery is identified, and under echocardiography guidance, the puncture location in the LV is chosen near the apex. A 3-0 polypropylene pledged purse-string suture is placed in the LV free wall just lateral to the LAD coronary artery.

18.5 Main Pulmonary Artery Hybrid Access for Delivery of Ductal Stent or Branch Pulmonary Artery Stent

The hybrid approach for hypoplastic left heart syndrome (HLHS), and its variants, combines stent implantation in the ductus arteriosus with bilateral branch pulmonary artery surgical band placement to maintain systemic perfusion and restrict pulmonary blood flow [38, 39] (Fig. 18.4).

This procedure is performed under general anesthesia through a median sternotomy without the need for cardiopulmonary

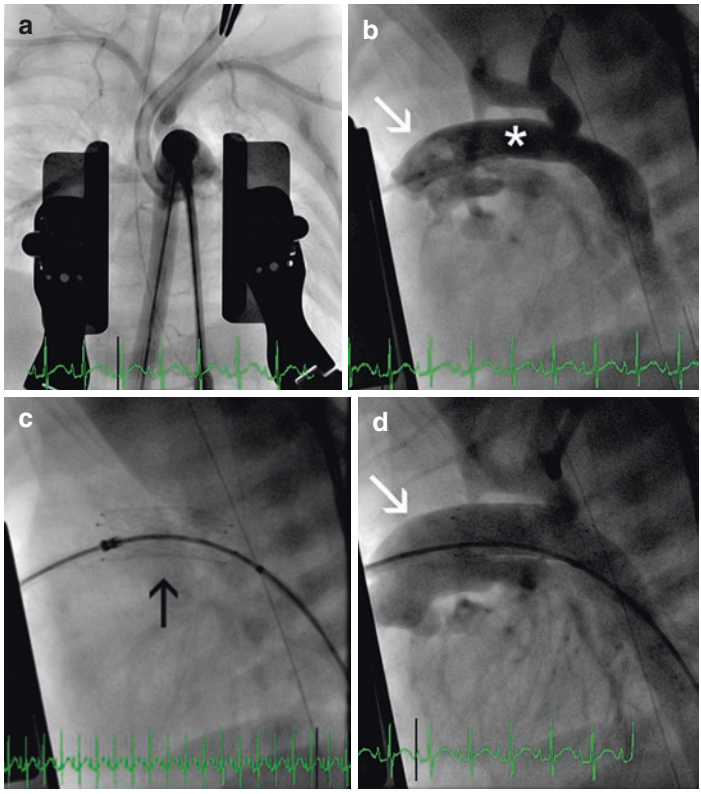

Fig. 18.4 Hybrid PDA stent implantation in HLHS. (**a**) AP projection demonstrates angiogram performed via mid line sternotomy directly into the ductus arteriosus, using a 4 F pigtail catheter inserted through the 6 F sheath inserted into the main pulmonary artery. (**b**) In the lateral projection, one can visualize access into the main pulmonary artery (white arrow) and detailed anatomy of the PDA (white asterisk). There is retrograde filling of the aorta, including a diminutive ascending aorta in the setting of aortic valve atresia. (**c**) A self-expanding EV3 Protégé stent has been implanted within the PDA. (**d**) Angiogram performed through the main pulmonary artery sheath (white arrow) post intervention in the lateral projection demonstrates good stent position covering the ductal tissue entirely and unobstructed retrograde filling of the arch

bypass. First, the bilateral pulmonary artery (PA) bands are placed before PDA stent implantation to minimize manipulation of the area and minimize risk of stent distortion. The bands are fashioned by cutting a 1- to 2-mm ring from a 3.5-mm Gore-Tex tube graft to wrap around each PA branch. For patients weighing less than 2.5 kg, a 3.0-mm graft is used instead. While the left band is placed immediately after the main PA bifurcation, the right band is positioned between the ascending aorta and the superior vena cava. The bands are attached to the adventitia, and the degree of tightening is based on patient size, caliber of the branch PA, systemic blood pressure and oxygenation response, as well as angiographic appearance. Typically, there is a 10-point increase in systolic blood pressure and a 10-point decrease in oxygen saturation.

After band implantation, a 5-0 polypropylene purse-string suture is placed in the main PA, avoiding pulmonary valve injuries in a too proximal position or laying in proximity to the PDA in a too distal position. Once the sheath in introduced, under angiographic guidance, the stent is positioned to completely cover the PDA, which typically extends from the left PA past the retrograde orifice of the transverse aorta. Angiography is then performed to confirm adequate stent position, with complete coverage of ductal tissue, following which the sheath is removed, the purse-string suture is secured and reinforced with a 5-0 polypropylene suture. Few centers prefer to perform PDA stent implantation percutaneously at a different time than bilateral pulmonary artery banding [40].

The final step is addressing the atrial septum, which can be done either by balloon atrial septostomy via peripheral venous access or through per-auricular access during the same hybrid procedure. A purse-string in the right atrium allows the introduction of a sheath through which the balloon is inserted in the atrium.

18.6 Right Mini-Axillary Thoracotomy

In patients with aortic re-coarctation after surgical aortic arch reconstruction or coarctation repair, balloon angioplasty is typically highly successful. However, there are times when vessel recoil or re-stenosis occurs, and then, surgical reoperation would be necessary. Coarctation stent implantation has become standard of care for the older child, but rarely is considered for small children. The major limitation of aortic stent implantation in infants and young children relates to the small diameter of the femoral and carotid vascular access to deploy a stent which can become adult size in the future. A hybrid access approach can overcome this limitation. A right mini-axillary thoracotomy can then be used to position a larger introducer sheath in the ascending aorta to deliver a balloon expandable stent of adult size capabilities in the descending aorta at the desired location. This less invasive thoracotomy can also be used in substitution to transhepatic cardiac catheterization for some interventional procedures on the right or left heart chambers, such as closure of atrial septal defects, mitral valve procedures, RVOT procedures, in patients with poor peripheral venous access, or obstruction of the inferior vena cava [41, 42].

The patient is positioned in left semi-lateral decubitus at 45° angle, with the right arm suspended at a right angle in a semi-flexion position. Pen marking of anterior and posterior axillary lines are done, with a third transverse line tracing from the lower scapular angle to the xiphoid appendage. An almost horizontal incision (3.5 cm) is performed in the mid-portion of the transverse line between the anterior and posterior axillary lines (bikini line in girls or more cephalad, nipple level, in boys). Using a muscle sparing technique, the third intercostal space is opened, and a retractor is placed. The pericardium is opened at 2 cm anterior to the phrenic nerve, and traction sutures are placed to expose the heart and vessels. A stabilizing 5-0 polypropylene suture is also placed in the apex of right atrial appendage for better aortic exposure.

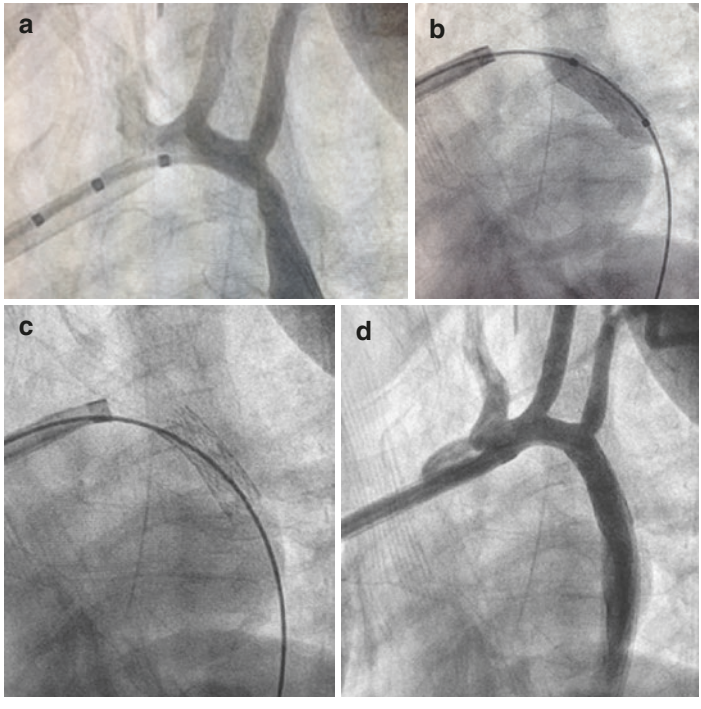

Fig. 18.5 Hybrid coarctation stent implantation in an infant. (**a**) In a lateral projection, angiography performed with pigtail catheter inserted via aortic sheath demonstrates severe postoperative coarctation. (**b**, **c**) Stent implantation is performed using a balloon expandable stainless steel stent. (**d**) Post-intervention angiogram performed via introducer sheath demonstrates significant angiographic improvement

Figure 18.5 illustrates an example in a 6-month-old infant with a history of coarctation repair in the neonatal period, who had had two prior interventions for coarctation. The patient presented with recoarctation again. A hybrid approach through a right thoracotomy to deliver a large stent, with adult size potential was proposed. The ascending aorta was exposed. After heparin was administered (150 IU per kg), a 5-0 polypropylene double purse-string suture was placed and a 7 F sheath was located into the ascending aorta under direct vision. On this occasion a stent

implantation was performed using a Palmaz Genesis 1910 stent mounted on a 7 × 20 mm Powerflex balloon. The lesion was balloon dilated at high pressure with resolution of the balloon waist and elimination of the 40 mmHg pre-intervention gradient. At 5 years of follow-up, the patient has been doing well with mild coarctation not yet meeting criteria for reintervention. The disadvantage of this approach versus reoperation is that there will be a need for repeat dilation of the stent over time for sure. Alternatively, percutaneous stent implantation with a smaller stent may allow for a successful palliation, but then rely upon ultra-high pressure angioplasty and unzipping capability of the stent to assure adequate diameter at long-term follow-up. If unsuccessful, reoperation would be required.

After the hybrid procedure, the catheter and sheath are both removed, the purse-strings are tied down, and the hemostasis is obtained. The pericardium borders are joined with two interrupted sutures, a chest tube is installed in the right pleura and placed under continuous negative pressure. The intercostal space and skin are closed in standard fashion.

In Fig. 18.6, we can see the location of the incision as imaged at the end of the procedure.

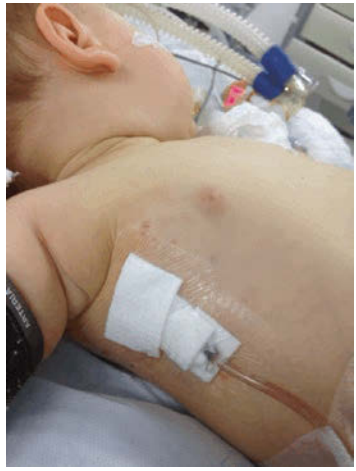

Fig. 18.6 Right axillary access incision. The photograph illustrates the appearance of the incision following hybrid stent implantation in the coarctation of the aorta in an infant shown in Fig. 18.5

18.7 Left Thoracotomy

Although some reports have shown that a left thoracotomy approach can be associated with respiratory complications due to single lung ventilation and chylothorax due to thoracic duct injury, this approach can also be of interest in cases with multiple sternotomies to avoid the risks of a redo surgery or when there is a contraindication for a secure percutaneous access for retrograde approach to address a lesion in the descending thoracic aorta (such as aortic stent implantation with no femoral arterial access) [43].

After double-lumen endotracheal tube placement, the patient is positioned in a right lateral decubitus position with the pelvis rotated to the left and the left arm suspended in an abduction and 90 grades flexed position. Essential landmarks need to be identified before proceeding with the incision, including the tip of the scapula, anterior superior iliac spine (ASIS), and axillary lines. Depending on the aortic stenosis level, the fifth or fourth intercostal space can be used. A 4–5 cm transverse incision is performed along the superior border of the rib, between the anterior and the posterior axillary line; using a muscle sparing technique, the entry intercostal space is identified, and the intercostal muscles are freed from the superior border of the rib, entering the pleural space.

After the thoracotomy is completed, a chest wall retractor is placed, right selective ventilation is established, and the left lung is collapsed and retracted. Under echocardiographic guidance, the entry site is chosen for direct cannulation in the descending aorta. The mediastinal pleura and descending aorta are dissected immediately superior and inferior to the entry site to have vascular control. A 5-0 polypropylene pledged purse-string is placed, so that the interventional cardiologist can finish the procedure. After successful device deployment, the catheter and wire are pulled back, and the purse-string is tightened. After reestablishment of bilateral lung ventilation, a chest drain is placed, and the thoracotomy is closed in the standard fashion.

18.8 Open Heart Access or Intraoperative Procedures

18.8.1 Intraoperative Diagnostic Angiograms (Completion or "Exit" Angiography)

This technique is most valuable to detect residual structural lesions after open heart surgery, allowing identification of patients who may benefit from either additional hybrid procedures (stent placement) or surgical reintervention, while the patient is still in the OR with an open chest. Examples include exit angiography after Blalock–Taussig shunt anastomosis, pulmonary artery unifocalization procedures, Glenn anastomosis, and pulmonary artery plasty or patch augmentation, among other procedures [44, 45].

As an example, following a bidirectional Glenn procedure in patients with previous hybrid approach for Stage I palliation for HLHS, branch pulmonary artery stenosis is not rare, given the detrimental effects of bilateral pulmonary artery band implants. If the oxygenation is not adequate immediately after weaning of the cardiopulmonary bypass, an exit angiogram can be performed through direct puncture of superior vena cava to demonstrate the anatomy of the pulmonary artery branches and the anastomotic site and possibly intervene on the spot with either patch augmentation or stent implantation via hybrid access.

18.8.2 Aortic Arch Obstruction: Hybrid Balloon Stent Implantation After Norwood Procedure During Stage II Palliation or Fontan Procedure

Re-coarctation associated sometimes with tubular hypoplasia may occur after Norwood procedure. Prompt relief of the obstruction is essential to preserve ventricular function and cardiac output. Balloon angioplasty is considered standard therapy and has high success rate. However, there are times when vessel recoil leads to a suboptimal result. Stent implantation is standard ther-

apy for vascular recoil. However, in infants the use of stents is limited given their inherent requirement for larger delivery sheaths, particularly when stents which have the ability to expand to an adult-size diameter are being considered. A hybrid alternative is to address this lesion at the time of surgery (bidirectional Glenn or Fontan procedure) with stent delivery under direct visualization or via direct puncture of the ascending aorta under fluoroscopic guidance.

Arterial catheters are inserted in the right radial artery and in the right femoral artery for pressure measurements during the procedure. The same access used for the surgical procedure is applied for the aortic re-coarctation treatment. A purse-string suture is placed in the ascending aorta. A portable digital C-arm (operating room) or biplane digital angiography is utilized (hybrid catheterization laboratory or hybrid operating room). The introducer sheath (sized according to the planned procedure) is placed into the proximal transverse aortic arch via the purse-string incision. The side arm of the sheath is utilized for serial angiography and aortic pressure measurements throughout the procedure. A right coronary artery catheter or other end hole catheter is advanced through the coarctation, and a guide wire is placed across the obstructed segment into the descending aorta. Stent implantation follows, often utilizing a large stent with capability of expansion to adult size, hand-crimped on the desired balloon. Angiograms are performed to ensure precise placement. After stent implantation, final angiography and hemodynamic evaluation is performed, measuring arterial pressures in radial and femoral arteries to assess the result of the procedure. The introducer sheath is then removed, and the purse-string suture is tied down in the aortic incision. The remainder of planned surgical procedure (bidirectional Glenn or Fontan procedure) is then completed with standard technique.

18.8.3 Pulmonary Valve: Intraoperative Balloon Valvotomy

Given the long-term advantages of having a functional valve after TOF repair, there is increasing consensus around advocating for valve sparing techniques. Among these strategies, intraoperative

balloon valvuloplasty has shown variable results in short a mid-term follow-up periods [46–48]. Risk factors for worse outcomes with this approach include age younger than 3 months, annular *Z*-scores less than −2.45, dysplastic and thickened valves, and suboptimal balloon stretching.

After trans-infundibular and trans-pulmonary incisions, Hegar dilators are used to measure the annular diameter, aiming to get a final diameter increase of 120–140% or a *Z*-score of zero. A limited sharp commissurotomy is performed before antegrade balloon dilation, to ensure that the radial transmission of stress completes the division of the fused/incomplete commissures, preserving the intrinsic support and function of the leaflets and the valve annulus.

18.8.4 Pulmonary Arteries and Pulmonary Veins: Intraoperative Stent Implantation

Intraoperative stenting of pulmonary arteries or pulmonary veins can be considered in the following circumstances: (1) Percutaneous stent delivery is difficult or limited (lack of vascular access, or inability to deliver a stent with adult size potential). (2) Cardiac surgery is being undertaken for another reason. (3) "Rescue" of patients with vascular complications after prior procedures, whether transcatheter or surgical.

Previously implanted stents are also subject to be redilated via a hybrid approach during concomitant surgery.

Anatomic definition of the lesion may be done via noninvasive imaging or angiography to demonstrate an area of obstruction in a region where surgical patch repair would be challenging. The stent and balloon sizes are selected based on measurements made during diagnostic imaging.

The stents are placed into the pulmonary arteries through either the main pulmonary artery (Fig. 18.7) or right ventricular outflow tract (RVOT), using a purse-string suture (where a sheath is installed under fluoroscopic guidance before going on CPB, on the beating heart). Intraoperative angiography may be performed, and to optimize the angiographic imaging, the opposing branch PA can be transiently clamped during the injection.

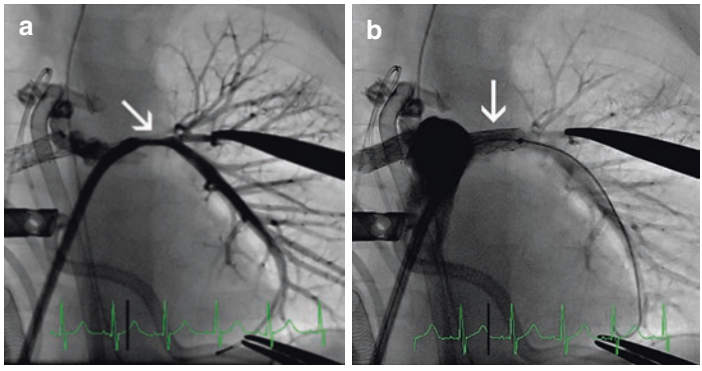

Fig. 18.7 Hybrid branch pulmonary artery stent implantation early postoperatively. (**a**) Main pulmonary artery angiogram performed via direct MPA hybrid access in a patient with Alagille's syndrome and bilateral severe branch PA stenosis. There is severe LPA stenosis (white arrow). (**b**) Angiogram following stent implantation demonstrates significant improvement in vessel diameter (white arrow). Note that bilateral hybrid stents have been implanted, which helped this patient come off ECMO support postoperatively

The stents can also be deployed under direct visualization by the surgeon while the patient is on cardiopulmonary bypass (CPB). During normothermic CPB, the stent can be placed in the pulmonary artery branch while the heart is electrically fibrillating. Commonly, a small incision is made in the right ventricular outflow tract or main pulmonary artery (or conduit), and small dilators are placed into the pulmonary artery to enlarge the lumen, so that a stent delivery catheter could be inserted. Furthermore, use of dilators helps to provide the surgeon with a sense of where the stent should be situated. The balloon catheters are placed without the use of a wire. The balloons are inflated to the manufacturer's recommended pressure for approximately 30 s. Placement under direct vision allows to place the stents in a manner such that they do not obstruct access to the contralateral pulmonary artery. Usually, the surgeon places the stent across the stenotic area, and the cardiologist selects the stent and inflates the balloon. Stents that are not believed to be in the optimal location can be easily removed.

For the pulmonary veins ballooning or stenting, a guide wire is introduced in the narrow pulmonary vein lumen and then a balloon (cutting balloons may be utilized) is introduced and dilated under direct vision. This will disrupt the hypertrophic intimal layer and show the surgeon which layer to excise. An extensive pulmonary venous "endarterectomy" extending into the intraparenchymal pulmonary veins can be performed or a re-expandable stent can be inflated in the vein. If a stent is placed in the pulmonary vein, it is prudent to place a few stitches securing it to the wall, to prevent embolization of the stent into the atrium and subsequently into the systemic circulation [49].

18.8.5 Intraoperative Transcatheter Closure of Blalock–Taussig Shunts or Major Aorto-pulmonary Collateral Artery

In patients with shunt-dependent pulmonary perfusion, or with significant aortopulmonary collaterals, in whom presurgical closure is not tolerated well due to profound desaturation, a combined single-hybrid approach is desirable [50, 51]. Moreover, surgical ligation of those vessels/shunts is often very difficult given their distal location, leaving the endovascular occlusion as an optimal approach to prevent excessive pulmonary blood flow during the procedure. The interventional cardiologist selects the suitable type of device (coils or vascular plugs) to be used depending on anatomical location, size, and length of the shunt or collateral. Likewise, depending on the location, vascular access is chosen centrally in the MPA or the ascending aorta or more distally in a branch artery. The intervention is performed under fluoroscopy or TEE guidance. Once the shunt is occluded, cardiopulmonary bypass is initiated, and the surgical procedure performed.

18.8.6 Intraoperative Delivery of Aortic Valve Prosthesis

The use of sutureless, rapid deployment valves is an alternative to surgical biological valve implantation in isolated or combined

aortic valve replacement in patients with aortic annulus size between 19 and 27 mm [52].

The usual steps for surgical aortic valve replacement are still required: sternotomy or minimally invasive surgical access, heparinization, extracorporeal circulation, aortic cross-clamping, myocardial protection and opening of ascending aorta, resection of diseased valves, and decalcification of the aortic annulus. Decalcification of the aortic annulus should be performed completely to avoid paravalvular leakage; however, extensive decalcification is not recommended due to the risk of annular ruptures and bleeding. Sizing is a crucial step in the management of sutureless and rapid deployment valves to avoid paravalvular leaks, central aortic regurgitation, dislodgement, and valve migration as well as annular rupture. Therefore, oversizing is neither necessary nor recommended. This strategy may lower the surgical risks by reducing the cross-clamp and extracorporeal circulation times [53, 54].

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Part III

Fetal Procedures

Fetal Interventions

19

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and Simone Fontes Pedra

19.1 Introduction and Clinical Scenarios

With evolving catheter and imaging technologies and better patient selection, fetal interventions have become an important therapeutic modality in last 20 years for some complex congenital heart diseases (CHD). These include aortic stenosis (AS) and evolving hypoplastic left heart syndrome (HLHS), HLHS with intact or highly restrictive interatrial septum (IAS), and pulmonary atresia (PA) or critical pulmonary stenosis (CPS) with intact ventricular septum (IVS) and evolving hypoplastic right heart syndrome (HRHS). In these clinical scenarios, a prenatal intervention may remodel cardiac morphology and function and result in improved pre- and postnatal outcomes, including an increased

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likelihood of achieving a biventricular (BV) circulation [1–3]. In this chapter, we review the indications, techniques, and current results of fetal cardiac interventions.

19.2 Anatomy, Physiopathology, Indications for Interventions, and Patient Selection

1. Critical AS and evolving HLHS [1–3]: AS is determined by echocardiographic visualization of a thickened, immobile aortic valve with turbulent, or decreased color Doppler flow. The Doppler-derived gradient should not be used to select patients because of frequently associated left ventricular (LV) dysfunction and endocardial fibroelastosis (EFE). Ideally, the LV diastolic length should have a *Z*-score >−2 at the time of diagnosis. Occasionally, we perform aortic valvuloplasty in smaller LVs (LV diastolic length *Z*-score between −2 and −3) not only with the hope to avert LV hypoplasia but also to ameliorate LV function and promote antegrade flow across the aortic valve. Evolving HLHS is diagnosed based on functional parameters such as reversed blood flow in the transverse aortic arch (TAA), left-to-right flow across the IAS, monophasic mitral valve (MV) inflow, and moderate-to-severe LV dysfunction in mid-gestation. Fetal aortic valvuloplasty should ideally be performed under 30 weeks gestational age (Fig. 19.1).
2. HLHS and intact or highly restrictive IAS [1–3]: A prenatal echocardiographic diagnosis of HLHS with either an intact IAS or a tiny (≤1 mm) atrial septal defect (ASD) or patent foramen ovale (PFO) and prominent flow reversal in the pulmonary veins should be made. Fetal atrial septostomy and ASD creation are ideally performed between 29 and 32 weeks gestation in order to endure until term (Fig. 19.2).
3. PA/IVS or CPS/IVS and evolving HRHS [1–3]: Patients should have a prenatal echocardiographic diagnosis of PA/IVS or CPS/IVS with the following features: membranous pulmonary atresia, with identifiable pulmonary valve (PV) leaflets or membrane, no or minimal systolic opening, and no or minimal color Doppler ultrasound flow across the pulmonary valve (PV); an intact ventricular septum; left-to-right shunting across

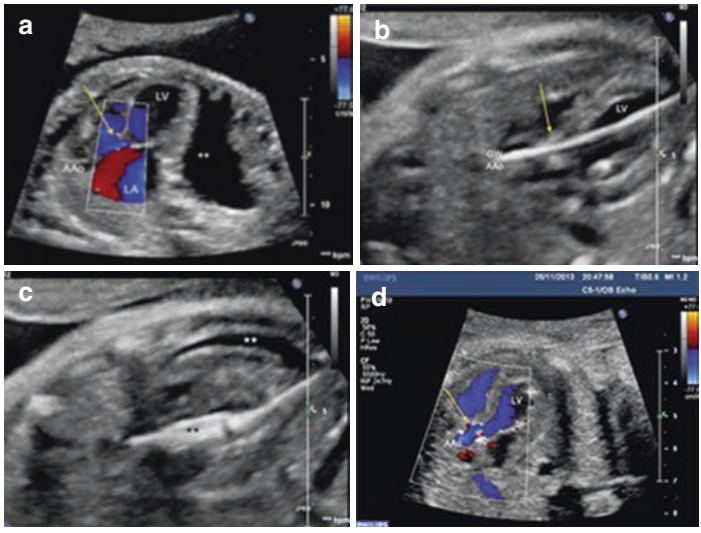

Fig. 19.1 Critical aortic stenosis in a 26 weeks gestational age hydropic fetus. (**a**) Pre-intervention echocardiographic assessment. Color flow mapping across the aortic valve (indicated by an *arrow*) showing tiny forward flow (*blue color*). The *red color* displays the flow into the right ventricle across the tricuspid valve. The LV is conspicuously dilated and dysfunctional. The fetus is in severe heart failure resulting in hydrops. *Asterisks* show ascites. (**b**) The guidewire is clearly seen in the ascending aorta, which proves the aortic valve (indicated by an *arrow*) was crossed successfully. (**c**) The balloon (*black asterisks*) is inflated across the aortic valve. There is some pericardial effusion (*white asterisks*) that required drainage after dilation. (**d**) Immediate results after aortic valvuloplasty. There is a significant improvement in antegrade flow across the aortic valve (indicated by an *arrow*) as shown by color flow mapping (*blue color* depicts a much wider vena contracta across the aortic valve). *LA* left atrium, *LV* left ventricle, *AAo* ascending aorta, *GW* guidewire

a patent ductus arteriosus (PDA); and right heart hypoplasia, with a tricuspid valve (TV) annulus *Z*-score ≤2 and an identifiable but qualitatively small right ventricle (RV) with no evidence of RV growth after 2–4 weeks of serial echocardiographic evaluation. Cases with fetal diagnosis of major coronary-to-RV fistulas should be excluded. Pulmonary valvuloplasty is performed between 24 and 30 weeks gestation (Fig. 19.3).

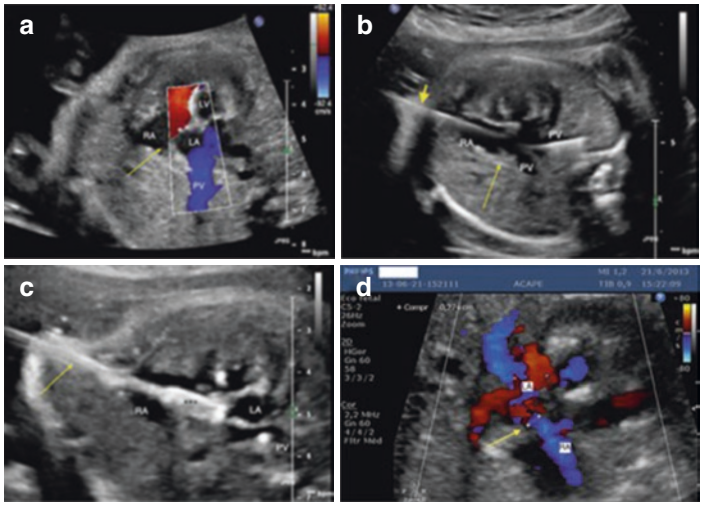

Fig. 19.2 Fetal atrial septostomy in a 29 weeks gestational age fetus with established hypoplastic left heart syndrome (**a**). Pre-intervention echocardiographic assessment. The LA is conspicuously dilated and the interatrial septum (indicated by an *arrow*) is almost intact. There is flow reversal to the pulmonary veins by color flow mapping (*blue color*). The *red color* displays the flow into the right ventricle across the tricuspid valve. The LV is hypoplastic, and diffuse endocardial fibroelastosis can be seen as bright hyperechogenic areas. (**b**) The interatrial septum (indicated by a *long and narrow arrow*) was traversed with the Chiba needle (indicated by a *short and broad arrow*), and the guidewire is seen in a pulmonary vein. (**c**) A 4 × 10-mm coronary balloon (marked with *black asterisks*) is inflated across the interatrial septum up to the burst pressure reaching 4.7 mm in diameter. Note that the Chiba needle (indicated by an *arrow*) is perpendicular to the plane of the interatrial septum. (**d**) Post-intervention echocardiographic assessment on the following day after fetal atrial septostomy. A 2.8-mm atrial septal defect was created within the atrial septum (indicated by an *arrow*). *LA* left atrium, *RA* right atrium, *PV* pulmonary vein, *LV* left ventricle

4. Critical AS, massive mitral regurgitation (MR), giant left atrium (LA), and hydrops [1, 2]: These fetuses have normal-sized LV and reversed flow in the TAA. Aortic valvuloplasty and atrial septostomy should be considered between 30 and 34 weeks gestation as a "salvage" procedure to diminish the risk of fetal loss due to conspicuous hydrops associated with pulmonary veins and right ventricular compression.

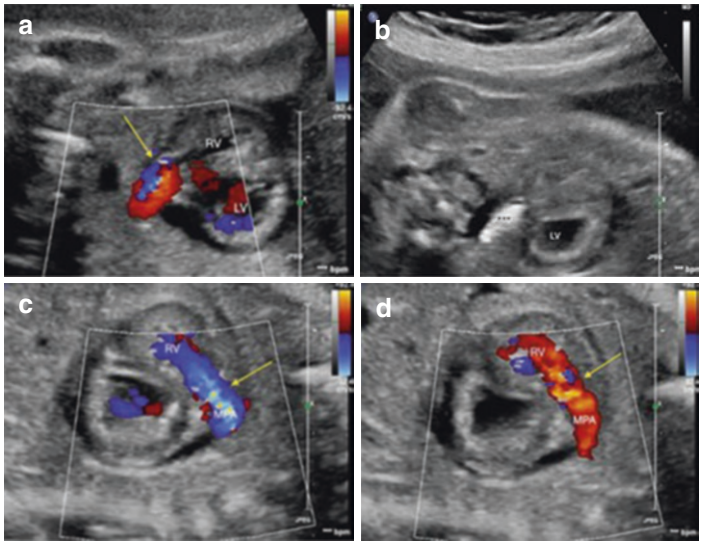

Fig. 19.3 Critical pulmonary valve stenosis in a 28 weeks gestational age fetus. (**a**) Pre-intervention echocardiographic assessment. Color flow mapping across the pulmonary valve (indicated by an *arrow*) shows tiny forward flow (in *blue color*). The *red color* depicts the retrograde flow across the ductus. The pulmonary valve annulus measured 4.0 mm. (**b**) A 4 × 10-mm coronary balloon (marked with *black asterisks*) is inflated across the pulmonary valve up to the burst pressure reaching 4.7 mm in diameter. (**c**) Immediate results after pulmonary valve dilation. Color flow mapping across the pulmonary valve (indicated by an *arrow*) shows significant improvement in forward flow across the valve (in *blue color*). (**d**) Color flow mapping across the pulmonary valve (indicated by an *arrow*) shows significant pulmonary insufficiency (in *red color*), which is a marker of effective dilatation. *RV* right ventricle, *LV* left ventricle, *MPA* main pulmonary artery

19.3 Pre-procedural Imaging and Planning

Fetal cardiac interventions should be performed by a multidisciplinary team. The fetal cardiologist is responsible for patient selection and pre- and post-procedural echocardiographic assessment. The fetal medicine specialist conducts fetal positioning and anesthesia and simultaneously controls the puncture needle and

the ultrasound probe. The interventionalists (usually two) handle the catheters and wires while the fetal medicine specialist holds onto the needle to keep its position during the procedure.

19.4 Technique (Step-by-Step), Materials, and Tips and Tricks

We perform such interventions under maternal conscious sedation and regional spinal blockade conducted by an anesthesiologist [1, 2]. An appropriate fetal lie is achieved by external version. Maternal positioning is kept with left uterine displacement. To promote uterine relaxation mothers are given nifedipine 20 mg TID for 48–72 h, starting 12–24 h before the procedure. An occasional large polyhydramnios is evacuated using a 15-cm-long 21-G Chiba needle (Cook Inc, Bloomington, IN, USA). If ideal fetal positioning cannot be attained by external manipulation, the procedure should be abandoned. We do not perform any interventions through a maternal abdominal wall incision and uterus exposure. After optimal fetal position is achieved, the fetus is anesthetized using a mixture of fentanyl (5–10 μg/kg), pancuronium (10–20 μg/kg), and atropine (20 μg/kg) given intramuscularly or in the umbilical cord using a 21–22-G Chiba needle [1, 2].

Cardiac access is attained through direct needle puncture of the fetal heart via the uterus and the fetal chest wall (Fig. 19.4). Under continuous two-dimensional ultrasound guidance, a 15-cm-long 17–18-G Chiba needle (with a stylet) is advanced to the target fetal cardiac chamber (LV, RV, or right atrium). The imaging plane is carefully adjusted to yield a picture in which both the entire needle length and the target cardiac chamber were included in the field of view (Fig. 19.4). A pre-marked system (a rapid exchange 10-mm-long coronary balloon premounted over a cutoff 0.014″ floppy tip guidewire) is advanced to the desired location. The needle, guidewire, and balloon shafts are premeasured and marked so that positioning within the fetal heart is known from external measurements rather than the ultrasound imaging alone. The balloon shaft is marked with sterile tapes so that no more than the full length of the balloon is extruded out of the Chiba needle tip when fully advanced. The wire is also fixed with sterile tapes

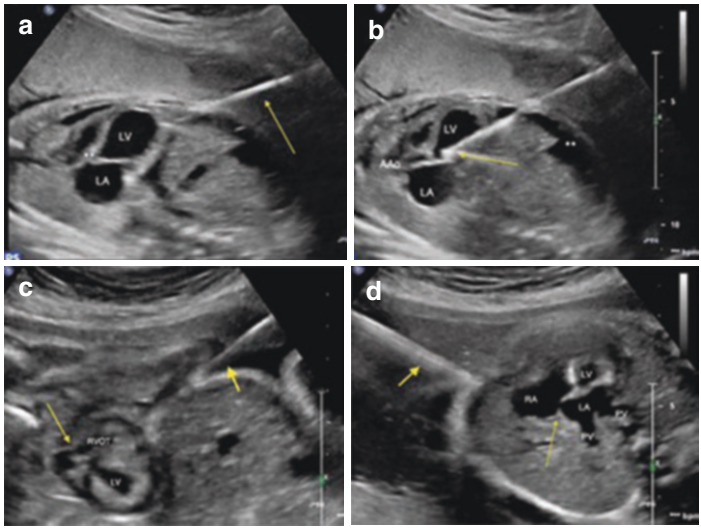

Fig. 19.4 Proper needle course, angulation, and positioning during the fetal cardiac procedures. (**a**) During fetal aortic valvuloplasty the tip of the needle (indicated by an *arrow*) is aimed at the aortic valve (marked with *asterisks*) in an imaginary line across the left ventricular apex toward the left ventricular outflow tract. (**b**) After perforation of the left ventricular apex, the tip of the needle (indicated by an *arrow*) is parked below the aortic valve so that the valve can be crossed with minimal manipulation. The *asterisks* indicate ascites in this hydropic fetus. The LA is conspicuously dilated (**c**). In fetal pulmonary valvuloplasty, the tip of the needle (indicated by a *short and broad arrow*) is aimed at the pulmonary valve (indicated by a *long and narrow arrow*) in an imaginary line across the right ventricular apex toward the right ventricular outflow tract. (**d**) During fetal atrial septostomy, the needle (indicated by a *short and broad arrow*) should course in a perpendicular angle toward the plane of the interatrial septum (indicated by a *long and narrow arrow*). *LV* left ventricle, *LA* left atrium, *RV* right ventricle, *RA* right atrium, *AAo* ascending aorta, *RVOT* right ventricular outflow tract, *PV* pulmonary vein

so that no more than 3–4 cm of the distal flexible wire straight tip extruded out from the balloon tip.

The LV or the RV is entered at the apex, with the needle course parallel to the outflow track directed at the stenotic/atretic semilunar valves (Fig. 19.4). In this way, the valves can be crossed

almost blindly, with minimal wire and catheter manipulation. For PV perforation, the same needle that was used for apex entry is advanced through the atretic PV. Occasionally, a transplacentary and/or subcostal transhepatic needle course is required to reach the desired location depending on the placenta and fetal positions. After stylet removal, the catheter system is introduced and advanced until the shaft mark reaches the proximal hub of the needle. Balloon positioning for inflation is based on the external aforementioned measurements and ultrasound imaging, with emphasis given to the visualization of the guidewire in the ascending aorta (for critical AS) or in the right pulmonary artery or descending aorta through the PDA (for PA/IVS) or in the left atrium (LA) or one of the dilated pulmonary veins (for atrial septoplasty). Balloons are inflated with pressure gauges to allow precise estimates of inflation diameters. Balloon diameters 10–30% larger than the aortic or pulmonary valve annulus are selected for valve dilation (Figs. 19.1 and 19.3). Two to four inflations are performed depending on the fetal clinical status.

For atrial septostomy, a 17-G Chiba needle with a greater internal lumen diameter is used in order to accommodate the profile of larger dilating balloons (the largest possible; usually 4 mm, expandable to 4.7 mm). Although we have not attempted to implant stents in the IAS, this may be achieved using special catheters specifically designed by the Boston group for fetal interventions [3]. The 17-G Chiba needle is advanced through the right atrium (RA) in a perpendicular course toward the IAS (Figs. 19.2 and 19.4). The same needle is used to perforate the IAS to gain access to the LA (Fig. 19.2). Once the tip of the needle is seen in the body of the dilated LA, the pre-marked system is advanced until the tape mark on the catheter balloon shaft reaches the proximal hub of the Chiba needle. At this point, the whole system is brought back as a unit until the balloon straddles the IAS. The balloon is inflated with enough pressure to achieve the maximum balloon diameter under the bursting pressure limit. A second puncture within the IAS is performed using similar techniques if the newly created ASD is judged to be too small to relieve left atrial hypertension. We have not performed stent implantation to the IAS because we favor the hybrid approach after birth.

After the valves or the IAS is dilated, the whole system (needle + balloon + wire) is withdrawn as a unit through the fetal cardiac wall and out of the fetal and maternal bodies to avoid shearing off the balloon from the catheter shaft. Small-volume-unit doses of epinephrine (1–10 μg/kg) and atropine are available for immediate fetal intracardiac injection to treat hemodynamic instability due to significant and persistent fetal bradycardia (<80–100 bpm for 3–5 min). Also a new 21–22-G Chiba needle should be readily available for pericardial drainage in case of tamponade (Fig. 19.1).

19.5 Pitfalls and Complications

Significant morbidity to the mothers is rare. On the other hand, fetal hemodynamic instability due to fetal bradycardia and hemopericardium is a common complication, especially in procedures that involve ventricular access. Fetal loss may happen, and although it is more commonly associated with hemodynamic instability and hemopericardium, other contributing factors such as fetal and maternal anesthetic issues and mechanical stimuli may also play a role. Premature labor may ensue as in any other fetal intervention.

19.6 How to Manage Complications

Given the high frequency of fetal bradycardia and significant hemopericardium, prophylactic atropine administration during fetal anesthesia, intracardiac therapeutic injection of epinephrine and atropine, and prompt pericardial drainage should be considered part of the standard of care in such interventions.

19.7 Post-procedural Care and Follow-Up

After the procedure, mothers are hospitalized overnight. The fetuses are assessed by ultrasound later on the same day and/or the following day before planned maternal discharge.

Echocardiography is performed at intervals determined by the primary fetal cardiologist.

It is recommended that these mothers give birth at the referral institution with a fully developed neonatal cardiology program. Although these fetuses may be delivered transvaginally, we believe that a C-section poses less stress on such fragile patients. They should be immediately transferred to the neonatal intensive care unit and started on a prostaglandin drip.

19.8 Expected Immediate Results and Postnatal Outcomes

A technically successful aortic or pulmonary valvuloplasty is defined as one in which a balloon is inflated across the valve, with unequivocal evidence of antegrade flow and/or new aortic/pulmonary regurgitation (AR or PR) as assessed by color Doppler echocardiography (Figs. 19.1 and 19.3). We have considered post-procedural AR and PI as a marker of effective dilatation of the aortic and pulmonary valves (Fig. 19.3). AR is well tolerated due to the low systemic vascular resistance determined by the placental circulation and the high-end diastolic left ventricular pressure and improves significantly or disappears until birth. A technically successful atrial septoplasty is defined as one in which there was unequivocal echocardiographic evidence of a newly created ASD at the conclusion of the intervention or on the following day (Fig. 19.2) associated with reduction in LA size and improvement in the pulmonary vein Doppler pattern (Fig. 19.5). The ASD size is determined by measuring the width of the color jet (vena contracta) (Fig. 19.4).

In general, a neonatal BV circulation is achieved in about 30–50% of fetuses who had undergone in utero aortic valvuloplasty. Usually these patients have a LV long-axis *Z*-score >0, a LV short-axis *Z*-score >0, an aortic annulus *Z*-score >3.5, a MV annulus *Z*-score >2, and a high-pressure LV defined by the presence of MR or AS with a maximum systolic gradient of ≥20 mmHg and milder degrees of EFE. Fetuses that have smaller LVs may also benefit from the procedure due to improved coronary flow

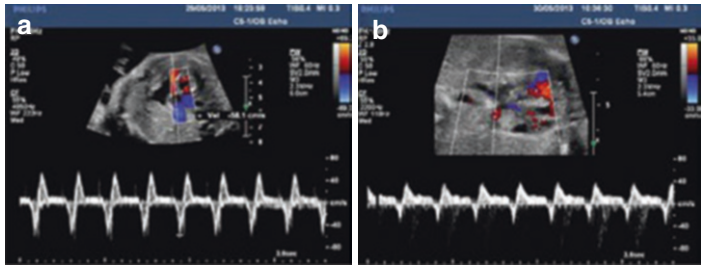

Fig. 19.5 Doppler tracing of the pulmonary veins pre- and post-fetal atrial septostomy in a fetus with hypoplastic left heart syndrome and almost intact interatrial septum. (**a**) Pre-intervention assessment. Bidirectional flow in the pulmonary vein with high reversal flow velocity (58 cm/s) during atrial contractions (negative wave below the baseline). (**b**) Post-intervention assessment on the following day. Triphasic flow pattern in the pulmonary veins (better diastolic filling) with improvement of the reversal flow velocity (less than 40 cm/s)

and preservation of myocardial function, which may have a positive impact on neonatal outcomes, regardless of the surgical strategy (Norwood vs. Hybrid) [1, 2]. In addition, promoting forward flow across the aortic valve in utero may theoretically help to minimize the neurodevelopmental abnormalities secondary to retrograde TAA perfusion. Moreover, progressive growth of the left heart structures during fetal life and over infancy resulting in an eventual BV repair has been observed our experience [1, 2] (Fig. 19.6). We have employed a staged strategy for such patients with fetal aortic valvuloplasty followed by a neonatal hybrid procedure ± balloon aortic valvuloplasty ± ASD enlargement. This approach works as a bridge to LV overhaul and BV repair later in infancy. Although postnatal LV diastolic dysfunction may be an issue in these patients, we still think that this is a lesser evil than the immediate and long-term morbidity and mortality of a univentricular pathway [1]. From January 2008 to September 2020, we have performed 41 procedures in 40 fetus with critical AS at a mean gestational age of 27.4 ± 2.7 weeks and with a mean weight of 1.2 ± 0.4 kg. Eleven had already milder degrees of LV hypoplasia and intervention. Technical success was achieved in 39/41 procedures and fetal demise occurred in three fetuses. Significant

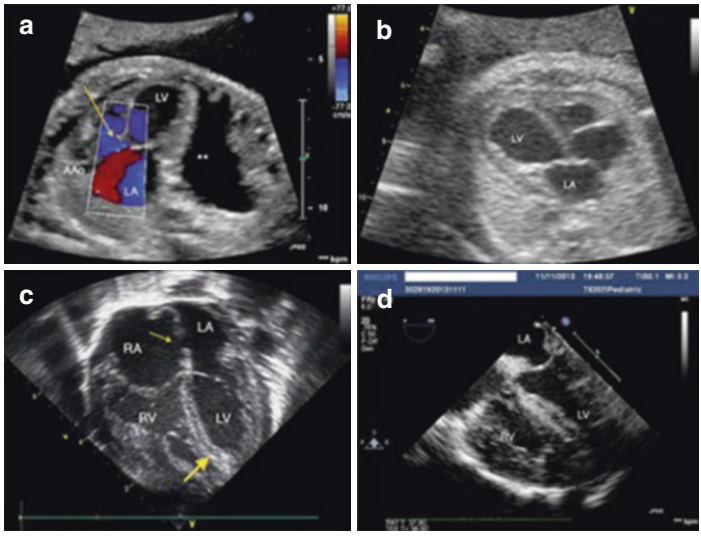

Fig. 19.6 Staged rehabilitation of the left ventricle in a fetus with critical aortic stenosis (same patient as in Fig. 19.1). (**a**) Pre-fetal intervention echocardiographic assessment at 29 weeks gestational age. Color flow mapping across the aortic valve (indicated by an *arrow*) showing tiny forward flow (*blue color*). The *red color* displays the flow into the right ventricle across the tricuspid valve. The LV is conspicuously dilated and dysfunctional. The LA is gigantic due to severe mitral regurgitation. The fetus is in severe heart failure resulting in hydrops. *Asterisks* show ascites. (**b**) Significant improvement of the LA size and heart failure at 32 weeks gestational age. No hydrops is seen. (**c**) Neonatal transthoracic echocardiogram after neonatal atrial septostomy, balloon dilation of the aortic valve followed by a hybrid procedure. Four chamber view. The LV is of borderline size (LV length *Z*-score −2.7) and still displays some endocardial fibroelastosis (indicated by a *broad and short arrow*). There is a 4-mm atrial septal defect indicated by a *long and narrow arrow*. (**d**) Intraoperative transesophageal echocardiogram performed after surgical left ventricular overhaul at the age of 9 months. After resection of the endocardial fibroelastosis layer, there is a significant improvement in the LV size (LV length *Z*-score −1.2). *RA* right atrium, *LA* left atrium, *RV* right ventricle, *LV* left ventricle, *AAo* ascending aorta

pericardial effusion requiring drainage and/or bradycardia was observed in 28 fetuses. Fetuses who survived were born at term after a programmed C section. Employing the staged strategy described above, we observed a ~50% survival rate (total of

18/37) at 1 year of age with most patients achieving a BV circulation (16/37; 6/10 of those with mild in utero LV hypoplasia; two babies are still in the interstage period). However, one patient developed severe diastolic dysfunction and pulmonary hypertension after the third year of age and is being considered for heart transplant.

Fetuses with critical AS, severe MR, and gigantic LA have in general a worse prognosis due to hydrops, fetal loss, or prematurity. However, in our experience, we observed a 66% survival rate (6/9) with all patients achieving a BV circulation and normal LV function after an initial hybrid procedure and LV overhaul at 9–12 months [2].

Fetuses with HLHS who undergo in utero ASD creation or enlargement are born with higher saturations and a more stable clinical initial course. However, surgical mortality after the Norwood operation remains higher than in HLHS patients who did not require in utero ASD interventions [3]. Whether the procedure performed in late gestation is efficacious in terms of preventing the development of secondary pulmonary vascular and parenchymal changes is debatable. From 2005 to 2020, we have created one or more ASDs using the aforementioned technique in ten fetuses with HLHS at a median GA of 32 weeks. All procedures were successful and babies were born at term. Emergent postnatal septostomy was required in seven patients. None of the patients survived after neonatal Hybrid or Norwood-Glenn at 6 months. Stent implantation to the IAS may result in larger ASDs in utero and better postnatal outcomes according to the Toronto and Boston experiences.

In utero pulmonary valvuloplasty for PA/IVS or CPS/IVS is more challenging from the technical standpoint due to the heavily trabeculated RV and a smaller RV cavity, which may be associated with a significant failure rate, especially at the beginning of the learning curve. Despite that, it seems that fetuses who undergo a successful intervention show a significant growth of the right ventricular structures from mid-gestation to late gestation when compared with control fetuses who did not undergo prenatal intervention and had univentricular outcomes after birth. From 2007 to 2020, we have performed 35 procedures in 32 fetuses with PA/

IVS ($n = 20$) or CPS/IVS ($n = 12$) at a mean age of 26.8 ± 2.3 weeks and a mean weight of 1.1 ± 0.3 kg. We had four failed attempts in fetuses with PA (first case and the remaining three at earlier GA <25 weeks). We had two deaths related to the procedure (main pulmonary artery perforation), two deaths in utero 1–2 weeks after the index valvuloplasty due to circular shunt in the setting of associated severe tricuspid regurgitation, and two additional neonatal deaths due to premature complications in other centers. The remaining 26 fetuses were born at term after a programmed C section. All underwent neonatal pulmonary valvuloplasty with most ($n = 20$) requiring ductal stenting [2]. At a mean follow-up of 36 ± 15 months, 21 showed significant growth of the RV structures achieving an eventual BV circulation (two required catheter closure of the ductal stent; the remaining showed spontaneous closure; ASD closure was required in four due to lower saturations). One and a half circulation was observed in four patients and one had a UV status during follow-up. We believe that RV rehabilitation in fetuses with PA/IVS or CPS/IVS and a hypoplastic RV is a continuous process that should be initiated in utero (the sooner the better), optimized in the neonatal period with repeat valvuloplasty ± ductal stenting and completed in the first 1–3 years of life, allowing time for the RV to catch up.

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Part IV

Step-By-Step Procedures: Valve Dilatation

Aortic Valvular Stenosis

20

Xiangbin Pan

20.1 Anatomic Description and Physiopathologic Factors

The normal aortic valve is trifoliate. The normal function of the valve depends on the well-developed aortic annulus and the proper relationship among the leaflets within the aortic root.

The aortic annulus of the patient with aortic stenosis (AS) is usually hypoplastic to some extent; the leaflets are thickened, and the commissures, to different degrees, are fused.

The anatomic types of AS include unicuspid, bicuspid, tricuspid, quadricuspid, and undifferentiated aortic valves.

Most AS is the bicuspid type, accounting for about 1–2% of cases worldwide and 67% of congenital AS. There are two types of bicuspid aortic valve: balanced (anatomically bicuspid) and unbalanced (functionally bicuspid). Two equal-sized cusps, combined with two Valsalva sinuses, form the anatomically bicuspid valve. However, the functionally bicuspid valve has three sinuses despite the open bicuspid, two of which exist next to a fused cusp

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formed basically by two different-sized cusps through an unopened commissure. The fused cusp is called an unbalanced bicuspid valve because it is larger than the valve opposite.

Tricuspid valves are seen in 25% of infants and about 40% of older patients who require treatment. Three leaflets vary in size. Most right coronary valves are hypoplastic; the leaflets are thickened and curled, and the commissures are fused. Unicuspid valves are often seen in newborns and present in about 10% of infants and 3% of the older children in whom treatment is indicated. The leaflets are thickened and fused; the valve has only one junction, and most of them lie at the one side. The rest lie at the center; the activity of the leaflets is limited, and, sometimes, the aortic annulus is hypoplastic.

AS leads to an obstruction in left ventricular ejection due to increased systolic blood pressure, prolonged ejection time, increased blood pressure, and decreased diastolic aortic pressure established as a transvalvular gradient. Initially, AS causes left ventricular pressure overload. Over time, the left ventricle adapts to the systolic pressure overload through a hypertrophic process that results in increased left ventricular wall thickness without dilatation of the left ventricular chamber (concentric hypertrophy).

Another compensatory mechanism is a lengthening of the ejection time at the expense of the diastole duration. Due to the adaptation, normal systolic left ventricular function is maintained. As hypertrophy progresses, the left ventricle becomes less compliant, left ventricular end-diastolic pressure increases, and left ventricular diastolic function decreases. Elevated end-diastolic pressure and shortened diastole duration limit coronary flow. Physical exercise or tachycardia can produce a maldistribution of coronary blood flow and subendocardial ischemia. Acute myocardial ischemia during exercise may cause ventricular arrhythmias and syncope or sudden death. On the other hand, increases in systolic blood, ventricular mass, and ejection time lead to increased consumption of oxygen by the myocardium. The increase in oxygen consumption and myocardial ischemia cause further deterioration of left ventricular function.

20.2 Clinical Scenarios

20.2.1 Critical Aortic Stenosis in Newborn

Newborns with critical AS suffer from low cardiac output and shock secondary to poor left ventricular function. These patients present with symptoms of heart failure such as pallor, tachypnea, tachycardia, air bubbles, and hepatomegaly. Usually newborns with critical AS need PGE infusion and intubation, and even some patents need urgent procedure.

Some newborns experience left ventricular endocardial fibroelastosis and fibrosis of the papillary muscles with mitral insufficiency due to subendocardial ischemia. Outcome is usually fatal in most of these patients with critical AS within the first weeks of life with medical treatment alone. Percutaneous balloon aortic valvuloplasty can be considered the first-line treatment for newborns with critical AS.

20.2.2 Aortic Stenosis in Older Children and Adolescents

Most patients with mild to moderate stenosis are usually asymptomatic, and the disease is diagnosed by a murmur. However, disease progression with symptom onset is common. In older children and adolescents with severe AS, the main symptoms are angina chest pain, syncope and dyspnea, or other symptoms of heart failure such as orthopnea, paroxysmal nocturnal dyspnea, and pedal edema. In the natural course of the disease, sudden death occurs in over 70% of patients with severe AS.

20.3 Indications and Patient Selection

The normal aortic valve area is about 2.0 cm^2/m^2. AS is considered mild when the area is less than 0.8 cm^2/m^2, moderate when the area is 0.5–0.8 cm^2/m^2, and severe when the area is less than 0.5 cm^2/m^2.

According to the peak systolic gradient, the degree of severity is considered mild when the gradient is less than 50 mmHg, moderate with a gradient of 50–79 mmHg, and severe with a gradient of 80 mmHg or higher.

Patients with mild AS rarely need treatment. However, AS may be progressive, and patients with mild disease may require treatment later in life. Usually, severe AS is an indication for treatment. When symptoms of syncope or heart failure develop, the prognosis changes dramatically. Therefore, regardless of the gradient in patients with these symptoms, treatment also is indicated.

20.4 Treatment Options

Neonates with critical AS and low cardiac output require resuscitation and administration of prostaglandin E_1. Establishing patency of the ductus arteriosus can restore adequate systemic blood flow and perfusion of vital organs. These patients should be sedated and intubated before balloon valvuloplasty is performed. Patients with significant aortic valve insufficiency in combination with mild to moderate stenosis may be carefully treated with afterload reduction, diuretic therapy, or both, although hypotension may occur. Inotropic drugs such as dopamine, dobutamine, and epinephrine may be indicated in cases of reduced cardiac output and decreased left ventricular systolic function. In critical AS, drugs that cause significant vasodilation should be avoided, because they may cause significant hypotension in the presence of a small aortic valve area. Patients with difficulty breathing and pulmonary edema benefit from intubation, positive pressure ventilation, and diuretic therapy.

Percutaneous balloon valvuloplasty was first described in 1983. With the improvement of catheter technology, it has become standard in patients with severe AS and can be safely performed with minimal morbidity. However, in patients with severely dysplastic valves and significant aortic regurgitation, balloon valvuloplasty is not the best choice. Surgical valvotomy is now rarely used except for in more complex valves, where simple balloon dilation is not sufficient. The Ross Procedure is another surgical

option that may be particularly beneficial for young children. The prosthetic aortic valve replacement is primarily reserved for patients in whom balloon valvuloplasty or surgical valvotomy has failed and significant aortic valve insufficiency has developed in association with left ventricular dilation or deterioration of left ventricular systolic function.

20.5 Preprocedure Imaging

Preprocedural echocardiography is the most important data. It can provide the following information: the morphology of the aortic valve, peak instantaneous and mean aortic valve gradient by Doppler, aortic valve annulus diameter and *z*-score, left ventricular dimensions, left ventricular shortening fraction and ejection fraction, and severity of aortic valve regurgitation, which are needed for a valvuloplasty indication. It should be noticed that the degree of aortic valve gradient may be underestimated because of low left ventricular ejection fraction. In addition, other lesions such as atrial septal defect, ventricular septal defect, aortic coarctation, mitral valve disease, and Shone complex can be identified by echocardiography. These lesions may influence the strategy of the operation, for example, the degree of aortic valve gradient may be overestimated because of patent ductus arteriosus (PDA) shunt; it is better to deal with PDA firstly for patients with mild AS.

20.6 Step-by-Step Technique

1. Preoperative preparation: Generally, to ensure adequate oxygen supply, cardiac catheterizations are conducted under general anesthesia to avoid restlessness and bleeding in infants or patients in poor clinical condition. Simultaneously, an external defibrillator and cardiopulmonary resuscitative drugs must be prepared to manage ventricular fibrillation or cardiac arrest during valve dilation due to severely impaired cardiac output. Soon after establishing vascular access, patients are given heparin, 100 IU/kg of body weight.

2. Establish vascular access: Wires and catheters can pass through the aortic valve from either the aorta (retrograde approach) or the left ventricle (antegrade approach). The retrograde approach is the most common method used for puncturing femoral artery. However, vascular complications may arise at the site of entry in the femoral artery in particular in newborns. The umbilical artery or surgically exposed right common carotid artery (or right axillary artery in newborns) is used. Occasionally, the guidewire cannot cross a severely stenotic aortic valve from the retrograde approach to the left ventricle. The antegrade approach is an alternative way by using a femoral venous access or an umbilical vein. Despite the advantage of reducing the risk of femoral arterial injury and aortic valve leaflet perforation, the antegrade approach can injure the mitral valve.

   A 4-Fr sheath is usually needed in newborns. In older children and adolescents, the access size depends upon the balloon to be chosen.
3. Hemodynamic assessments: Hemodynamic assessments include measurement of pressure gradients across the aortic valve and aortic angiography. By simultaneously obtaining pressure in the left ventricle and ascending aorta, the pressure gradient across the aortic valve is measured most accurate. Alternatively, distal pressure can be measured using a cannula in the radial artery.

   Generally, an aortic angiography is performed using a pigtail or side hole catheter placed just above the aortic sinuses. The details of the valve anatomy and assessing the aortic annulus diameter between the hinge points of the valve leaflets were defined by the 40° left anterior oblique and straight posterior-anterior projection (Fig. 20.1). Aortography is not performed in patients with hemodynamically unstable states such as in cases with hypotension and poor left ventricular function. Transthoracic echocardiography is used to observe aortic valve morphology, quantify the degree of AS, and measure the aortic annulus diameter (Fig. 20.2).

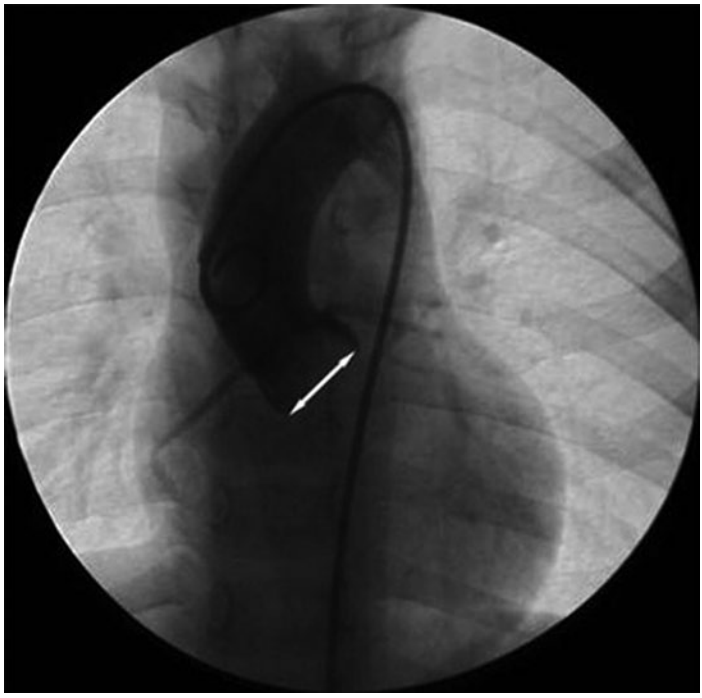

Fig. 20.1 The aortic valve annulus is measured by aortography in posterior-anterior projection. The double-headed arrow indicates the aortic annulus diameter between the hinge points of the valve leaflets

4. Select the appropriate balloon: Balloon valvuloplasty-induced aortic regurgitation can be the result of commissural avulsion and cusp tear or perforation. It can make further surgical reinterventions more difficult. Oversized balloons are a risk factor for aortic regurgitation. The balloon-to-annulus ratio should be less than 1:1. Start with a balloon diameter of about 80% of the aortic annulus and increase its size by 1 mm.

   Concerning length, an ideal balloon length should allow safe straddling of the aortic valve without overlapping the mitral valve chordae. We often choose a 20-mm balloon length in newborns and 30 mm in older children and 40 mm in adolescents.

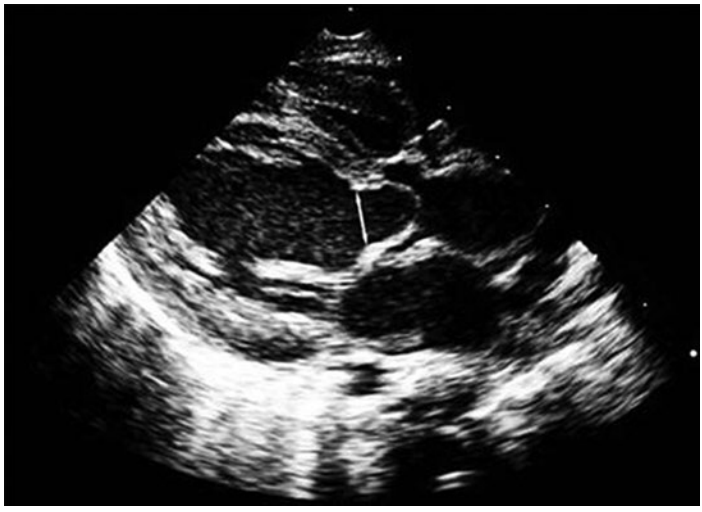

Fig. 20.2 Echocardiographic measurement of the aortic valve annulus between the hinge points of the valve leaflets (double-headed arrows) in the parasternal long-axis view

5. Techniques of valvuloplasty: The techniques of valvuloplasty include single- and double-balloon valvuloplasty.

   (a) Single-balloon valvuloplasty technique: With the help of an angiographic catheter (Judkins right, multipurpose catheter), cross the stenotic aortic valve by using a hydrophilic guidewire (0.014″ standard coronary wires in neonates, 0.018″ hydrophilic J-tipped wires in small kids, 0.035″ J-tipped wires in older kids and adolescents). Special care should be paid to avoid any force applied by the wire over the aortic cusps. Another important point is to be sure that you are not in the coronary artery with the wire.

   The angiographic flow jet on the ascending aortogram may be used as a guide.

   Exchange the hydrophilic wire for another wire (0.014″ coronary wire in newborns, 0.035″ standard guidewire in children, J-tipped 0.035″ extra stiff guidewire in adolescents) that will be used for the angioplasty. The wire is

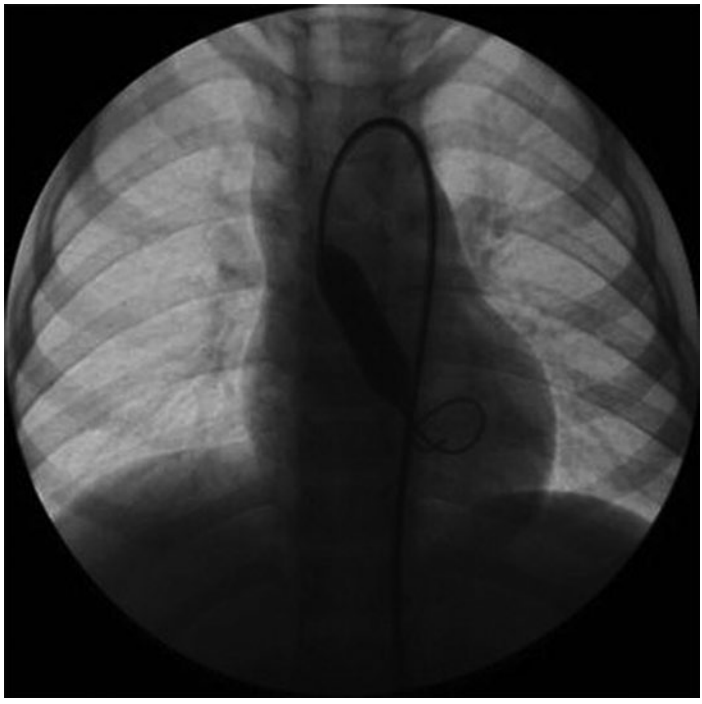

Fig. 20.3 Single-balloon technique in posterior-anterior projections

placed in the LV apex along the septum, anterior to the mitral valve chordal apparatus.

Advance a balloon valvotomy catheter over the guidewire, straddle the valve into the correct position, and inflate with a pressure of 4–7 ATM until the balloon waist disappears (Fig. 20.3). In newborns hand inflation is enough. Each inflation-deflation period lasts no more than 5–10 s. To preserve balloon stability across the aortic valve during inflation, a technique called temporary rapid pacing arrests mechanical systole to decrease the chance of balloon migration. The technique is performed by putting a bipolar pacing catheter in the right ventricular apex and using VVI pacing at a rate of 220–240 impulses per min-

ute during the balloon inflation. Ultimately, such pacing can increase the success rate of the procedure.

(b) Double-balloon valvuloplasty technique: Except for the use of two separate arterial catheters to cross the aortic valve retrograde, the double-balloon technique is identical to the single-balloon approach. Each balloon has a similar diameter and length so the ratio of the sum to the valve annulus diameter is about 1:3. The two balloons are positioned similarly across the aortic valve and inflated simultaneously (Fig. 20.4). Studies [1] have suggested that the double-balloon valvuloplasty technique provides improved gradient relief by producing deeper tears in the lines of commissural fusion than can be obtained with a single bal-

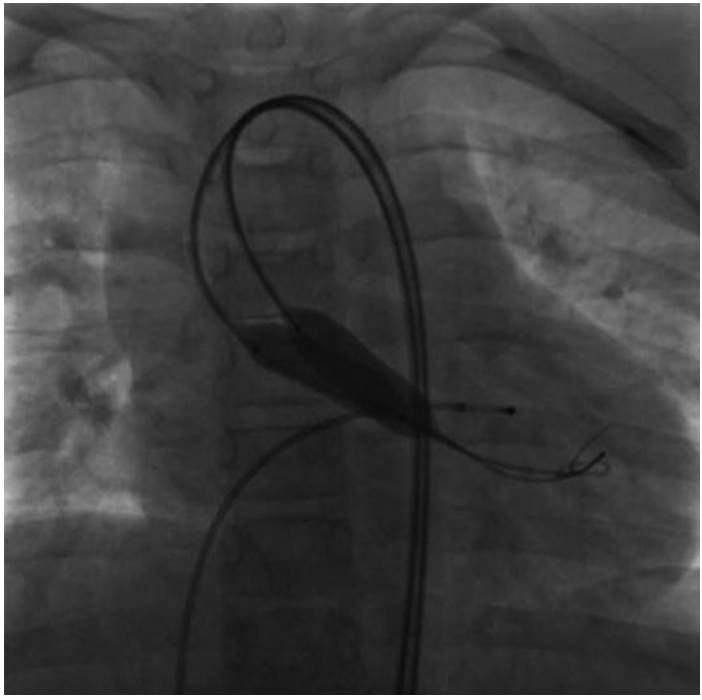

Fig. 20.4 Double-balloon technique in posterior-anterior projections

loon. In addition, two smaller balloons can be introduced to and removed from the femoral arteries more easily and, presumably, with less vessel trauma than a single large balloon. Third, the double-balloon technique extends the range of annulus sizes amenable to balloon dilation. It allows effective dilation of an aortic annulus ≤31 mm in diameter. Finally, inflation of two smaller balloons side by side has a smaller risk of completely occluding left ventricular outflow than inflation of a single larger balloon.

6. Postoperative evaluation: Hemodynamic assessment is conducted again using angiography and echocardiography. The ideal surgical result is effective relief of stenosis (residual gradient <30 mmHg) without moderate to severe aortic regurgitation.

20.7 Materials

Different balloon and guidewire products are available:

1. Tyshak and Tyshak II (NuMED, Inc.) are often used in aortic valvuloplasty. Except for the balloon diameters of 21 and 24 mm, they provide a wide range of balloon diameters, from 4 to 30 mm, with 1-mm increments up to 25 mm. Because it has a thicker shaft and wire, Tyshak can better resist the left ventricular ejection power and is therefore preferred in older children. On the other hand, Tyshak II is preferable in infants in whom the lowest possible introducer profile is important. Tyshak Mini usually applies to neonates; it provides balloon diameters between 4 and 10 mm and needs introducer sizes of only 3–4 Fr. But it can only be guided by 0.014-in. wires.
2. VACS II is manufactured in Germany by Osypka. It has balloon diameters of 4–30 mm, with 1-mm increments up to 18 mm, except for 11 and 13 mm. Above 18 mm, the increments are 2 mm. It is a low-profile balloon that may have an advantage in infants.
3. The size of the wire depends on the inner lumen of the balloon catheter. Coronary guidewires can be useful for Tyshak Mini balloons in neonates. The Terumo™ coronary wires with

floppy and soft tips can be recommended. When the inner lumen of the balloon catheter is between 0.018 and 0.028″, the 0.018″ SV 5™ Straight Wire (Cordis Company), with its short floppy tip, gives excellent support to the balloon. If a 0.035-in. guidewire fits into the inner lumen of the balloon catheter, the 0.035-in. Teflon-coated THSF wire from Cook Company can be recommended.

20.8 Expected Results

Generally, the criteria for a successful valvuloplasty are a more than 50% decrease in pressure gradient across the aortic valve, an increase of more than 25% in the aortic valve area, and no significant aortic regurgitation. A residual gradient of <30 mmHg is usually aimed.

20.9 Suggestions for Procedural Success

1. Operation approach selection: The retrograde and antegrade approaches are the two main techniques, with the retrograde approach being the most common. However, the retrograde approach may lead to difficulty in the guidewire's ability to cross a severely stenotic valve, and vascular complications may arise at the site of entry of the femoral artery. The antegrade approach using transseptal antegrade access to the aortic valve reduces the risk of arterial complications and more easily facilitates the passing of the guidewire through critical stenosis. If, after 20–30 min, the valve has not been successfully crossed or femoral arteries are tortuous or small, the antegrade approach should be considered.
2. The biggest challenge of valvuloplasty is passing wires through AS uneventfully. The commissure between the left and the noncoronary cusps is the most common area of opening of the valve, and the guidewire-catheter system should be pointed in that direction in left anterior oblique view. Another method is that when the catheter is within the turbulent flow coming out

of the aortic valve it thrills (vibrates), we can keep the catheter in that position and try with the wire.
3. Using the right coronary catheter to pass the guidewire through the aortic valve can be helpful. When the aortic valve cannot be crossed using the retrograde approach and injury of the mitral valve is a concern, a combined approach of passing through the aortic valve using the antegrade approach and snaring the wire in the aorta should be considered.
4. Displacement of the balloon before it is fully inflated leads to poor valve dilation and may injure the valve leaflets or surrounding tissues. To minimize and prevent balloon movement during dilation, we choose rigid shaft and wire and adopt the double-balloon and rapid right ventricular pacing technique.

20.10 Pitfalls

Occasionally, a guidewire in the left coronary artery might look like it is in the left ventricle, particularly on posteroanterior projection. The guidewire should be confirmed before advancing the catheter in the posteroanterior and lateral positions. The guidewire should be advanced gently and pulled into the catheter to avoid injuring the valve or damaging the coronary arteries. Balloon rupture may lead to air embolism. We usually use a one-third contrast and two-thirds normal saline solution to carefully remove all air from the balloon. It is important to prevent balloon movement and avoid reaching the rated burst pressures of valvuloplasty balloons.

20.11 Complications

Balloon aortic valvuloplasty has been shown to be efficacious in terms of gradient relief and lack of aortic regurgitation. However, a dilated balloon may completely block the blood flow in the aorta, prevent blood ejection of the left ventricle, and induce fatal ventricular fibrillation, left ventricular systolic dysfunction, and asystole. The early mortality rate is about 4%, and the complica-

tion rate appears to be related to age of the patient and type of lesion. Neonates have a higher rate of complications and worse midterm outcomes than older children after balloon aortic valvuloplasty. Aortic regurgitation is a potentially serious complication. About 15% of patients experience moderate or severe aortic regurgitation after balloon valvuloplasty. The incidence of vascular complications remains a concern after aortic balloon valvuloplasty, especially in newborns in which femoral artery access is used. In addition, transient bradycardia and left bundle-branch block, premature beats, mitral valve tears, and falls in systemic pressure during balloon inflation can occur.

20.12 Management of Complications

Many of these complications are now considered preventable with current catheterization technology and experience. The double-balloon valvuloplasty technique using smaller, less traumatic catheters reduces obstruction to the left ventricular outflow tract during balloon inflation. It can reduce the incidence of some kinds of arrhythmia during surgery. In addition to greater gradient relief, some reports show that it has lower aortic regurgitation than single-balloon valvuloplasty. In general, the retrograde approach is associated with the least number of complications. However, the guidewire should be advanced gently to prevent damage to the mitral valve.

To reduce complications of percutaneous balloon valvuloplasty, a hybrid balloon valvuloplasty through the ascending aorta via median sternotomy is performed in infants with severe congenital valvular AS [2] (Fig. 20.5). Its obvious advantages include ample size of the arterial sheath and balloon, effective dilation of the valve, no peripheral vascular complications, less exposure to radiation, cardiac compression under direct vision, and extracorporeal circulation, if needed. We recently improved the hybrid procedure by designing a new sheath (Fig. 20.6) specifically for interventional treatment using an ascending aorta approach. Compared with percutaneous arterial sheath, the new sheath can

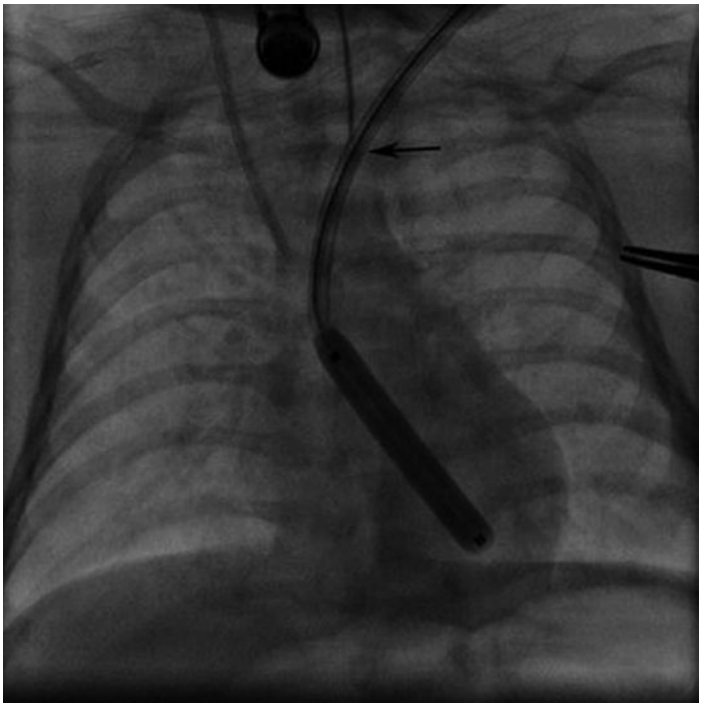

Fig. 20.5 Patients underwent tracheal intubation under general anesthesia on supine position in the hybrid operation room. After median sternotomy, the wire and balloon advanced into the aorta by an arterial sheath which was inserted into the aorta (the *arrow* pointed to the arterial sheath)

directly and quickly lead the guide wire to the aortic valve orifice thanks to an 120° angle elbow in head end, which greatly reduces time for guidewire attempts to pass through the aortic valve, thereby effectively avoiding damage to the aortic wall, aortic valve, and coronary artery. In addition, the sheath is rendered more stable by two holders, and cardiopulmonary bypass can be established quickly if required thanks to an adapter, thereby increasing reliability of patient's rescue. With the new sheath and increased treatment experience, we gradually achieved the goal of performing the procedure under echocardiography guidance with-

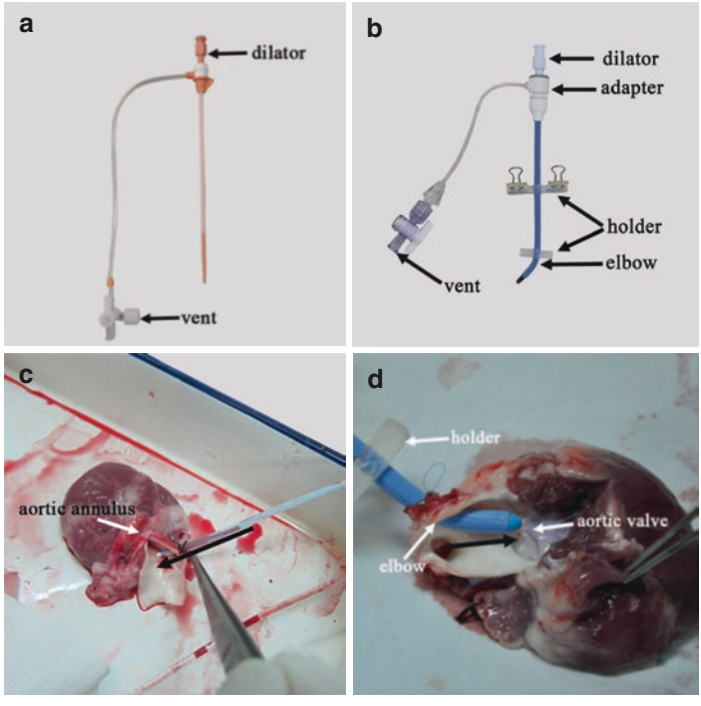

Fig. 20.6 Appearance of percutaneous (**a**) and new (**b**) arterial sheath. The new sheath has a 120° angle elbow in head end, two holders, an adapter, a dilator, and a vent for exhaust. In animal experiments, percutaneous arterial sheath was almost perpendicular to the ascending aorta (**c**, black arrow). It will lead the guide wire to aortic wall and then the wire turned to the direction of aortic valve. Unlike percutaneous sheath, the elbow of new sheath allowed to directly and quickly lead the guide wire to the aortic valve orifice (**d**, black arrow), which will reduce operation time and damage to the aortic wall

out fluoroscopy in an ordinary operating room. Echocardiography provides a more intuitive image than fluoroscopy and, more importantly, allows safe and effective treatment of patients with severe CAS in most hospitals without a hybrid operation room as is commonly the case in developing countries.

20.13 Postprocedure Care

Patients should be carefully transferred to the intensive care unit after aortic valvuloplasty. Patients who had an intraoperative antegrade approach angioplasty need to remain supine in bed for 12 h. Patients who had an intraoperative retrograde approach angioplasty need to remain supine in bed for 24 h. Changes in the patient's sense of well-being should be closely observed. Breath rate, blood pressure, and pulse should be measured once every 30 min, and body temperature should be measured once every 4 h. After 6 h, vital signs should be taken every hour; after 72 h, vital signs should be taken every 4 h. Patients should be strictly observed for signs of orthostatic hypotension and arrhythmia. Patients must avoid strenuous exercise for 72 h after surgery. To prevent limb necrosis caused by lengthy compression of the femoral artery or vein, the dorsalis pedis artery pulse should be taken regularly.

20.14 Follow-Up

Congenital AS is a lifelong disease, and the families of these patients should be told about the palliative nature of the procedure. All patients should be regularly followed using electrocardiography and echocardiography in 1, 3, 6 months, and each year after the procedure. Examination ergometry and Holter in older children and adolescents are helpful to judge the cardiac function and rhythm.

Percutaneous balloon aortic valvuloplasty is favored for its low procedural mortality, but it has high rates of reintervention (15–65%) in long-term follow-up. Aortic valve morphology and the diameter of the annulus have been associated with procedural success. Maskatia and colleagues [3] found that after initial percutaneous balloon valvuloplasty, 65% of patients were able to avoid repeat valvuloplasty, 61% avoided aortic valve replacement, and 87% avoided death or heart transplantation 15 years after initial percutaneous balloon valvuloplasty. Patients with postoperative gradients >25 mmHg or a lower baseline left ventricular shortening fraction experienced worse outcomes.

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Pulmonary Valve Stenosis

21

Tingliang Liu and Wei Gao

21.1 Anatomic Description and Physiopathology

The most common pathologic description of a stenotic pulmonary valve is a "dome-shaped" configuration of the pulmonary valve.

The fused pulmonary valve leaflets protrude from their attachment into the pulmonary artery as a conical, windsock-like structure.

The size of pulmonary valve orifice varies from a pinhole to several millimeters, most usually central in location, but can be eccentric.

Pulmonary valve ring hypoplasia and dysplastic pulmonary valves may be present in a small percentage of patients.

Pulmonary valve dysplasia is characterized by thickened, nodular, and redundant valve leaflets with minimal or no commissural fusion, valve ring hypoplasia, and lack of poststenotic dilatation of the pulmonary artery.

The obstruction is mainly related to thickened, myxomatous immobile pulmonary valve cusps and valve ring hypoplasia.

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21.2 Clinical Scenarios

Two clinical scenarios may occur: (1) critical pulmonary stenosis that occurs at birth and (2) pulmonary valve stenosis in infants, children, and adolescents.

1. Critical pulmonary valve stenosis
   Patients are cyanotic at birth and require prostaglandin infusion to maintain pulmonary blood flow. These subjects are usually in the intensive care unit and are often intubated.
2. Pulmonary valve stenosis beyond neonatal period
   These patients are mostly asymptomatic, unless severe stenosis is present. The lesion is detected because of a cardiac murmur on routine clinical examination. With growth, moderate stenosis may cause symptoms, including fatigue, chest pain, limited exercise tolerance, and mild cyanosis. Patients with severe stenosis may have different degrees of cyanosis and are predisposed to right heart failure.

21.3 Indication for Treatment and Treatment Options

1. Critical pulmonary valve stenosis
   This is an urgent procedure.
   Sometimes a low gradient associated to a right ventricular dysfunction may be found.
2. Pulmonary valve stenosis beyond neonatal period
   It is generally accepted that the indication for balloon pulmonary valvuloplasty is a pulmonary valve stenosis with an echocardiographic systolic peak instantaneous gradient of >60 mmHg (that correlates to a peak-to-peak invasive gradient ≥30–40 mmHg) or clinically significant pulmonary valvular obstruction in the presence of right ventricle dysfunction.
   It is reasonable to perform pulmonary valvuloplasty on a patient with pulmonary valve stenosis who meets the above

criteria in the setting of a mild to moderate dysplastic pulmonary valve.

It is suggested that balloon dilation should not be undertaken for pulmonary valve stenosis with severe right ventricular dysplasia, significant valve annulus hypoplasia, or severe pulmonary hypoplasia. In these cases, surgical operation should be the first choice.

21.4 Pre-procedural Imaging

Echocardiographic studies are most useful in the evaluation of pulmonary valve stenosis. Thickening, doming of pulmonary valve leaflets, and the poststenotic dilatation of the pulmonary artery can be well visualized by two-dimensional (2D) echocardiographic views.

The valve annulus can also be measured, and such measurements are very helpful in the selection of balloon diameter during balloon dilation.

In cases with hypoplastic or relatively hypoplastic RV, measurements and function of the tricuspid valve are needed. In fact in newborns with critical pulmonary valve stenosis and when the TR annulus is <-2 SD, stent implantation in the ductus arteriosus may be needed.

Qualitative and semiquantitative evaluation of the RV function is obtained.

Pulsed, continuous wave, and color Doppler evaluation can be used to confirm the diagnosis and the degree of obstruction. Using the modified Bernoulli equation, the peak instantaneous gradient can be calculated:

$$\Delta P = 4V^2$$

where ΔP is the pressure gradient and V is the peak Doppler flow velocity in the main pulmonary artery. However, the peak instantaneous Doppler gradient may overestimate the peak-to-peak catheter gradient, presumably related to a pressure recovery phenomenon.

21.5 Technique (Step-by-Step)

1. Candidates selection
   The diagnosis and assessment of pulmonary valve stenosis are made by the usual clinical, radiographic, and echocardiographic data. An informed consent is obtained from the parents or the patients.
2. Sedation and anesthesia
   The procedure is usually performed under sedation. General anesthesia with endotracheal ventilation is needed in newborns, while it can be avoided beyond the neonatal period.

   Heart rate, blood pressure, respiration, and pulse oximetry are continuously monitored throughout the procedure.
3. Vascular access
   The most preferred entry site for balloon dilation is the percutaneous femoral venous route. A sheath is inserted into the vein depending on the age and size of the patient (in newborns usually a 5-F sheath). In some high-risk patients an arterial line may be used to continuously monitor the arterial blood pressure. After sheath placement, heparin is given in the dose of 100 IU/kg, to keep the activated clotting time above 200 s.
4. Right ventricular angiography
   Hemodynamic assessment is done routinely, and the peak-to-peak catheter gradient across the pulmonary valve is assessed. Biplane right ventricular cineangiograms (posterior-anterior with cranial angulation and lateral views) are performed using Berman angiographic catheters or pigtail catheters to confirm the site of obstruction, to measure the pulmonary valve annulus, and to evaluate the function of right ventricle. The annulus is measured as the distance between the hinge points of the valve in both views in systole (Fig. 21.1).
5. Positioning of the guidewire
   An end-hole catheter is advanced across the pulmonary valve and the tip of the catheter positioned into the distal left (preferable) or right pulmonary artery.

   Usually a Judkins right coronary or a cobra catheter is used. To allow the catheter to enter the RV, usually a pre-shaped stiff

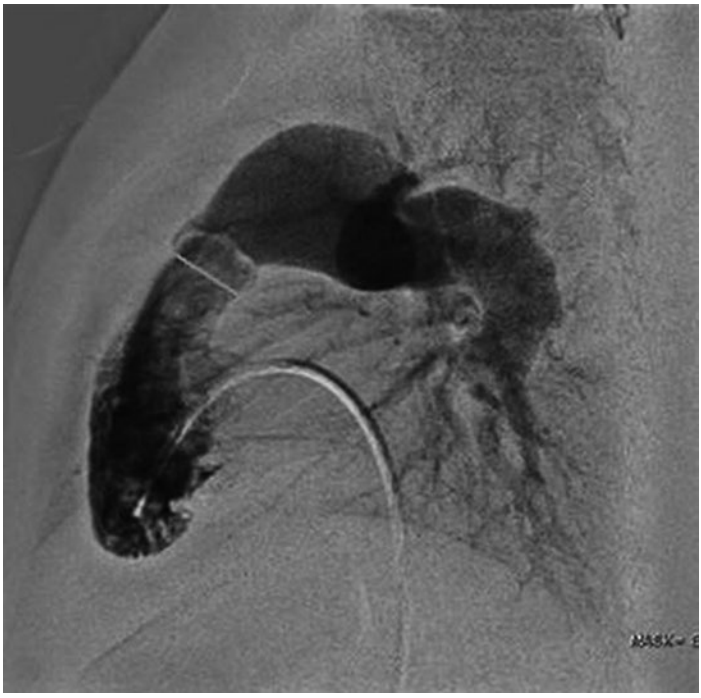

Fig. 21.1 Right ventriculogram in left lateral view showing pulmonary valve stenosis; thickened, domed pulmonary valve; and poststenotic dilatation of the main pulmonary artery. The white line shows the length of the pulmonary valve annulus

end of a straight standard guidewire is needed to guide the catheter across the tricuspid valve.

Alternatively a 4- or 5-F end-hole multipurpose catheter can be used. It is advanced up to the right atrium. Then a loop is created over the free wall of the right atrium with a gentle push. The next step is to rotate clockwise in order to direct the end of the catheter toward the tricuspid valve. Finally a gentle pull is applied in order to allow the catheter to slip in the right ventricle.

Then the catheter is oriented toward the RVOT with a clockwise rotation and a gentle pull. The catheter is passed through the valve.

Usually, this part of the procedure is not easy because of hypertrophy of the RV.

Floppy- and soft-tipped guidewires can be used to assist the catheter crossing of the pulmonary valve. Great attention is to be paid when using hydrophilic wires. In fact even a floppy-tipped wire can perforate.

In newborns a 0.014″ coronary wire or a 0.018″ hydrophilic wire can be used and positioned either distal in the pulmonary artery or in the descending aorta through the ductus arteriosus.

In older children 0.035″ wire can be chosen and placed distally in the pulmonary artery bed.

Wire position is crucial for the procedure!

A J-tipped, exchange length, stiff guidewire is advanced through the catheter already in place, and the catheter is removed. The selection of the exchange wire diameter is dependent upon the selected balloon catheter.

In newborns when the passage through the valve is a pinhole, predilation using a coronary balloon (diameter 2.5–3 mm; length 15–20 mm) may be needed.

6. Selection of balloon catheter
The inflated diameter of the balloon is selected in accordance with the diameter of the pulmonary valve annulus. Usually a balloon diameter 1.0–1.2 times the annulus is chosen. The length of the balloon should be 20 in newborns and infants, 30 mm in children, and 40 mm in adolescents and adults. Longer balloons may impinge upon the tricuspid valve, causing heart block or tricuspid valve regurgitation. An adequately sized femoral sheath must be inserted for the introduction of the balloon catheter.
7. Balloon dilation
The selected balloon angioplasty catheter is advanced over the guidewire and positioned across the pulmonary valve. The body landmarks, such as the ribs, sternum, or other fixed landmarks, are used for this purpose. A frozen video frame of the

right ventricular cineangiogram displayed on the screen is used as a road map.

The balloon is quickly inflated with diluted contrast material (1 in 4) (Fig. 21.2). The inflation pressure is gradually increased up to the manufacturer's recommendation (Fig. 21.3), and then the balloon is quickly deflated. The duration of inflation is kept as short as possible, usually just until after the waist disappears.

If the balloon is not appropriately centered across the pulmonary valve or moves during the inflation, the position of the catheter is readjusted and balloon inflation repeated. Usually a

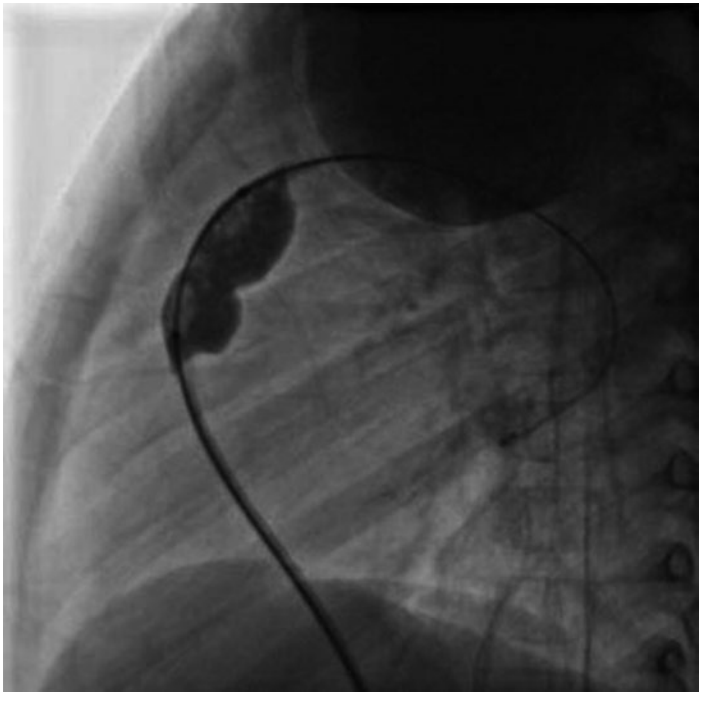

Fig. 21.2 Left lateral view of a partially inflated balloon catheter positioned across the pulmonary valve. As the balloon is inflated, a waist appears at the site of the pulmonary valve

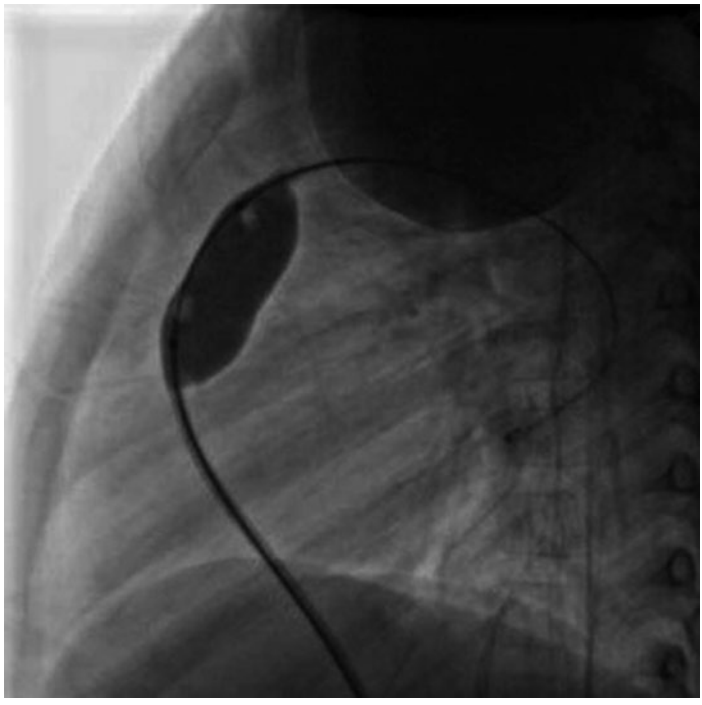

Fig. 21.3 Left lateral view of the balloon waist completely disappeared

gentle pull can be applied during full inflation in order to counteract the push forces of the right ventricle systole.

One additional balloon inflation may be performed after satisfactory balloon inflation has been achieved, to ensure adequate valvuloplasty.

8. Post-dilation protocol
   A multitrack catheter may be used to record the pressure gradients across the pulmonary valves in older patients.

   In newborns, a multipurpose catheter can be advanced over the 0.014″ guidewire as used for angiography by using a Y-connector.

   If the result is not satisfactory (peak-to-peak valve gradient usually in excess of 50 mmHg), it is important to understand

why this occurs: Is there any muscular infundibular reaction? Is there a residual valvular gradient? Is there a vascular hypoplasia and severe pulmonary valve dysplasia?

If the problem is related to residual valvular stenosis, then the balloon inflation is repeated until a satisfactory reduction in the gradient is detected (obviously do not exceed 120% balloon to annulus ratio).

In adolescents and adults, if a satisfactory balloon inflation is not achieved due to a small balloon to annulus ratio, then a double balloon technique may be considered.

Some patients with a dysplastic pulmonary valve cannot be treated by balloon dilation and are candidates for surgery.

A final RV angiography is usually performed.

Echocardiography can be used to evaluate the mobility of the pulmonary valve leaflets, to measure the gradient after dilation, to detect infundibular stenosis, and to discern any complications such as tricuspid insufficiency and pericardial effusion.

21.6 Materials (Balloon Angioplasty Catheters)

The catheterization lab armamentarium is ever changing and varies not only over time but also from institution to institution. A variety of balloon angioplasty catheters can be used for balloon dilation such as Tyshak-II, Opta Pro, Z-Med, Z-Med II, Powerflex, ev3, and Cristal, depending on the availability at a given institution. The following characteristics of the angioplasty balloon should be taken into consideration:

1. Sheath requirement
2. Balloon length. Long balloons can improve stability during inflation, but take longer to inflate and deflate fully, and may impinge upon the tricuspid valve, causing tricuspid insufficiency. Short balloons can cause less straightening of curved cardiac or vascular structures and thus may cause less injury at the tips of the balloon, but it may be difficult to maintain the balloon centered across the pulmonary valve annulus during inflation.

3. Balloon compliance (degree of balloon stretch at a certain pressure). Compliant balloons can be inflated easily and will apply less pressure to the stenotic segment. Noncompliant balloons can be inflated to a relatively high pressure and have a more predictable maximum inflation diameter compared to compliant balloons.

In neonates or infants we generally use 20-mm-long compliant balloons (e.g., Tyshak-II). It has a low profile; the deflated balloon can cross a 4–5-French sheath. The working pressure of this kind of balloon is about 3.5–4 ATM. In order to avoid balloon rupture, a gauge should be used to apply a specific amount of inflation pressure. Thirty- and 40-mm noncompliant balloons, such as Cristal angioplasty balloons, can be used in children and adolescents, respectively. The balloon diameter usually varies from 12 to 20 mm, the inflation pressure is up to 6–8 ATM, and 7–9 Fr sheaths can be used.

When the pulmonary valve annulus is too large to dilate with a single balloon (≥25 mm) or the patient's femoral vein is small, valvuloplasty with simultaneous inflation of two balloons across the pulmonary valve can be performed (Fig. 21.4). It may reduce the incidence of complications. Single balloon dilation can cause complete obstruction of the right ventricle and then cause systemic hypotension. However, during the double balloon procedure, the right ventricular output may continue in between the balloons and cause less hypotension. Though the double balloon technique involves an additional femoral venous puncture site, relatively small shaft sizes can be used. When two balloons are utilized, the following simplified formula may be used to calculate the effective balloon size:

$$Effective balloon diameter = 0.82\left(D_1 + D_2\right)$$

where D_1 and D_2 are the diameters of the balloons used. It is better to use balloons of the same size, because two same size balloons can be easily maintained at the site during inflation.

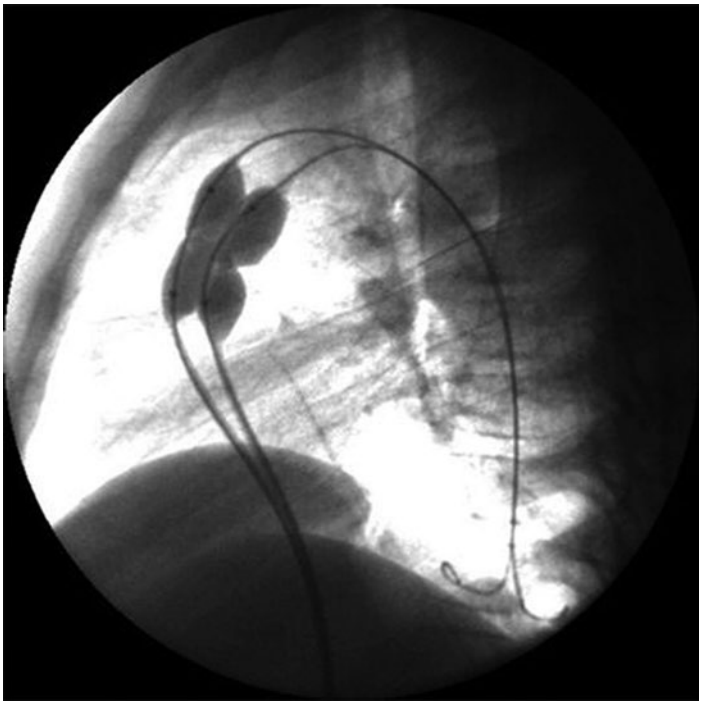

Fig. 21.4 Left lateral view of two balloon catheters positioned across the pulmonary valve showing waisting of balloons produced by the stenotic valve

21.7 Expected Results

The peak-to-peak invasive pressure gradient between the pulmonary artery and right ventricle should be reduced to less than 20–25 mmHg after balloon pulmonary valvuloplasty.

Short-term and long-term results of both balloon pulmonary valvuloplasty and surgery have low mortality rates and appear to be successful in relieving the acute transpulmonary gradient (peak-to-peak valve gradient <30 mmHg). In comparison to surgery, there is a relatively higher prevalence of restenosis after balloon dilation, and the re-intervention rate for pulmonary ste-

nosis is higher after balloon dilation. But the prevalence of pulmonary insufficiency and ventricular arrhythmias is higher after surgery.

21.8 Tips and Tricks

1. Even in mild or moderate pulmonary valve stenosis, very fast progression in infancy or early childhood has been shown. Patients with severe stenosis should undergo treatment even if they are clinically well. For children with moderate or severe pulmonary valve stenosis, it is better to do balloon pulmonary valvuloplasty early on.
2. Make sure that the guidewire does not pass through the tendinous chords and papillary muscles of the tricuspid valve before advancing the balloon catheter. Once the balloon catheter has been advanced across the chords and papillary muscles of the tricuspid valve, it can cause chordae rupture and severe tricuspid valve insufficiency. Berman wedge catheters can be used to cross the tricuspid valve, in order to avoid damage to the tricuspid valve.
3. The ratio of contrast to saline differs for each type of balloon. For small balloons less than 4 mm in diameter, the syringe should be highly concentrated with contrast, nearly 100%, so it can be easily visible by fluoroscopy. A typical mixture for a 5–10-mm balloon is one-third contrast and two-thirds saline, and for balloons > than 10 mm, 1 in 4 diluted contrast. A high concentration of contrast will increase the viscosity of the fluid and therefore the inflation and deflation times of the balloon.
4. At times it may be difficult to position an appropriately sized balloon angioplasty catheter across the severely stenotic pulmonary valve, especially in neonates. In such instances we use smaller, 2–4-mm-diameter balloon catheters initially to predilate then use larger, more appropriately sized balloon catheters.
5. If the balloon diameter is small compared to the pulmonary valve annulus, only a subtle waist is produced and the dilating force is small. The dilating force can be increased by increasing the inflation pressure, but this puts the balloon at risk for

rupture. Another choice is to use a larger balloon. If the balloon diameter is too large compared to the stenosis, the waist will be very tight, and the dilating force will be large. In this setting, the annulus is at greater risk of injury. So intermediate waists are the most effective and safest. Visualizing the waist and reacting appropriately is one of the most important skills in interventional cardiology. As a general rule, if the waist is less than 75% the diameter of the proximal and distal balloon, it is too tight; the balloon should not be fully inflated, and the dilation should be done with a smaller balloon.

21.9 Pitfalls

The balloon may not be truly across the pulmonary valve during balloon inflation. It is important to ensure that the balloon is indeed across the valve. The waisting of the balloon may be produced by supravalvular stenosis or infundibular constriction. When in doubt, centering the balloon at various locations across the right ventricular outflow region may become necessary.

21.10 Complications

Balloon pulmonary valvuloplasty is a safe and effective treatment of moderate and severe pulmonary valve stenosis. The complication rate of balloon pulmonary valvuloplasty is low based on a large number of reports. The presence of complications is common in neonates or infants with the most severe pulmonary valve stenosis.

Transient bradycardia, premature beats, hypoxia (in the presence of an atrial septal defect), and a fall in systemic pressure during balloon inflation have been reported. These abnormalities return rapidly to normal following balloon deflation.

Complete right bundle branch block, transient or permanent heart block, ventricular arrhythmia, femoral venous obstruction, injury of the tricuspid valve, pulmonary regurgitation, balloon rupture at high balloon inflation pressures, cardiac arrest, and

cardiac tamponade, though rare, have been reported. Some of these complications may be unavoidable. However, meticulous attention to the technique and the use of the appropriate diameter and length of the balloon and avoiding high balloon inflation pressures and short inflation/deflation cycles may prevent or reduce the complications.

Injury of the tricuspid valve is most frequently caused by strained passage of the catheter between the tendinous chords and papillary muscle of the tricuspid valve. It may also be associated with the use of very long balloons. Serious insufficiency of the tricuspid valve may even require plastic surgery.

The clinical significance of severe pulmonary regurgitation caused by balloon dilation may increase with age. Therefore these patients need cardiologic surveillance. Mild pulmonary insufficiency can be well tolerated by patients. Severe pulmonary insufficiency requires pulmonary valve replacement.

Contractile subvalvular stenosis of the right ventricular outflow tract has been reported. Though the obstruction of the pulmonary valve has been improved after the balloon pulmonary valvuloplasty, the reflexive subvalvular stenosis leads to increased right ventricular pressures. β-Blockers may be used (e.g., in young children: propanolol 1 mg/kg/day). The patients usually recover in 3–6 months.

Recurrent stenosis is rare; however, a second balloon pulmonary valvuloplasty may be indicated before surgery is scheduled. If the anatomic substrate (dysplastic valves without commissural fusion, supravalvular pulmonary artery stenosis, or severe fixed infundibular obstruction) is the problem, surgical treatment may become necessary.

21.11 Post-procedural Care

Newborns may need intensive care unit monitoring and PGE infusion for some days even after balloon angioplasty.

An electrocardiogram and an echocardiogram are performed following the procedure. Electrocardiographic and echocardiography

evaluation at 1, 6, and 12 months after the procedure and yearly thereafter is generally recommended.

Regression of right ventricular hypertrophy on the electrocardiogram following balloon dilatation has been well documented. Echocardiography plays an essential role in the follow-up of patients with pulmonary valve stenosis. The Doppler gradient is generally reflective of the residual obstruction and is a useful and reliable noninvasive monitoring tool.

21.12 Follow-Up

The short-, intermediate-, and long-term follow-up have demonstrated a high procedural success and low complication rate. The results of long-term observations confirm the usefulness and effectiveness of balloon valvuloplasty in the treatment of pulmonary valve stenosis, even in patients with dysplastic valves. Therefore, balloon pulmonary valvuloplasty can be considered as the treatment of choice for patients with pulmonary valve stenosis.

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22 Pulmonary Atresia and Intact Ventricular Septum

Marhisham Che Mood and Mazeni Alwi

22.1 Introduction

Pulmonary atresia with intact ventricular septum (PAIVS) is an uncommon defect characterized by complete obstruction of the right ventricular outflow which may be in the form of an imperforate membranous valve or muscular atresia of the infundibulum. It is usually associated with varying degrees of right ventricle (RV) cavitary hypoplasia due to muscles overgrowth. A peculiar association is the presence of RV-coronary connections, particularly in those with muscular atresia and severe RV hypoplasia. The coronary circulation may be RV-dependent in a small minority.

Transcatheter therapy with perforation of the atretic valve and balloon dilation is the preferred choice of management in those with membranous atresia. Patent ductus arteriosus (PDA) stenting may be performed concomitantly in patients who are likely to require additional pulmonary blood flow e.g. modified Blalock Taussig (BT) shunt on account of small RV cavity size.

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22.2 Anatomic Description

The principal anatomic lesion in PAIVS is complete obstruction of the RV outflow, which may be in the form of an imperforate membranous valve, seen in about 60–70%, or muscular atresia of the entire right ventricular outflow tract (RVOT, infundibulum). Although the designation suggests a simple solitary lesion, a major feature of the disease is the spectrum of RV cavitary hypoplasia that ranges from mild to very severe, with near obliteration of RV cavity by overgrowth of muscles (Fig. 22.1) [1]. However in the rare cases with associated severe Ebstein's anomaly, the RV (and RVOT) is thin-walled and markedly dilated (Fig. 22.1c). RV-coronary connections are a peculiar association in PAIVS, seen most often in those with diminutive RV cavity.

22.2.1 Major Anatomic Subtypes

1. **Imperforate membranous valve**
   In this group, the valve leaflets are completely fused, leaving an imperforate membrane. The infundibulum is usually well developed and smooth-walled, though occasionally abnormal muscle bundles may cause additional, fixed subvalve stenosis. The RV cavitary hypoplasia is often of mild to moderate degree, either with all three parts (inlet, infundibulum, and apical) present, i.e. "tripartite", or quite commonly the RV is "bipartite" where there is near-obliteration of the apical part by muscles while the remaining two are well developed (Fig. 22.1a, b). The atretic membranous valve is usually a thin membrane which has a mild or moderately hypoplastic annulus. In contrast, the pulmonary artery root and its sinuses are usually well developed, seen angiographically "cupping" over the membranous valve and infundibulum (Fig. 22.2a, b). The main pulmonary artery and its branches are also well developed. Occasionally, the valve is thick and immobile with the pulmonary artery root and sinuses being poorly developed (Fig. 22.2c, d). In a small minority, the RVOT is markedly

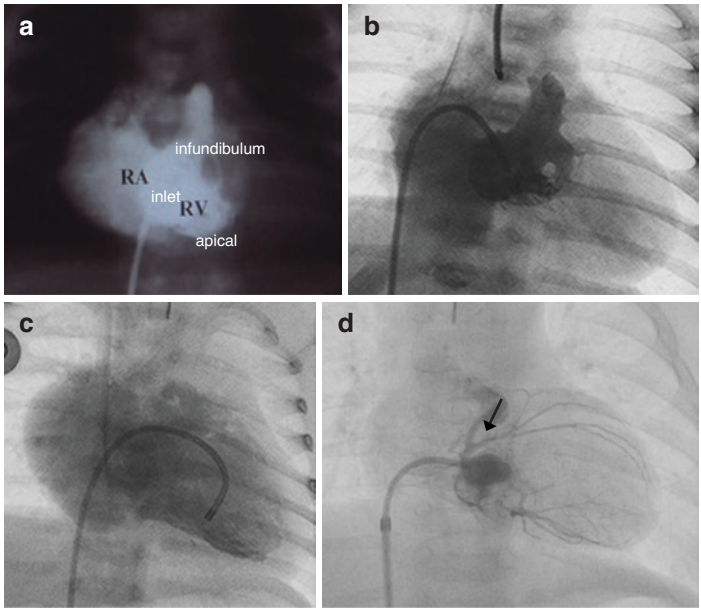


Fig. 22.1 Angiographic morphology of the RV in PAIVS. (**a**) Mild RV cavitary hypoplasia with well-developed infundibulum, inlet and apical parts, "tripartite" RV. There is moderate TR, and the RA is dilated. (**b**) "Bipartite" RV—the inlet and infundibulum are well developed, but the apical part is almost obliterated by muscles. There is also moderate TR. (**c**). PAIVS with severe TR from a dysplastic tricuspid valve. Markedly dilated and thin-walled RV and RA. (**d**). Severe hypoplasia of the RV cavity with only the inlet part present, i.e. "unipartite". RV-coronary connections to both the left and right coronary systems. Reflux of contrast into the aortic root (arrow). No TR. *RA* right atrium, *RV* right ventricle, *PAIVS* pulmonary atresia with intact ventricular septum, *TR* tricuspid regurgitation

dilated such that the usual tunnel like configuration is lost with the small valve and small annulus capping the dome-like infundibulum (Fig. 22.2d). This is usually seen in association with severe Ebstein's malformation of the tricuspid valve.

The presence of RV-coronary connections is less common in this group.

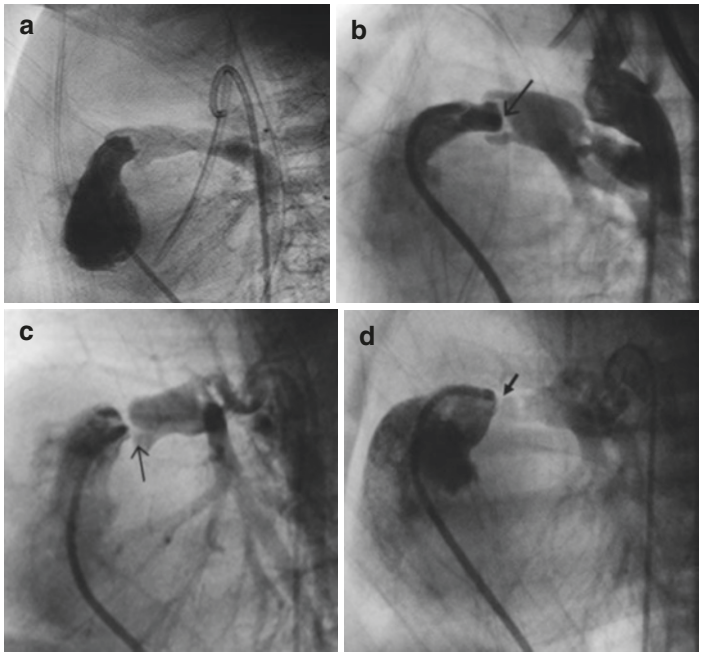


Fig. 22.2 The RVOT, valve and pulmonary artery. Simultaneous angiograms in the RVOT and descending aorta opposite the PDA are performed to outline these structures. (**a**) Well-developed infundibulum, thin membranous valve and mildly hypoplastic annulus. The main pulmonary artery and sinuses are well developed, "cupping" over the valve membrane and annulus. (**b**) Similar anatomy as in (**a**), but the valve membrane is "slightly" thicker (arrow). (**c**) Very thickened valve plate (arrow). The sinuses of the pulmonary artery root are less well developed. (**d**) Markedly dilated RVOT in a patient with severe TR from associated Ebstein's malformation of the TV. The pulmonary annulus is small. The pulmonary artery root and sinuses are poorly developed (arrow). *RVOT* right ventricular outflow tract, *PDA* patent ductus arteriosus, *TR* tricuspid regurgitation, *TV* tricuspid valve

The morphologic subtype illustrated by Fig. 22.1a, b is suited for valve perforation and balloon dilation, having the best chance of achieving 2 ventricle or 1½ ventricle circulation. It will be the focus of this chapter.

2. **Muscular atresia**
   Patients with complete or near-complete muscular atresia of the infundibulum generally have severely hypoplastic RV cavity where only a small inlet is present ("unipartite"), the rest obliterated by muscles. The RV is often markedly hypertensive, and RV-coronary connections are commonly present (Fig. 22.1d). In 5–10%, the coronary circulation is RV dependent because of atresia or severe stenoses of the proximal coronary arteries. This subgroup of PAIVS is managed as hearts with single-ventricle circulation as the RV is too small to function as an effective pulmonary pump.

The tricuspid valve is not uncommonly dysplastic with varying degrees of regurgitation. Severe Ebstein's malformation with markedly dilated, thin-walled right atrium (RA) and RV is a well-recognized association.

The PDA in PAIVS tends to resemble that of isolated PDA, arising from the distal arch and connecting onto the dome of the main pulmonary artery (MPA) away from the orifice of the proximal left pulmonary artery (LPA). Though they are usually elongated, they are seldom tortuous. However, as in other cyanotic heart disease especially those with muscular atresia, the PDA may have a more complex morphology and insert onto the proximal LPA causing stenosis of this branch.

22.3 Pathophysiology and Clinical Presentation

Because of complete RV outflow obstruction and major aortopulmonary collateral vessels being rarely present, the pulmonary circulation is duct-dependent. There is an obligatory right to left shunt through the patent foramen ovale (PFO); hence, cyanosis at birth—linked temporally to ductal constriction—is the commonest presentation. However, in the current era, diagnosis is often made antenatally during foetal cardiac screening. As in lesions with duct-dependent pulmonary blood flow, severe cyanosis, acidosis, and circulatory collapse may occur with rapid closure of the

PDA. The RV is markedly hypertensive especially when the tricuspid valve is competent. Marked RV hypertrophy and small RV cavity cause reduced RV compliance and may lead to persistence of right to left shunt even after RV pressure is normalized following balloon dilation. Severe tricuspid regurgitation may cause marked enlargement of the right atrium. In the presence of RV-dependent coronary circulation, ischaemia/infarction may occur when the RV is decompressed following pulmonary valvotomy (surgery) or balloon dilation.

22.4 Treatment Options, Indications and Patient Selection

Management strategy should be formulated according to the likelihood of 2 ventricle (or 1½ ventricle) circulation vs. single-ventricle physiology, and this is based on the RV anatomy at presentation [2].

Patients with definite muscular atresia of the infundibulum and diminutive RV should be directed towards the Fontan track at the outset, a largely surgery-based management beginning with the BT shunt at diagnosis. Catheter interventional therapy is limited to PDA stenting as alternative to surgical shunt and balloon atrial septostomy should be performed at the same time.

In patients with well-developed infundibulum where the atresia is limited to a membranous imperforate valve, the goal is to establish unobstructed antegrade flow into the pulmonary vascular bed, abolish RV hypertension, reduce tricuspid regurgitation, promote RV growth and, in the occasional patients to disrupt RV-coronary connections and restore normal coronary perfusion. The final objective is to achieve biventricular circulation or at least 1½ ventricle circulation in those whose RV fails to grow adequately.

Today catheter intervention with radiofrequency assisted valvotomy and balloon dilation (RFV-BD) is the preferred method of opening the valvar atresia and establishing antegrade flow to the pulmonary arteries. The use of a stiffer-tipped coronary guidewire

normally used for chronic total occlusion (CTO) in coronary revascularization therapy is a reasonable alternative to radiofrequency (RF) wire [3]. PDA stenting may be performed at the same time in patients whose RV is deemed small and likely to require augmentation of pulmonary blood flow following RFV-BD. Assignment of individual patients towards the single ventricle vs. 2 ventricle track is based on the initial echocardiographic assessment and cardiac catheterization.

22.5 Pre-procedure Imaging

Echocardiography provides detailed information for the initial planning of management. An important parameter in echocardiographic assessment of PAIVS is the size and morphology of the RV, whether the patient is a likely candidate for RFV-BD, i.e. a membranous atresia with well-developed infundibulum or one destined for the Fontan track, i.e. muscular atresia of the infundibulum and diminutive RV cavity.

TV *Z*-score and TV/MV annulus ratio provide a semi-quantitative measurements of RV size. Doppler echocardiography provides additional information on the degree of tricuspid regurgitation and estimate of the RV systolic pressure.

Finally, branch pulmonary artery size and confluence are assessed. More importantly, evaluation of the PDA morphology should be noted for purposes of PDA stenting if this forms part of the management. Large RV-coronary connections may be visible on colour Doppler, but its delineation can only be achieved with angiography.

Angiography provides a more precise information and is essential in the formulation of a management strategy. Accurate assessment of the nature of pulmonary atresia, the infundibulum, the pulmonary valve annulus and the PDA morphology is obtained as part of the interventional procedure. We recommend this to be performed in all patients, even those destined for the Fontan track particularly for a detailed evaluation of RV-coronary connections.

22.6 Technique, Materials, Tips and Tricks

1. Ideally, the patient should weigh >2.5 kg to prevent access-related complications. The procedure is done under general anaesthesia with ICU backup for post-procedure recovery. Biplane fluoroscopy is essential and echocardiography should be on stand-by in the event of suspected tamponade.
2. PGE1 infusion should be stopped 4–6 hours prior to the procedure to make the PDA more suitable if stenting is to be contemplated but that may not always be possible. Arterial and venous access is obtained with 4 F and 5 F sheath, respectively. If heparin administration is a routine practice, it is preferable to give this after RFV-BD. This may limit the effect of misperforation outside the heart. A 4 F pigtail catheter is placed in the aorta opposite the PDA for angiogram of the pulmonary artery and PDA.
3. Initial hemodynamic and diagnostic evaluation: Baseline aortic, RA and RV pressures and arterial blood gases are obtained. [**Tip**: Catheterizing the RV for the initial pressure recording, angiography and subsequent manipulation for valve perforation is often challenging because of the usually dilated RA from tricuspid regurgitation and the frequently small, shallow RV cavity. This may be facilitated by placing a 5 F Mullins' sheath (Cook Inc, Bloomingdale, IN 47404, USA) in the RA. The tip of this long sheath naturally faces tricuspid valve, enabling easy passage of catheter into the RV (Fig. 22.3a, b)]. We recommend a 4 or 5 F Judkins Right (JR) catheter for both the diagnostic and interventional purposes. As the RV cavity is usually small, hand injections would normally suffice. RV angiogram is performed in the antero-posterior (AP) and lateral projections to assess the size of the RV and morphology of the RVOT (Figs. 22.1 and 22.2). If RV-coronary connections are present, multiple plane RV angiograms should be performed to delineate them, together with aortic root angiography to exclude RV-dependent coronary circulation (RVDCC). If RVDCC is present or suspected, valvotomy and balloon dilation is contraindicated.

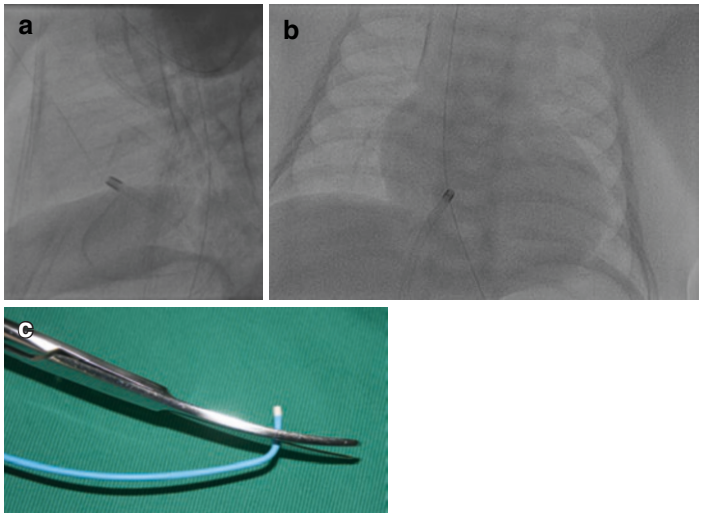


Fig. 22.3 (**a**, **b**) A 5 F Mullins' sheath placed in the RA has its tip facing the TV, facilitating quick access to the RV, and provides catheter stability during angiography and wire perforation of the valve. (**c**) Cutting the distal tip of a JR catheter facilitates manoeuvring of catheter into the RVOT for positioning beneath the valve. Shallow RV cavity and heavy trabeculations often make this manoeuvre difficult. *RA* right atrium, *TV* tricuspid valve, *RV* right ventricle, *RVOT* right ventricular outflow tract

4. A selective angiogram in the infundibulum is performed by manoeuvring the JR catheter tip into the RVOT. This is done simultaneously with aortography at the PDA level to opacify the PDA, the main pulmonary artery and its root. The size of the pulmonary annulus and thickness of the valve membrane/plate can be measured. In favourable cases, the pulmonary root and sinuses "cup" over the membranous atretic valve and infundibulum (Fig. 22.2a, b). [**Tip**: Difficulties may be encountered in manoeuvring the JR catheter into the RVOT because of the shallow RV cavity and heavy trabeculations. This can be facilitated by cutting off the distal tip of the catheter but retaining the primary curve (Fig. 22.3c)].

5. **Valve perforation:** The same JR catheter is now used to deliver the perforation wire. The catheter tip is placed underneath the valve membrane/plate. To ensure correct placement and avoid misperforation and tamponade, fine adjustments of position and checking with small volumes of contrast should be made.

   (a) **Perforation with radiofrequency (RF) wire: equipment and materials**

   The Baylis radiofrequency system (Baylis Medical Company, Mississauga, ON, Canada) with its generator and wire is the most widely used system for valvotomy. It consists of radiofrequency puncture generator (RFP 100 or RFP 100A) and the Nykanen RF wire. The active tip diameter of the wire is 0.016″ and the body outer diameter is 0.024″.

   As part of the system, ProTrack™ microcatheter is used with the RF wire for easy exchange to guidewire once perforation is accomplished. However, many operators prefer to use the wire on its own. Usually it is possible to advance a medium stiffness coronary guidewire, e.g. Choice PT Extra Support (Boston Scientific 8600 NW 41 Street Miami, FL 33166 USA) alongside the perforating wire which is subsequently removed once the former is anchored in a distal pulmonary artery branch (Fig. 22.4g). Graded balloon dilation is then performed.

   A Y-connector is attached to the JR catheter to prevent blood loss and for hand shot angiograms via the side port. The RF wire is inserted and its tip advanced towards the valve. Once the position is confirmed with small volumes of contrast, the generator is activated at usual setting of 5 Watts (W) for 2 seconds. In most cases, this will be sufficient. However, with a thick valve plate an energy of 10–15 W may be needed. The wire is carefully pushed under lateral fluoroscopy and the generator is turned off once the wire is across the valve. Aortogram opposite the PDA is performed to ensure that the wire is in a pulmonary

artery lumen (Fig. 22.4e, f). A misperforation outside the heart or pulmonary artery would be demonstrated (Fig. 22.6b). The wire following the outline of the heart in the pericardial space is also suggestive of misperforation. The RF wire is replaced with a coronary wire of medium stiffness and parked either into one of the branch pulmonary arteries (Fig. 22.4f) as mentioned above or through the PDA into the descending aorta. The JR catheter is then removed and replaced by 3 mm coronary balloon for the initial dilatation [**Tip**: 5 F JR guiding catheter with lumen of 0.058″ is used for smooth delivery of balloon catheter as otherwise balloon and wire may loop in the right atrium—especially if this chamber is markedly dilated—in the attempt of pushing the balloon across the valve through the small perforation (Fig. 22.4h)]. Final balloon dilation is carried out with a Tyshak Mini balloon (Numed Canada Inc., 45 Second St West Cornwall, ON K6J 1G3) 120–150% larger than the annulus size (usually a 6–8 mm diameter balloon) (Fig. 22.4i).

(b) **Perforation with stiffer tipped coronary wire (CTO)**
Alternatively, in the more straight-forward cases, perforation can be accomplished with stiffer-tipped coronary wires that are generally reserved for recanalization of chronic total occlusions in coronary intervention, e.g. the Conquest-Pro wire (Asahi Intecc Co. Ltd., 3-100 Akatsuki-cho, Seto, Aichi 489-0071, Japan) (Fig. 22.5).

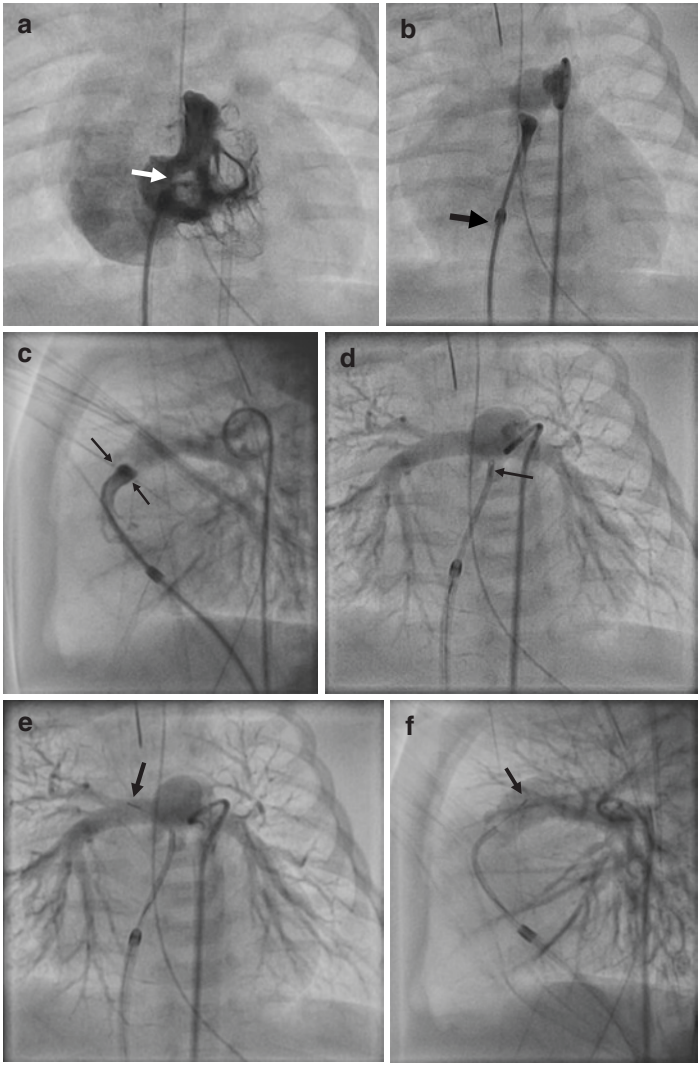
The same guidewire may then be used for balloon dilatation (and PDA stenting if indicated). Simplifying the procedure is attractive, but mechanical perforation with a stiff wire requires more force than RF wire, hence a higher risk of misperforation. In some cases, just a firm forward push of the stiff wire against the valve will not effect perforation. It is advisable in all cases to combine a gentle forward push with a rotary, "drilling" motion of the wire (rotating the wire with a torquer) (Fig. 22.5).

6. Subsequent hemodynamic data is obtained and RV angiogram is done. In patients with borderline RV size, PDA stenting can be done at the same time if PDA is less than 2 mm, and morphology is favourable.

22.7 Expected Results

The expected outcome of a successful procedure is establishment of forward flow into the pulmonary artery, normalization of RV pressure and marked improvement in oxygen saturation in the

Fig. 22.4 The procedure step-by-step. (**a**) RV angiogram in AP projection using a diagnostic 4 F JR catheter. A small, bipartite RV—only the inlet and infundibulum are present. The apical part is nearly obliterated by muscle bundles. The infundibulum is well developed but ends blindly with an imperforate, membranous valve. Arrow indicates tricuspid valve which is in an open position. (**b**, **c**) JR catheter negotiated the RVOT and is positioned beneath the imperforate valve (coaxially over a 5 F Mullins sheath, thick arrow). Near simultaneous hand injections in RVOT and aorta opposite the PDA. The RVOT has gone into spasm. Thin membranous valve. The MPA with well-developed sinuses "cup" over the doming valve. Thin arrows indicate PV annulus. (**d**) Aortography opposite the PDA in AP projection showing tip of JR catheter gently held against dome of membranous valve. Active tip of RF wire ready for perforation (arrow). (**e**, **f**) Successful perforation. RF wire tip in RPA confirmed by hand injection of dilute contrast (arrows). (**g**) Over the same JR catheter, a 0.014″ coronary guidewire of moderate stiffness and straight tip (thick arrow) is passed alongside RF wire (thin arrow), to be anchored in a distal RPA branch. The RF wire and diagnostic JR catheter are then withdrawn, to be replaced with a 5 F JR guiding catheter. (**h**) Graded balloon dilation, initially with a 3 mm × 20 mm coronary balloon over a JR guiding catheter (arrow). (**i**) Final balloon dilation with a 6 mm × 20 mm Tyshak balloon. The PDA was electively stented at the end of the procedure in view of the small RV size. *RV* right ventricle, *AP* antero-posterior, *JR* Judkins right, *RVOT* right ventricular outflow tract, *PDA* patent ductus arteriosus, *MPA* main pulmonary artery, *PV* pulmonary valve, *RF* radiofrequency, *RPA* right pulmonary artery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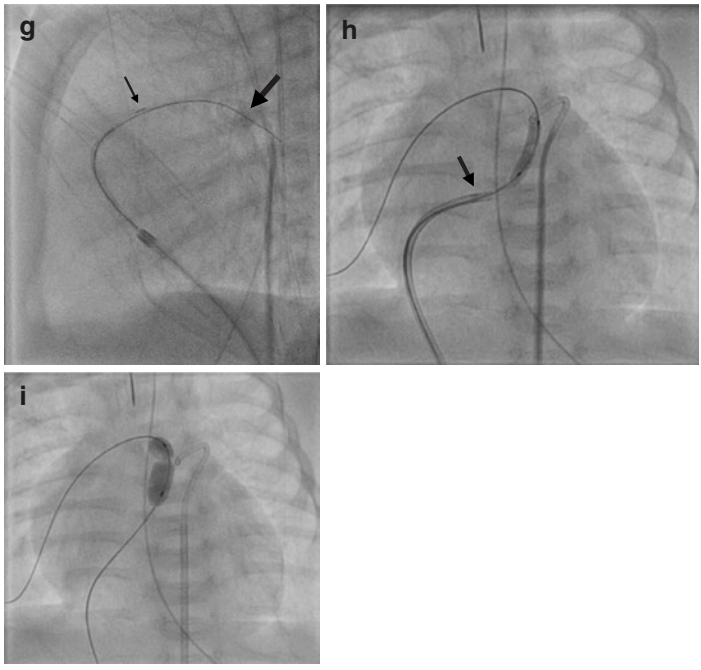

Fig. 22.4 (continued)

short term. Immediate full saturation is not the usual as reduced RV compliance from the RV hypertrophy and the often relatively smaller RV cavity continue to cause some right to left shunting across the PFO even when the RV systolic pressure is normal. In the longer term, regression of muscular hypertrophy can be expected to progressively improve the RV cavity and compliance.

Tricuspid regurgitation—unless due to Ebstein's malformation or dysplasia of the valve apparatus—is also expected to improve. RV pressure may remain elevated and possible reasons for that include insufficient dilation, infundibular spasm or the presence of fixed subvalve stenosis. In patients with RV-coronary connections, RV angiography will show immediate disappearance of these vessels if the RV has been adequately decompressed [4]. If the PDA is also stented at the same procedure, pulmonary overcirculation is common and diuretics may be required for a few weeks.

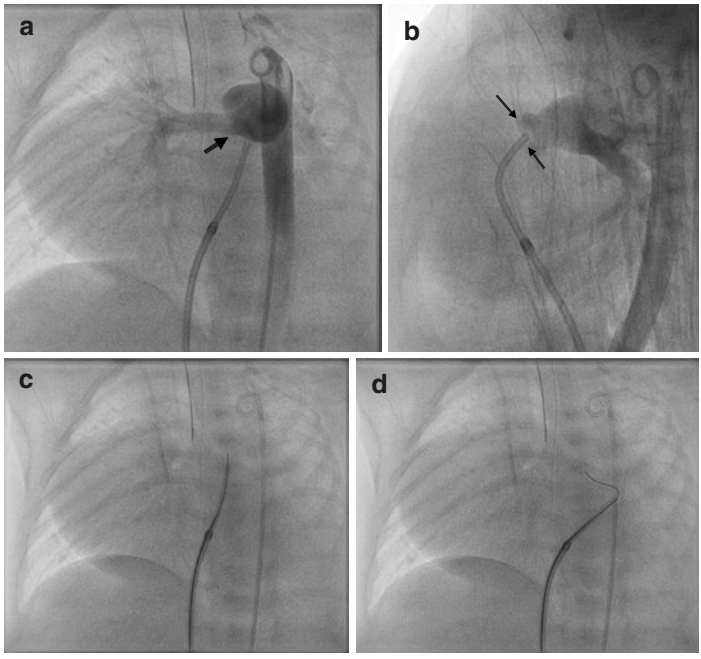

Fig. 22.5 Perforation using a CTO wire (Conquest-Pro, 9g tip-load). (**a**, **b**) MPA is opacified by contrast injection in descending aorta opposite the PDA in left anterior oblique (LAO)-cranial and lateral projections showing tip of 4 F JR diagnostic catheter positioned beneath the membranous valve. This is better seen on the lateral view (arrows). In the other view, the dilated MPA at the bifurcation partially overlaps with the sinuses of the MPA root (arrow, **a**). The cardiac silhouette shows a markedly dilated RA due to severe TR. A 5 F Mullins sheath is used coaxially to facilitate crossing of TV and stable positioning of JR catheter in RV. (**c**) Tip of Conquest-pro wire advanced forward to perforate valve membrane. (**d**) Guidewire tip failed to perforate the valve, buckled and displaced the position of JR catheter tip. (**e**) Successful perforation with combination of gentle forward push and "drilling" motion using a torquer. (**f**) Final dilation with 6 mm × 20 mm Tyshak balloon. *CTO* chronic total occlusion, *MPA* main pulmonary artery, *PDA* patent ductus arteriosus, *LAO* left anterior oblique, *JR* Judkins right, *RA* right atrium, *TR* tricuspid regurgitation, *TV* tricuspid valve, *RV* right ventricle

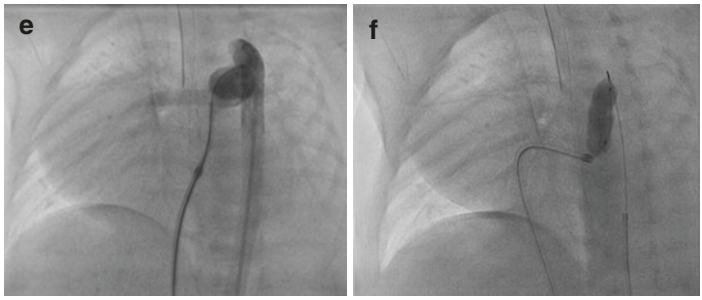

Fig. 22.5 (continued)

22.8 Pitfalls, Complications and Their Management

1. The major specific complication of this procedure is misperforation of wire outside the heart or main pulmonary artery. Fortunately, most misperforations do not lead to tamponade. To avoid misperforation, it is vital that the catheter tip and wire is correctly positioned beneath the valve as described above. Once perforation has been effected, aortography opposite the PDA to opacify the pulmonary arteries is performed to confirm that the wire is the pulmonary artery lumen before balloon dilation. Misperforation tends to occur in patients with dilated RVOT and small annulus (Fig. 22.6). Retrograde perforation via the PDA is may be an alternative [2].

   Rapid haemodynamic deterioration suggests tamponade which can be confirmed by echocardiography. Emergency pericardiocentesis may need to be performed, but if the patient is stable, surgery is preferable. Drainage of hemopericardium in a neonate may lead to more inadvertent punctures of the RV. It is preferable to withhold heparin until valve perforation is successful and balloon dilation is completed.
2. If valvotomy and balloon dilation is to be performed in a patient with major RV-coronary connections, it is imperative to ensure that the coronary circulation is not RV-dependent by performing detailed angiography to exclude proximal stenoses or ostial atresia of the coronary arteries. RV decompression following balloon dilation may lead to myocardial infarction.

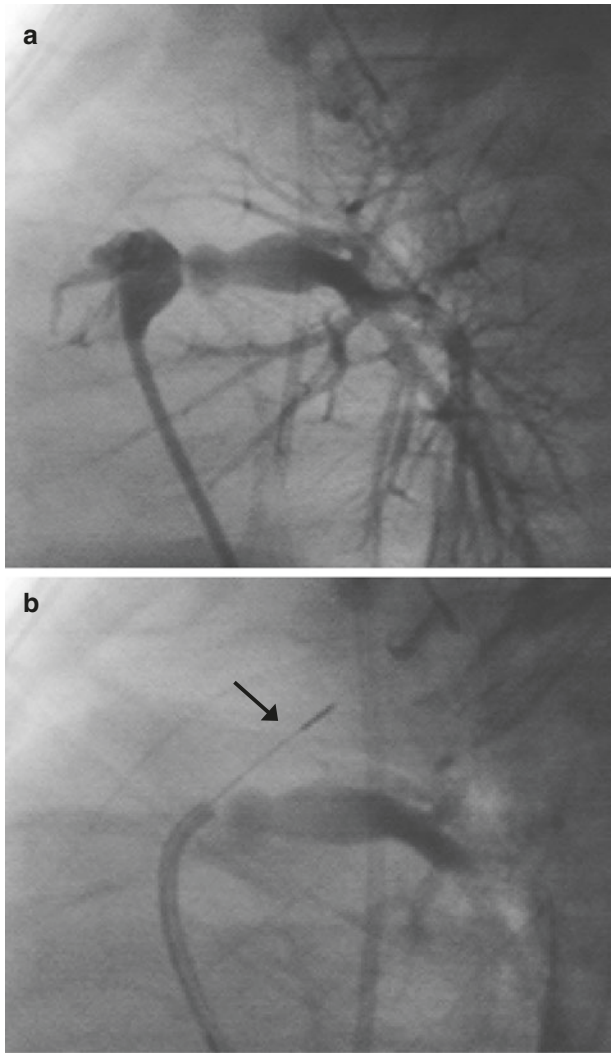

Fig. 22.6 Misperforation. (**a**) RVOT angiogram in lateral projection in a patient with associated Ebstein's anomaly, severe TR. The RVOT is markedly dilated, with a small valve annulus and pulmonary artery root. (**b**) RF wire has perforated the RVOT wall anteriorly. Contrast injection in descending aorta opposite the PDA showed that the RF wire is outside the pulmonary artery (arrow). *RVOT* right ventricular outflow tract, *TR* tricuspid regurgitation, *RF* radiofrequency, *PDA* patent ductus arteriosus

3. Guidewire or catheter manipulation in the PDA may lead to spasm and severe hypoxia. In this situation PGE infusion should be restarted. PDA stenting should be performed if this is ineffective. A soft-tipped coronary wire should be used to cross the PDA for stent implantation.
4. Even in patients with a fair sized RV cavity, RV dysfunction may occur following a successful balloon dilation. Stiff and poorly contracting RV may cause poor haemodynamics, hypoxia and metabolic acidosis ("suicidal RV"). If this is not responsive to supportive measures including re-starting PGE1, PDA stenting is recommended.
5. Severe or significant pulmonary regurgitation may occur following balloon dilatation. This, if combined with severe tricuspid regurgitation, a large PDA and PFO may lead to the rare phenomenon of "circular shunt" causing severe cyanosis. This requires surgical repair of the tricuspid ± pulmonary valves and reduction of the interatrial communication.

22.9 Post procedure

The patient is transferred to ICU for monitoring arterial blood pressure and blood gases. If the patient remains hemodynamically stable and had good results, early extubation is aimed. Some babies will need inotropic support overnight. Patients with smaller or bipartite RV in whom PDA stenting was not done need to be carefully monitored as once the duct closes they may become hypoxic, necessitating PGE infusion to be recommenced and re-intervention (ductal stenting or BT shunt). Routine echocardiogram is performed the next day to assess any residual gradient across RVOT and to exclude pericardial effusion.

22.10 Follow-up

Clinical progress and echocardiographic examination are important parameters for monitoring. Growth of the RV, degree of residual obstruction, tricuspid regurgitation and shunting across inter

atrial communication are the main areas of focus. Residual obstruction which could be valvar or subvalvar may increase with time, requiring re-intervention, either repeat balloon dilation or surgical reconstruction of the RVOT depending on the pathology. Patients with well-developed RV are unlikely to need re-interventions in the short and medium term.

Patients with borderline RV size need close monitoring for growth of RV and level of cyanosis. They may be candidate for bidirectional Glenn shunt to off-load the RV (1½ ventricle circulation).

In early adult life, progressive tricuspid and pulmonary regurgitation may require surgical intervention in patients who have done well following the initial catheter intervention.

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23 Percutaneous Transcatheter Balloon Mitral Commissurotomy

Raghavan Subramanyan,
Anuradha Sridhar, and Deepak Thakur

23.1 Introduction

Mitral valve (MV) dilatation by balloon catheter also known as percutaneous transcatheter mitral commissurotomy (PTMC) is now recognized as the treatment of choice to relieve rheumatic mitral stenosis (MS) in patients with suitable valve anatomy. Dr. Kanji Inoue first developed PTMC in 1982. Since then, a large worldwide experience has accumulated showing both immediate and sustained improvement. This chapter describes a practical approach to patient selection, work-up, technique, materials, limitations, and complications in patients undergoing PTMC.

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23.2 Anatomic Description and Physiopathology

Rheumatic heart disease (RHD) still represents a substantial health burden in developing nations. MS is the most common late sequel. It is due to a combination of leaflet thickening, commissural fusion, and chordal thickening/fusion. The obstruction may be either predominantly valvular or subvalvular. Calcification is common and may involve the leaflets, commissures, annulus, or the chordal apparatus. With reduction in the mitral valve area (MVA), diastolic filling of the left ventricle (LV) is maintained by a high trans-mitral pressure gradient. This leads to a rise in the left atrial (LA), pulmonary venous, and pulmonary arterial (PA) pressures. Atrial fibrillation is common, especially with increasing age, and is associated with thromboembolism in nearly 20% patients.

Like surgical mitral valvotomy, PTMC increases MVA by splitting the fused commissures to produce dramatic relief of MV obstruction and improved hemodynamics. The MVA obtained by PTMC is similar to that achieved by surgery, with comparable rates of restenosis. The characteristics of an ideal valve for PTMC are severe MS, fusion of both commissures, relatively mobile leaflets, no calcification, relatively mild fusion or thickening of the subvalvular apparatus, and no mitral regurgitation. Normal sinus rhythm and thrombus-free LA are also important for safe PTMC.

23.3 Indications and Patient Selection

Selection of a patient for PTMC is based upon both clinical and anatomical factors (Table 23.1). It is usually indicated in a symptomatic patient with MVA <1.5 cm^2 and favorable valve anatomy. In general, PTMC is contraindicated if the MVA is >1.5 cm^2 or if there is LA thrombus, absence of commissural fusion, commissural calcification, more than mild mitral regurgitation, or associated valvular/coronary disease which

Table 23.1 Indications for PTMC in clinically significant (moderate or severe) mitral stenosis (valve area ≤1.5 cm^2)

Recommendations	Class[a]	Level[b]
PTMC is indicated in symptomatic patients without unfavorable characteristics[c] for PTMC	I	B
PTMC is indicated in any symptomatic patient with contraindication or high risk for surgery	I	C
PTMC should be considered as initial treatment in symptomatic patients with suboptimal anatomy but no unfavorable clinical characteristics[c] for PTMC	IIA	C
PTMC should be considered in asymptomatic patients without unfavorable clinical and anatomical characteristics[c] for PTMC and • High thromboembolic risk (history of systemic embolism, dense spontaneous contrast in LA, new onset or paroxysmal atrial fibrillation) and/or • High risk of hemodynamic decompensation (systolic pulmonary pressure >50 mmHg at rest, need for major noncardiac surgery, desire for pregnancy)	IIA	C

Adapted from Baumgartner H et al. [1], Eur Heart J 2017;38: 2764 (with permission from Oxford University Press)

[a]Class of recommendation

[b]Level of evidence

[c]Unfavorable characteristics for PMC can be defined by the presence of several of the following characteristics. Clinical characteristics: old age, history of commissurotomy, NYHA Class IV, permanent atrial fibrillation, severe pulmonary hypertension. Anatomical characteristics: echocardiographic score >8, Cormier score 3 (calcification of mitral valve of any extent as assessed by fluoroscopy), very small mitral valve area, severe tricuspid regurgitation.

will benefit from surgery [1]. PTMC is also not advisable if the predominant obstruction is due to subvalvular thickening rather than due to commissural fusion.

23.4 Pre-procedural Evaluation

Trans-thoracic echocardiography (TTE) is the most useful imaging tool for obtaining comprehensive information about the MV. The severity and extent of fusion, thickening, shortening,

calcification, thrombus, and mitral regurgitation are all meticulously studied [2]. Many semi-quantitative scoring systems of the MV pathology have been used to predict the immediate outcome after PTMC and thereby select the appropriate patient. In the most commonly used method, leaflet mobility, leaflet thickness, leaflet calcification, and subvalvular thickening are graded in severity from 1 to 4. The sum of the four grades is called Wilkins mitral valve score. A score of less than 9 with not more than mild MR is considered most suitable for PTMC [3]. The Cormier score identifies three grades of severity based on the presence of anterior mitral leaflet calcification and severity of subvalvular disease [4]. Neither of these two scores however quantifies commissural fusion or commissural calcification. As splitting of the fused commissure is essential for relief of MS, commissural fusion and calcification must be studied in detail. In the Echo Score Revisited system of grading leaflet movement (3 points), commissural fusion (3 points), and subvalvular disease (3 points) are taken into account besides the severity of MS (2 points). A total score of 6 or more points indicates unfavorable anatomy for PTMC [5]. With a higher MV morphology score and/or moderate degree of MR one should balance the risk of PTMC and surgical open mitral commissurotomy (OMC). PTMC is still an option if the patient is a high-risk candidate for OMC. Trans-esophageal echocardiography (TEE) is necessary to exclude thrombus in the LA, especially in the appendage. It is desirable in all patients before PTMC and mandatory in patients with atrial fibrillation or previous embolism, and in patients with sub-optimal TTE images.

23.5 Balloon Catheters for PTMC

Inoue's method using an inflatable nylon-latex balloon catheter (NLBC) is the most widely used technique [2] and is described in detail below. Two types of NLBC are commonly available: Inoue balloon catheter (Toray, Tokyo, Japan) and Accura balloon catheter (Vascular Concepts Limited, Hallstead, United Kingdom). The NLBC is designed to expand initially in its distal portion, and then sequentially in its waist and proximal part. The low compliance in

its waist allows the balloon to take a dumb-bell shape centered in the narrowest part of the mitral valve and dilate it to the desired diameter (Fig. 23.1). The Inoue balloon catheter is a triple lumen catheter having three ports, two vent holes and vent tube. The 12 Fr shaft allows the use of contrast in a dilution of 1:4 for inflation. The balloon diameters that can be achieved with the Inoue balloon range from 22 to 26 mm (+4 mm) depending upon the volume of inflation. The Accura balloon catheter (11 Fr shaft) is available in sizes of 23–26 mm (+3 mm) and has two lumen and two ports. There are no vent tubes or vent holes. The contrast dilution recommended for Accura balloon is 1.6–1.8. It does not allow seepage of contrast between the layers and is economical as compared to the Inoue balloon catheter. The Blue Arrow PBMV

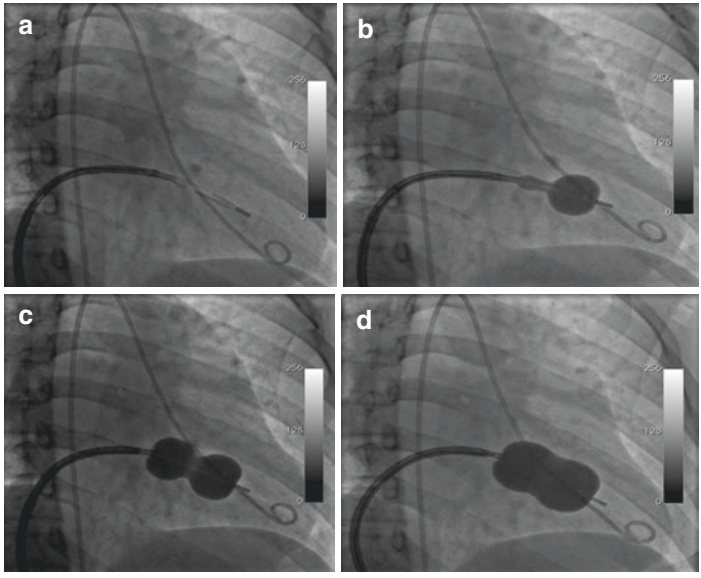

Fig. 23.1 Fluoroscopy in right anterior oblique view: (**a**) Ideal position of the balloon catheter on entry pointing toward the LV apex; (**b**, **c**) inflation of the distal balloon followed by positioning across the mitral valve and inflation of the proximal balloon; (**d**) full inflation and dilatation of the valve orifice to the desired diameter

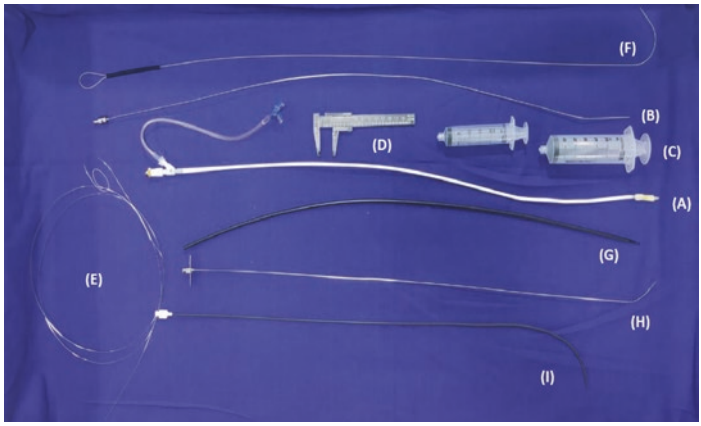

Fig. 23.2 Hardware for PTMC: (**a**) Nylon-Latex balloon catheter, (**b**) central metallic tube, (**c**) balloon inflation syringe, (**d**) caliper, (**e**) coiled steel guidewire, (**f**) J-shaped spring wire stylet, (**g**) septal dilator, (**h**) Brockenbrough needle, (**i**) Mullins sheath dilator

balloon catheter (SYM Shenzen Shineyard medical devices Limited, Shenzen, China) is a three-port PTMC balloon catheter very similar to Inoue balloon catheter [6]. The hardware required for PTMC are shown in Fig. 23.2.

23.6 PTMC Procedure

1. PTMC is most commonly performed from the percutaneous femoral vein approach. Usually the patient is under conscious sedation, but sick or unstable patients are preferably anesthetized.
2. Once the baseline catheterization data have been obtained, trans-septal puncture (TSP) is done observing usual precautions. A pigtail catheter is placed in the aortic root to indicate the level of the aortic valve. TSP is performed using Brockenbrough needle below the aortic root midway between the aorta anteriorly and spine posteriorly in left anterior oblique view (LAO). Too low a puncture can make subsequent entry into the MV difficult and must be avoided.

Heparin (5000–10,000 units) is administered once the LA is entered. The LA and LV pressures are recorded simultaneously for the gradient (Fig. 23.3a).

3. The 0.028″ coiled steel wire guidewire is passed through the Mullins sheath or dilator into the LA. The femoral as well as septal puncture sites are then dilated with the 14-Fr long stiff dilator.
4. Selection of appropriate balloon diameter for dilatation is essential for optimum results. The most commonly employed method relates MV diameter to the patient's height (diameter = 10 + height (cm)/10), although the relationship is not linear. A slightly more objective method measures the

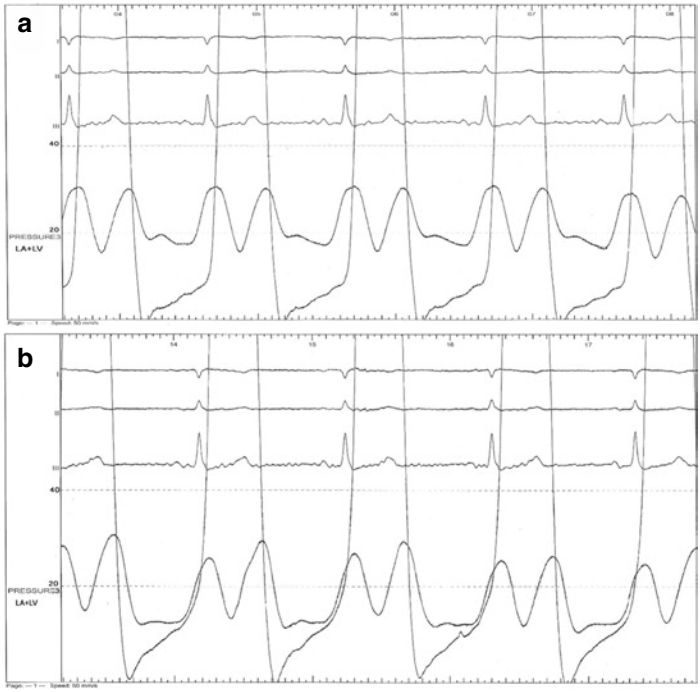

Fig. 23.3 Simultaneous LA and LV pressure tracings before (**a**) and after PTMC (**b**), showing abolition of the end-diastolic gradient and marked reduction in the mean diastolic gradient across mitral valve with successful PTMC. (Pressure recording: courtesy Dr. Rajagopal S)

minimum MV annulus diameter on TTE in apical four-chamber or two-chamber view in systole [2].

5. Initial dilatation is always done with a balloon diameter 2 mm less than the selected maximum diameter. In case the valve morphology is not entirely favorable, one should start with a balloon diameter of 4 mm smaller than the estimated maximum diameter.
6. The NLBC is stretched by insertion of the central metallic tube and then introduced over the guidewire into the femoral vein. It is then advanced carefully into the RA and across the atrial septum into the LA. At this point, the stretching tube is pulled back to make the NLBC more flexible in its distal part. A 80-cm 0.038-in J-shaped spring-wire stylet is used to manipulate and direct the tip of the NLBC toward the MV. Partial inflation of the distal part of the balloon may also help. Once the NLBC enters the LV, the distal part of the balloon is inflated so that it does not fall back into the LA. The catheter is then pulled back gently so that the waist of the balloon engages the orifice formed by the fused MV leaflets. The balloon is then fully inflated. The dumb-bell shape of the balloon ensures a stable position in the orifice and avoids inflation in the subvalvular area (Fig. 23.1).
7. After each dilatation, two-dimensional TTE is performed to assess the commissures, MVA, and mitral regurgitation. It is also advisable to measure the transmitral pressure gradient using the balloon catheter and the arterial catheter (Figs. 23.3 and 23.4).
8. Subsequent dilatations are done with 1 mm increments. The desired end-point may be one or more of the following without worsening of MR: (1) abolition of the gradient by pressure measurement (Fig. 23.3b), (2) one or both commissures are fully opened, (3) MVA 1.50 cm^2 or greater. Increase in MR to more than mild grade is a definite end-point to avoid catastrophic MR.
9. LV angiogram and pressure tracing (Fig. 23.4a, b) are usually done to assess MR if any, although color Doppler echocardiography may be sufficient.
10. The NLBC should be stretched again by the metallic tube to facilitate removal and to avoid trauma to the tissues.

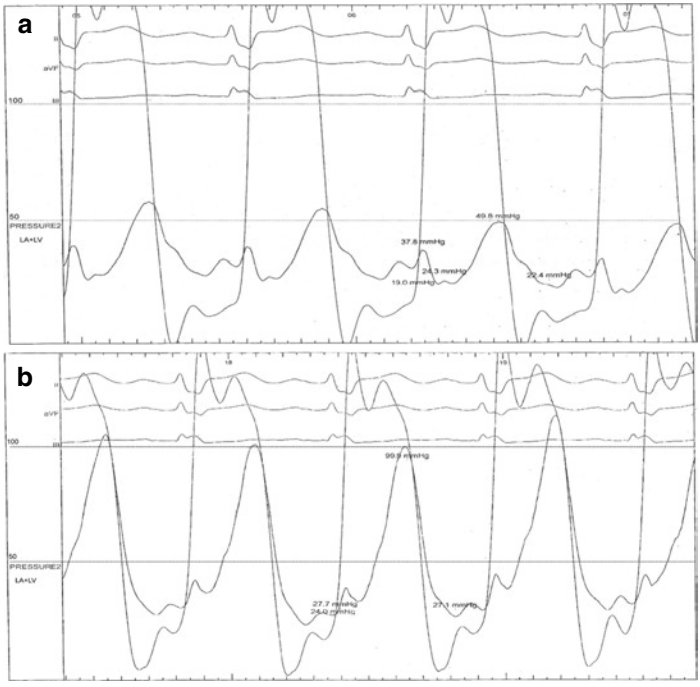

Fig. 23.4 Simultaneous LA and LV pressure tracings before (**a**) and after PTMC (**b**), showing reduction in the diastolic gradient across mitral valve. However, the high LA pressure and large V waves indicate severe acute mitral regurgitation. (Pressure recording: courtesy Dr. Rajagopal S)

23.7 Alternate Methods and Newer Techniques of PTMC

Conventional large-diameter cylindrical polyethylene valvuloplasty balloons can also be used to perform PTMC, but these catheters are stiff and difficult to maneuver. It involves placing a long stiff guidewire in the LV across the MV and then advancing the balloon catheter over the wire. A larger MVA can be obtained with two balloons being inflated simultaneously over two wires or in a monorail fashion on a single guidewire (Multitrack system, Numed Co). The retrograde trans-arterial approach has also been

used to do PTMC with either one or two balloons. The metallic mitral commissurotome developed by Cribeir is also as effective as the Inoue method in relieving MS. Being re-usable, it is more economical. However, the commissurotome is a large and stiff device. The alternate methods of PTMC are technically demanding and more commonly associated with complications.

23.8 PTMC in Special Situations

Pregnant patients with MS Pregnant patients with severe MS can develop clinical deterioration and pulmonary edema. It is associated with a higher maternal mortality during labor. Both surgical commissurotomy and PTMC can relieve MS during pregnancy but the latter method is associated with better maternal and fetal outcome. If indicated, PTMC should be performed by experienced operators in the second or third trimester with minimal radiation and proper shielding.

Mitral re-stenosis PTMC is effective if the mechanism of restenosis after previous surgical commissurotomy or PTMC is predominantly commissural fusion rather than subvalvular fusion. The evaluation, selection, and technique for PTMC in re-stenosis is similar to de novo MS.

Juvenile rheumatic MS PTMC can be safely performed in children with rheumatic MS with excellent immediate and late results. The indications are similar to those in adults.

Congenital MS Congenital MS represents a wide spectrum with varying involvement of the commissures, leaflets, subvalvular apparatus, and the mitral annulus. The results of PTMC therefore are predictably variable. It can be performed in selected cases with appropriate monitoring for MR.

MS with LA thrombus Presence of an LA thrombus involves a high risk of embolism during PTMC, and open commissurotomy

is preferred. If surgery is not acceptable or possible, PTMC can be undertaken with special precautions: administer effective anticoagulation for 8–12 weeks and confirm by TEE the absence of thrombus in the LA. Care is taken to minimize the manipulation of the wire and catheter in the LA. A modified over-the-wire method is used in which the guidewire is placed in the LV across the stenotic valve, and the NLBC is advanced over the wire. This reduces the movement of the NLBC in the LA, especially in the region of the LA appendage and roof. A lower than usual puncture site in the septum is advisable in this approach, to achieve a more direct course toward the mitral orifice.

23.9 Tips and Tricks for Successful PTMC

The key factors for a safe and successful PTMC are awareness of pitfalls, meticulous technique, and cumulative experience. Experienced operators have found the following tips and tricks useful:

1. Selection of a suitable patient with favorable valve anatomy is perhaps the single most important element. When PTMC has to be done for a less favorable valve anatomy, the target valve area should be realistic and MV disruption must be avoided.
2. The femoral vein is the recommended approach for PTMC. The entry site into femoral vein should be clearly >2–3 cm below the inguinal ligament to allow easy passage of the stiff dilator and catheter. Access through the internal jugular vein has been used in patients with severe scoliosis and distorted septal anatomy, as well as in those ileo-femoral vein or inferior vena caval occlusion [7].
3. The inter-atrial septum (IAS) is ideally punctured in the fossa ovalis, avoiding the muscular upper part. A high location of TSP makes manipulation of the NLBC and entry into the MV difficult. A low puncture carries the risk of injuring the coronary sinus. The septal anatomy is often distorted in MS due to atrial dilatation and bulging of the septum. A mid-posterior

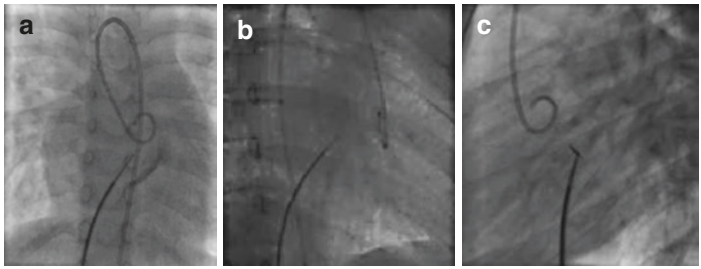

Fig. 23.5 Fluoroscopy of inter-atrial septum in various views for trans-septal puncture: (**a**) LAO view (left anterior oblique), (**b**) RAO view (right anterior oblique), (**c**) lateral view

puncture is preferred for PTMC, as it provides a favorable working height in the left atrium and a coaxial plane with the mitral valve. Fluoroscopy in multiple planes and continuous pressure monitoring are recommended for safe trans-septal catheterization. Left anterior oblique (LAO), right anterior oblique (RAO), and lateral projections are commonly utilized (Figs. 23.5a–c). The fossa ovalis can be visualized by staining the septum with 1–2 mL of contrast injected through the Brockenbrough needle held with the tip against the IAS (Fig. 23.5c). In difficult cases, delayed LA angiography can help visualize the septum.

4. TEE allows direct imaging of the soft tissue structures that are relevant to TSP and provides accuracy and reassurance for an optimal puncture. This technique of TSP is being increasingly used now. The bicaval view (90°) in TEE details the superior and inferior extent of IAS while anterior and posterior extents are best seen in the short-axis (30°) and four-chamber (0°) views. With 3D software and TEE, an *x*-plane image displays both bicaval and short-axis views of IAS simultaneously [8]. This is extremely helpful in puncturing the IAS at the desired location (Fig. 23.6).
5. Newer imaging techniques that can facilitate TSP include intracardiac echocardiography, three-dimensional echocardiography, computed tomography–derived three-dimensional augmented fluoroscopy, real-time magnetic resonance imag-

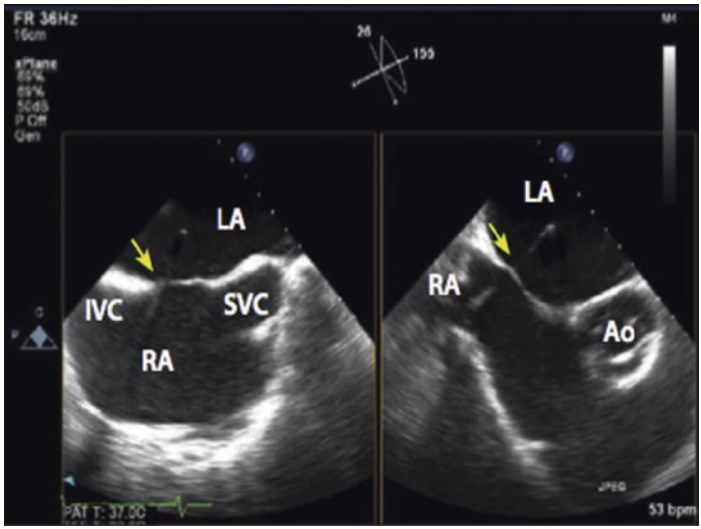

Fig. 23.6 *X*-plane trans-esophageal echocardiography showing inter-atrial septum simultaneously in two planes. *LA* left atrium, *RA* right atrium, *Ao* aorta, *SVC* superior vena cava, *IVC* inferior vena cava. (Adapted from Singh GD et al. [8], with permission from Elsevier)

ing, and rotational angiography [9–12]. But the advantages and recommendations still remain to be evaluated. Fusion of different imaging modalities is also gaining popularity for TSP. Currently however multi-plane fluoroscopy with or without TEE is the method of choice for safe TSP during PTMC.

6. Entrapment in the pulmonary vein or in the IAS is best avoided by placing the NLBC deep in the LA with a generous curve before removing the guidewire. This position makes subsequent manipulation and entry into the LV easier by gradual withdrawal of the spring stylet. It also avoids entry into the left atrial appendage, which is the usual site for LA thrombus.
7. The most common method of finding the MV orifice with the tip of the NLBC is to gradually withdraw it so as to straighten the curved course in the LA and orient it along the MV to

apex axis. This is best done with fluoroscopy in the RAO view (Fig. 23.1). On close observation, the NLBC is seen to move back toward the LA roof in systole. In diastole, the tip of the NLBC characteristically dips toward the MV orifice, especially if the distal balloon is slightly inflated. The trick is to push the catheter forward in diastole while at the same time withdrawing the spring stylet. Both timing and coordinated movement are extremely important in this maneuver, which consists of a combination of floating toward the orifice along with pushing and sliding the NLBC over the spring stylet. Sometimes the spring stylet in its original J shape may not align properly from the site of septal entry to the MV orifice. Changing the shape of the spring stylet is then helpful. If the LA is quite dilated, a large smooth curve is preferred, while a smaller J curve is used in a small LA. In cases with very large LA, entry may be achieved by forming a loop against the atrial wall. Occasionally, one may have to do a new septal entry at a different point to get better orientation to the mitral orifice.

8. Sometimes, it is extremely difficult to cross the stenotic MV with the balloon catheter. This may be due to very severe MS, markedly dilated LA or distorted IAS. Different methods have been used to overcome this problem [13]. In the modified over-the-wire technique, a diagnostic AR-1 catheter is used to cross the mitral valve over a 0.035″ hydrophilic 260-cm long Glidewire. The Glidewire is then exchanged with a 0.035″ Amplatz super-stiff guidewire over which the PTMC balloon catheter is advanced [14]. A double-loop technique has been used successfully to cross the mitral valve in difficult patients [15]. The J-shaped stylet and anti-clockwise rotation can be used to make a double loop of NLBC in LA, after which it can advance to the LV. Veno-arterial loop techniques have also been utilized to support the advance of the NLBC across the stenotic MV [16, 17]. This involves advancement of a Glidewire through the NLBC, trans-septal sheath or a diagnostic catheter into the LV and aorta. The aortic end of the wire is then fixed either manually or by a snare, so that the guidewire provides support to the balloon catheter. Rapid snare sliding is a modification of the veno-

arterial loop technique, described in a case with multiple failed attempts to cross the MV. A 0.025″ hydrophilic 260 cm Terumo (Terumo Corporation, Japan) guidewire was passed through the NLBC and snared in the ascending aorta and exteriorized. A 5 Fr Launcher guide catheter was placed in the LV apex on the snared wire. An arterial clamp was placed on the snared guidewire at the guide catheter end. The 0.025″ wire was rapidly pulled at the NLBC end making the wire taut, which slid the NLBC across the MV into the LV [18].

9. It is essential to avoid MV trauma and regurgitation. The most important trick here is to select the appropriate-sized balloon and dilate the valve stepwise, beginning with a smaller-than-target diameter and increasing it by 1–2 mm at each step. Proper positioning of the NLBC in the LV is critical and is best done in the RAO view. The NLBC should form a smooth curve and point toward the LV apex (Fig. 23.1a). An abnormal bend or vertical direction of the tip or abnormal shape of the balloon on inflation indicates entry into and entanglement in the chordal apparatus (Fig. 23.7a). The next step is to move the NLBC back and forth on the stylet to confirm a free position in the central part of the MV apparatus. Partial inflation of the distal balloon at this stage will prevent it from falling back into the LA.
10. Look carefully for signs of severe subvalvular pathology such as failure to advance the catheter despite the tip being in the MV orifice or cogwheel resistance to movement of the catheter. Abnormal shape of balloon during inflation (Fig. 23.7b–d) may indicate entrapment in the mitral chordae and is more likely to occur with severe subvalvular pathology.
11. Avoid damage to the IAS by stretching the NLBC both on introduction and on withdrawal, and by making a gentle loop in LA during inflation in the MV. It is advisable to pull back the stiff portion of the guidewire before the catheter comes out of LA, thus leaving only the coiled soft portion across the IAS.
12. PTMC must be monitored by echocardiography and by pressure-gradient measurement after each inflation to know the endpoint, and for early recognition of complications such as MR or perforation (Fig. 23.4).

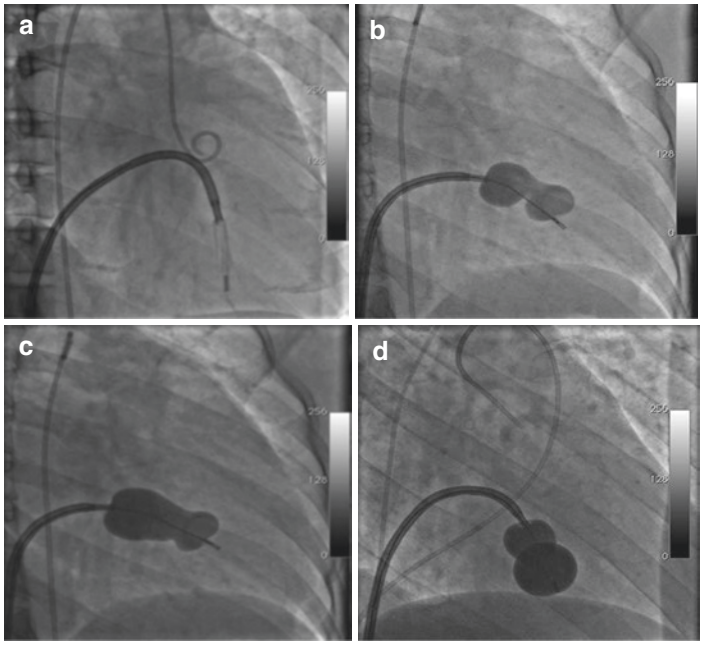

Fig. 23.7 Fluoroscopy in right anterior oblique view: (**a**) Improper position of the balloon catheter pointing inferiorly and posteriorly into the chordae; (**b**, **c**) distortion of the distal and proximal parts of the balloon by subvalvular apparatus; (**d**) both distal and proximal parts of the balloon inflated in the LV below the valve orifice. These clues indicate entrapment of the balloon catheter

23.10 Expected Results

Immediate hemodynamic improvement indicates successful valvotomy and is assessed during PTMC by mean LA pressure, pressure gradient across mitral valve (Fig. 23.3a, b), and severity of MR (Fig. 23.4a, b). Increase in the MVA measured by two-dimensional echocardiography is a strong indicator of successful PTMC. In the majority of patients with favorable characteristics, the valve area increases by 100%. Doppler-derived measures of

MS immediately after PTMC do not correlate well with the clinical outcome. Older age, smaller valve area, previous commissurotomy, higher mitral valve morphology score (>8), valve calcification, and baseline MR are predictors for poor immediate outcome [2].

Event-free long-term survival after successful PTMC refers to survival with freedom from repeat PTMC, mitral valve replacement, cardiac death, and high NYHA functional class. Restenosis refers to a decrease in MVA or loss of the initial gain in MVA on late follow-up. At 10 years, the event-free survival has been reported to be about 79% in younger patients [19] and 56% in slightly older patients [20]. The MV characteristics also affect the late results of PTMC. In young subjects with an MV score of ≤8, the event-free survival and freedom from restenosis were 66% and 65%, respectively, at 15 years. If the MV score was >8 at the time of PTMC, the event-free survival and freedom from restenosis were much lower (9% and 8%, respectively, at 15 years) [19].

23.11 Complications and Management

Significant complications or failed valvotomy has been reported in 1.0–15.0% patients [2]. Some increase in MR can occur after PTMC due to splitting of the fused commissures. Commissural MR of mild or moderate degree is often tolerated well without the need for valve replacement. Severe MR can occur in 1.5–10.0% due to disruption of the leaflet or rupture of subvalvular apparatus and is likely to need either immediate (<1.0%) or early MV replacement.

Cardiac perforation and hemopericardium (incidence 0.5–10.0%) can occur during IAS puncture or during manipulation of the balloon catheter. Management consists of protamine administration, volume expansion, and if necessary, pericardiocentesis or surgical repair. Local vascular site complications, embolic complications, and death are uncommon (0.5–5.0%). Major complications and sequelae are more common among patients who had inadequate immediate result of PTMC.

23.12 Summary of Critical Points for Safe and Successful PTMC

- Careful evaluation and selection of case
- Choice of appropriate balloon diameter
- Stepwise dilatation with 1 mm increment in balloon diameter
- Well-defined end-points: abolition of gradient, MVA >1.50 cm^2, worsening MR
- Careful watch for complications
- Meticulous technique and experience

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Part V

Step-By-Step Procedures: Vessel Treatment

24 Stent Implantation in Patients with Pulmonary Arterial Stenosis

Andreas Eicken and Peter Ewert

24.1 Introduction

Pulmonary arterial branch stenosis may occur congenitally or after surgical procedures. Patients of all ages (newborns to adults) with congenital heart disease may need pulmonary arterial interventions. All sections of the pulmonary arterial tree may be involved: the main pulmonary artery or its origin, the central branch pulmonary arteries, and the segmental pulmonary arteries. In newborns and young infants with pulmonary arterial stenosis after surgical procedures involving the pulmonary arteries, balloon angioplasty may be employed as first-line treatment, if repeated surgery does not seem adequate [1]. In these small patients, large stents dilatable to adult vessel diameter cannot be implanted since large stents on large balloons need large sheaths.

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However, balloon angioplasty may not always lead to an acceptable flow increase of the stenotic lung segment and even in this difficult patient group stent implantation may be the only solution. In general, stent implantation is the preferred first-line treatment in patients with pulmonary arterial stenosis, in whom a stent can be implanted, which can be expanded to an adult size diameter. In patients with tight segmental pulmonary arterial stenosis, balloon dilatation with cutting balloons may be indicated [2].

24.2 Anatomic Description and Pathophysiology

Normal pulmonary vessels have an adequate caliber and an appropriate distensibility, and they provide active support to forward blood flow by their Windkessel function. Pathologic vessels can be stenotic at different levels of the vascular tree, hypoplastic in segments, or finally can be compressed by other structures like the aorta or the airways. Hypoplasia can be due to flow restriction by a localized stenosis, or due to an intrinsic vessel disorder like in Williams-Beuren syndrome. A generalized vessel abnormality is present, whenever an aortopulmonary collateral has been unifocalized surgically in a patient with pulmonary atresia and MAPCAs. From its embryologic origin, these collaterals are more systemic—than pulmonary arteries.

A severe vessel stenosis (<50% diameter of the adjacent normal pulmonary artery) leads to a detectable systolic pressure gradient, which can be measured by a catheter pullback across the stenosis. In a biventricular circulation, if several lung segments are stenotic, a pressure rise in the subpulmonic ventricle (usually the right ventricle) is seen. Doppler echocardiography helps to locate and quantify the flow acceleration across a stenotic vessel, and if tricuspid regurgitation is present, the systolic pressure in the right ventricle (RVP normal <30 mmHg) can be assessed. After noninvasive measurement of the patient's blood pressure, the pressure ratio between the right ventricle (RVP) and the aorta

(AoP) can be assessed. In patients with univentricular hearts, the source of pulmonary blood flow is important. After first-stage palliation (for example, Norwood operation in hypoplastic left heart syndrome), a right ventricular to pulmonary artery (Sano shunt) or an aorto-pulmonary shunt (modified Blalock–Taussig shunt) ensures pulmonary blood flow. In impeded pulmonary blood flow, the arterial oxygen saturation may be lowered to critical values. Again, Doppler echocardiography is helpful to show a flow acceleration between the systemic and pulmonary circulation. The flow acceleration may not be transferred directly into a systolic pressure gradient in mmHg. In patients with univentricular circulation, the RVP:AoP ratio naturally is not helpful. After a cavopulmonary shunt in univentricular circulation (PCPC, partial cavopulmonary; and TCPC, total cavopulmonary connection), the pressure gradient is of less importance, since there is no subpulmonary pumping chamber, and the pressure gradients are, if at all present, very low. The pre-stenotic pressures may rise, and this may lead to severe symptoms (cyanosis due to veno-venous collaterals, effusions, "failing Fontan" circulation). In these situations, treatment decisions are based on the patient's clinical status.

24.3 Indication for Stent Treatment in Pulmonary Arterial Vessel Stenosis

Biventricular circulation treatment indication:

- RVP:AoP pressure ratio >66% (if RV function is impaired, an intervention may be indicated at lower RVP:AoP ratios)
- Significantly abnormal (right to left, R:L) pulmonary arterial flow distribution assessed by perfusion scans or by cMRI (normal is R:L = 60:40%), significantly abnormal is a perfusion distribution of 80% or more to one lung
- Systolic pullback gradient >20 mmHg
- Angiographic stenosis with <50% lumen diameter compared to the normal adjacent pulmonary vessel

Univentricular circulation treatment indication:

- Arterial desaturation (<65% SaO_2) early after an aorto-pulmonary—or right ventricle to pulmonary artery shunt operation
- Arterial desaturation after a cavopulmonary shunt operation in the early postoperative period
- Elevated central venous pressures with signs of congestion
- Signs of impaired pulmonary arterial blood flow even in mild angiographic stenosis

24.4 Pre-procedural Imaging

Echocardiography is the most important imaging modality in congenital heart disease. For direct assessment of the pulmonary arterial tree, however, echocardiography often is of limited value, since the peripheral vessels cannot be seen. Despite this limitation, echocardiography can detect pulmonary arterial stenosis at the pulmonary bifurcation and may help to quantify the pressure burden and function of the subpulmonary ventricle. MRI may depict the pulmonary arterial tree, helps to quantify right ventricular function, and offers flow quantification (R:L ratio). A CT scan offers high-resolution images of the pulmonary arteries even if stents or pacemakers have been implanted previously.

24.5 Stent Implantation

A complete cardiac catheterization with pressure assessment and catheter pullback at the site of the stenosis is performed. Following angiographic depiction of the anatomy, a guidewire is advanced over the stenotic region and positioned distally into the pulmonary artery. The target lesion should be depicted with only minimal foreshortening and, if ever possible, in a biplane approach with perpendicular X-ray angulations. Meticulous measurements of the vessel diameter and the length of the stenosis are necessary.

A balloon stent assembly (either premounted or manually crimped) is chosen with a balloon diameter equal to the diameter of the adjacent normal pulmonary vessel. A suitable sheath (coronary guide catheter in newborns or infants), or a long sheath is advanced. If possible, this sheath is advanced over the wire to pass the stenosis. The balloon/stent assembly is then advanced over the wire to the target region. Then the sheath is withdrawn, and small contrast injections are performed through the sheath allowing the stent position to be adjusted. Then the balloon/stent is inflated, the balloon is withdrawn, and the result is documented by angiography. If feasible, a repeated catheter pullback may be performed and the RVP:AoP ratio acquisition repeated to document an adequate hemodynamic effect of the procedure in patients with biventricular circulation. In some lesions, it may be helpful to perform an interrogation of the stenosis with a low pressure balloon catheter to exactly locate the site of stenosis and to assess the length of the stenotic vessel [3–10].

24.6 Stents

Balloon-expandable stents provide the best immediate result regarding vessel diameter increase, lack of recoil, safety to avoid dissection, or vessel rupture. If covered stents are chosen, larger sheaths are mandatory, which may not be feasible in small patients. In newborns and small infants, stent implantation into the pulmonary arteries may be a life-saving procedure, for example, after a Norwood operation (stenotic modified BT shunt or stenotic Sano shunt). Sometimes these patients are on ECMO due to postoperative cyanosis. If the stent needs to be delivered through an aorto-pulmonary shunt, it may be necessary to use an arterial access (4 F sheath, for example, Terumo Radifocus introducer II 25 cm; Terumo Deutschland GmbH, Eschborn, D) through which only coronary stents can be delivered. This sheath is stiff and cannot be advanced into the pulmonary arteries through the shunt; hence, the premounted coronary stent has to be advanced carefully to the target without being covered. In our unit, we use the cobalt chrome Coroflex blue neo premounted

stent (B.Braun AG, Melsungen, Germany) for this indication. These stents can be expanded to a maximum diameter of 5 mm. Later on, the stents have to be removed surgically, cut open longitudinally, and widened with a patch, or they must be cracked open with ultra-high pressure balloons. A 5 F Terumo slender sheath in combination with a short 5 F coronary guide catheter (65 cm, Cordis, Miami Lakes, FL, USA) may be used for this purpose to have optimal forward push and the possibility of angiographies for final stent positioning.

In larger infants, or if a venous access can be used, stents from the Cook Formula™ family (Cook Medical; Bloomington, Indiana, USA) have proven to be very helpful recently (Fig. 24.1). The Formula™ 418 6 × 12 mm stent can be implanted on a 0.018″ guidewire through a 5 F long sheath or through a 6 F coronary guide catheter. Shorter 5 F (65 cm) and 6 F (55 cm) coronary guide catheters are available and very helpful due to their flexibility and Judkins right configuration. The Formula™ 418 stent may be expanded up to 12 mm before it breaks. The Formula™ 535 (8 × 12 mm stent can be expanded to 14 mm, and the Formula™ 10 × 20 mm stent can be expanded to 16 mm, without significant foreshortening) stents are delivered over a 0.035″ guidewire through 6 F and 7 F long sheaths, respectively [11–17].

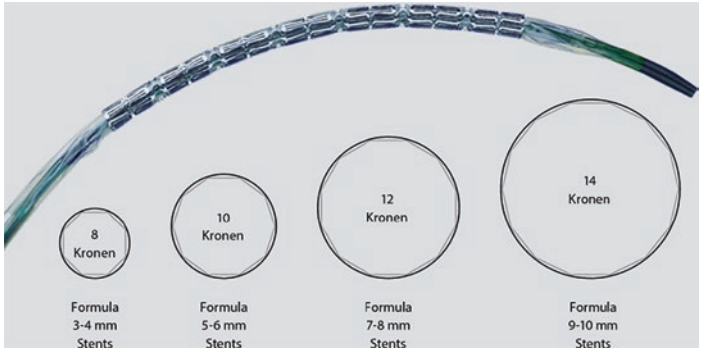

Fig. 24.1 Premounted Cook Formula™ stents (Cook Medical; Bloomington, Indiana, USA) mounted on 6, 8, and 10 mm balloons. Kronen = stent crowns

In older children and adults, stents dilatable to an adult size vessel (>20 mm) are available. The Max LD stent (Medtronic, MN, USA) is available in lengths between 16 and 36 mm and has an open cell stent design. The Mega LD stents (Medtronic, MN, USA) may be delivered on smaller balloons and through smaller sheaths, but their radial forces are less. The Andra xl stent (Andramed Reutlingen, D) has a hybrid stent design (open and closed cells) and is available in lengths from 13 to 57 mm. Another frequently used stent is the Cheatham platinum stent (Fig. 24.2) (Numed Hopkinton NY, USA) which has a closed cell design and is available in lengths between 16 and 45 mm. This stent is also available as a covered stent.

A special issue is the treatment of a pulmonary arterial bifurcation stenosis with bilateral narrowing of the pulmonary arterial branches. To overcome the problem of obstructing the contralateral vessel by stent implantation, a simultaneous implantation of two stents in each side through two long sheaths can be performed (Figs. 24.3, 24.4, and 24.5).

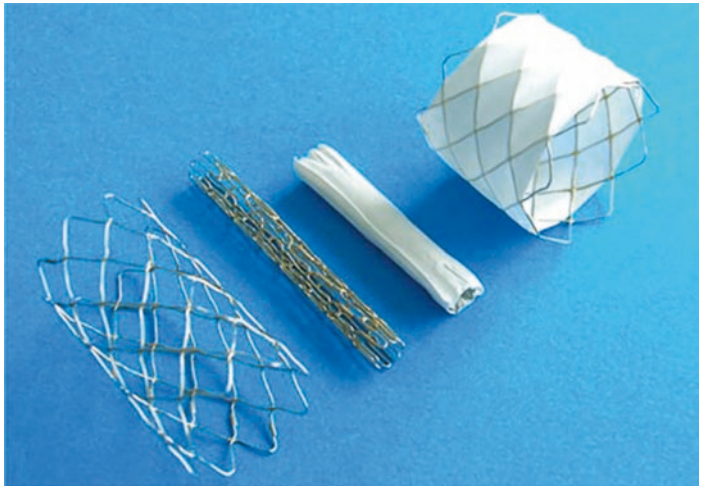

Fig. 24.2 Cheatham 8z platinum stent, on the right an expanded bare metal and on the left an expanded covered 8z Cheatham platinum stent

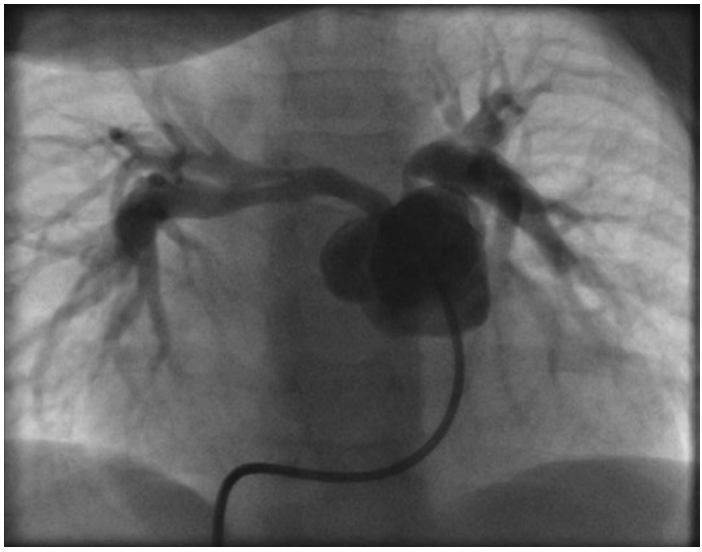

Fig. 24.3 One-year-old child with common arterial trunk 6 months after implantation of a 14 mm Contegra graft (RV-PA) now with RVP 88/0/12, PAS 20/13/15, PAD 21/12/14, AoP 105/46/66 (RVP:AoP ratio 88%)

The disadvantage, however, is that the creation of a double lumen in the pulmonary arterial trunk might be an obstacle for further catheter interventional treatments like percutaneous pulmonary valve implantation. An alternative treatment concept is the creation of a Y-stent, in which two stents are placed in y-fashion though their meshes directly into the pulmonary bifurcation (Figs. 24.6, 24.7, and 24.8).

24.7 Expected Results

In isolated circumscript pulmonary arterial stenosis, stent implantation can result in complete relief of pressure gradients and to a normalization of flow distribution. In multiple stenosis, an attempt to treat the most severe and proximal stenosis should be tried first. Since in these cases even very small vessels are involved, cutting balloon dilatation and stenting techniques can be combined. The

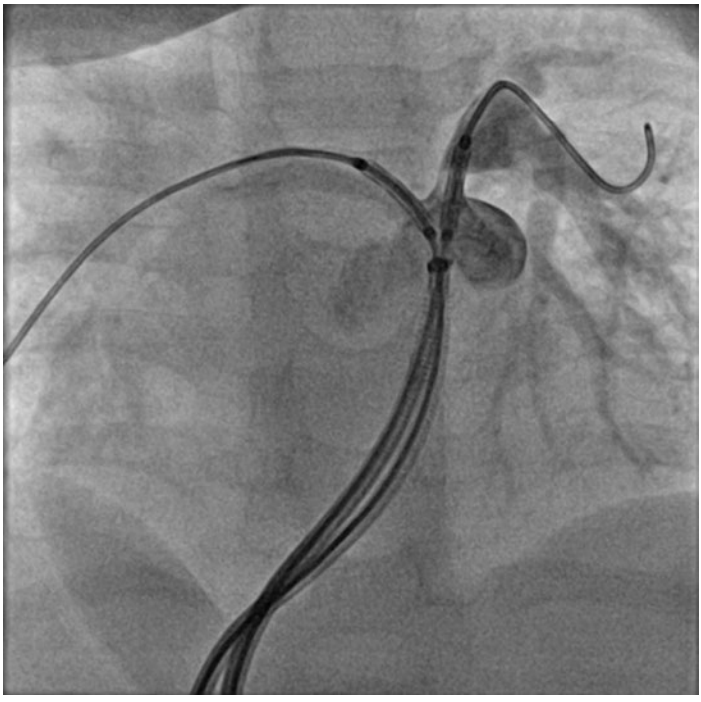

Fig. 24.4 Two 6 F Terumo 45 cm long sheaths (via both femoral veins) were advanced on 0.035″ guidewires into the right ventricular outflow tract. A 6 × 12 mm Formula™ 535 stent was positioned into the origin of the right pulmonary artery and a 8 × 12 mm Formula™ 535 into the origin of the left pulmonary artery. The stents were inflated simultaneously

overall result, however, is variable. Assessment of a beneficial therapeutic effect can be challenging if stenosis relief can only be achieved in segments of a generally pathological multi-stenosed pulmonary vascular bed as in patients with pulmonary atresia and VSD with MAPCAs after unifocalization procedures. If unobstructed flow is directed to a small pulmonary vascular bed, pulmonary hypertension may result due to pressure and volume overload. Transient pulmonary hyperperfusion may also occur. In generalized pulmonary arteriopathy with stiff and hypoplastic vessels, transcatheter interventions (as any interventions) are of very limited effect.

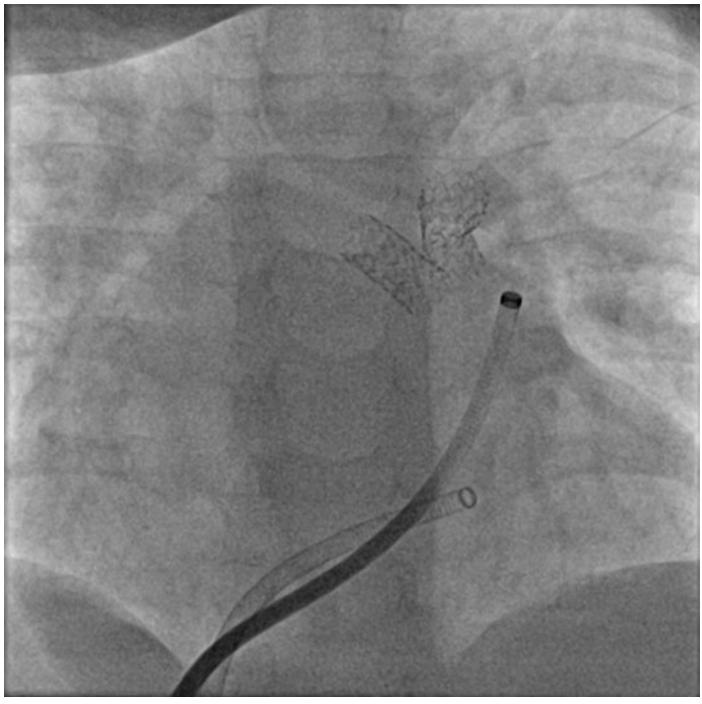

Fig. 24.5 Result after inflation of both stents. RVP was 58/2/16, PAD 29/14/16, AoP 119/54/67 (RVP:AoP ratio 48%)

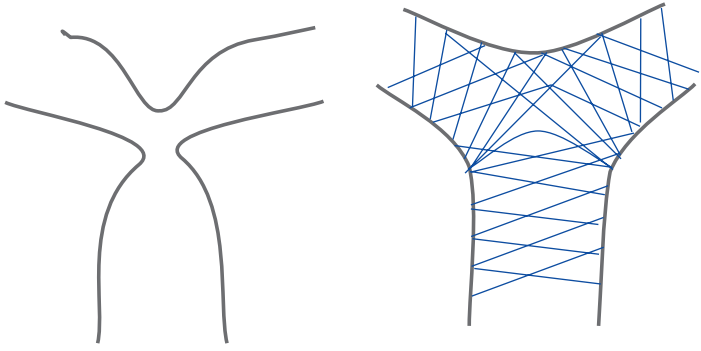

Fig. 24.6 Schematic drawing of a bifurcation stenosis pre and post stent dilatation

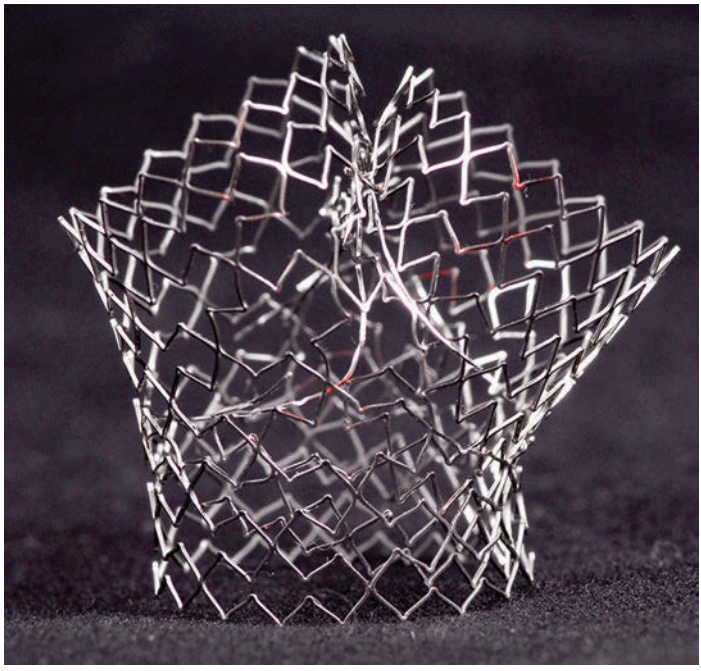

Fig. 24.7 Two 26-mm Max LD stents (Medtronic MN, USA) form the y on the bench

24.8 Tips and Tricks

24.8.1 Challenging Anatomies

In smaller patients, it is sometimes difficult to enter the left pulmonary artery with a long sheath for safe stent placement. A wedge balloon catheter of the corresponding sheath size can be helpful. The wedged catheter withstands a certain pull and, thus, may facilitate the advancement of a long sheath across the pulmonary stenosis. Another trick for difficult PA-entry with a long sheath is to use a 4- or 6-mm PTA balloon dilatation catheter

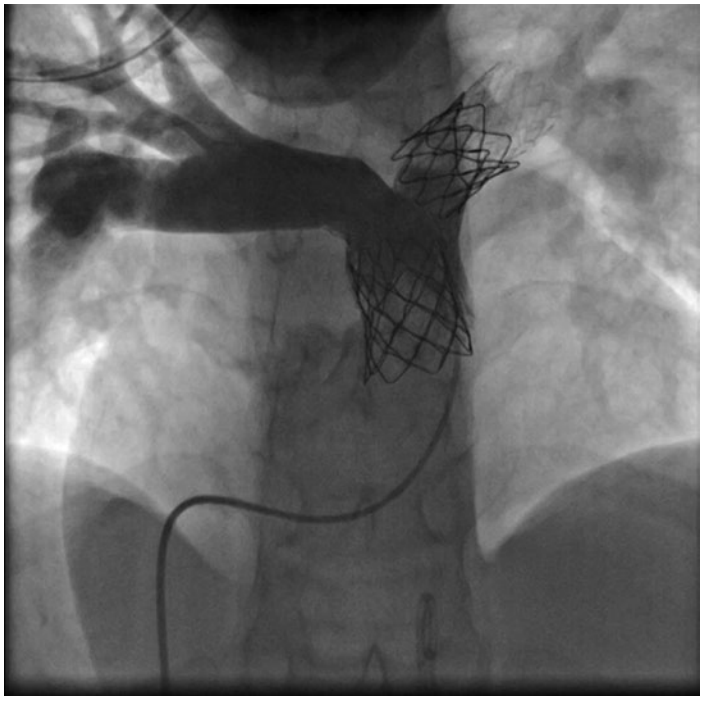

Fig. 24.8 A 15-year-old boy (58 kg, 174 cm) after surgical Fallot (TOF) correction with a transanular right ventricular outflow tract (RVOT) patch at 7 months of age and after implantation of a bare metal CP stent (22 mm) into the left pulmonary artery. Although no gradient is measured at the pulmonary bifurcation, bilateral stenosis is present. A 57-mm Andra xxl was implanted into the LPA. After opening to the RPA, a 47-mm Andra xxl stent was implanted into the RPA, and the entrance to the LPA was opened with an 18-mm balloon. Finally, a Melody valve (Medtronic, Minneapolis, MN, USA) was implanted with a 22-mm delivery balloon

(Mustang™ Boston Scientific, 60 mm) and inflate and deflate this balloon while advancing the long sheath into the pulmonary artery.

A difficult anatomy can exist whenever small left and right pulmonary arteries are connected to a dilated conduit, especially on the left side combined with enlarged right ventricle and

atrium. No possibility to get guiding from the anatomic structures. Thus, telescoping is helpful: long sheath, catheter, microcatheter, and wire in combination. When smaller balloons or stents should be placed, telescoping with a long sheath in combination with a guiding catheter can be helpful to bring the stent into position.

24.8.2 Choosing the Right Balloon Diameter

Care has to be taken to avoid undersizing of balloons when the vessels pump during the cardiac cycle. The best orientation is to measure the largest diameter usually in end systole.

Pre-dilation or sizing of the stenosis might be useful whenever a funnel-shaped stenosis is present.

In many cases, a circumscript stenosis can be revealed if a sizing with a low-pressure balloon is performed.

24.9 Pitfalls

- *When the distal end of the stent-carrying balloon extends to a small vessel*, the balloon with the stent can be pushed back during inflation leading to stent misplacement or dislodgment—in this scenario, select the shortest possible balloon for stent delivery.
- *Implanting a stent mounted on an undersized balloon due to miss-sizing*—often in very compliant stenosis with systolic–diastolic change in diameter. If the stent dislodges, try to keep wire position, remove the balloon and exchange it for a larger one in order to reposition the stent.
- *Extra hard stenosis* which cannot be dilated after stent placement will result in a suboptimal result with an incompletely expanded stent. Whenever an extra hard stenosis cannot be ruled out, a pre-dilation with a cutting balloon should be performed prior to stent placement.

24.10 Complications and Their Management

24.10.1 Dissection

- Dissection of balloon-dilated vessels might be partially acceptable in order to have a lasting effect of dilation.
- However, if larger layers of the vessel wall are floating partially free in the lumen, this may cause obstruction.
- In these cases, an attempt may be justified to re-apposition the layer to the wall by careful balloon inflation with a low-pressure balloon.
- Otherwise, stenting of the affected vessel may be mandatory.

24.10.2 Vessel Rupture

- Vessel rupture can cause bleeding into the interstitial lung tissue, into the bronchi, and into the pleural space. It may be immediately life threatening.
- Considerable bleeding may occur, and blood in the bronchi may impede an effective gas exchange.
- If possible, the ruptured vessel should be obstructed with a low-pressure balloon. Mandatory additional steps are sufficient volume supply, antagonizing of any anticoagulation and positive pressure ventilation.
- If bleeding from the bronchi cannot be controlled, selective ventilation by a double-lumen orotracheal tube can be indicated.
- Once the bleeding is controlled by these measures, the patient should be ventilated, and relaxation should be considered to avoid reactivation of bleeding by coughing.
- Surgical closure of the ruptured vessel may be successful if the bleeding site is surgically accessible.
- In desperate situations, partial pneumonectomy can be indicated.

24.10.3 Stent Embolization

- Stent embolization can be caused by undersizing the target vessel or by overestimating the stiffness of the stenotic vessel.
- And it can occur as a consequence of stent misplacement, for example, when the balloon dislodges during implantation.
- An attempt can be made to introduce a slightly larger balloon and load the stent on it by careful inflation.
- Once the stent is captured on the balloon, repositioning can be successful in some cases.
- In dislodged large stents, which have moved back into the pulmonary trunk, the stent can be maneuvered on a larger balloon even back through the tricuspid valve and then be "parked" in the IVC at a localization of adequate diameter.
- Smaller, softer stents can sometimes be removed through a large sheath after snaring.

24.11 Post-procedural Care

- After effective treatment of severe vessel stenosis in patients with high pulmonary artery pressures, respiratory discomfort can be an early sign of a reperfusion edema in the capillary bed behind the dilated vessel.
- In many centers, the administration of heparin for about 24 h and the prescription of low-dose aspirin is performed; however, there is no evidence-based data supporting it.

24.12 Follow-Up

- Interventional success can rarely be controlled by direct visualization of the treated vessel segment during follow-up.
- Thus, indirect signs have to be monitored.
- By echocardiography, right ventricular systolic pressure estimation, and quantification of tricuspid valve regurgitation, velocity is very valuable.

- Measurements of post-interventional flow distribution can best be achieved by MRI.
- Rarely, it may be necessary to perform a scintigraphy for this purpose. Flow velocities in front and behind a stent can help to detect in-stent stenosis by MRI.
- In patients with cyanosis, a relief in pulmonary vessel obstructions can lead to an increase in arterial oxygen saturation measured noninvasively by transcutaneous oximetry.

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Aortic Coarctation

25

Raul Ivo Rossi Filho
and João Luiz Langer Manica

25.1 Anatomic Description and Physiopathology

Aortic coarctation comprises roughly 7% of all known congenital heart defects, with an approximate frequency of 0.04% of live births. It is usually a discrete stenosis in the region of the ligamentum arteriosum. Rarely it can occur in the ascending aorta or the abdominal aorta. It may be associated with diffuse hypoplasia of the aortic arch and isthmus, sometimes associated with duct-dependent circulation. Isolated aortic coarctation may occur in 82% of cases and is the most common form detected in adults.

25.2 Clinical Scenarios

The clinical manifestation depends basically on the degree of obstruction and the importance of associated lesions. Aortic coarctation is diagnosed very often in asymptomatic adolescents or adults in the context of investigation for systemic arterial hypertension. It can also be found in routine medical screening in

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children with abnormal lower limb pulses or dorsal heart murmur. On the other hand, in the newborn it can present as a life-threatening situation with refractory heart failure just after closure of the arterial duct, requiring urgent intervention.

25.3 Indications for Treatment of Aortic Coarctation

Systemic hypertension with resting pressure gradient between upper and lower limbs greater than 20 mm of mercury (mmHg).

1. Demonstration of severe aortic coarctation by spiral computerized tomography, magnetic resonance imaging or angiography.
2. Presentation with congestive heart failure with or without associated cardiac lesion as may be the case in neonates and infants.
3. Mild aortic coarctation with:
   (a) Abnormal blood pressure response to exercise.
   (b) Left ventricular dysfunction.
   (c) Symptoms of exercise intolerance.
   (d) Associated lesions such as coronary artery disease and aortic insufficiency.
   (e) Exercise gradient of more than 20 mmHg.
   (f) Presence of left ventricular hypertrophy and/or left ventricular diastolic dysfunction.

25.4 Pre-procedural Imaging

Echocardiogram is the gold standard tool for diagnosis and indication for intervention based on Doppler assessment of flow in the descending aorta.

Magnetic resonance imaging (MRI) and computed tomography (CT) scans give us morphological aspects not available with echocardiographic evaluation and are of paramount importance for an adequate assessment and planning of the procedure. They

also play an important role in the evaluation of complications during follow-up.

Care must be taken not to increase radiation exposure in patients submitted to repeated CT scans. MRI has the advantage of the absence of radiation; however, metallic artifacts can preclude its use in patients previously treated with steel stents.

Three-dimensional rotational angiography is a very useful tool for aortic coarctation. A single acquisition can demonstrate the size of the ascending aorta, features from the aortic arch hardly imaged in two-dimensional studies like the presence of pseudoaneurysms in patients previously submitted to percutaneous or surgical procedures. The use of 3D-RA in this context reduces the number of necessary acquisitions as well the radiation exposure increases the diagnostic accuracy in patients with complex anatomy.

25.5 Surgical Treatment

Surgical treatment for aortic coarctation was first described by Crafoord in 1945 and improved the prognosis of the involved patients. Recoarctation rates vary from 8% to 35% depending on the surgical technique used and the time of follow-up. Surgical treatment is currently the gold standard for aortic coarctation in newborns and young children weighing less than 15 kg.

25.6 Balloon Angioplasty

Initial reports on balloon angioplasty demonstrated good results in patients who previously underwent surgical repair, despite increased incidence of reintervention in patients with long tubular narrowing, isthmus hypoplasia, or mild obstruction. The increased incidence of wall damage resulting in aneurysm formation after balloon dilatation of native aortic coarctation raises controversy regarding the employment of this simple technique.

25.6.1 Indications

1. Native discrete coarctation of the aorta without associated hypoplasia of the transverse arch and/or the isthmus.
2. Recurrent coarctation of the aorta following previous surgery or intervention—balloon angioplasty is the therapy of choice.
3. Occasionally in sick neonates or infants less than 3 months of age, balloon dilation may be indicated as palliation because of severe left ventricular dysfunction or surgery being associated with high risk.

25.6.2 Technique

1. General anesthesia.
2. Access: femoral artery—percutaneous puncture. Rarely, femoral venous or carotid or brachial or axillary arterial approach may be needed.
3. Anticoagulation: Heparin 50–100 IU/kg right after obtaining arterial access.
4. From the femoral arterial approach, a multipurpose catheter is used to cross the coarctation with the help of a soft tipped guidewire.
5. Guidewire is positioned in the ascending aorta, and then the end-hole catheter is exchanged for an angiographic catheter (for example, Pigtail or Multitrack).
6. Hemodynamic measurements are performed—aortic pressures: ascending, and descending. The advantage of using the Multitrack catheter is that the guidewire position can be maintained while repeated pullback measurements are made.
7. Aortography in left anterior oblique, right anterior oblique, and lateral projections is performed using any of the previously mentioned angiographic catheters. The projection has to be variable and depends on the anatomy. Recently, three-dimensional (3D) rotational angiography has been successfully employed in aortic coarctation to avoid multiple injections (Fig. 25.1).

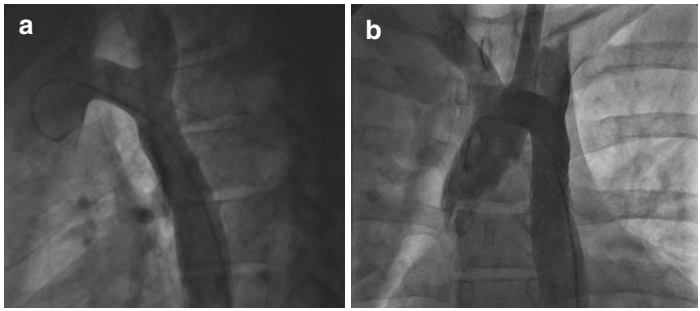

Fig. 25.1 Stent implantation with ideal result. (**a**) Angiography depicts a bare stent implantation which is properly apposed to the aortic wall. (**b**) Postero-anterior view depicts absence of aortic wall damage

8. Measurements (lateral projection is most commonly used): diameters of transverse arch distal to the brachiocephalic artery, distal to the left carotid artery and distal to the left subclavian artery, minimum diameter of the coarctation, aorta below the coarctation and the descending aorta at the level of diaphragm (Fig. 25.2).
9. The stiff exchange wire (such as Amplatz super-stiff) is positioned across the coarctation with the soft J-curve in the ascending aorta or in the right or the left subclavian artery (depending on the anatomy).
10. Selection of the balloon catheter:
    (a) Balloon diameter should not exceed the diameter of the aorta above the coarctation or at the level of the diaphragm.
    (b) It should not also exceed three times the diameter of the coarctation.
    (c) Low-profile balloon catheters should be used (such as Tyshak balloons). Low-pressure balloons may be effective in younger children, while high-pressure balloons are more effective in older children and patients with recurrent coarctation.

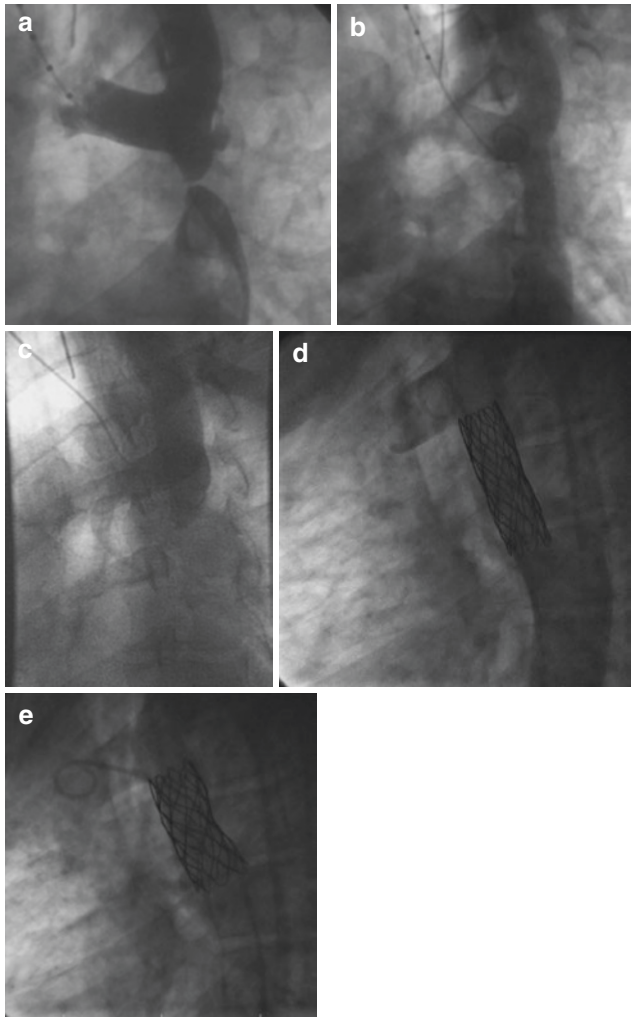

Fig. 25.2 Examples of uses of covered stents. (**a**) Atretic coarctation with aortic wall aneurysm. (**b**) LAO aortogram showing the final result of a covered stent placement which allowed adequate relief of the obstruction and also treated the aortic wall injury. (**c**), "Acquired" aortic atresia best imaged through a catheter placed via right radial artery. (**d**), Immediately after a covered stent placement which was (**e**), redilated at a later time with a larger balloon to best fit the aorta

11. Preparation of the balloon catheter: flush the guidewire lumen and remove the air from the balloon with syringe by creating vacuum.
12. Over the wire, exchange the angiography catheter for the balloon catheter.
13. Place the balloon at the level of the coarctation. Inflate the balloon with diluted contrast material (25% contrast + 75% saline). Appearance of the waist on the balloon indicates the site of coarctation. The balloon should be inflated till the waist disappears. Balloon should be kept inflated for approximately 10–30 s. After this time, the balloon should be deflated as quickly as possible. Additional balloon inflation is not recommended in the most cases, but may be required if the balloon slips during inflation or the waist has not been completely abolished.
14. After all the contrast material is removed from the balloon, it should be withdrawn through the sheath (continuous negative pressure is applied on the balloon lumen to diminish its profile).
15. Exchange wire position should be maintained.
16. Multitrack catheter is inserted over the wire to the ascending aorta.
17. Repeat aortography in the same projection as prior to balloon dilation to check the anatomical result of dilation. Measure the diameter of coarctation.
18. Repeat the hemodynamic measurements—pressures in the ascending and the descending aorta with pullback method.

25.6.3 Expected Results

1. Systolic pressure gradient less than 10 mmHg.
2. Increase diameter of aorta at the level of the coarctation.

25.6.4 Hints

1. In infants less than 3 months of age, balloon dilation can only be recommended as palliation when these patients have severe left ventricular dysfunction or are at a high risk for surgery. It

should be recognized that in this group of patients the restenosis rate is higher.
2. If crossing the coarctation with guidewire from femoral artery proves difficult or impossible, try to cross it from above (through an axillary or brachial artery approach).
3. Femoral artery pressure monitoring through the side port of the arterial sheath is very helpful in immediate assessment of dilation result.
4. Rapid right ventricular pacing may be useful for stabilization of the balloon position during inflation, particularly in older patients.
5. An Indeflator is useful to control and monitor the balloon pressure, but manual inflation can be performed with low-pressure balloons. This depends on individual operator's experience.
6. Avoid manipulation of the tip of catheters or guidewires in the dilated area or losing guidewire position and then trying to recross the dilated lesion.
7. In older patients, the risk of wall complications increases, so stent implantation should be considered as the primary treatment or covered stents should be available.

25.6.5 Pitfalls

1. In patients with large collaterals, the guidewire and the diagnostic catheters may pass easily into the collaterals instead of the coarctation.
2. Measurements needed to determine the size of the balloon should be accurate as errors in measurements may lead to complications.

25.6.6 Limitations

1. Patients with coarctation of the aorta coexisting with marked transverse aortic arch hypoplasia should be referred for surgery.

2. Patients with tubular or diffuse coarctation of the aorta and patients with aortic isthmus hypoplasia should be treated with stent implantation, especially in the older age group.

25.6.7 Main Complications

1. Aortic wall dissection:
   (a) Small dissection: additional balloon inflation for longer time (approximately 1–2 min). Repeat CT or magnetic resonance scan to follow the progress of the dissection and, if necessary, implantation of a bare or covered stent.
   (b) Larger dissection: implantation of a bare or covered stent during the same procedure.
2. Small aneurysm:
   (a) Repeat CT or magnetic resonance imaging scans to follow the progress of the aneurysm.
   (b) If necessary (when the diameter increases or there is a spiral aneurysm), implantation of a stent may be indicated.
3. Larger or increasing aneurysm:
   (a) Immediate implantation of a covered stent.
4. Other complications include aortic rupture (emergency surgery or covered stent implantation), femoral artery damage (thrombolysis or surgical repair).

25.6.8 After the Procedure

- Antihypertensive treatment (same as before the procedure).
- CT scan or magnetic resonance imaging assessment before discharge if there have been any complications during the procedure or 1 year later if the procedure was uncomplicated.

25.7 Stent Implantation

First reported in 1991 [1], stenting aortic coarctation has proven to be an effective procedure for both residual and native lesions, providing excellent immediate relief of the obstruction and con-

tinuing to provide beneficial effects at medium-term follow-up, mainly in patients weighing more than 20 kg.

The bare stents most frequently used are the Palmaz® stent (Cordis Corporation, Miami, USA), Palmaz-Genesis® (Cordis Corporation, Miami, USA), CP stents (NuMed Inc., Hopkinton, NY, USA), and Andrastent (Andramed GmbH, Germany).

The use of a stent covered with a layer of expanded polytetrafluoroethylene (e-PTFE) to treat aortic coarctation was first described in 1999 in a patient with coexistent aneurysm of the aortic wall. Case reports and small series contributed to augment the spectrum of patients that might benefit from this approach. Currently its use is also accepted in extremely severe aortic coarctations, in association with patent ductus arteriosus, previously implanted conduits, patients with inflammatory disease and long segment stenosis, advanced age, aortic wall disease (Marfan and Turner syndromes), acute aortic rupture after primary bare stenting as a bail-out situation, associated dilation of the ascending aorta, patients with an irregular aortic wall and those previously treated with the use of surgical patches. Some authors recommend its use in all cases of adolescent and adult with aortic coarctation or recoarctation, although this is not yet common practice.

The CP stent (NuMed Inc., Hopkinton, NY, USA) is a regular bare CP stent, which is involved with an expandable sleeve of ePTFE. It is available in lengths from 16 to 45 mm and can be dilated up to a maximal diameter of 24 mm. The Advanta V12 LD stent (Atrium Medical, NH) is a low-profile covered stent that is marketed in three lengths (29, 41, and 61 mm). It is pre-mounted on 12-mm, 14-mm, and 16-mm balloons and can be dilated to a maximal diameter of 22 mm. However, recent reports of Advanta stent collapse and necessity of stent redilation and new stent implantation denote lower radial force of this device, and it was no longer indicated for the treatment of aortic coarctation. Recently, the BeGraft Aortic Stent (Bentley InnoMed GmBH, Hechingen, GE) which is a premounted ePTFE-covered balloon-expandable Cobalt Chromium stent has been successfully described in the literature for the treatment of aortic coarctation. Balloon sizes range from 12 (9 F) to 24 mm (14 F) and stent length ranges from 19 to 59 mm.

The need for larger sheaths to implant covered stent still limits its use in the pediatric population. Additional advances in the development of materials, such as bioabsorbable stents, will expand the indications for percutaneous treatment of aortic coarctation in children.

25.7.1 Indications

1. Dilatable stenosis but which recoiled after balloon angioplasty
2. Tubular or long segment coarctation
3. Coarctation coexisting with hypoplastic isthmus
4. Recurrent coarctation following surgery or intervention resistant to balloon angioplasty

25.7.2 Technique

The procedure follows the same steps from balloon angioplasty.

1. The size of the balloon is chosen to equal that of the distal arch at the level of the origin of the subclavian artery. If hypoplasia of the distal arch is present, the diameter of the transverse arch is used.
2. A super-stiff guidewire is usually positioned distally into the right subclavian artery or ascending aorta. Some authors recommend the use of left subclavian artery to deliver the stent in specific situations in accordance with coarctation anatomy.
3. When a near atretic aortic coarctation is found, predilation of the aortic segment using small-size balloons is sometimes necessary to allow a large Mullins sheath to cross the obstruction. A radial artery approach can be necessary to cross from above a pinhole orifice and snare a guidewire to perform an arterio-arterial loop for posterior insertion of the long sheath from the femoral artery.

4. The same approach can be used in atretic coarctations in which a radiofrequency perforation is performed to make way for covered stent implantation.
5. The long sheaths should be 1 or 2 F larger than the sheath needed for the balloon when bare stents are implanted and 3–4 F larger for covered stents. Usually, long sheaths ranging from 8 to 14 F are used.
6. The balloon catheter is chosen to be longer than the stent length, and some authors prefer to crimp the stent in a partially inflated balloon in order to assure opening of the stent from its extremities. The length of the stent should be adequate to cover the lesion and (if needed) to treat isthmus hypoplasia. Careful measurement is paramount to avoid jailing the brachiocephalic arteries, especially when using covered stents.
7. The balloon is then manually inflated up to the pressure recommended by the manufacturer, which is usually up to 4–6 atm.
8. Angiography is performed during and after stent placement through the side arm of the sheath or by using a pigtail to assess the result and rule out aortic dissection or rupture.
9. Pressure measurements above and below the stent and pump angiograms are recorded after the procedure. The sidearm of the Mullins sheath can be used to measure the final gradient.
10. Hemostasis is achieved by manual compression, vascular closure devices, or even by surgical repair.
11. All patients should be on antibiotic prophylaxis (cephazolin for 24 h).

25.7.3 Tips and Tricks

The key cornerstones of stent implantation in coarctation of the aorta are:

1. Pre-implantation assessment: we need precise delineation of the anatomy of the lesion and very accurate measurements of the arch.

2. Once the anatomy of the lesion is known, one can assess its compliance with a low-pressure balloon. This subject is very controversial because the near-totality of the coarctation expands at 4–6 atm and predilation can cause loss of the highly necessary tight waist, which helps to secure the stent in place. It can also be associated with late aneurysm formation. On the other hand, it may be helpful to identify patients with pseudocoarctation.
3. Implantation technique: Stent stability is a must. It can be obtained simply by implanting a stiff wire far into the right brachial artery or using overdrive pacing to obtain a cardiac standstill (highly necessary in transverse arch lesions or in hyperdynamic circulation; such is the case in aortic regurgitation). Another technique that can be used is to create a radial-femoral arterial rail, which will provide absolute control of the balloon.
4. The stent and balloon unit: The choice of the balloon can affect the outcome. Balloon-in-balloon (BIB®) balloons (NuMed, Hopkinton, NY, USA) are excellent and are devised for patients with aortic coarctation. Their ability to partially expand the stent can avoid malposition. It also precludes the sharp edges of some stents impinging into the aortic wall. However, they add bulk, and sometimes, it is necessary to trade additional safety for a lower profile. Do not be concerned about using regular balloons to implant stents in the aorta. Careful and slow inflation also help avoid stent malposition.

   Hand crimping the stent is a technique that many of us still use. Crimping a bulky stent in a low-profile balloon requires care and patience. After choosing the correct spot, hand crimp it slowly using rolling movements of your fingers and, at the same time, press the stent against the balloon's shaft. Do not forget to pass a wire through the balloon beforehand. Failing to do so may lead to compression of the balloon's lumen. Some also use a cardiac tape to finish the crimping with more pressure. Covered stent manipulation calls for dry gloves to avoid separation of the thin-glued ePTFE layer from the stent.
5. Position control: This is most commonly obtained using the sidearm of the long sheath. When using covered CP stents,

care must be taken to avoid peeling off the ePTFE layer when the tip of the sheath is too close to the stent. Control angiography can also be obtained through a second arterial catheter from the radial artery, or into the ascending aorta with a Berman catheter via a transeptal puncture.

6. In younger patients, surgical cut-down of the femoral, iliac, or carotid artery may be needed for the introduction of the sheath.
7. Because of the need for a large sheath, preparation of the femoral access with special devices (for example, Perclose) is useful for hemostasis after the procedure.
8. In order to prevent femoral injury, Bruckheimer suggested serial approach to stent implantation. The use of a small balloon reduces the size of the delivery system required and is followed by serial dilations of the implanted stent. Care must be taken not to increase the risk of stent slippage at implantation due to balloon underestimation.
9. Avoid stent overdilation. It may predispose to neointimal hyperplasia.
10. Positioning the bare stent across the left subclavian artery orifice does not diminish flow through it and therefore is not contraindicated. However, the close proximity of the origin of the left subclavian artery to the coarctation area can be a problem when a covered stent is needed. Tsai et al. came up with an elegant solution for this problem, when they managed to perforate the e-PTFE layer and created a hole through the stent using the stiff end of a guidewire previously placed into the left subclavian artery. All of this trouble can be avoided with a pre-procedure CT or MRI scan of the intra- and extracerebral vessels which will demonstrate the presence of a left vertebral artery that can be supplied by the basilar system.
11. Sometimes, it is helpful to advance the sheath over the balloon taking care not to push the stent forwards.
12. Femoral artery pressure monitoring through the side port of the arterial sheath is very helpful in immediate assessment of dilation result.
13. In severe coarctation, staged stent dilation over a period of several months is an acceptable alternative method to avoid

aortic wall complications such as dissection or aneurysm formation.
14. Patients with coarctation of the aorta coexisting with transverse aortic arch hypoplasia may need to be dealt with by implantation of more than one stent.
15. Age for stent implantation should preferably be greater than 10 years.

25.7.4 Limitations

1. Implantation in younger patients (neonates and infants) should be performed only in exceptional circumstances (critical and life-threatening situations, in cases not suitable for balloon angioplasty, or when there is early surgical recoarctation after extensive arch reconstruction with foreign material).
2. It has been described that there is a possibility of breaking stents previously implanted in neonates with staged ultra-high pressure balloon dilatations and new stent implantation in order to prevent deformation of the struts.
3. Breakable stents have been reported in recent literature despite no studies showing follow-up of these stents in growing children (CCI 2016).

25.7.5 Complications

25.7.5.1 Aortic Wall Complications

Bare stents reduced but did not abolish aortic dissection or aneurysm formation in comparison with balloon or surgical aortoplasty. Previous studies reported that the incidence of this kind of complication ranged from 0% to 16% and was more common in older patients and those with tight native or complex coarctation.

The use of covered stents clearly decreased the incidence of aortic dissection or rupture. However, few cases of this kind of complication are still reported in the literature. Cystic medial necrosis is an important risk factor for this complication.

Aortic aneurysm or acute aortic rupture must be dealt with the implantation of a covered stent. During bail-out situations, an inflated balloon in the site of the injury can be a life-saving approach while covered stent is prepared for implantation.

25.7.5.2 Technical Complications

Stent migration is the most frequently encountered technical complication occurring in up to 5% of bare stent implantation. It is unlikely to occur during long-term follow-up and has never been reported during the deployment of covered stents. This phenomena is probably related to the development of the NuMed BIB® (NuMed, Hopkinton, NY, USA), which provides better control of the stent position, and to previous experience with bare stents, which demonstrated some risk factors for stent migration that can be avoided during covered stent implantation (balloon catheter larger than the aorta proximal to the coarctation site and the use of undersized balloon diameter in cases of pseudocoarctation). The use of adenosine or overdrive pacing during deployment has already been described during bare stent implantation to avoid stent migration.

Balloon rupture with inadequate stent expansion may be prevented by avoiding kinking of the balloon/stent assembly by the use of newer stents with softer ends and by the use of BIB systems.

The possibility of side branch occlusion during covered stent implantation is one concern, especially because occlusion of the spinal artery can lead to paraplegia. However, as the spinal artery usually originates below the diaphragm, occlusion of the spinal artery is unlikely to occur except in cases of stent embolization—a fact that, until now, was never reported in the literature.

25.7.5.3 Access-Related Complication

Acute arterial occlusion is a concern in patients in the first year of life.

Bleeding, local hematoma, and arteriovenous fistula are not uncommon due to the use of large sheaths. Some authors routinely recommend surgical cut-down, mainly in children submitted to covered stent implantation, in order to avoid those complications. Another useful approach associated to diminished

incidence of access-related complications is the use of vascular closure devices. Manual compression is not prohibitive for arterial hemostasis and, if well performed, is related to a low incidence of vascular complications.

25.8 Restenosis

Restenosis is rarely seen and redilation is almost completely limited to occurrence as part of a planned serial procedure due to severe aortic coarctation, somatic growth, or less frequently, due to neointimal hyperplasia.

25.9 Stent Fracture

Stent fracture is described in the literature after CP or Palmaz stent implantation. It could predispose to lumen obstruction due to two factors: loss of the structural integrity and neointimal hyperplasia. Moreover, the fractured strut can be associated to aortic wall disruption at the level of the fracture margins or to distal strut embolization, despite there is no reported case of such a complication in the literature. Therefore, some authors suggest that when it occurs, particularly when associated to stent instability and even without lumen obstruction, another stent implantation is recommended (Fig. 25.3).

25.10 Post-procedural Care and Follow-Up

Patients are discharged after 48 h. Although there is no evidence supporting the use of aspirin (dose of 3–5 mg/kg once daily), some authors advocate its use for 6 months after stent implantation. Patients are advised to avoid physical activity for 30–60 days.

Outpatient follow-up consists of clinical assessment, including blood pressure and the need for antihypertensive medication, 12-lead electrocardiogram, chest X-ray, and transthoracic echocardiogram at 1, 6, and 12 months, and annually thereafter. An

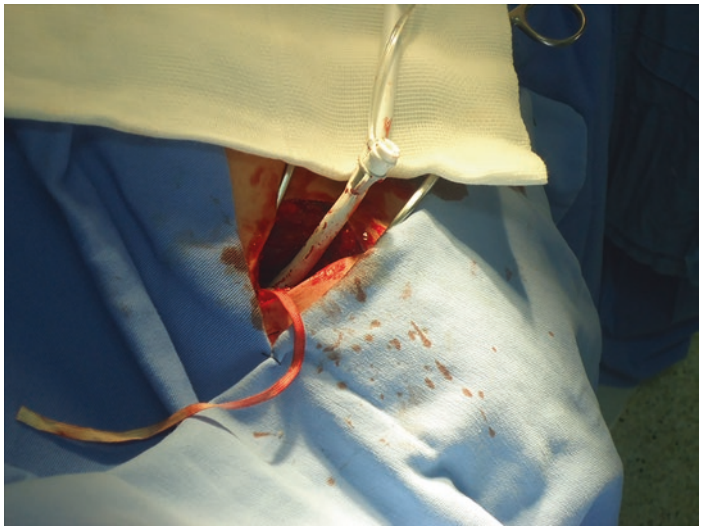

Fig. 25.3 Axillary artery dissection in an adult with bilateral femoral artery occlusion who needed a covered stent implantation for post-dilatation aneurysm. The sheath size is 14 F

exercise test can be performed at 3 months and then at 12 months after the procedure. Spiral computed tomography is recommended between 6 and 12 months after the procedure; however, in complex cases, it can be performed 30 days after the procedure.

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26 The Role of Transcatheter Interventions in Middle Aortic Syndrome

Diego Porras

26.1 Introduction

Mid-aortic syndrome (MAS) is characterized by obstructive lesions of the aorta distal to the aortic isthmus and proximal to the iliac bifurcation (mid-aorta), regardless of the etiology (Fig. 26.1). Involvement of at least one branch of the abdominal aorta was seen in 85% of patients, and 15% of patients had complete obstruction of at least one branch of the abdominal aorta. The most common branches of the abdominal aorta involved were the renal arteries (69%), followed by the celiac artery (60%), and the superior mesenteric artery (60%) [1]. MAS is the most common clinical syndrome associated with stenotic aorto-arteriopathy in children [2].

Severe MAS is associated with significant morbidity and mortality. Data regarding the natural history of MAS are limited to early reports which showed that approximately half of untreated patients died at a mean age of 34 years, with less than 20% survival reported after age 40 years [3]. While in the modern era the majority of patients present with severe hypertension on routine

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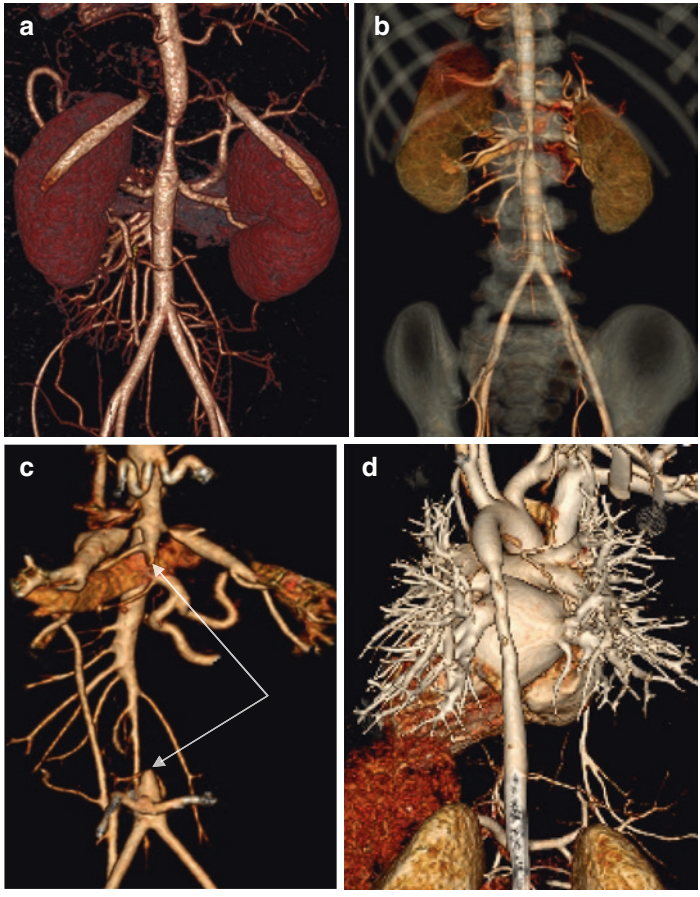
screening during childhood and are largely asymptomatic, MAS can present during infancy and childhood with a hypertensive emergency. Signs and symptoms include severely elevated upper extremity blood pressures in the presence of end-organ failure and their sequelae, including acute heart failure, stroke, acute renal failure, and intestinal ischemia. Medical management of renovascular HTN caused by MAS is one of the cornerstones of therapy, however, in the setting of severe anatomic obstruction of the mid-aorta and the renal arteries, it can have limited success [1, 2]. Invasive intervention, including surgery or transcatheter interventions, is often necessary in order to address the anatomic obstruction(s), so that adequate blood pressure control can be achieved. Other goals of invasive intervention are to relieve symptoms and to reverse or prevent end-organ damage. Surgical techniques are varied and are generally associated with good long-term results, although some of the procedures are quite complex and

Fig. 26.1 Anatomic characteristics of mid-aortic syndrome. Three-dimensional reconstructions of CTAs in patients with obstruction at different levels of the mid-aorta. (**a**) Dorsal view of supra-renal obstruction. (**b**) Frontal view of abdominal aortic obstruction that starts several millimeters above, and extends several millimeters below, the take-off of the renal arteries (intra-renal). There is also severe, proximal stenosis of the right renal artery and atresia of the left renal artery. The left kidney is smaller than the right kidney, consistent with atrophy secondary to chronic hypoperfusion. (**c**) Dorsal view in a patient with atresia of the infra-renal abdominal aorta. The aorta has a long segment of atresia (whit arrows) starting below the take-off of the renal arteries and gets reconstituted distally be collaterals. There is also severe bilateral renal artery stenosis. (**d**) Dorsal view of long-segment, irregular stenosis of the proximal and middle thirds of the descending thoracic aorta, starting several millimeters distal to the aortic isthmus and extending for several centimeters behind the left atrium of the heart. (**e**) Frontal view of abdominal aorta obstruction starting at the level of the renal arteries and extending several millimeters. There is bilateral renal artery stenosis and an atrophic right kidney. (**f**) Dorsal view of a diffusely hypoplastic mid-aorta, typical of mid-aortic syndrome in Williams syndrome. There is diffuse hypoplasia of the aorta starting in the distal third of the thoracic aorta, with the a relatively long-segment of more severe narrowing in the distal thoracic aorta. The abdominal aorta continues to be hypoplastic, although it resumes a slightly larger lumen. There is also bilateral proximal renal artery stenosis

may be associated with significant morbidity [1, 4–6]. Balloon angioplasty and stenting have been used as a palliative options to avoid surgery on the developing aorta. Most reports show encouraging results, though limitations include technical failures, iatrogenic tears and dissections, aneurysms, and re-stenoses [1, 7, 8].

Management of MAS is complex, requiring a multidisciplinary approach. A combination of medical management and invasive strategies is often employed in any single patient and can result in



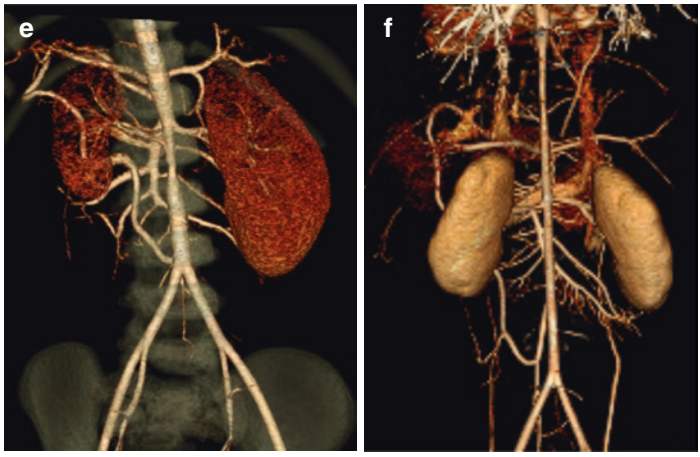


Fig. 26.1 (continued)

adequate control of HTN and preservation of end-organ function, even in patients with severe MAS and those who present at an early age [1].

26.2 Factors to Consider in the Invasive Management of MAS

26.2.1 Associated Diagnoses

Approximately half of the patients diagnosed with MAS have an underlying associated diagnosis, most commonly Williams syndrome, familial supravalvar aortic stenosis (SVAS), neurofibromatosis-1 (NF-1), and Alagille syndrome. The vascular phenotype in each of these syndromes can be quite characteristic (Fig. 26.2). Even in patients without the classical stigmata of these syndromes, mutations in the genes associated with them have been reported in a significant proportion of patients with so-called *idiopathic* MAS [9]. There is still much to learn about the etiology of MAS. Currently, multiple heterogeneous conditions are often grouped together under one diagnosis of MAS. Although they

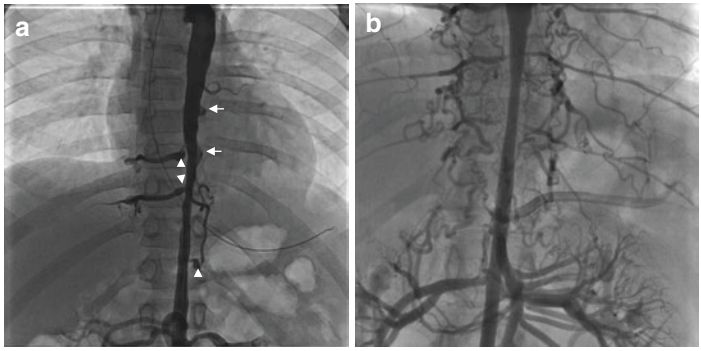


Fig. 26.2 Angiograms showing vascular phenotypes associated with specific etiologies of mid-aortic syndrome. (**a**) Antero-posterior view of the aorta in a patient with neurofibromatosis type 1 (NF-1) and long-segment obstruction of the mid-to-distal thoracic aorta. Notice the irregular lumen of the affected segment of aorta, as well as the aneurysms seen along its length (white arrows). Also, notice the small aneurysms seen in some of the enlarged intercostal arteries (white arrow heads). These findings are characteristic of MAS associated with NF-1. (**b**) Antero-posterior view of the aorta in a patient with Williams syndrome and long-segment narrowing of the distal thoracic aorta. Notice that the aorta becomes narrow at the level of the aorta that is behind the most caudal portion of the cardiac silhouette and slowly tapers in diameter with its most narrow portion being at the level of T11, then slowly increasing in diameter distally. However, the aorta remains diffusely hypoplastic, which is typical of MAS associated with Williams syndrome. Bilateral proximal renal artery stenosis can also be appreciated. (**c**) Frontal view of a patient with tuberous sclerosis and severe narrowing of the descending thoracic aorta. (**d**) Oblique view of the distal thoracic and proximal abdominal aorta in a patient with idiopathic MAS. Idiopathic MAS has variable phenotypes. In this patient, the lumen is very irregular, and there are several aneurysms in the aorta and a large aneurysm of the celiac artery (white arrow), which becomes atretic after the aneurysm. There is also severe narrowing of the proximal superior mesenteric artery and the renal arteries. (**e**) Lateral view of the abdominal aorta in a patient with Takayasu's arteritis, showing mild narrowing of the aorta starting above the take-off of the celiac artery and extending to the level of the take-off of the superior mesenteric artery. There is atresia of the celiac artery (white arrow) and mild narrowing of the superior mesenteric artery. (**f**) Frontal view of the abdominal aorta in a patient with Alagille syndrome. There is mild narrowing of the aorta at the level of the take-off of the renal arteries, and there is severe stenosis of the right renal artery (white arrow). The narrowing in this artery is characteristic of patients with Alagille and MAS. While the narrowing of the aortic branches in other etiologies of MAS characteristically involves the ostium and very proximal segment of the artery, in Alagille syndrome, it tends to spare or is very mild at the level of the ostium but then becomes severe in the proximal third of the artery. The left renal artery is nearly atretic and is only seen on selective angiograms (not shown; see Fig. 26.6b)

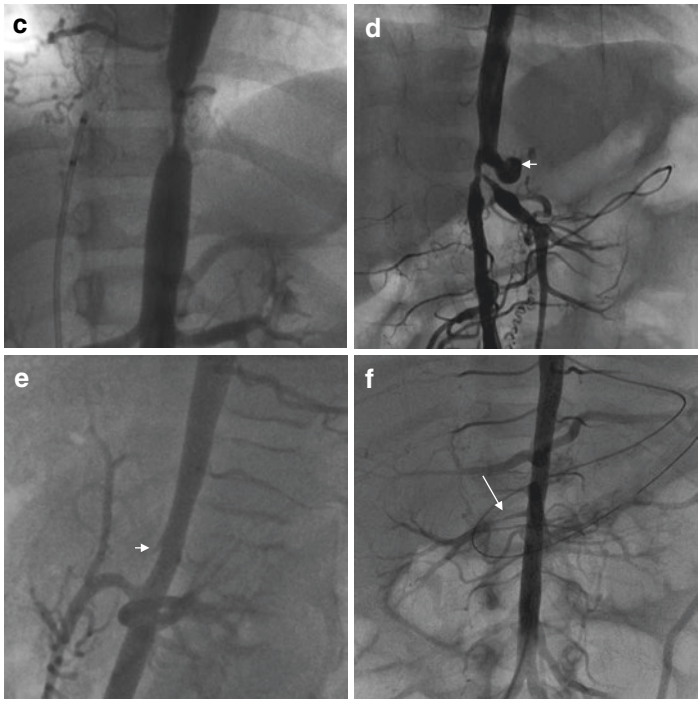


Fig. 26.2 (continued)

may share similar aortic lesions, it is important to keep in mind that the underlying or associated diagnoses may play a role in the pathophysiology, natural history, prognosis, and response to therapy. For example, the diagnosis of NF-1 has been shown to be a risk factor for vascular complications of endovascular interventions [1]. Similarly, patients with active vascular inflammation at the time of intervention may be at higher risk for vascular complications, and controlling the inflammatory process before intervening is preferable. As more is learned about each specific etiology of MAS, specific management strategies to optimally approach each diagnosis will likely emerge.

26.2.2 End-Organ Function

1. Heart
   (a) *Left ventricular hypertrophy* is relatively common in children with MAS and resolved in the majority of the patients, although this resolution can take several years (median >2 years) [1].
   (b) *Systolic ventricular dysfunction* on the other hand is less common in this population and, when present, should be considered an emergency. It is mostly seen in advanced cases and is a major indication for anatomic intervention.
2. Kidneys
   (a) *Chronic renal insufficiency*: While advanced chronic kidney disease is not highly prevalent in patients with MAS, mild to moderate decreases in GFR are relatively common [1, 10]. Although this level of renal compromise is rarely clinically significant, it may progress and become an important source of morbidity.
   (b) *Acute kidney injury and failure*: Approximately 15% of patients with MAS present in a hypertensive emergency, often including acute renal failure, sometimes requiring renal replacement therapy. In our experience performing catheterization procedures in MAS patients, we have not had any cases of clinically significant contrast-related acute kidney injury after endovascular procedures. However, we still consider all patients with MAS to be at high risk for this complication and take every precaution to prevent it.
3. Brain
   Neurovascular involvement in patients with MAS has been described, including moya-moya disease. Early diagnosis of a neurovascular abnormality in patients with MAS is very important and may affect their management, since interventions to aggressively lower the blood pressure may result in acute cerebral hypoperfusion and stroke if the patient has stenotic lesions in the cerebral vasculature. Therefore, it is very

important to obtain detailed cerebral arterial imaging to rule out such lesions in all patients with MAS before interventions are undertaken.

26.2.3 Anatomic Characteristics

The anatomic characteristics of the obstructive lesions in MAS can be quite variable (Figs. 26.1 and 26.2). It is important to describe the anatomy in detail, including:

1. Type of obstruction: discrete narrowing, slowly tapering lesion, diffuse hypoplasia, irregular lumen, presence of calcifications on the aortic wall, concentric or eccentric wall thickening, presence of aneurysms.
2. Site of most proximal involvement and length of mid-aorta involved.
3. Site of most severe obstruction within each lesion.
4. % stenosis: minimal luminal diameter compared to the diameter of the aorta that appears normal above and below the lesion.
5. Branches involved: including the degree of stenosis of each and which part of the vessel is involved (ostial narrowing, proximal, mid or distal third, etc.).

It is also important to note the pressure gradient across the area of obstruction. However, it must be kept in mind that in patients with well-developed collateral circulation, the pressure gradient may be low or absent despite the presence of severe mid-aortic obstruction.

26.3 Invasive Management of MAS

When considering indications for invasive management in patients with MAS, there are no widely accepted or validated evidence-based guidelines. Multiple factors need to be considered [1, 10, 11], including:

1. Anatomic substrate: patients with mid-aortic stenosis ≥60% are highly likely to require invasive management, whereas patients with stenosis <40% are unlikely to require such management for the mid-aortic obstruction itself [1]. Even if the degree of stenosis of the mid-aorta is mild, invasive management of severe branch stenoses may be required.
2. Response to medical therapy: HTN refractory to medical management or significant side effects from antihypertensive medications.
3. Evidence of end-organ damage despite medical management of hypertension.
4. Debilitating symptoms like claudication or abdominal angina.

The choice of invasive strategy depends on the anatomy, patient age, and condition. At our center, we have tended to favor early intervention, which often means using percutaneous techniques as palliative measures with the goal of allowing growth and preservation of end-organ function until the patient is of an age and size that is more amenable to definitive corrective surgery.

26.4 Role of Transcatheter Interventions in the Management of MAS

The roles of transcatheter intervention in the management of MAS can be divided into supportive roles and primary therapies.

1. **Supportive Roles of Transcatheter Interventions in the Management of MAS**

   (a) *Stabilization of the unstable patient*
   Patients presenting with hypertensive emergency and end-organ dysfunction, including acute heart failure and acute renal failure, often need to be stabilized before any type of surgical intervention. These patients can present with severe left ventricular dysfunction and pulmonary edema, requiring mechanical ventilation and even extracorporeal membrane oxygenator support. They can often also require

acute renal replacement therapy. Transcatheter interventions can help improve the anatomic substrate, allowing for acute relief of obstruction resulting in reduction in ventricular afterload, improved renal perfusion, improved response to medical management of hypertension, and ultimately, improvement of end-organ function. This strategy can allow for delay of surgery until the patient is a much better surgical candidate and may even allow for the patient to grow so that they can later undergo a single, definitive surgery. This situation is encountered in patients presenting acutely with severe obstruction or even complete occlusion of the abdominal aorta and/or the renal arteries. In these patients, the aorta and the renal arteries can be recanalized, balloon dilated, and stented, as seen in Fig. 26.3. It is likely that patients presenting acutely with complete obstruction of the aorta or the renal arteries are

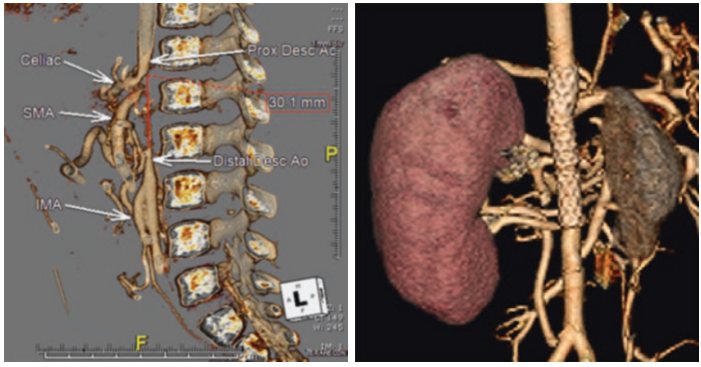


Fig. 26.3 (**a**) Three-dimensional reconstructions of a CTA in a patient with idiopathic MAS presenting with atresia of the abdominal aorta extending from immediately after the take-off of the celiac artery to just proximal to the inferior pole left renal artery. There was also atresia of the superior mesenteric artery (which filled retrograde through collaterals), the right renal artery and the superior pole right renal artery. (**b**) Dorsal view of a three-dimensional reconstruction of a CTA after the patient underwent recanalization, balloon dilation, and stenting of the abdominal aorta, as well as recanalization and balloon dilation of the superior pole left renal artery. The right kidney was atrophic and was later removed through laparoscopic nephrectomy

more amenable to recanalization because the complete occlusion may be a combination of severe stenosis and acute thrombosis, as opposed to patients who have complete occlusion of the aorta but present with no symptoms, in whom it is likely that the occlusion is chronic and may be less amenable to recanalization using transcatheter techniques.

(b) *Palliation of severe obstructive lesions to delay surgery and allow for somatic growth*
Patients presenting during early childhood with severe hypertension that is refractory to maximal medical management are often not considered good surgical candidates because of their size. If an aorto-aortic bypass is employed, for example, it would need to be replaced when the child outgrows the graft in the future. In this situation, transcatheter strategies can be used to palliate the obstruction, allowing for improved response to medical management and for somatic growth until the child reaches a size at which a more definitive corrective surgery can be carried out. Techniques include angioplasty with high-pressure balloons, cutting balloons, and/or stents. Often, this approach requires serial procedures to be able to reach a certain anatomic goal and to treat any recurrence or progression of disease.

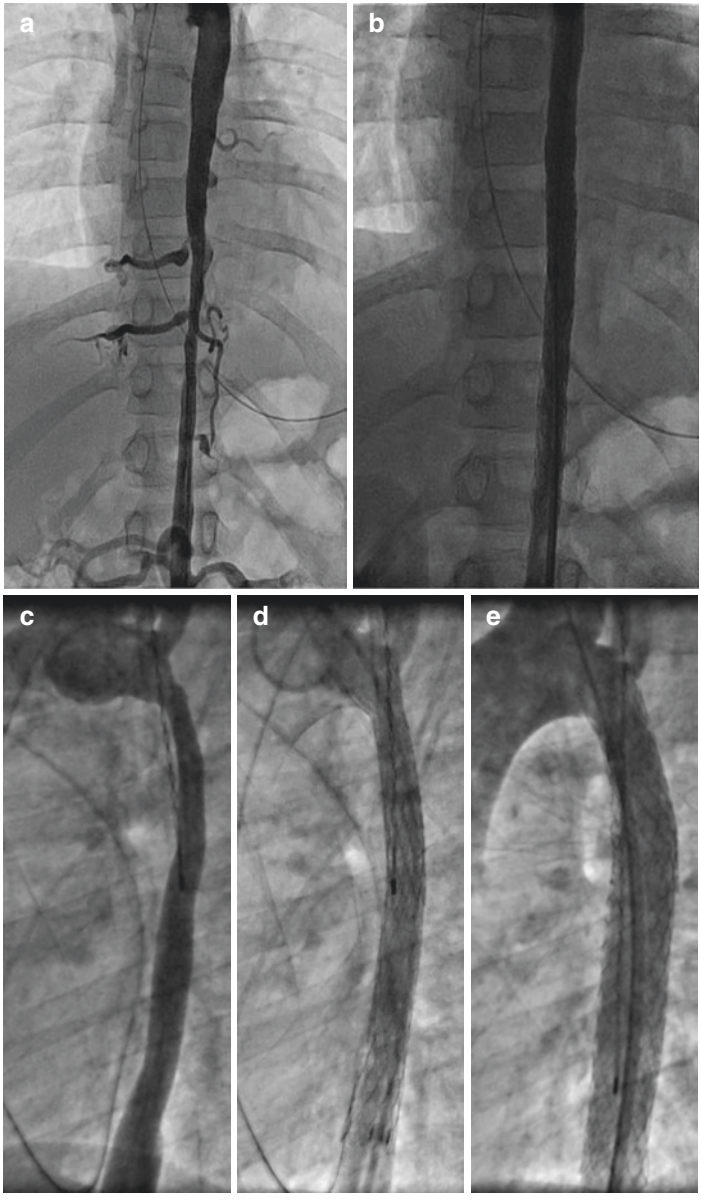
(c) *Re-intervention on prior surgical sites*
Patients who have undergone surgery for MAS may present with discrete lesions at prior surgical sites, which tend to respond quite well to balloon angioplasty. Other post-surgical complications that can be treated using transcatheter techniques include acute thrombosis of grafts, which can be treated with mechanical clot extraction techniques and/or direct infusion of thrombolytic agents for acute lesions. Chronic thrombotic occlusion (CTO) wires and standard balloon and stent angioplasty can prove useful for more chronic lesions. Often, a combination of these techniques is necessary to address thrombotic lesions.

2. **Role of Transcatheter Interventions as Primary Therapy in MAS**

   (a) *Thoracic aorta lesions*

   Lesions in the thoracic aorta represent a type of MAS lesion for which a transcatheter approach is particularly appealing for several reasons. Surgery usually requires a thoraco-abdominal bypass graft, which has several downsides, including that it generally is performed under atrio-femoral or aorto-femoral cardiopulmonary bypass and, in general, is a relatively complex operation involving thoracic and vascular surgery teams. The grafts used are usually synthetic and will not grow with the patient, so the surgery will generally be delayed until an "adult-sized" graft can be employed, leaving the patient with significant stenosis while awaiting surgery. On the other hand, transcatheter interventions, like primary stenting of the lesion (Fig. 26.4) can give an excellent acute result in a much less invasive manner. While it will require multiple catheterizations to gradually increase the size of the stent as the child grows, each intervention is relatively simple, requiring a single-night observation period in the hospital in most patients. Other reasons that this anatomic location is quite amenable to stenting include the fact that there are less major branches of the aorta in this region, and the fact that the stents are protected inside the thoracic cage, making them less vulnerable to deformation or crushing, which can be a problem when using balloon expandable stents in the abdominal aorta during childhood. Some important

Fig. 26.4 Use of stents to treat MAS involving the thoracic aorta. (**a**, **b**) Frontal views of the thoracic aorta in a patient with neurofibromatosis type-1 before (**a**) and after (**b**) use of bare metal stents to treat the narrowing. (**c**–**e**) Lateral views showing staged primary stenting of the thoracic aorta in a patient with idiopathic MAS involving the proximal and middle thirds of the thoracic aorta before (**c**), after the initial stenting procedure (**d**) and after the final dilation of the stents 3 months later (**e**)


a
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e

factors to keep in mind when considering the use of balloon-expandable stents for thoracic aortic lesions in MAS include:

- Use stents that are able to easily reach diameters of 16–20 mm in the future. While placing a "premounted stent" requires a smaller introducer sheath and is technically less demanding, these stents will only be able to reach 11–12 mm. The adult aorta is much larger than this, and therefore, the stent will invariably create an iatrogenic, highly resistant lesion in the future. We prefer using robust stainless steel stents like the Palmaz XL because of the risk of stent fracture in the thoracic aorta, which makes other stents like the Genesis XD and the Cheatham-Platinum stents less ideal. Covered stents are appealing because they may be less prone to in-stent stenosis and may be safer in terms of aortic wall trauma causing acute issues. However, there is no data to support their superiority in this situation, and they have the downside of excluding side branches, which in the thoracic aorta may have the potential of causing acute spinal ischemic injury, at least theoretically (see below). We therefore reserve the use of covered stents for patients that have aneurysms or a tear, or in patients that have had recurrent in stent stenosis after use of bare metal stents.
- Spinal cord perfusion should be kept in mind: bare metal stents should not obstruct flow to side branches, even if the side branches are jailed. However, it is important to keep spinal perfusion in mind and, when possible, avoid covered stents that may exclude the artery of Adamkiewicz. This artery usually arises from a left posterior intercostal artery between T9 and T12. Digital subtraction angiography and CTA prior to the procedure are useful techniques to identify this artery and to better understand spinal perfusion in these patients.

(b) *Discrete renal artery lesions*
Renal artery lesions seen in MAS are usually proximal lesions, involving the ostium of the renal artery and the very proximal renal artery, but rarely extend past the proximal third of the renal artery. These lesions respond very well to cutting balloons and high-pressure balloons (Fig. 26.5). Stents are also often used and have been shown to be effective. Often, these lesions are quite resistant; however, it is important to ensure the waist on the balloons are fully resolved before a stent is placed. We usually perform balloon dilation with high-pressure balloons up to the diameter of the distal "normal" renal artery. If there is a waist of the balloon that does not resolve at ~12 ATM, we use an appropriately-sized cutting balloon (based on the size of the waist on the prior balloon) and then re-dilate with the high-pressure balloon to ensure the waist is resolved. If the waist is resolved, but there is not enough angiographic improvement in the stenosis, we place a bare metal balloon-expandable stent. Usually, the preferred stent in this location is a coronary stent, ideally a drug-eluting stent. If the vessel is >4 mm, then a pre-mounted biliary stent usually works well.

(c) *Discrete lesions of the splanchnic arteries*
Patients with MAS frequently have discrete stenosis of the origin and proximal portion of the celiac artery and/or the superior mesenteric artery. The inferior mesenteric artery can also be involved, but is spared in most patients with MAS. It is rarely indicated to treat these lesions because there is significant redundancy in these circulations. We have found that if one of the three splanchnic vessels is widely patent (celiac, SMA, or IMA), the patients rarely have any issues with intestinal perfusion. Therefore, we rarely address these lesions surgically or in the catheterization laboratory. If necessary, transcatheter techniques similar to those used for renal artery stenosis (see above) are very effective.

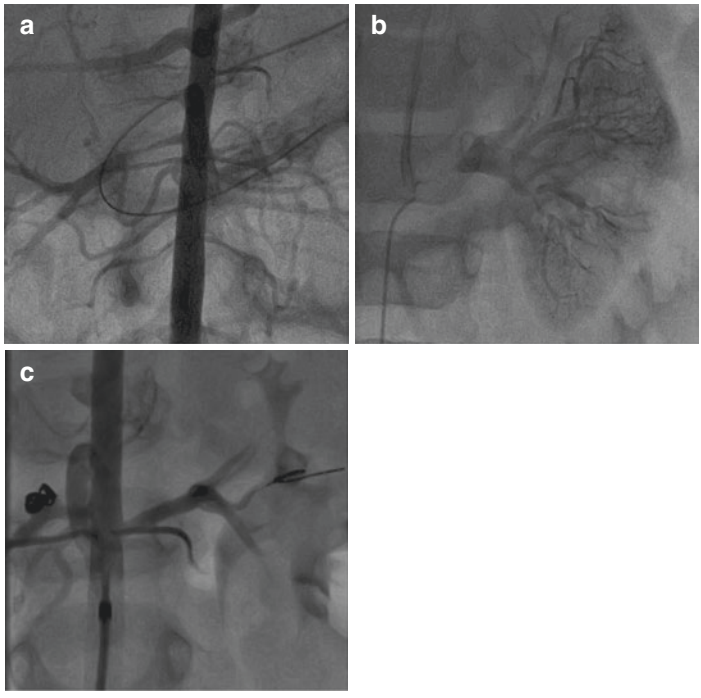

Fig. 26.5 Renal artery stenosis in patient with Alagille syndrome and mild narrowing of the abdominal aorta. (**a**) Frontal view of abdominal aorta angiogram showing mild narrowing of the abdominal aorta between the take-off of the superior mesenteric artery and the right renal artery. There is severe narrowing of the proximal right renal artery, which is long segment and appears to spare the ostium and very proximal segment. The right renal artery assumes a more normal caliber in its mid-to-distal thirds. The left renal artery is not seen until injected selectively (**b**) and has near-atresia of its proximal and middle thirds with a more normal caliber distally. (**c**) Frontal view of angiogram after balloon angioplasty of both renal arteries, using cutting and high pressure balloons. Note that the right renal artery has a coil on its superior aspect. This coil was used to close an aneurysm that resulted as a complication of the balloon angioplasty. Both renal arteries appear widely patent, with no significant obstruction

26.5 Complications of Transcatheter Therapies in MAS

Percutaneous interventions on the mid-aorta have been shown to be effective in relieving obstruction in the acute setting. However, they are associated with a relatively high incidence of restenosis and reintervention [1, 2, 8]. The mechanisms of re-obstruction remain poorly understood. In the case of restenosis after stenting, the major mechanisms responsible appear to be in-stent stenosis, lack of growth of the stented area, and stenosis of the segment of aorta immediately distal and/or proximal to the stents (Fig. 26.6).

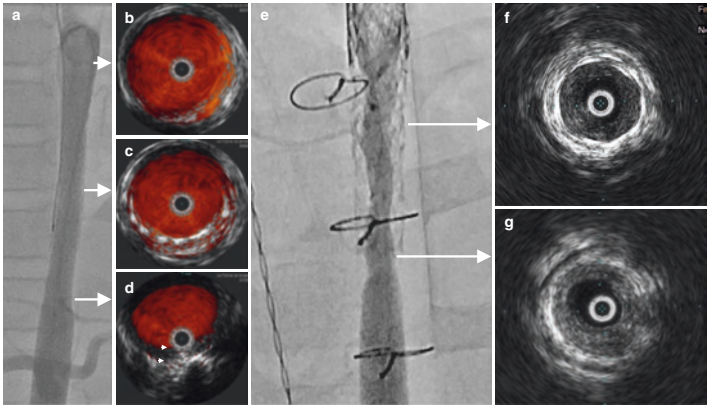

Fig. 26.6 Follow-up angiography and intravascular ultrasound (IVUS) 1 year after stenting of the thoracic aorta in two patients with MAS. (**a**–**d**) Patient with idiopathic MAS who underwent primary staged stenting of the thoracic aorta. A. Frontal view of angiogram showing the stented thoracic aorta. (**b**) IVUS of the aorta above the level of the stent, with no significant narrowing, or wall thickening. (**c**) IVUS of the stented portion of the aorta, with no narrowing and no significant in-stent stenosis. (**d**) The aorta immediately distal to the stent in the aorta. There is mild narrowing at this level secondary to mild media/intima thickening as can be appreciated on the IVUS (between white arrow heads). This represents only mild disease distal to the stent, with no hemodynamic consequence. (**e**–**g**) Patient with Williams syndrome-associated MAS involving the mid-thoracic aorta. (**e**) Frontal view of angiogram showing stented thoracic aorta with severe in stent stenosis as seen angiographically and confirmed by IVUS (**f**), as well as media/intima thickening and luminal narrowing distal to the stent as confirmed by IVUS (**g**). This is one common problem when using stents in patients with MAS and Williams syndrome. There is a high incidence of in-stent stenosis in mid-term follow-up, as well as new vessel wall disease distal to the stent

Percutaneous interventions can be associated with severe complications, including vascular tears, development of aneurysms at sites of prior angioplasty, and even death [7, 8]. The importance of operator experience and expertise cannot be overstated. In addition to available surgical backup, the presence of at least two experienced operators for every procedure is recommended.

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27 Reopening of Peripheral and Central Arteries and Veins

Henri Justino and Athar M. Qureshi

27.1 Anatomic Description and Physiopathology

Vascular occlusion can affect all vessel types, including systemic arteries, systemic veins, pulmonary arteries, pulmonary veins, and portal veins. The most common cause of vessel occlusion in children and young adults is thrombosis, particularly in systemic veins and systemic arteries, usually secondary to the placement of intravascular catheters. However, other mechanisms exist, including iatrogenic postoperative occlusions following anastomosis or patch angioplasty, infection, and inflammation. Finally, a unique form of postnatally acquired pulmonary artery and aortic occlusion can occur by the mechanism of closure of the ductus arteriosus.

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27.2 Clinical Scenarios

The most common clinical scenarios leading to the consideration of performing recanalization of occluded vessels are symptoms associated with venous congestion (e.g., facial or extremity edema), chylothorax (a common complication of superior vena cava or innominate vein obstruction), or arterial insufficiency. Another common scenario is the asymptomatic individual who requires restoration of vascular access routes for future catheterizations or for the placement of indwelling lines.

27.3 Indications and Patients Selection

Indications for catheter-based recanalization procedures for *acute* or *subacute* thrombotic vascular occlusions include failure of medical therapy (e.g., after an appropriate anticoagulation or thrombolytic regimen) but should also be considered as a primary form of therapy in cases where anticoagulation is relatively contraindicated. Patients with *chronic* thrombotic occlusions, or *non*-thrombotic occlusions (e.g., postsurgical), should not be subjected to inappropriate attempts at anticoagulation therapy but should rather be offered transcatheter recanalization as a primary therapy, provided the clinical sequelae warrant the procedural risk.

Appropriate patient selection depends, in part, on operator experience. While smaller patient size certainly complicates recanalization procedures, we have nonetheless successfully recanalized occluded vessels in many newborns, including premature infants down to a weight of 1100 g. The most favorable cases are those with a relatively short-segment occlusion in a single vessel, with patency of peripheral vessels allowing catheter access for the recanalization procedure itself. However, even patients with multiple occlusions, including occlusions of the vascular access sites themselves, can be considered candidates. These more complex patients require a great deal more time and effort, and experience at ultrasound-guided vascular access may be indispensable in these cases.

27.4 Treatment Options

Acute thrombotic occlusions, whether arterial or venous, could be treated primarily with anticoagulation, and in life- or limb-threatening cases, thrombolytic therapy should be considered, either systemically or locally. Surgical thrombectomy could be considered in cases of localized occlusion but is more invasive and not likely to be more successful than a catheter-based approach. Chronic occlusions, particularly if long segment, are much more complex to treat surgically and could require bypass grafts (which are prone to reocclusion and are particularly problematic in young children due to their inability to grow in length or diameter). In postsurgical acquired occlusions or occlusions secondary to constriction of ductal tissue, either surgical or catheter-based techniques may be viable options.

27.5 Preprocedural Imaging

Recanalization procedures are among the most challenging and time-consuming cases performed in the cardiac catheterization laboratory. Because of the tremendous demands that these procedures pose in time and equipment, methodical preprocedural planning is extremely important. While careful planning may not always be possible (e.g., some occluded vessels are encountered unexpectedly during failed attempts at vascular access), most cases of vascular occlusion are known or at least suspected. Preprocedural imaging does not need to clarify every detail regarding the occlusion but should grossly identify the location and extent of occlusion and should allow planning of the access site and approach to recanalization. Conventional angiography, which has far higher spatial and temporal resolution, will be performed at the time of the catheterization and will provide greater detail about the lesion.

Most vascular occlusions can be readily detected by ultrasound examination. The extent of occlusion, number of affected vessels, and degree of collateralization can usually be ascertained. When

ultrasound is unable to characterize the occlusion sufficiently, CT (computed tomography) or MR (magnetic resonance) angiography will typically reveal the vascular anatomy of the occlusion reliably. We do not routinely obtain CT or MR angiography in all patients and generally resort to these modalities when ultrasound is unable to characterize the occlusion sufficiently to allow planning of the procedure. It is important to note that in patients with preexisting metallic stents in whom a reintervention is being contemplated, MR is generally not helpful in imaging those vessels due to the susceptibility artifact created by the stents.

27.6 Technique (Step by Step)

The recanalization technique involves the following steps:

1. Choosing the optimal vascular access site
2. Obtaining vascular access
3. Crossing the total occlusion
4. Establishing a wire loop, if necessary
5. Thrombectomy, if necessary
6. Balloon angioplasty
7. Stent implantation, if necessary

27.6.1 Choosing the Optimal Vascular Access Site

One of the most critical steps in determining success or failure during recanalization is planning of the route of access. Many occlusions can be treated from a prograde (in the direction of normal blood flow) or a retrograde (against the direction of normal blood flow) approach. When either approach is available, the general rules for choosing the access site *from which to begin recanalization* are as follows:

1. The site should be *appropriately far* from the occlusion to allow a sufficient length of sheath or catheter to be intravascular so as to provide the necessary support for advancing guidewires.

2. The vessel being accessed should represent the *straightest path* to the occlusion, so that the vector of force during advancement of guidewires will be optimally directed at the occlusion.
3. In infants and young children, the *larger vessel* should be chosen if possible in order to reduce the risk of inducing vessel trauma from vascular access.
4. The approach (prograde or retrograde) having the occlusion cap that most resembles a *concave beak* on the surface should be chosen, as this will facilitate engaging the guidewire into the occlusion cap.

In many situations, the preferred access site will not satisfy all of the above criteria; thus, the operator will need to exercise judgment in deciding which of these competing criteria to prioritize. Lastly, it is important to note that the criteria above outline a method for thinking about choosing the access site from which to *initially attempt to cross the lesion* for recanalization. This does not imply that the vessel chosen will be the one to handle the largest sheath for the purpose of introducing balloons or stents.

27.6.2 Obtaining Vascular Access

Once the optimal access site is selected, vascular access may be obtained percutaneously using ultrasound guided by landmarks only. In cases where thrombolytic therapy is anticipated, ultrasound guidance may be preferred in order to avoid multiple passes of the needle and in order to visualize entry only into the anterior wall of the vessel rather than resorting to transfixation of the vessel. Ultrasound guidance also allows access in sites where anatomic landmarks are not commonly used or available (e.g., directly entering the superficial femoral vein at the level of the mid-thigh in cases of ipsilateral common femoral vein occlusion or direct percutaneous entry into the portal or splenic veins).

27.6.3 Crossing the Total Occlusion

Routine hemodynamic assessment is performed when indicated. In most cases we will delay heparin administration until wire recanalization is complete, if appropriate, in order to avoid bleeding into false tracts during attempts at wire passage across the lesion. In older children or those with relatively large peripheral vessels and with an entry site sufficiently far from the occlusion, we will generally enter the vessel directly with the smallest suitable sheath (usually 4 F), ideally one with a radiopaque band at the tip. In very small infants or when the peripheral vessel being entered is too small or is insufficiently far from the occlusion to accommodate the sheath, we will first enter the vessel with a soft Nitinol guidewire (0.014″ or 0.018″) and advance the wire as far as possible, using a coaxial micropuncture dilator set (e.g., 4 F Micropuncture set, Cook Medical, Bloomington, IN) over the wire to provide additional support, sometimes resorting to using only the innermost dilator if the combined coaxial dilator will not advance across the occlusion. If the wire does not appear to follow a desired anatomic course, then we assume it has entered a collateral or become extravascular and confirm this with angiography *over the wire* by using only the outer micropuncture dilator or a long 18 or 20 G peripheral intravenous catheter, with contrast injected through a Y-adaptor (Tuohy-Borst).

The technique of recanalization of an occluded vessel begins with crossing the lesion with an appropriate guidewire. In some cases of acute thrombosis, crossing of an occluded vessel may be accomplished directly with a catheter. Unless the lesion is immediately adjacent to the site of entry, the wire used for percutaneous access is not generally employed for recanalization. Selection of a guidewire for crossing the initial lesion is therefore an important initial step. As a general rule, guidewires should be attempted in order of increasing stiffness and increasing likelihood of exiting the vascular space (i.e., the softest and least traumatic guidewire that could reasonably be expected to cross the lesion should be tried first). Our first choice of guidewire is usually a hydrophilic all-purpose wire such as a 0.018″ or 0.035″ Glidewire (Terumo

Medical, Somerset, NJ), a 0.035″ Roadrunner wire (Cook, Bloomington, IN), or 0.018″ V-18 Control wire (Boston Scientific, Natick, MA). Whenever possible, an exchange-length wire should be used right at the outset. If a short wire (e.g., 145 cm) is inappropriately chosen to begin recanalization, the operator may find that the wire advances too far across the occlusion, only to later discover that there is insufficient length of wire outside the body to allow advancing of an appropriate catheter over it. We prefer a wire with a gently angled tip in order to negotiate a tortuous path toward the occlusion cap. However, a completely straight wire may be used, if the following conditions exist: a straight course from the access point to the occlusion cap, the absence of other vessels nearby that the wire would engage preferentially instead of the occluded vessel, and an ideally shaped concave beak at the occlusion cap. When these conditions exist, a completely straight wire may actually be advantageous, as the vector of force will be directed through the central core of the occluded vessel, whereas an angled wire will direct the vector of force toward the vessel wall and may hinder wire advancement across the lesion.

Gradually stiffer and/or lower-profile hydrophilic wires may be used if the initial wire fails to cross the occlusion. We use a variety of wires designed for chronic total occlusions (CTO) such as the following 0.014″ (0.36 mm) wires, in order of increasing risk of entering the subintimal space or the extravascular space altogether: ASAHI Fielder XT, Pilot, ASAHI Confianza (Abbott, Abbott Park, IL); if these wires fail, the most aggressive CTO wires we resort to are the Victory (Boston Scientific) and ASAHI Astato (Abbott). When these approaches fail, we have used on a number of occasions the stiff end of a 0.035″ Glidewire but only if the portion of the vessel requiring recanalization is very straight, given the inability of the stiff end of a wire to negotiate any significant curves. With any of these wires, the operator should attempt to use gentle advancement of the wire first and only proceed to applying more force if gentle advancement fails. In addition to carefully choosing an appropriate wire, the operator must also choose an appropriate catheter to advance wires through. A straight catheter may be appropriate if it is needed to add support

to a wire that is being advanced in the straight portion of a vessel and may be very helpful in preventing the wire from developing multiple S-shaped bends during wire advancement. However, when directional control over the wire advancement is needed, a low-profile curved catheter with a high degree of torque should be used; we commonly use a 4 F JR2 or JR3 catheter or 4 F Terumo JB1. Preloading the catheter within an appropriate long sheath is very helpful at this stage, as gradual advancement of the catheter through the lesion can be followed by gradual advancement of the long sheath. Another very helpful technique is to load a long catheter (e.g., 100 cm 4 F JR3) inside an appropriate caliber shorter guide catheter (in this case, a 55 cm 5 or 6 F JR or multipurpose guide catheter). As a general rule, a given catheter requires a guide catheter of 1–2 F sizes larger. This combination of a diagnostic catheter within a guide catheter is ideal because both the catheter and the guide can be individually turned, and the two will have a near ideal taper between them. This combination is also useful when a relatively straight course is not present.

We would consider techniques such as the use of a transseptal needle (curved or straightened, as appropriate) only as a last resort when all other guidewires have failed to cross the lesion. We have also successfully used radiofrequency wires to perforate across long-segment occlusions that could not be crossed by any other means. However, these techniques require an intimate knowledge of the surrounding anatomy, as there is a higher likelihood of exiting the vascular space and causing injury to adjacent anatomic structures.

As soon as the lesion is crossed, the wire used for crossing should be exchanged for a more supportive wire. In infants or when only small diameter vessels are being recanalized, we use an exchange-length supportive 0.014″ wire, e.g., ASAHI Grand Slam or Ironman (Abbott) for anticipated balloons of ≤5 mm in diameter, or a 0.018″ Platinum Plus (Boston Scientific) or SV-5 wire (Cordis, Santa Clara, CA) for anticipated balloon diameters of ≤10 mm. For larger patients or recanalization of vessels >10 mm in diameter, we prefer using an Amplatz Super Stiff 0.035″ guidewire (Boston Scientific).

27.6.4 Establishing a Wire Loop, If Necessary

When feasible, we strongly prefer exteriorizing the soft end of the wire using a snare and creating a through-and-through wire loop, and we typically clamp the soft end of the wire to the drape outside the body for added security. The added time required to exteriorize the wire will be more than compensated for in time saved during the procedure and in assurance of a stable wire position. Another advantage of an exteriorized wire loop is that sheaths exist on both sides of the occlusion, providing the convenience of performing angiography through the side arm of either sheath throughout the procedure and the ability to monitor the pressure on both sides of the occlusion without having to advance a catheter across the lesion each time, thus saving a great deal of time. With an exteriorized wire loop, it is possible to select the larger of the two vessels to accommodate the larger sheath necessary for the intervention. For example, when recanalizing a left innominate vein, we might create a venovenous loop between the left brachial vein and a femoral vein; in this instance, we might favor the brachial vein as the initial access site for recanalizing the occlusion using a 3 or 4 F sheath but would favor the femoral vein as the site for placing the 7 F or larger sheath for balloon and stent delivery. When an exteriorized wire loop is not possible or practical, we will lodge the soft end of the wire in the most remote and most harmless vessel that is appropriate (e.g., when recanalizing an occluded SVC from the femoral venous approach, the soft end of the wire would be more safely located deep in a peripheral vein of the arm rather than in the jugular vein in order to avoid neurological injury from wire manipulations).

27.6.5 Thrombectomy, If Necessary

In cases of acute thrombotic occlusion, there may be abundance of fresh clot. It is important to be able to distinguish fresh thrombus from other causes of vessel stenosis or occlusion. Failure to recognize thrombus might result in repeated futile angioplasties

with resulting vessel trauma and ultimately failure to establish flow through the lesion. Extraction of thrombus can be accomplished using numerous techniques, including manual aspiration catheters in various sizes (e.g., Fetch 2 Aspiration Catheter, Boston Scientific, or Pronto Catheter, Vascular Solutions, Minneapolis, MN) and various systems for mechanical thrombectomy and fragmentation, such as AngioJet (Bayer Healthcare) and Indigo (Penumbra, Alameda, CA). In addition, acute thrombotic occlusions may necessitate infusion of thrombolytic agents such as tissue plasminogen activator (tPA), either peripherally (i.e., systemic) or site-directed using an infusion catheter or an ultrasound pulse-assisted thrombolytic system (Ekos, Boston Scientific).

27.6.6 Balloon Angioplasty

Balloon angioplasty is almost always required during recanalization procedures (possible exceptions include cases of fresh thrombosis where simple thrombus extraction may restore vessel patency). As a general rule, the more chronic the occlusion, the more likely the lesion will be resistant to standard angioplasty. In such cases, a high-pressure noncompliant balloon is required. It is mandatory to eliminate the waist on a given balloon before proceeding to a larger diameter balloon. For lesions measuring <8 mm in diameter, we often utilize cutting balloons (Wolverine or Flextome, Boston Scientific) to treat resistant lesions if they are undilatable despite high-pressure angioplasty. For lesions >8 mm in diameter (or for resistant lesions within previously placed stents), we resort to noncompliant ultra-high-pressure balloons such as Dorado, Conquest, or Atlas (Bard, Murray Hill, NJ). Balloon angioplasty should be performed in a very gradual process, gradually increasing the balloon diameter as appropriate, reassessing the result with an angiogram after each angioplasty. If significant vessel wall trauma is encountered after angioplasty, it may be more prudent to accept a partial result rather than to dilate with larger balloons that may result in worsening dissection or even rupture.

27.6.7 Stent Implantation, If Necessary

Total occlusions, particularly if chronic, commonly necessitate stent placement, as balloon angioplasty alone may not provide long-term patency [1, 2]. In children, self-expanding stents are almost never utilized: balloon expandable stents must be used because they allow for future redilation to accommodate somatic growth. However, there are important exceptions when balloon-expandable stents should not be used: in areas of where there is extreme flexion that could lead to stent fracture (e.g., the common femoral artery or vein) and areas where there is prominent bony compression (e.g. subclavian vein as it passes between the clavicle and first rib at the thoracic outlet). It is imperative that stents be selected so that they can be redilated to an adult size for the vessel in question (at least 18 mm for the central pulmonary arteries) and >22 mm for the inferior and superior venae cavae and thoracic aorta. If important side branches will be crossed during stent implantation, open cell stents should ideally be utilized (e.g., Mega or Max LD, Medtronic, Minneapolis, MN) to facilitate enlargement of the cells jailing the ostium.

27.7 Materials

Most materials required for recanalization procedures have been described above. In addition, biplane fluoroscopy is essential to ensure that the target path is approximated in two nearly orthogonal views. We generally perform these cases using general anesthesia in children due to their length and complicated nature, but moderate sedation may be used in adults. During thrombectomy with AngioJet, bradycardia, and even asystole, may result. A pacing catheter should therefore be readily available. Blood products may become essential during thrombectomy (as thrombus removal may engender significant blood loss) or in the event of vessel rupture. In some instances, covered stents should be on hand (if not commercially available, then self-fabricated covered stents may be necessary). In addition, surgical backup may be required for

certain cases (e.g., recanalization of atretic aortic segments). Often as these are tedious and long procedures, the availability of a second skilled operator can facilitate the procedure and shorten procedural time.

27.8 Expected Results

Acute procedural success during recanalization procedures can be commonly achieved, as long as the operators are persistent and willing to change course if initially unsuccessful, such as approaching the occlusion from a different vascular access site if prolonged attempts appear fruitless. Long-term success, however, depends on numerous factors, such as the adequacy of the treatment of the occlusion itself, the adequacy of the inflow and outflow vessels on either side of the occlusion, the adequacy of thrombus removal, and the patient's compliance with long-term anticoagulation. We have experienced higher success rates with stenting than with balloon angioplasty alone when treating chronic total occlusions. Stent placement in a young child carries the disadvantage of committing the patient to numerous repeat procedures to redilate the stent in order to accompany somatic growth. However, avoidance of a stent at the initial procedure may result in an early reocclusion that may be even more difficult to treat the second time. Therefore, deciding whether or not to place a stent should be based on the likelihood of long term patency from angioplasty alone (i.e., if the acute result looks excellent, we may avoid a stent, but if there is acute recoil or severe luminal irregularity with poor flow, we will generally place a stent) and the risk to the patient and level of difficulty of a reintervention if there is reocclusion. Certainly, if there is reocclusion after angioplasty alone, we will generally implant a stent at the time of reintervention. Long-term patency after stenting has been high in our experience as long as there is adequate inflow and outflow from the lesion, with complete stent coverage of the occluded segment.

27.9 Tips and Tricks

An important tip during recanalization is to use soft wires initially and only resort to gradually stiffer wires if necessary, in order to avoid exiting the vascular space. Once the vascular space has been exited, the false tract can be very difficult to avoid, which may require abandoning that approach in favor of attempts to cross the lesion from the other side of the occlusion. It is paramount that balloons not be used for dilating a wire tract unless the operator is certain that the wire is intravascular. The ability to snare the soft wire tip from the other side in order to create a through-and-through wire loop is an ideal way to confirm that the wire is indeed intravascular. If the wire cannot be snared despite multiple attempts, angiography will often confirm that the wire tip is actually subintimal or, rarely, completely extravascular. This is generally not cause for alarm, as a wire perforation alone will usually not result in significant bleeding [3]. However, if the operator fails to recognize that the wire is extravascular and proceeds to balloon angioplasty, the result could be catastrophic, particularly in the arterial system. An additional important tip is to be cognizant of whether the area undergoing recanalization has been previously operated upon: postoperative occlusions are at less risk of bleeding because of abundant adhesions surrounding the vessel.

The operator must pay careful attention to surrounding vascular structures that may play a role in the occlusive process, as in the May–Thurner syndrome where the right common iliac artery compresses the left common iliac vein.

Lastly, perhaps the most important tip during venous recanalization procedures is the following: success is much more likely if at the end of the case, the *flow of contrast through the lesion is brisk* and if *collaterals are no longer prominently opacified*. Persistent opacification of the collateral network is surely a sign that the treated lesion itself is not the path of least resistance for blood flow, which will predispose to reocclusion. We do not believe in intentionally occluding collateral vessels in order to encourage flow through an inadequately treated lesion: to do so would potentially harm the patient and would leave no path for flow if the lesion were to reocclude (Figs. 27.1, 27.2, and 27.3).

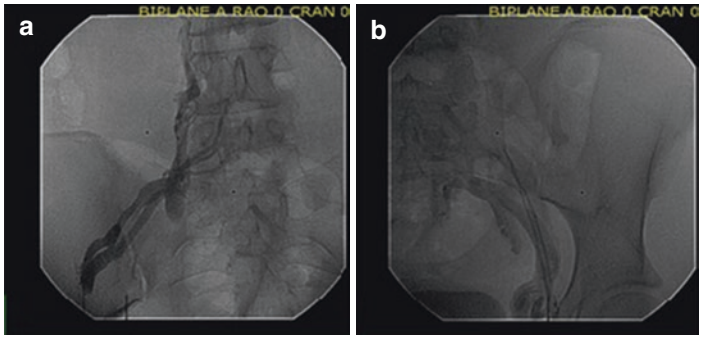

Fig. 27.1 Angiograms performed in the right femoral vein (**a**) and left femoral vein (**b**) show total occlusion of the common iliac veins bilaterally as well as of the infrarenal inferior vena cava, with numerous venous collaterals seen

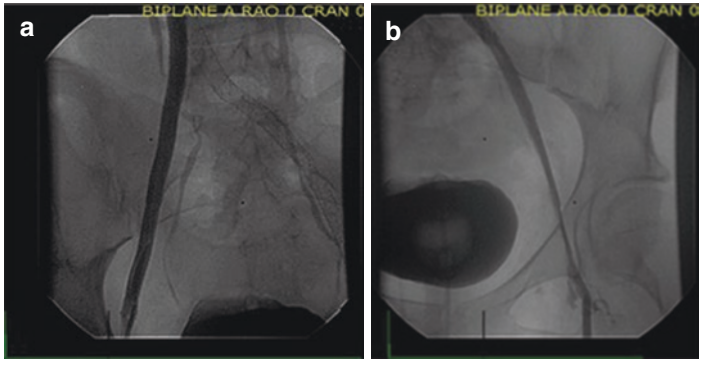

Fig. 27.2 Angiograms performed after placement of stents in the right (**a**) and left (**b**) external and common iliac veins, showing restored patency of these veins, with very little opacification of the collateral veins

27.10 Pitfalls

Pitfalls to be avoided include inadequate preprocedural planning and incomplete understanding of the occlusion and surrounding anatomy. Careful study of noninvasive imaging prior to the procedure is extremely important in this regard. As stated above,

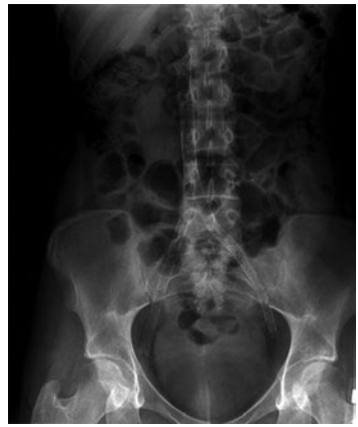

Fig. 27.3 Abdominal X-ray showing the configuration of the stents in the external and common iliac veins bilaterally entering jointly into stents placed in the infrarenal inferior vena cava

balloon angioplasty is contraindicated until there is certainty that the wire has crossed the occlusion and has entered the vascular space on the other side of the occlusion.

27.11 Complications

Complications include subintimal wire passage, which is generally benign, or complete perforation of the vessel wall. As long as no bulky catheters or balloons are advanced across the perforation, bleeding should be negligible. Failure to recognize that the wire is extravascular could result in an uncontrolled tear during angioplasty. Stenting could result in stent malposition or frank embolization. In cases with abundant fresh clot, emboli could result in end-organ damage such as pulmonary embolism or stroke. Thrombectomy with AngioJet is well reported to potentially cause hypotension, bradycardia, and even asystole, particularly if close to the heart. Thrombectomy systems that result in significant erythrocyte lysis, such as AngioJet, will also cause hemoglobinuria, which could lead to renal failure. Contrast nephropathy and radiation injury are always potential concerns during long and complex recanalization cases.

27.12 How to Manage Complications

Vascular rupture should be managed according to the severity: if a small and nonessential blood vessel is torn, coil occlusion may effectively stop the bleeding and may allow the case to proceed despite the complication. Rupture of a major blood vessel, however, must be treated expeditiously: anticoagulation should be reversed (if appropriate), blood products administered, and inflation of an appropriately sized balloon in the lumen of the blood vessel may temporarily halt the bleeding, as long as this does not drastically impact the cardiac output or perfusion of the affected organ. Placement of a covered stent may be necessary, or placement of an occlusion device at the site of the tear, and these options should be weighed carefully against an urgent surgical exploration.

Stent embolization can be managed by recapture of the stent with redeployment in the target lesion if possible or intentional deployment in a remote area. Removal of stents from the body using snares can be extremely challenging, particularly for large diameter stents, and requires the use of very large and braided sheaths.

Distal clot embolism should be readily treatable with aspiration catheters or other thrombectomy systems.

Bradycardia or asystole can occur during AngioJet thrombectomy. These complications can be avoided by simply limiting the duration of each pass of the AngioJet catheter to no more than 10 s at a time (we often use 5–6 s only) and can be managed with epinephrine and temporary ventricular pacing [4].

Hemoglobinuria and contrast nephropathy are generally treatable with abundant fluid administration.

Bleeding secondary to thrombolytic therapy with tPA or other agents must be managed according to the specific mechanism of action of the agent in question and require that the operator be familiar with the half-life and reversal mechanism of the thrombolytic agent. For instance, tPA has a half-life of only about 5 min and can be reversed by administration of fresh frozen plasma, cryoprecipitate, and an antifibrinolytic.

27.13 Postprocedural Care

Unless there are major contraindications to anticoagulation, we recommend treatment with subcutaneous low-molecular-weight heparin for at least 6 months, usually with additional antiplatelet therapy with aspirin. After 6 months, we generally continue with aspirin only.

27.14 Follow-Up

Postprocedural imaging is mostly accomplished with Doppler ultrasound but can be supplemented with MRI (as long as there are no stents in the region of interest) or CT angiography. For upper body venous recanalizations, we commonly perform surveillance conventional venograms from a peripheral upper extremity, as these do not require sedation and provide a quick and easy way to follow the status of the recanalized lesion. We have a low threshold to return to the catheterization laboratory for additional interventions, believing that it is far easier to re-intervene when restenosis is moderate, rather than allowing the lesion to completely reocclude. In our experience, if patency can be maintained for a few months, then long-term patency is almost assured, as late reocclusion is quite uncommon.

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28 PDA Stenting in Duct-Dependent Pulmonary Circulation

Kothandam Sivakumar

28.1 Ductal Stenting for Pulmonary Circulation

Surgical *aortopulmonary shunts* palliate neonates with duct-dependent pulmonary circulation. The surgical problems included prolonged mechanical ventilation and intensive care stays, bleeding and transfusions, frequent use of multiple inotropes, pulmonary complications, sepsis, and injury to surrounding structures like phrenic nerve, recurrent laryngeal nerve, and thoracic duct. *Ductal stenting* (DS) provides a nonsurgical attractive alternative option to surgical aortopulmonary shunts. On follow-up after both the procedures, there is a progressive fall in oxygen levels due to intimal ingrowth within the ductal stents and fibrointimal peel formation and thrombus within surgical shunts. About 5–20% of patients suddenly die on follow-up due to shunt or stent thrombosis. While the surgical shunts offer a longer palliation of few years, DS gives longevity of only 6–12 months. This difference in duration of palliation will influence patient selection for DS [1].

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28.2 Anatomic Description and Physiopathology

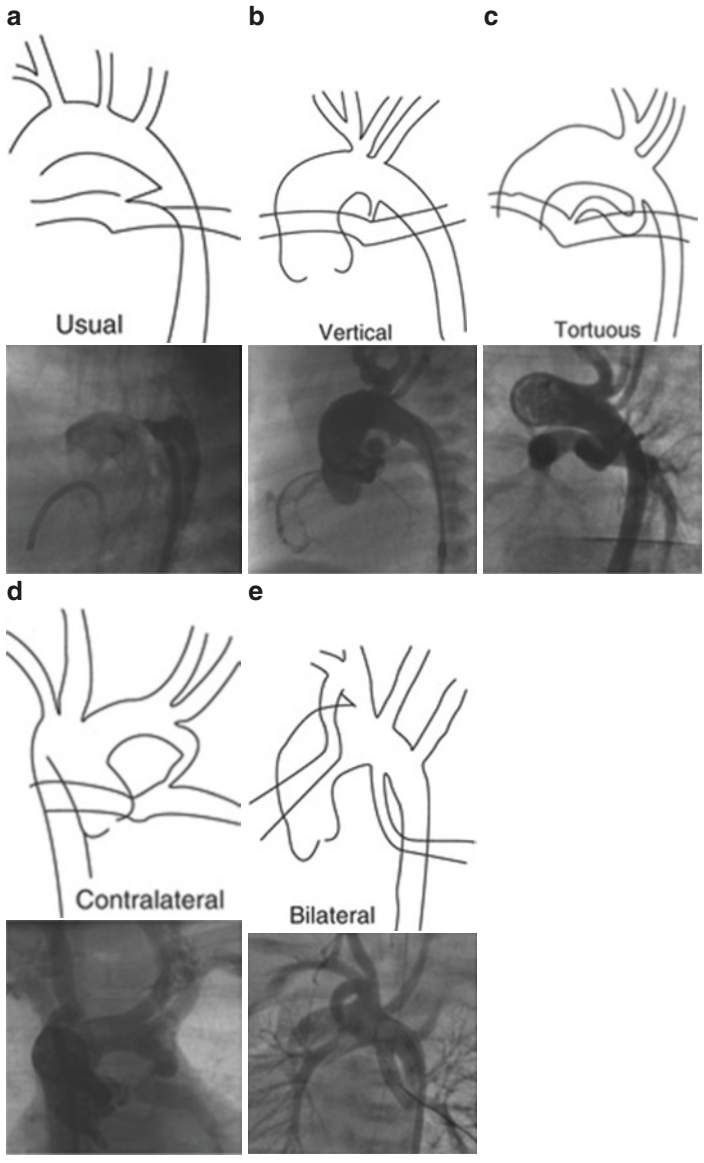
28.2.1 Anatomy of Ductus Arteriosus

In patients diagnosed to have patent ductus arteriosus, the shape of the duct may vary from conical or tubular or window-like, but aortic origin of the duct is almost always constant and is beyond the origin of the last subclavian artery. In contrast, in pulmonary atresia, the origin of the duct varies and may have a more proximal origin from the undersurface of the aortic arch [2]. In few instances, the duct may arise from the contralateral innominate or subclavian artery and rarely may be bilateral connecting to non-confluent pulmonary arteries (Fig. 28.1).

28.2.2 Pathophysiology

Early neonatal withdrawal of prostaglandins leads to swelling of the ductal intimal cushions, ductal constriction, and obliteration of the lumen. Functional closure of duct occurs in first 2–3 days; anatomical closure with fibrous tissue occurs later. Duct closure

Fig. 28.1 Different ductal morphology: The ducts in pulmonary atresia can be in various morphologic forms. Type I duct, usual form: (**a**) The duct arises from the junction of the arch to descending aorta and courses anteriorly to insert in confluence of pulmonary arteries. Type II duct, vertical form: (**b**) The duct arises proximally from the undersurface of the aortic arch and courses vertically down to the confluence. Type III, tortuous form: (**c**) In this commonest morphological form, the duct takes a C- or S-shaped bend before inserting in the confluence. Type IV, contralateral form: (**d**) The duct arises opposite to the side of the aortic arch from either the contralateral innominate artery or subclavian artery. Type V, bilateral form: (**e**) Rarely, ducts can be bilateral, each duct will insert into ipsilateral pulmonary artery. In most of these patients, the pulmonary arteries are not confluent and are separated from each other


a
b
c
Usual
Vertical
Tortuous
d
e
Contralateral
Bilateral

in pulmonary atresia results in severe hypoxia. In a few patients, ducts remain patent for few weeks or few months to maintain pulmonary circulation, possibly due to hypoxia, which postpones duct closure. Fetal circulation in pulmonary atresia is different from normal by allowing only a smaller blood flow form the combined ventricular output to pass through the duct. This results in smaller size of ducts in fetuses with pulmonary atresia, which may predispose to earlier closure in some, despite hypoxia.

28.2.3 Different Anatomical Lesions

Congenital heart lesions with critically reduced pulmonary blood flows in neonatal period that depend on the duct patency to maintain pulmonary circulation can be grouped as follows [1].

28.2.3.1 Group A: Pulmonary Atresia in Biventricular Hearts

This group includes pulmonary atresia associated with tetralogy of Fallot (TOF), double-outlet right ventricle, and transposition of great arteries. The anatomy is suited for a later biventricular repair with extra cardiac valve homograft or xenograft *conduits*.

28.2.3.2 Group B: Pulmonary Atresia in Univentricular Circulation

This group includes pulmonary atresia associated with single ventricles, unbalanced atrioventricular canals, double-outlet right ventricle with non-routable ventricular septal defects, pulmonary atresia with intact ventricular septum, right ventricle-dependent coronary circulation, and complex heterotaxies. They are palliated later by bidirectional Glenn and Fontan surgeries.

28.2.3.3 Group C: Transient Inadequacy of Pulmonary Circulation

1. Neonates having pulmonary atresia with intact ventricular septum and critical pulmonary stenosis continue to have inadequate antegrade pulmonary flows even after a successful neonatal pulmonary valvotomy for a few weeks to months.

2. Functional pulmonary atresia occurs before regression of high fetal pulmonary vascular resistance in neonates with severe forms of Ebstein's anomaly, right ventricular cardiomyopathy, Uhl's anomaly, severe forms of tricuspid valve dysplasia with regurgitation and very large fetal rhabdomyoma which obliterate right ventricular cavity. In both these groups, longer ductal patency is needed.

28.3 Clinical Scenarios

A few case studies highlight the different presentations of the various groups.

Case Study 1
After a fetal diagnosis of TOF with pulmonary atresia in the sixth month of gestation, a 3.2-kg baby was electively admitted after birth for observation in neonatal unit. His echocardiogram confirmed the fetal diagnosis, closing vertical duct and confluent pulmonary arteries measuring 4 mm each. After 36 h, *prostaglandin E1* (PGE1) infusion was started for hypoxia. After discussion with cardiac surgeons, an elective DS with a 4-mm coronary stent was done on the fourth postnatal day with a coronary guide catheter advanced through right ventricle into the aortic arch from right femoral venous access. The acute angulation of the vertical duct from the undersurface of the aortic arch did not permit wiring the duct from femoral artery (Fig. 28.2). He later underwent elective conduit repair at 8 months of age.

Case Study 2
A 10-day-old neonate weighing 2.2 kg with severe cyanosis was diagnosed as TOF, pulmonary atresia and small confluent pulmonary arteries measuring 3 mm each. Shunt surgery in small pulmonary arteries and low body weight carries high risks. DS was performed with 3.5 mm coronary stent through femoral arterial access (Fig. 28.3). Elective conduit repair was done at 1 year of age.

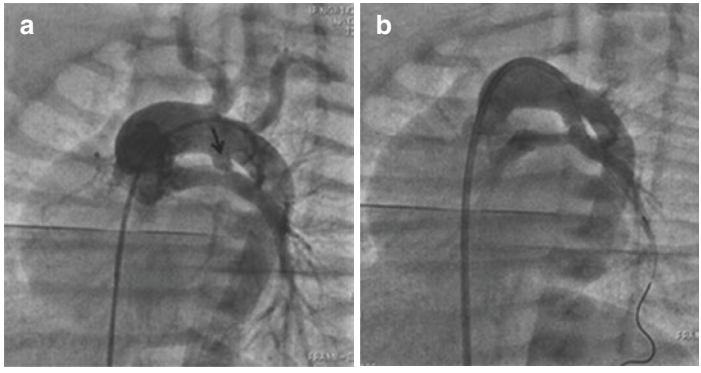


Fig. 28.2 Aortogram in shallow left anterior oblique view with a pigtail catheter advanced from femoral vein through the right ventricle into the left aortic arch (**a**) shows a vertical duct (*arrow*) arising from undersurface of the aortic arch opposite to the right innominate artery. In such cases, advancing a guidewire into distal branch of the left pulmonary artery and DS is done more easily (**b**) from favorable angle through the transvenous route

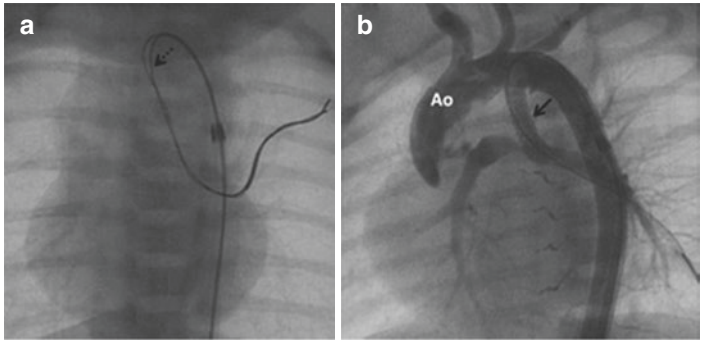


Fig. 28.3 Through a 4 F long sheath, a coronary guidewire was advanced (**a**) through a vertical duct. An additional buddy wire (*dotted arrow*) was advanced to facilitate the passage of the stent through the acute angulation of vertical duct. (**b**) After stenting, aortogram in shallow left anterior oblique view confirmed the stenting (*arrow*) of the entire length of the duct. *Ao* aorta

Case Study 3

A 2.8-kg neonate antenatally diagnosed with *Ebstein's anomaly* of tricuspid valve presented with severe cyanosis after birth needing mechanical ventilation. There was functional pulmonary atresia, no significant antegrade pulmonary blood flows, and right aortic arch. After stabilizing with PGE1, he was weaned off the ventilator. His continued dependence on PGE1 warranted DS with a 4-mm coronary stent on the 14th postnatal day (Fig. 28.4). The improving right ventricular function normalized antegrade pulmonary blood flows after 2 months. The stent was patent for 7 months, and his oxygen saturations were above 95% throughout his childhood.

Case Study 4

A 10-day-old neonate had *critical valvar pulmonary stenosis* with pulmonary annulus measuring 7 mm, right to left shunt through the foramen ovale, and hypoplastic right ventricle with tricuspid valve *Z*-score of −2. Severe hypoxia persisted after balloon pulmonary valvotomy, even though the right ventricular pressures are

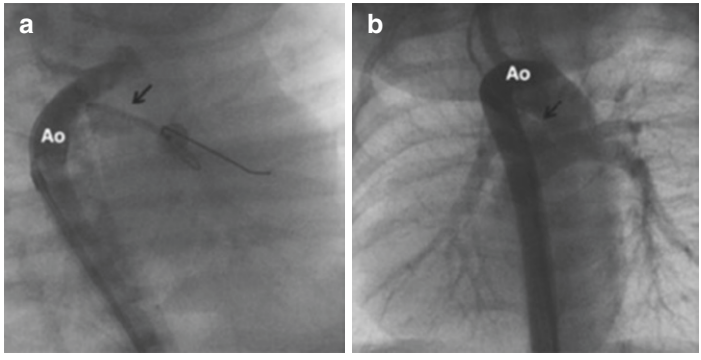


Fig. 28.4 Angiogram through a 4 F long sheath (**a**) in shallow right anterior oblique projection was done to check the position of a 4-mm stent (*arrow*) placed on a coronary guidewire through the duct advanced into the left pulmonary artery. After stenting, a repeat aortogram (**b**) showed good filling of the pulmonary arteries through the stented duct (*arrow*)

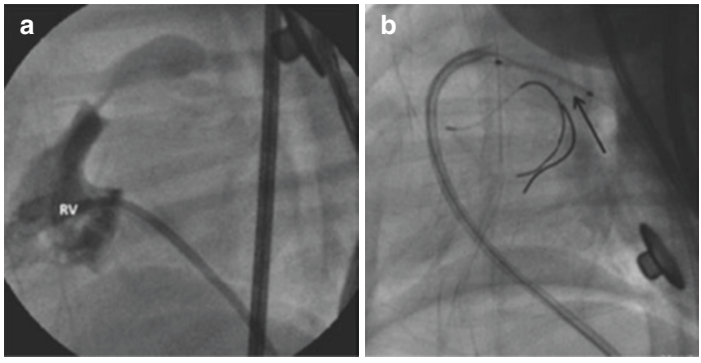


Fig. 28.5 Right ventricular (*RV*) angiogram in lateral view (**a**) demonstrates thick pulmonary valve with a narrow contrast jet into the pulmonary artery indicating severe stenosis which was dilated with a balloon. The persistent hypoxia due to inadequate antegrade pulmonary flows was an indication for ductal stent (*arrow*) done through a guide catheter (**b**) introduced from femoral vein into the pulmonary artery. A coronary guidewire was advanced from the guide catheter into the right pulmonary artery to guide the proximal extent of the ductal stent

reduced from 120 to 45 mmHg. His closing ductus was stented through a guide catheter advanced through the venous end into the main pulmonary artery (Fig. 28.5). His ductal stent remained patent for 1 year.

Case Study 5

Pulmonary atresia with intact ventricular septum was diagnosed in a 4-day-old neonate with severe hypoxia. The tricuspid valve *Z*-score of −2 was favoring a decision for balloon pulmonary valvotomy. However, there were extensive myocardial sinusoids communicating freely with the coronary arteries and *right ventricle-dependent coronary circulation* (Fig. 28.6). DS with a 4-mm coronary stent was done from the femoral artery.

Case Study 6

A 45-day-old infant with single ventricle, pulmonary atresia, common atrioventricular valve with no regurgitation, presented

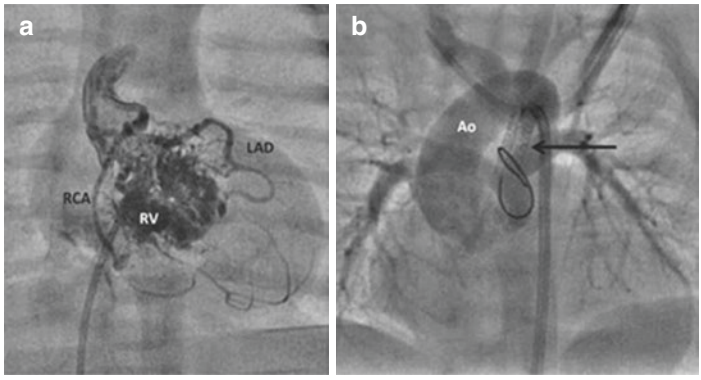


Fig. 28.6 Right ventricular angiogram (**a**) shows a hypoplastic right ventricle (*RV*) filling multiple sinusoids which fill the right coronary artery (*RCA*) and left anterior descending interventricular artery (*LAD*) indicative of right ventricle-dependent coronary circulation (RVDCC). This precludes decompression of the right ventricle with a pulmonary valvotomy. After DS, aortogram from the femoral arterial access (**b**) fills the well-formed pulmonary sinuses and pulmonary arteries through the stented duct (*arrow*)

with severe hypoxia. A long duct from the contralateral right subclavian artery was very narrow at the pulmonary end. An elective DS with a 3.5-mm stent maintained adequate oxygenation till his bidirectional Glenn surgery at 1 year of age (Fig. 28.7).

28.4 Indications and Patients Selection

28.4.1 Group A: Pulmonary Atresia with Biventricular Physiology

As conduit repair is deferred beyond infancy in order to place a larger conduit, a longer initial neonatal palliation is desired. In comparison with surgical shunts with 3.5 or 4 mm grafts, DS offers a palliation lasting only for 6–12 months. When the recent drug-eluting stents are used, longer palliation is achieved. In this group, DS is more advantageous than surgical shunts especially in:

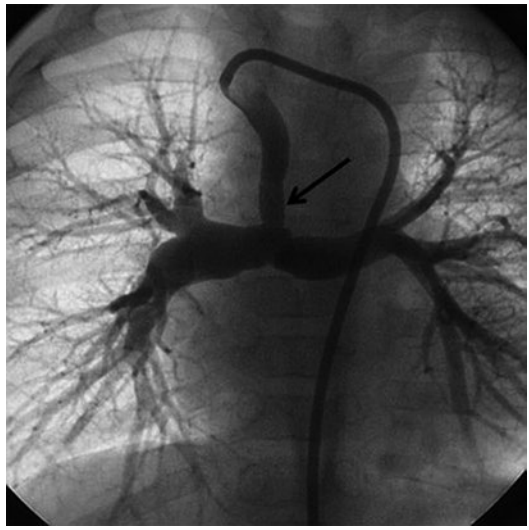


Fig. 28.7 Injection through a guide catheter passed from the left aortic arch into the right subclavian artery shows the long contralateral duct which inserts into the confluence. The narrowing in the distal insertion of the duct was stented (*arrow*) with a 3.5-mm stent

1. High-risk surgical candidates: low birth weight below 2.5 kg, syndromes like trisomy 21, comorbidities like bronchopneumonia, lung disease of prematurity, other organ malformations.
2. Small pulmonary arteries. Surgical shunts on very small pulmonary arteries are complicated by frequent shunt site narrowing. The distal hilar pulmonary artery narrowing after shunt is difficult to repair during the corrective surgery. In contrast, narrowing of confluence after DS is easily approachable in corrective surgery.
3. In non-confluent pulmonary arteries with Type V bilateral ducts, extensive reconstruction of the confluence on cardiopulmonary bypass is needed during shunt surgery. Bilateral DS can defer this reconstruction by few months.
4. If surgical experience is low in neonatal shunts, elective DS may be performed in low-risk candidates also.

28.4.2 Group B: Univentricular Hearts with Pulmonary Atresia

Both DS and surgical shunt increase the pulmonary blood flow and cause ventricular volume overload. Prolonged ventricular volume overload and resultant dysfunction complicate Glenn surgery. The high pulmonary artery pressures and resultant high pulmonary vascular resistance also complicate Glenn circulation. In this subset, early Glenn shunt avoids prolonged exposure of the pulmonary vascular bed to aortic flows. For this short-term palliation, DS is more ideal than surgical shunt as it avoids surgical morbidity. The ductal stent will induce fibrosis and stenosis in the confluence, which can be repaired during Glenn shunt as the confluence is easily repaired during stent removal.

28.4.3 Group C: Transient Need for Ductal Patency

Group C lesions need ductal patency for a few weeks or months until the right ventricle becomes adequate to maintain pulmonary circulation. In this subset, a temporary palliation of maintaining ductal patency is needed in neonatal period. The ductal stent needs to stay patent in these patients only for periods up to 1 year.

28.5 Treatment Options

28.5.1 Group A: Tetralogy of Fallot with Pulmonary Atresia Suited for Biventricular Conduit Repair

Surgical shunts offer longer palliation lasting 2–3 years compared to DS, which is adequate only till 6–12 months. However drug-eluting stents may increase longevity of palliation.

DS is preferred in

1. High surgical risk candidates
2. Small pulmonary arteries
3. Non-confluent pulmonary arteries
4. Institutions with high neonatal surgical morbidity

Surgical shunt is preferred in

1. Low surgical risk neonates
2. Preexistent confluence stenosis at the duct insertion site

28.5.2 Group B: Pulmonary Atresia in Univentricular Hearts

DS provides adequate palliation till Glenn shunt performed at 6–7 months of age. However, surgical shunt is preferred in patients with confluence stenosis, where surgery should include confluence plasty that often requires neonatal cardiopulmonary bypass.

28.5.3 Group C: Transient Neonatal Dependence on Duct

Prolonged PGE1 infusion for 2–6 weeks as an option is associated with (1) high costs due to prolonged intensive care stay, (2) venous thrombosis and sepsis, and (3) drug adverse effects, namely gastric antral hyperplasia and hyperosteosis of bones, (4) uncertainty about how long to continue the infusion. In this group, surgical shunt leads to uncontrolled poorly tolerated pulmonary blood flows. DS is ideal in this group as it shortens the hospital stay and provides adequate duration of palliation.

28.6 Preprocedural Imaging

Echocardiogram is the most vital imaging tool to record the following features:

1. Duct—morphology, origin and insertion, length, and diameter at aortic and pulmonary end
2. Aortic arch—side of the aortic arch and arch branches for axillary or carotid arterial access
3. Pulmonary arteries—mediastinal and hilar pulmonary artery sizes, stenosis of confluence at duct insertion site
4. Intracardiac anatomy—differentiate groups A, B, and C
5. Ventricular systolic function, atrioventricular valve annulus size and function, aortic root diameter, and aortic valve function
6. Venous anomalies for transvenous approaches
7. Interatrial communication and need for balloon septostomy

28.7 Advanced Three-Dimensional Imaging

Many centers adopt contrast computed tomography to get a precise delineation of the duct, its origin and insertion, its angulation in a three-dimensional perspective, tortuosity and number of curves to be manipulated, choice of vascular access—axillary or carotid instead of femoral artery. The concern about additional contrast load on renal function and additional radiation dose should be balanced against the pre-procedural planning advantages. But this technology should be employed only in institutions with advanced multislice scanners which can acquire good resolution images within fraction of second to avoid artifacts caused by breathing, cardiac motion, and motion of the patient. Issues of improper bolus tracking of the contrast can be avoided by acquiring a couple of additional runs of imaging within a short time.

28.8 Technique (Step-by-Step)

- PGE1 infusion should be stopped at least an hour, but still kept in-line, before DS to allow ducts to be well constricted. This carries the risk of inducing hypoxia before the start of the procedure. As the fear of stent embolization due to lack of ductal constriction is more in short straight ducts than in long tortuous ducts, PGE1 can continue till vascular access in the latter.
- A dose of **aspirin** 3–5 mg/kg is given before the procedure. In patients who present in emergency, aspirin and clopidogrel bolus doses can be administered through nasogastric tube immediately after completion of the procedure before removing the vascular sheaths.
- **Hypothermia** is avoided by use of warm air blower (3 M Bair hugger), draping sterile warm linen, and warming saline and contrast before use.
- Cross-matched packed red cells are reserved for procedural blood loss.
- Intubation and general anesthesia are institutional preference especially if axillary or carotid arterial access is anticipated. Conscious sedation with ketamine or dexmedetomidine and continuous supervision by intensivist with spontaneous ventilation is also possible in the majority of stable neonates.
- Boluses of intravenous fluids counter PGE1-related hypotension and facilitate quick vascular access.
- 4 or 5 F vascular access is obtained from femoral artery for initial aortogram with a high-flow pigtail catheter.
- 100 U/kg Heparin is given after vascular access; additional doses in prolonged procedures are given empirically every hour or guided by activated clotting time which is maintained above 180 s at all times.
- For showing the pulmonary arteries and their confluence, aortogram should be done in shallow left anterior oblique projection with cranial angulation (LAO 20° Cranial 20°) in Type I–III ducts (Figs. 28.2 and 28.3). In ducts originating from right arch, RAO 20° Cranial 20° projection is chosen (Fig. 28.4).

- Ductal origin, course and insertion, morphology, length, and diameter are delineated in lateral view (Fig. 28.1c) in Type I–III ducts. In Type IV and V ducts, aortogram is done in anteroposterior view (Fig. 28.7).
- There are operator preferences about choice of vascular access. In Type II and III ducts, which arise from undersurface of aortic arch, axillary or carotid access can provide a direct access.
- When axillary or femoral access is chosen, neonates can be positioned in table with foot up and "flip screen" option can be enabled to facilitate the procedure. When DS is carried out from these vessels, long catheters and guidewires that emerge from neck and axilla can be better handled if the neonates are positioned with their feet facing up.
- Being unconventional vascular access, axillary access can be facilitated by use of ultrasound vascular probes. In some instances, a guidewire placed from femoral artery into the axillary artery can serve as a fluoroscopic guide to insert the puncture needle from neck or axilla (Fig. 28.8).
- In Type II and III ducts, the tip of the pigtail or Simmonds catheter from femoral arterial access is cut to a J shape and reinserted into the aortic arch to engage the duct. A Judkins

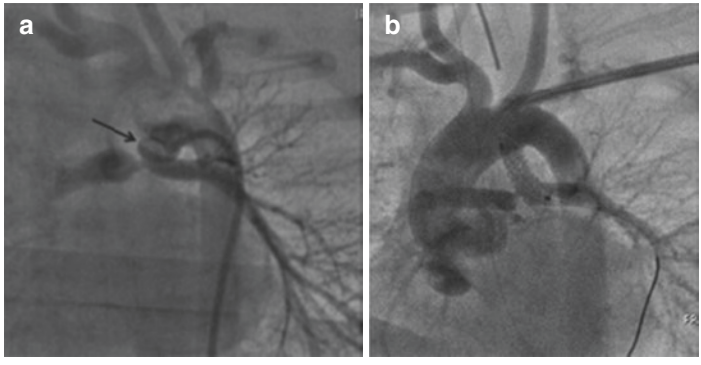


Fig. 28.8 Femoral access angiogram (**a**) with an end-hole catheter shows a tortuous Type III duct (*arrow*) which needed axillary artery access for successful ductal stenting with a 4-mm coronary stent (**b**). The tip of the guidewire is parked in the lower lobe branch of left pulmonary artery

right coronary catheter is preferred to cannulate Type I, IV, and V ducts.

- A 0.014″ coronary extrasupport guidewire with floppy J tip is advanced along the curvature of the duct into the pulmonary artery. A Y connector (Touhy Borst) controls the blood loss. The guidewire beyond the floppy tip should be advanced well into the pulmonary artery.
- If the guidewire fails to advance beyond the floppy tip, a **microcatheter** can facilitate further advances of the guidewire. This maneuver helps especially in vertical tortuous ducts.
- The stenting is done in most ducts through long 4 F sheaths but sometimes with 5 F Judkins right coronary guide catheter.
- Premounted bare-metal **coronary stents** are chosen, length of stent is chosen based on echocardiography and angiography. Care should be taken to stent entire length of the duct and not to leave any ductal portion unstented.
- After positioning the guidewire that straightens the duct, if the hemodynamics and oxygen saturations remain stable, a coronary balloon of known length and markers on either side is advanced into the duct (Fig. 28.9). A repeat angiogram is done, and the length of the duct is assessed in comparison to the balloon length.
- 3.5-mm-diameter stents are chosen in patients under 2.5 kg and 4 mm stents in patients over 2.5 kg. In bilateral Type V ducts supplying each lung, 3–3.5 mm stents are used. In patients in Group C, where a short-term patency is desired, 3–3.5 mm stents will suffice.
- When the stent is advanced from femoral access in Type II and III ducts, the stent may get pushed proximally into the aortic arch rather than through the duct. In such instances, an additional buddy wire (Fig. 28.3) will facilitate advancing the stent into the duct.
- Rapid inflation of the stent using **inflation device** ensures complete expansion of the stent.
- Angiogram is repeated to confirm that the entire duct length is stented.

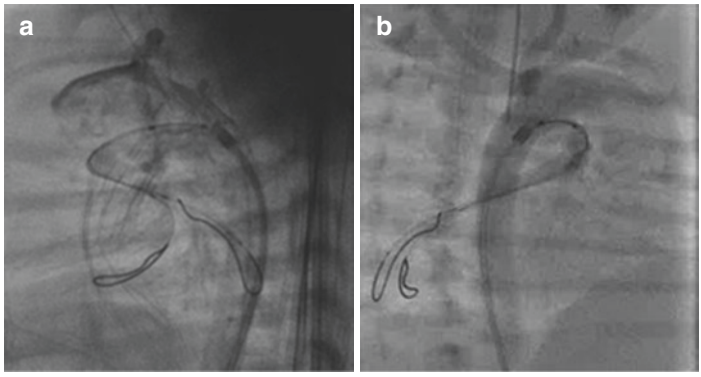


Fig. 28.9 Aortogram was repeated in lateral (**a**) and right anterior oblique (**b**) projections after an uninflated 1.5 mm × 10 mm balloon with two markers at either ends to indicate the length is placed in the duct. When the contrast fills the duct, the balloon length is compared to the straightened duct length to decide on the ultimate stent length. While using this uninflated balloon to measure the ductal length, care should be taken to select the balloons with markers at both ends. Some coronary balloons of smaller diameters will have only a single central marker

- After hemostasis, **heparin** infusion is continued for 24–48 h in a dose of 15–20 units/kg/h. Oral or nasogastric feeds are started at earliest opportunity. Antiplatelet drugs aspirin (3–5 mg/kg/day) and **clopidogrel** (1 mg/kg/day) are given daily.
- Oxygen saturations and hemodynamics are monitored for 48 h in intensive care before discharge from the hospital.

28.9 Materials

- Catheters: 4 F pigtail catheter, 4 F Judkins right coronary catheter, 5 F Launcher Judkins RCA guide catheter (Medtronic Co).
- Microcatheters: 0.014″ Finecross (Terumo Corporation) or Corsair (Abbott) microcatheters, 0.018″ lumen Cantata micro-

catheter (Cook Medical), 0.021″ lumen Progreat microcatheter (Terumo Corporation).
- Guidewires: 0.014″ balanced middle weight coronary guidewire (Abbott), Choice PT extrasupport wire (Boston Scientific).
- Long sheaths: 4 F Boston Children's Hospital sheath (Cook Medical).
- Stents: 3.0, 3.5, 4.0, and 4.5 mm diameter coronary stents Driver (Medtronic Co) and Vision (Abbott) of varying lengths.

28.10 Expected Results

Type I, IV, and V ducts are cannulated with Judkins right coronary catheter, and DS is often successful [3]. The oxygen saturation rises immediately to high 80s or low 90s. Acute runoff of aortic blood into the pulmonary artery may cause brief self-limiting systemic hypotension. Entire length of the duct should be stented. If the pulmonary or aortic end of the duct is left unstented, these ends narrow and close causing severe hypoxia. If angiogram shows any unstented portion of the duct, an additional overlapping stent is deployed to cover the entire ductal length.

In Type II and III ducts, a *cut pigtail* from the femoral arterial access often cannulates the aortic end of the duct and helps in wiring the duct. If the floppy guidewire fails to advance to a deeper position in the pulmonary artery, an additional guidewire as a buddy or a microcatheter will assist in advancing the guidewire. In a few patients, a transvenous catheter which advanced through the ventricle into the ascending aorta may prove useful to cannulate and wire the duct. This transvenous cannulation of the duct sometimes is favored as an attractive option as the duct origin forms an acute angle with the undersurface aortic arch. However, the drawbacks are its potential to create tricuspid and aortic valve insufficiency which might compromise hemodynamics in sick hypotensive neonates. The catheter may press on the atrioventricular conduction tissue and lead to transient heart block at the most

inappropriate moment. If all these attempts fail, alternative options of percutaneous ipsilateral *axillary artery access* or ipsilateral *carotid artery* entry through a cutdown exposure are attempted. As the ductal aortic origin is directly opposite to these arteries, cannulation is often easily done (Fig. 28.8). In spite of trying all these vascular access, about 5% of Type II and III ducts may be difficult to stent due to the tortuosity.

28.11 Tips and Tricks

- Even though transvenous ductal stenting helps in Type II and III ducts, hypotension and bradycardia may be anticipated for abovesaid reasons.
- Coronary stents can be expanded beyond their nominal diameter with higher inflation pressures.
- Covering entire duct with stent is very vital. If there is any unstented ductal tissue, an additional stent of the same diameter is overlapped to cover the entire duct (Fig. 28.10).
- Systemic hypotension immediately after DS due to large aortic run off into pulmonary artery is managed by dopamine infusions.

28.12 Pitfalls

1. Acute systemic hypotension, often managed with dopamine infusion and fluids.
2. Sudden increase in pulmonary blood flows resulting in congestive heart failure, managed by small doses of frusemide intravenously and restriction of fluids.
3. Unilateral pulmonary hyperperfusion due to preferential blood flows into one lung. Angiogram with end-hole catheters into the aortic end of the ductal stent may occasionally cause preferential flows into one lung (Fig. 28.11). In such instances, if echocar-

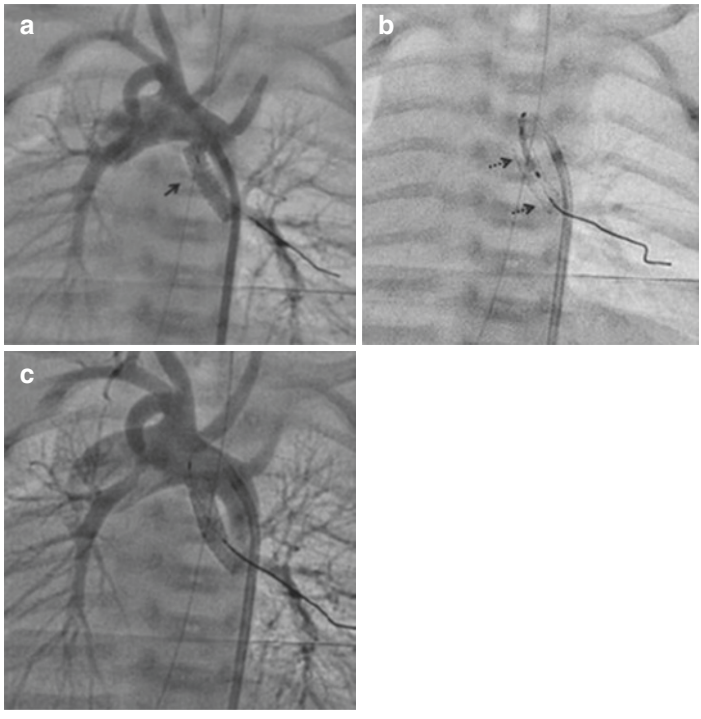


Fig. 28.10 In a Type V bilateral duct in a neonate with single ventricle, pulmonary atresia, aortogram (**a**) after stenting of the left-sided duct from the undersurface of the aortic arch shows that the aortic end of the duct is uncovered by the stent (*arrow*). The guidewire is still in place. A second stent is overlapped (**b**) into the previous stent shown in *dotted arrows*. Final angiogram (**c**) confirmed that the entire duct length is covered by the stent. The second duct from right innominate artery was stented subsequently in the same setting

diogram shows continuous flows into both pulmonary arteries, no immediate further interventions are needed.

4. Loss of femoral arterial pulses is often managed with heparin continued for 48 h.
5. Pulmonary hyperperfusion after DS is relatively easier to manage than an overflowing surgical shunt.

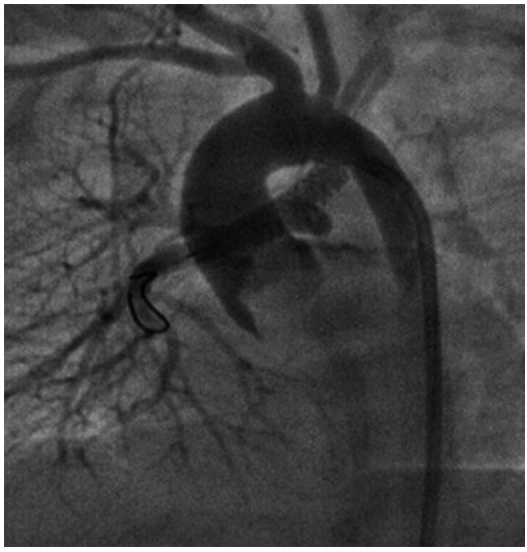


Fig. 28.11 Unilateral right lung hyperperfusion after ductal stenting in a patient with tetralogy of Fallot with pulmonary atresia

28.13 Complications

Immediate complications include failure to cannulate the ductus, ductal spasm, acute stent thrombosis, refractory hypotension, and heart failure due to overflowing ductal stent, dissection of the duct, stent embolization, groin hematoma, unilateral lung flows with complete lack of flows to the other lung (Fig. 28.11) and femoral arterial pulse loss.

Late complications include subacute stent thrombosis, progressive intimal ingrowth and stent restenosis, stenosis, or disconnection of the confluence of the pulmonary arteries.

28.13.1 How to Manage Complications

1. Type II and III ducts may prove difficult for DS from femoral artery. Alternative approach is through axillary or carotid

arteries. If the procedure fails, PGE1 should be restarted, and shunt surgery should be organized.

2. Acute *stent thrombosis* may be prevented by preprocedural aspirin therapy. It manifests as acute hypoxia a few minutes to hours after the procedure (Fig. 28.12). Additional dose of heparin is given immediately. If the guidewire is still in place, a coronary balloon is advanced into the stent to mechanically push the thrombus into the pulmonary artery. If sheaths are already removed, thrombolysis is done with streptokinase (2000 units/kg body weight bolus followed by infusion of 1000 units/kg/h) or tissue plasminogen activator (1 mg/kg bolus). These infusions may sometimes lead to uncontrollable groin site hematoma.
3. Acute *ductal spasm* is prevented by gentle guidewire manipulations and expedited stent deployment. If there is severe hypoxia, PGE1 should be restarted.
4. Acute *lung hyperperfusion and heart failure* occur with an oversized ductal stent and should be avoided. It presents with hypotension, respiratory distress, and tachypnea. It is managed with fluid restriction, diuretics, and dopamine. In extreme cases, if there is an alternative pulmonary blood flow, Amplatzer vascular plug IV (Abbott) may be used to close the stented duct (Fig. 28.13).

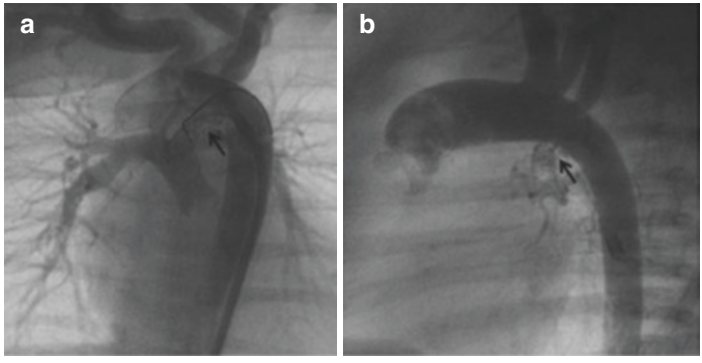


Fig. 28.12 Aortogram in shallow left anterior oblique view (**a**) shows successful stenting (*arrow*) of a Type III duct from undersurface of aortic arch in a patient with d-transposition of great arteries, large ventricular septal defect, and pulmonary atresia. After removal of the guidewire, repeat aortogram shows complete thrombotic occlusion (**b**) of the stent

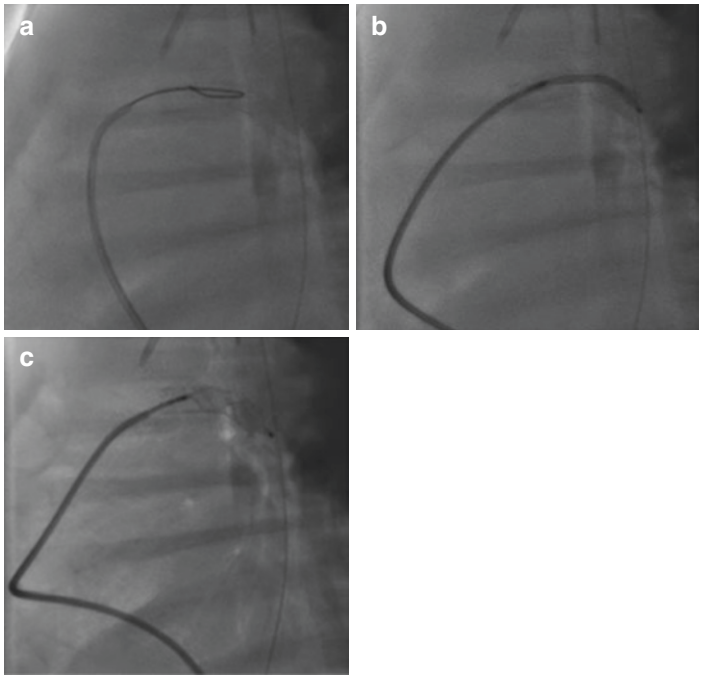


Fig. 28.13 A ductal stent which led to abundant pulmonary blood flow and aortic steal causing heart failure, coronary hypoperfusion, and ventricular dysfunction is crossed by a J-tipped floppy wire (which ensures that the wire courses through the lumen rather than through the struts), exchanged to a 4 F diagnostic catheter, that allows deployment of a small two-lobed vascular plug into the stent.

5. *Dissection* of the duct is identified by contrast staining of the ductal tissues. If the guidewire position is stable, a dissected duct should be stented quickly with a long stent to cover the entire ductal length. If guidewire position is unstable, it should be withdrawn and surgery should be planned (Fig. 28.14).
6. *Stent embolization* occurs in a non-constricted duct. It may also occur if PGE1 is continued during DS. Stent often embolizes into the pulmonary artery. If the guidewire is still in place through the lumen of the stent, it should not be removed. The child should be operated immediately and may need cardiopulmonary bypass. The retained guidewire

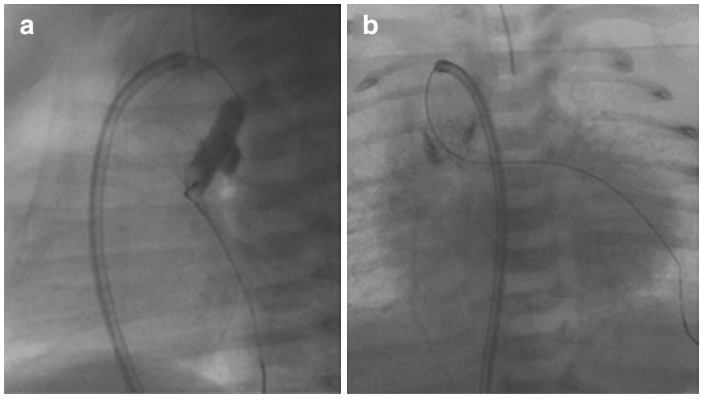


Fig. 28.14 Dissection of the duct during stent positioning (**a**), shown by contrast staining of the periductal tissues. This is immediately addressed by quick stent deployment (**b**). The staining of the ductal tissue with radiographic contrast disappears soon. Often the stent patency is not affected by this dissection and staining of the periductal tissues with contrast

will prevent too distal stent embolization beyond the hilar branches. In selected instances, balloon can be reintroduced into the embolized stent in the pulmonary artery, inflated to grip the mobile stent and bring it back to ideal position (Fig. 28.15). If the stent can be brought back to the duct again, a repetition of embolization can be prevented by intravenous paracetamol (10 mg/kg brief injection) or indomethacin (0.2 mg/kg brief injection) may induce spasm of the duct .

7. In patients with severe *confluence stenosis*, ductal stent will selectively protrude into either side leading to unilateral hyperperfusion. Such a duct can be cannulated with two wires from two different vascular access, and inverted Y-stenting can be performed to permit blood flows into both the lungs (Fig. 28.16).
8. *Circular shunts* result from repetitive continuous flow of blood from major blood vessels to cardiac chambers without traversing any capillary bed. Following pulmonary valvotomy for pulmonary atresia and intact ventricular septum

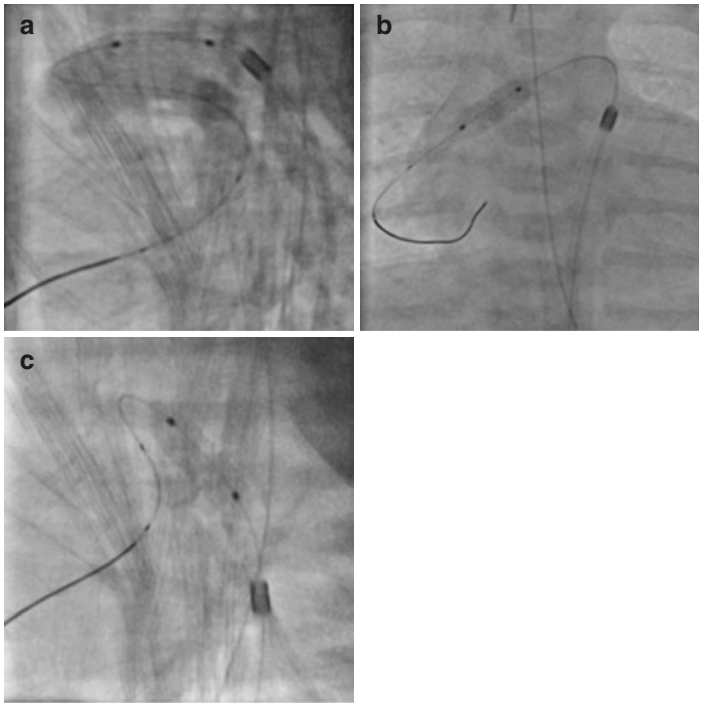


Fig. 28.15 A DS embolizes distally still over the wire into the main pulmonary artery after deflation of the balloon. Further manipulations lead to deeper advancing of the deployed stent into the right pulmonary artery. The stent was grabbed again with the balloon, brought back to the duct and redeployed

followed by DS, blood that flows from descending aorta to main pulmonary artery leaks back to right atrium through the leaking pulmonary and tricuspid valve to reenter systemic circulation through the patent foramen ovale (Fig. 28.17). This leads to severe hypoxia by depriving blood to pulmonary capillaries. Mechanical ventilation with high oxygen concentration, pulmonary vasodilator like nitric oxide or intravenous sildenafil to facilitate fall of fetal pulmonary vascular resistance are the common strategies in this self-limiting situation.

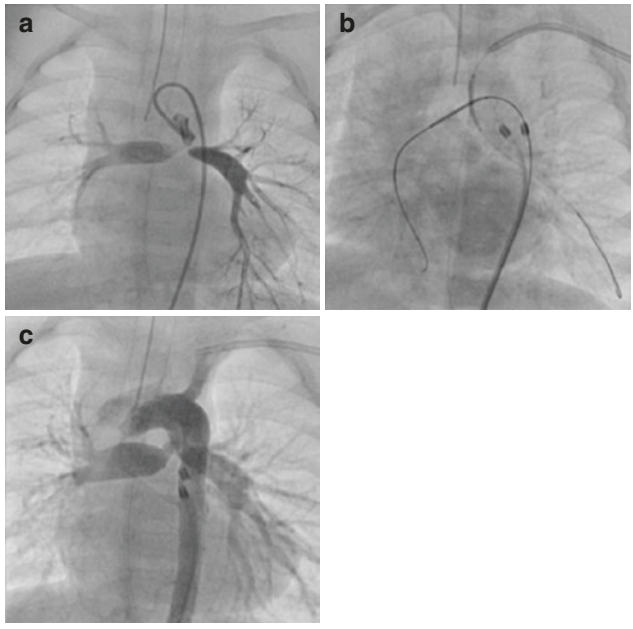


Fig. 28.16 Severe confluence stenosis often precludes DS. However, surgical repair will often require cardiopulmonary bypass to repair the confluence. In neonates who are poor-risk candidates, passage of two guidewires through different vascular access (one left axillary and one femoral artery) and Y stenting will result in symmetric perfusion of both lungs

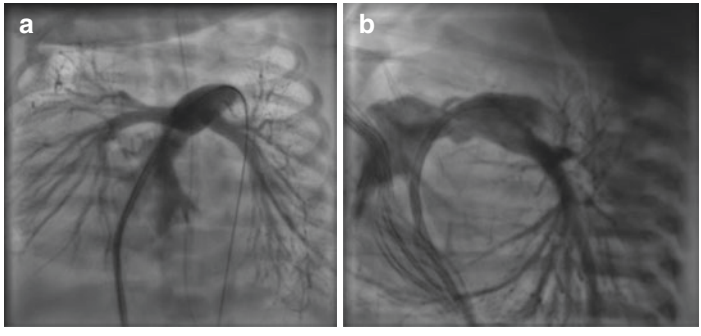


Fig. 28.17 DS was performed after pulmonary valvotomy through a transvenous guide-catheter advanced into the descending aorta. An injection in the main pulmonary artery after DS shows severe pulmonary regurgitation that later leaks through the tricuspid valve as evident from the marked cardiomegaly

9. In some of the vertical and tortuous ducts, axillary approach alone may not guarantee a successful cannulation of the duct with the stent (Fig. 28.18). If the guidewire enters a branch pulmonary artery with a tortuous loop, the stent may not track the entire duct. In such cases, the guidewire should be withdrawn to reenter the contralateral pulmonary artery with minimal looping.
10. In ducts that are too long and curved, single stent will straighten the duct and invariably protrude into the branch pulmonary arteries (Fig. 28.19). In such cases, an elective plan should be made to deploy two stents, initial stent to cover the distal pulmonary end and final stent to cover the aortic end.

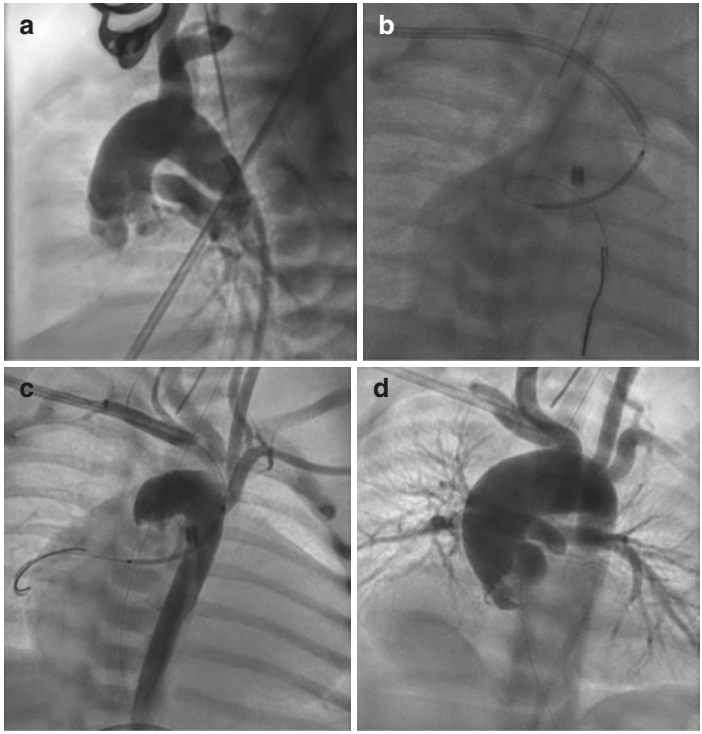


Fig. 28.18 A tortuous duct originates from undersurface of aortic arch with an S-shaped bend shown on aortogram. A right axillary artery access with guidewire parked deep in the left pulmonary artery fails to allow a stent to advance through the tortuosity. The duct is rewired to park the distal tip in right pulmonary artery, and this facilitates successful DS

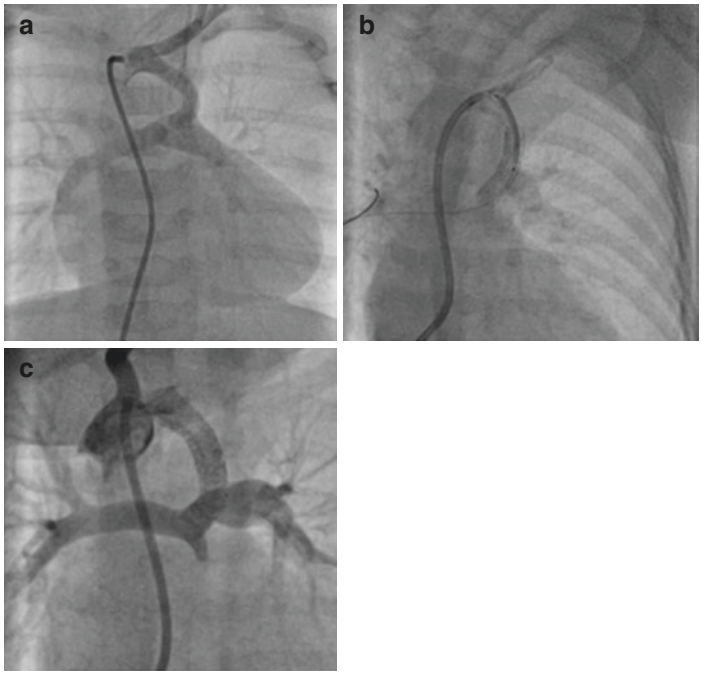


Fig. 28.19 A Type IV duct from left innominate artery is too long and curved to permit a single stent. Two stents are electively planned and deployed sequentially to retain the curvature of the duct

11. Some stents develop *accelerated instent restenosis* within a couple of months (Fig. 28.20). In such instances, rewiring the DS, balloon dilating the instent restenosis and restenting with another stent (preferably a drug eluting stent) may be needed.
12. Drug eluting stents are currently used in many neonates and infants for varied indications including DS. Even though serum sirolimus levels have been detected to be in immunosuppressive levels, use of a single stent up to 24 mm length has been shown to lead to drug levels well below the immunosuppressive range [4]. Patency of drug eluting stent is longer preserved than bare-metal stenting.

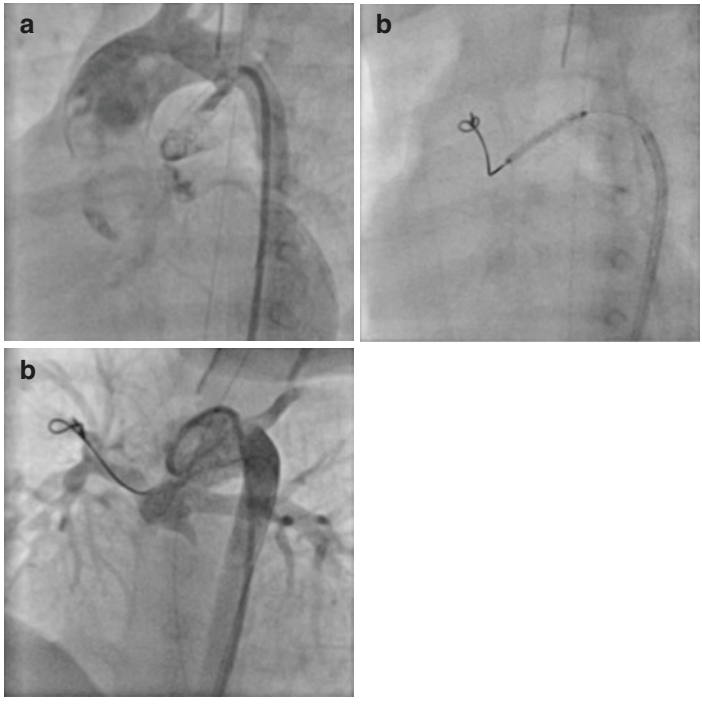


Fig. 28.20 Severe instent restenosis within 2 months leads to severe hypoxia. The stent was rewired from arterial access, ballooned with small coronary balloons to create path for an additional stent and finally restented with a drug-eluting stent

28.14 Postprocedural Care and Follow-Up

Aspirin and clopidogrel are continued till the next planned surgery in Group I and II patients. The patients are followed up monthly for oxygen saturations, clinical evidence of patency of ductal stent (continuous murmurs). Serial echocardiogram should document flows in the ductal stent, symmetric perfusion of both lungs, adequate growth of hilar pulmonary arteries and ensure lack of distortion and disconnection of the confluence. In Group I patients, surgery is delayed as long as clinically tolerated to ensure

a better body weight during surgery. In Group II patients, once the patient reaches about 5–6 months of age and somatic growth is adequate, they should be electively taken up for Glenn surgery. In Group III patients, where only transient ductal patency is desired, antiplatelets are stopped once ductal stent patency is no longer needed based on clinical evaluation.

Every ductal stenting procedure should be discussed in detail with the cardiac surgeons, and the decision to stent the duct should be taken after these discussions. In some very tortuous long ducts, it may be difficult to get a stable guidewire position, and this might result in procedural failure. The chances of failure in these tortuous ducts may be minimized by using microcatheters which will facilitate to advance the guidewire deeper into the pulmonary artery. It may be prudent to send some very tortuous long ducts direct to surgery. In the presence of severe confluence stenosis, the ductal stenting is contraindicated as it will facilitate ductal flows only to one lung and cut off the blood flows to the other lung, unless clear plans are made to overcome the confluence stenosis in a multidisciplinary discussion.

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29 PDA Stenting in Duct-Dependent Systemic Circulation

Dietmar Schranz

29.1 Anatomic Description and Physiopathology

Impaired systemic circulation caused by a morphological or functional incompetent systemic ventricle is completely or partially dependent on a subpulmonary positioned ventricle with a right to left shunting patent duct. The most common causes for duct stenting to guarantee a right-to-left shunt are congenital heart defects with "left-sided" hypoplastic structures ranging from severe aortic valve stenosis, hypoplastic left heart (Shone)-complex (HLH-C) to hypoplastic left heart-syndrome (HLH-S), but also severe aortic coarctation and some defects with associated interrupted aortic arch (see Chap. 38). Additionally, patients with a suprasystemic pulmonary arterial hypertension of variable genesis might benefit from duct resuscitation followed by stenting in order to create a reverse Potts shunt physiology.

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29.2 Clinical Scenarios

Independent of the reason of a disease, any neonate presented with tachypnea is severely ill. However, the symptom of high ventilation rate is often missed. Therefore, newborns with significant left-sided congenital heart malformations have the *disadvantage* for late detection and often misinterpretation as sepsis. Wherefore, a congenital heart defect with duct-dependent systemic blood flow has always be excluded in a tachypneic newborns. Prenatal fetal echocardiography helps to avoid postnatal heart failure or even cardiogenic shock caused by unperceived duct obstruction. Regarding a prophylactic therapeutic strategy in which prostaglandin infusion is employed, a low-dose infusion (5–10 ng/kg/min) is safe, but a high dose (25–50 or even 100 ng/kg/min) might be dangerous. A high dose is only indicated as a last resort in an emergency to treat severe duct obstruction, which is precisely what is the domain of percutaneous canal stenting. Therefore, for the employment of a prophylactic treatment regimen, a prenatal or immediate postnatal diagnosis is needed. Relevance for treating newborns in advance, before any clinical symptoms are obvious, highlights the need for sophisticated medical education.

29.3 Indications and Patient Selection

The outcome of newborns with hypoplastic left heart (HLH) is determined by many factors; particularly by the first-step palliative surgical procedure performed, independently of whether it is a Norwood procedure, Sano modification, or a challenging biventricular repair. The indication for duct stenting in a patient with duct-dependent systemic blood flow could be prostaglandin-refractory duct obstruction or as a part of an elective therapeutic strategy. In the past, about 10% of newborns seemed to have an unfavorable duct morphology for stenting, mostly in term of the junction of the duct to the descending aortic arch [1]. This feature was even more pronounced when balloon-expandable stents were the only option for duct stenting. Use of a large introducer sheath, the need for a trans-ventricular access through the tricuspid and

pulmonary valves, and the associated hemodynamic instability did not forgive any further interruption of the flow, no matter how brief, but complete, which always was always the case when the balloon was inflated for stent expansion. From a technical point of view, the use of new self-expandable stents, that can be delivered via a 4-F sheath makes duct stenting in duct-dependent systemic blood flow easy and feasible in almost all newborns, regardless of morphology or hemodynamics.

29.4 Treatment Options (See Also Chap. 39)

Duct stenting combined with bilateral pulmonary artery banding (bPAB), and if necessary, interatrial septum manipulation, was developed as a hybrid method [2]; this is offered as an alternative first-step approach at a number of centers worldwide. However, the Hybrid Step I approach is mostly used in newborns with high-risk HLH-S or HLH-C [3, 4]; some obtained results seem to indicate a lack of experience, if exclusively used in high-risk patients [5].

29.5 Pre-procedural Considerations/Imaging

Clinical condition, heart rate, oxygen saturations, and blood pressure measurement at all extremities have to be related to the neonatal diagnosis of HLH-S or HLH-C, which is obtained by echocardiography (ECHO) in a standardized manner; transverse and longitudinal planes with special emphasis on demonstrating the four-chamber view, outflow tracts, and a three-vessel view using two-dimensional (2-DE) color flow, and spectral Doppler interrogation. An additional focus of the echo examination needs to be on the pulmonary vein and systemic vein flow, the quality of interatrial communication, tricuspid and systemic ventricular function, as well as the duct morphology, in terms of its length and width and its relationship to the pulmonary branch arteries and descending aortic arch (Fig. 29.1). Blood flow obtained by pulse Doppler measurements of the cerebral anterior artery as

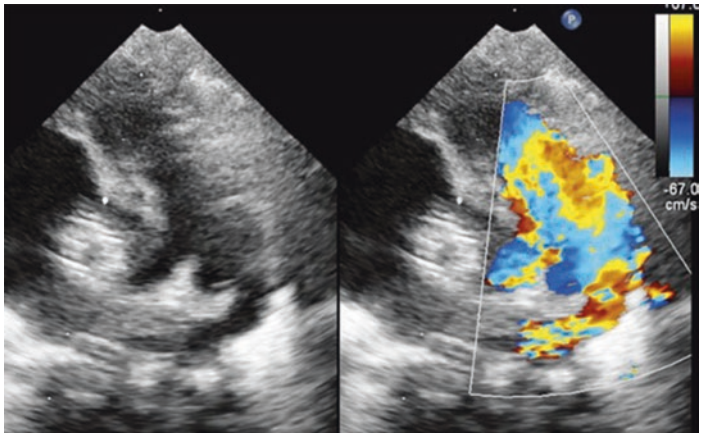


Fig. 29.1 Echocardiography (ECHO) evaluation of duct morphology, in terms of width, length, and touristy and its relationship to the pulmonary branch arteries and descending aorta

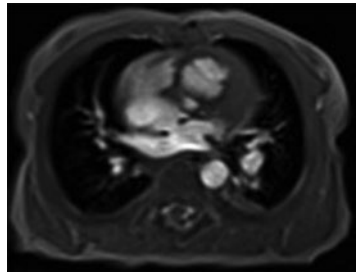


Fig. 29.2 Magnetic resonance imaging (MRI) analyzing pulmonary vein morphology, atrial communication, and aortic arch malformation

well as abdominal truncal flow belongs also to a complete ECHO update before and after DA stenting.

Percutaneous duct stenting is based on the pre-procedural imaging in particular ECHO; magnetic resonance imaging (MRI), even performed in only a sedated patient, is indicated to answer any open questions before Stage I; in particular, regarding detailed duct and aortic arch or pulmonary vein morphologies (Fig. 29.2).

Further considerations are summarized in Table 29.1.

Table 29.1 Consideration of elective duct-stenting in duct dependent—SBF

1.	Prophylactic anesthesia (intubation + ventilation) is a **high-risk** factor
2.	Duct stenting needs to be planned on ECHO and/or MRI -Imaging
3.	Duct-stenting has to be performed on local facilities (Sinus-Superflex-DS?)
4.	Decision for trans-venous or -arterial access best prior to catheterization.
5.	Beginners should opt for both accesses (intervention & diagnostic reasons)
6.	Should duct-stenting be performed as a single or part of multiple approaches?
7.	Local groin anesthesia after **analgo-sedation**; 4(5) F vein, 4 F arterial access
8.	Delineation of the DA in context of DA-DAO and DA-PA /LPA
9.	Stent-length for covering of both DA ends; stent-width 1–2 mm > DA + DAO!
10.	Precise stent placement by markers; gastric tube, arterial MP + wire/ PA-JK
11.	Angiographies (hand-injections?) angulated in RAO 30° and lateral 90°
12.	Obstructed DA should preferentially by stented by Balloon-expandable stents
13.	Non-obstructed DA only self-expandable stents allow safe placement
14.	Analysis of DAO/CoA prior to, but in any case after!!! (Cath and ECHO)
15.	Intra-and post-interventional medication (heparin, continuous PGE1 infusion)

Cath catheterization, *CoA* aortic coarctation, *DA* arterial duct, *DAO* descending aorta, *ECHO* echocardiography, *JK* Judkins catheter, *LPA* left pulmonary artery, *MP* Multipurpose catheter, *MRI* magnetic resonance imaging, *PA* pulmonary artery, *RAO* right anterior oblique

29.6 Technique (Step-by-Step)

The technique of duct stenting has to be performed in context of the aim of the approach. There are two scenarios: (a) the DA has to support Qs in part, as it might be required in a physiology with a reverse Potts shunt in patients with PAH. (b) Patients with hypoplastic left heart complex (HLHC) require a variable Qs support

of the subpulmonary ventricle depending on the amount of the remaining antegrade flow. Therefore, duct stenting can be part of a hybrid procedure performed by a transpulmonary access during an open chest, beating heart surgery, immediately after bilateral PAB placement, or as an elective transcatheter approach in a spontaneously breathing, in only sedated newborn, with or without additional manipulation of the atrial septum [2, 4]. A complete or almost fully duct-dependent systemic blood flow in a neonate needs stents with a diameter of 7–9 mm; also in premature babies with a body weight of 1000–1500 g. In contrast, resuscitated ducts utilized for creating a reverse Potts shunt circulation need a (stent) diameter in relation to the provided support of the systemic blood flow considering a still acceptable hypoxemia. In contrast to duct stenting in HLHS as a part of a hybrid approach, stent diameters providing a reverse Potts physiology need in any case to be smaller than the diameter of the descending aorta. Infants with PAH, refractory to specific PAH treatment, are mostly sufficiently palliated by a 4- to 5-mm-wide coronary stent placed within the duct [6]. A sufficient support of Qs is usually achieved, if a syncope is avoided or a low chronic or intermittent left ventricular preload compensated. Older children or young adults with decompensating PAH benefit from a pulmonary-DAO (descending aorta) shunt or stented duct with a diameter ranging from 50% to 90% of the width of the descending aorta. The use of covered stents is recommended with the potential to re-dilate during the follow-up, when both the pulmonary and systemic circulation did adapt to the acute and newly created "parallel" circulation.

Technical details of duct stenting in HLHS are summarized in Table 29.2 and illustrated in Figs. 29.3, 29.4, 29.5, 29.6, 29.7, and 29.8. Percutaneous duct stenting by *femoral artery access as a part of the Giessen Hybrid approach is summarized in* Chap. 39. Additionally, indication, timing, and technique of atrial septum manipulations are described.

Elective duct stenting needs to be well prepared, which includes any step of the interventional catheterization, as well as potentials of complications; therefore, the stock has to have sufficient catheter materials. After analgo-sedation and local groin anesthesia, usually the femoral vein and artery are punctured and

Table 29.2 Duct stenting In DA-SBF How to do

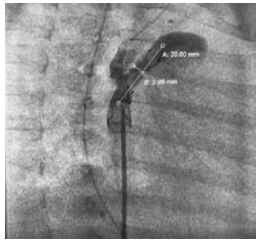
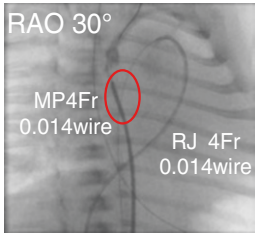
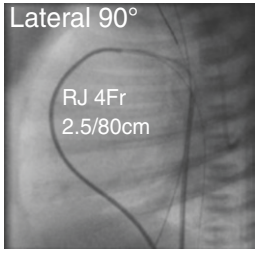
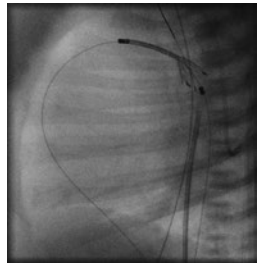
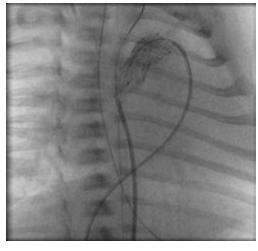
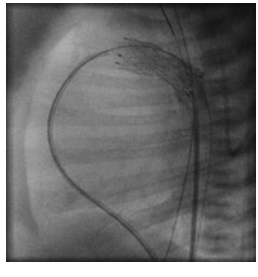
A	
Step 1 (Fig. 29.3): Bi-plane angulation RAO 30° and lateral 90°; Guided by RAO 30°: Placement of a 4 F MP together with 0.014″ BMW coronary wire (hemostat-ventil!) in DAO little distal of the DA insertion for delineation of DA-DAO junction by contrast medium injection per hand! Decision making for trans-venous or arterial DA stenting! Measurement of the duct morphology (approximate length and width)	 **Fig. 29.3** Arterial Duct delineation in RAO 30°
Step 2: Starts with AP-plane for placing a second coronary-wire per 4 F right Judkins catheter (RJ; 2.5 curved) from IVC across the TV, RV, PA trough the DA in the DAO: then back to the RAO 30° (Fig. 29.4); wire-MP-cross, point of provided distal DA stent placement! Exchange of the BMW to S' port stiff wire through the RJ for stenting.	 **Fig. 29.4** Arterial Duct/ DAO junction RAO 30°
Step 3: Proofing of wires and catheters positions in RAO 30° and lateral 90°planes (Fig. 29.5) including gastric tube as marker	 **Fig. 29.5** Arterial Duct/ DAO junction in lateral projection

Table 29.2 (continued)

B	
Step 4 (Fig. 29.6): after decision for a transvenous approach, the SSF-DS (8 × 20 mm) is already advanced over the stiff coronary wire in the desired duct; the 4 F delivery system is partially removed and the distal end of the stent is already opened, MP in DAO. BMW-wire in aortic arch for guiding exact stent placement	 **Fig. 29.6** Partial Sinus-Superflex-DS Stent expansion
Step 5: Full stent-expansion - from the aortic (RAO 30°) to the pulmonary end (lateral 90°) - is slowly and controlled performed with MP + wire in the DAO as a marker. Expansion of the SSF-DS doesn't interrupt the DA flow. The optimal stent placement is shown in both, favored planes! RAO 30° (Fig. 29.7) visualizes the stented duct and its relation to the DAO-AOA-junction; the fully covered duct at the pulmonary end is shown on the lateral plane (Fig. 29.8)	 **Fig. 29.7** Full Expansion of SSF-DS in RAO 30°
Step 6 Angiographic + hemodynamic re-evaluation, with focus on the stent expansion and the aortic isthmus area! Prostaglandin infusion is continuously applicated in a dosage of 2–5 ng/kg/min for further 24–48 h; ASS is not necessary, but careful post-procedural clinical and échocardiographie observation.	 **Fig. 29.8** Expanded SSF-DS in 90° lateral projection

a 4-F Terumo® sheath is placed within both vessels. For free crossing of the tricuspid valve, an in-floated 4 F open-end balloon catheter is advanced within the right ventricle (RV); there is no need to go to pulmonary artery or across the duct. It is easier to advance a soft coronary guidewire (BMW, Abbott, Wetzlar, Germany) through the 4-F wedge catheter in the pulmonary artery (PA), through the arterial duct, directly into the descending aorta (DAO). Alternatively, a 4-F RJ (2.5 curve) can be used for exchanging the wedge catheter or for the initial steps of the procedure. When the RJ is placed within the PA, DA, or DAO, a second stiff coronary wire (S'port, Abbott) is additionally and in parallel positioned to the already advanced soft coronary wire in the DAO; after double wire placement the soft wire is removed. The RJ can also be used for pullback pressure monitoring from the DAO to the PA over the remained stiff coronary wire by utilizing a hemostat valve; lastly, the RJ is exactly positioned close to the pulmonary end of the arterial duct. Further, the utilized hemostat valve allows to perform angiographies by manual injection of contrast medium, if necessary.

A step-by-step approach for duct stenting utilizing *femoral vein access* is given in Table 29.2a, b and Figs. 29.3 and 29.4. See also the text.

Percutaneous, transcatheter duct stenting is performed by a femoral vein or an arterial access. Depicted is the transvenous procedure, in which a 4-F multipurpose catheter (MP) is additionally placed in the femoral artery through a 4-F Terumo® sheath; the MP serves for blood pressure monitoring and delineation of the duct–aortic junction by repetitive angiographies per hand-injection of contrast medium as well as a marker in lateral 90° and right anterior oblique (RAO) 30° planes (see Figs. 29.3, 29.4, and 29.5).

Duct stenting by the femoral vein access is best guided by angiographies preferentially performed via a 4-F multipurpose catheter positioned at the aortic duct junction and guided by a BMW coronary guidewire. Biplane, 30° right anterior RAO, and 90° lateral are the favored planes depicting the arterial duct. Biplane imaging also reduces the procedure time and improves safety. As mentioned before, the diameter of utilized stents should be at least 1–2 mm bigger than the minimal measured duct

diameter, but in any case 7–9 mm in a newborn with complete duct-depending systemic blood flow and further exceeding the diameter of the DAO. Considering a possible stent embolization from the arterial duct to the DAO, the recommendations are of particular importance in newborns with an interrupted aortic arch. In case of utilizing a pre-packed, self-expandable Sinus-Superflex-Duct Stent (SSF-DS, OptiMed, Karlsruhe, Germany), the 4 F delivery system needs to be carefully flushed at the wire and side-arm port; the covered ensembled stent system can easily be advanced through a short 4 F sheath and exactly positioned within the duct, if guided by a stiff S'port wire, which is placed over the tricuspid valve, right ventricle, PA within the DAO. The MP together with the BMW guidewire serves also as a marker for stent positioning and during expansion. Slow pulling back of the stent-covered delivery system enables safe and full stent expansion with exact positioning under fluoroscopy control; rarely, a short cine scene is necessary for sufficient visualization of very thin struts of the open-cell designed stent. The lateral 90° plane serves for controlling the stent position at the PA end of the duct and the RAO 30° plane observing the exact stent position in relation to the junction of the arterial duct and DAO. Any step of the procedure can additionally be controlled by short contrast injection through the MP. Following full deployment of the self-expandable Nitinol stent, the delivery system needs to be carefully removed avoiding stent de-positioning; therefore, a short time period is needed for full expansion of the nitinol material at a temperature of 37 °C. Open-cell nitinol devices have facilitated duct stenting with non-obstructed wide-open lumen. Prostaglandin had not been stopped during the procedure. Far from it, we let run PGE1 in a dosage of 5–10 ng/kg/min continuously for further 24 (48) h together with a continuous infusion of heparin (300 U/kg/day). The PGE1 and heparin strategy was based on the hypothesis that the stretched and the foreign stent material-irritated duct will have a lower immediate constriction rate (radial force of the nitinol is limited) and may be associated with less hyperproliferation reaction in long-term. Therefore, we also do not recommend a cyclo-oxygenase inhibitor (ASS) for anti-aggregating purposes; and if, we recommend clopidogrel in a dosage of 0.2 mg/kg once

per day, a dosage, which was evaluated in the "Picolo®" study for neonates [7]. Considering a wide single-stent lumen, without kinking or residual obstruction and without over-stenting the descending aortic arch, usually we do not use any antiaggregating drug; only neonates, who received one or two additional stents in telescope technique for covering the duct in total, receive long-term treatment of with clopidogrel. Clopidogrel treatment is stopped 1 week before comprehensive Stage II or any other follow-up surgery; it is replaced by continuous infusion of heparin or enoxaparin-natrium 100I.E./kg as a daily single dose application. In general, the open-cell SSF-DS device with its low radial force has the advantage of delivering through a 4-F short sheath from the venous as well as arterial side with smooth expanding within the arterial wall of the duct, but despite these properties, there is always a risk of stent destabilizing during and after positioning; Figure 29.9 depicts a sling-fixed SSF-DS slightly mal-positioned before, migrated too far within the pulmonary trunk.

Therefore, before the stiff S'port wire is carefully removed after re-advancing the RJ catheter to the pulmonary end of the already stented duct, the appropriate stent position has to be evaluated by angiographies performed through the pulmonary positioned RJ and aortic placed MP. The junction of the descending aortic arch in relation to the stented duct, as well as the appropriate positioning of the stent in relation to the pulmonary trunk and left PA has to be carefully defined. It is not only important that the

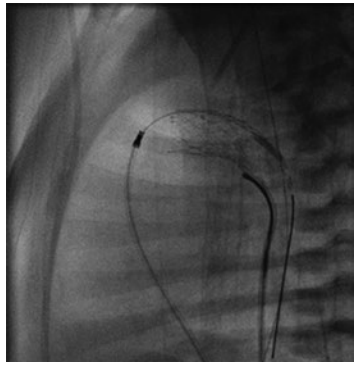


Fig. 29.9 Lateral view showing a second stent implantation, while the first stent is held by a snare avoiding embolization

duct is fully covered by the stent, but also the retrograde access to the aortic arch carefully analyzed excluding any significant obstruction. In most patients, there is no need to over-stenting the retrograde perfused descending aortic arch; but if, the aortic isthmus area needs to be carefully analyzed after stent placement; one additional reason why we recommend percutaneous duct stenting and preferred as an elective approach. The MP catheter still in ensemble with the BMW guidewire placed within the DAO allows easily such re-evaluation by pressure measurements as well as angiographies. Placement of an additional stent within the isthmus is based on pullback pressure measurements and results of an angiography (see also Chap. 39). Noninvasive systolic and diastolic blood pressure measurement, never mean blood pressure measurement at the right arm, if aberrant right subclavian artery is excluded, is the most important value to estimate the coronary and cerebral perfusion and perfusion pressures. It therefore makes sense to carry out noninvasive and invasive pressure measurements while still in the cath-lab; the PAP via RJ, the DAO-pressure by the MP and the non-invasive measured right-arm blood pressure, in order to exclude any significant pressure gradients across the stented duct and the aortic isthmus, respectively. Any kinking or slight obstruction within the SSF-DS should be re-dilated by 8 × 20 (30) mm Tyshak®-Mini or Sterling® balloon catheter, both are also advanceable through a 4-F sheath. The need for telescope stenting by placing two or three stents in series for fully covering the duct tissue has some additional careful considerations. At first, stents have to be overlapped like stent-in-stent. The stents do not have to impair the blood flow or force further kinking or compression of any stent; again, the self-expandable open-cell designed stents have, as mentioned, has a weak radial force, which cannot avoid any stent constriction or even collapse. However, if the first placed nitinol stent is kinked and not expanded by re-ballooning, a second nitinol or balloon-expandable stent can be used for effective treatment. Considering the duct–DAO junction, and the need for crossing the descending aortic arch by a duct stent, it should be avoided to place the second stent that the retrograde blood flow to the brain and coronaries is impaired by the struts of two stents. Lastly, the procedure of duct stenting is then finished, when the

hemodynamic and clinical condition of the patient remain stable, which takes time sometimes hours or perhaps days; in other words, the interventionalist knows best if he/she has achieved an optimal or only a sufficient result. The first 8 days after the procedure needs a continuous clinical monitoring with heart rate, SaO_2, and blood pressure monitoring as well as echocardiographic re-evaluations.

29.7 Materials

In the past, duct stenting was almost exclusively performed with balloon-expandable stents. Today, despite PGE1 treatment, only an obstructed arterial duct is stented utilizing a balloon-expandable stent. In such a case, we prefer to use a premounted Formula® stent with a width of 7 or 8 mm, which can even easily be advanced through 5 F sheath by assessing the femoral vein; in case of an obstructed duct, the Formula stent can also be advanced without a long guiding sheath; removing of a deflated balloon after stenting of a stenosed vessel has is less likely to cause the stent to slip, than stenting a large, un-obstructed vessel. In general, balloon-expandable stents have been replaced by self-expandable stents, when a usually non-obstructed large duct needs to be stented. Self-expandable stents allow stenting of large, non-obstructed vessels with low risk and high success rate, the blood flow is not interrupted as this is the case utilizing balloon-expandable stents. This has to be considered, in particular in HLHS with very tiny ascending aorta.

The Sinus-Superflex-DS TM (SSF-DS; Opti-Med, Karlsruhe, Germany) stent is certificated for duct stenting. The SSF-DS® is a self-expandable, open-cell nitinol stent available with diameters from 4 to 9 mm and stent lengths of 12, 15, 18, 20, 22, and 24 mm. All stents are covered by a delivery system, which can be advanced through a 4-F sheath. The use of a long sheath is not necessary, which further reduced the risk of duct stenting. The need only for a 4-F sheath allows duct stenting in newborns safely not only by a femoral vein but also by femoral artery access; by this, the approach of duct stenting is enormously facilitated.

In summary, neonatal duct stenting with duct-dependent systemic blood flow is performed by a SSF-DS TM, which has a CE (Conformité Européene) mark for its specific indication of duct stenting in newborns in particular with HLHS and HLHC; the self-expandable stent design with its limited radial force is preferentially used in wide, non-obstructed ducts. In obstructed or resuscitated arterial ducts, balloon-expandable stents are preferentially used. Our best experience is made by utilizing a Formula® (Cook) pre-mounted stent, available in lengths of 12–20 mm and mostly sufficient diameter of 7–8 mm, advanceable through a 5-F introducer sheath.

29.7.1 Currently Used Materials

- Puncture needle (Vygon, Aachen, Germany, 2 F arterial set) 4 F sheath (Terumo®)
- 4-F wedge, balloon open tip catheter (Cordis®, Hamburg, Germany)
- 4-F right Judkins catheter (Cordis®, Hamburg, Germany); 4 F Multipurpose catheter (Cordis®, Hamburg, Germany); Hemostat valve
- 0.035-in. wire for introducing the catheters through the vessel sheath (Cordis®, Germany)
- 0.014-in. coronary floppy wire (balanced middle weight, Abbott®, Wetzlar, Germany)
- 0.014-in. super stiff (support, S sport wire, Abbott®, Wetzlar, Germany)
- SinusSuperFlex-DS™ (Optimed®, Karlsruhe, Germany)
- Formula™ (Copenhagen®, Denmark)

In addition, a sufficient stock of materials is needed not only for the procedure of duct stenting itself, but, more importantly, for handling any possible complications, as snares and long sheaths in different sizes.

29.8 Expected Results

Percutaneous duct stenting in duct-dependent systemic blood flow can be performed with a mortality rate of less than 1% [8]. Performed as a straightforward procedure, the duct might be stented with a fluoroscopy time of less than 10 min utilizing a venous access, and less than 5 min by an arterial access. However, the results are dependent on the institutional experience, the general therapeutic strategy, and the materials used. In this context, it should be mentioned that experience is defined by the know-how to manage complications. The goal of any procedure should be an optimized outcome, with less complications. Complications can happen, but should not be a surprise; the team needs to be well-prepared dealing with all potential problems in advance. In term of complications and final results, duct stenting should not be started in complicated, high-risk scenarios.

29.9 Tips

29.9.1 Tip 1: Preparation

Before taking the parents'/carers' written consent, an exact plan, detailed catheter strategy should be prepared, best based on a self-performed echocardiography.

Remaining open questions, concerning pulmonary vein connection, single- or multiple-vein stenoses or unusual duct or aortic arch morphologies, can be answered by additional MRI imaging.

Detailed morphological knowledge prior to the catheter approach avoids any surprise during catherization.

29.9.2 Tip 2: Avoidance of Hemodynamic Instability

Patients with duct-dependent systemic blood flow need a high pulmonary vascular resistance or immediate and sufficient bilat-

eral PB. Anesthesia and controlled ventilation as a prophylactic tool might be dangerous may be more than the percutaneous transcatheter procedure itself. Therefore, catheterizations for duct stenting are usually performed in a spontaneously breathing patient. In context of a hybrid approach, duct stenting is an elective procedure; it is not must as long as low-dose PGE1 infusion prevents a duct obstruction. Duct stenting should usually be performed following surgical pulmonary banding; analgo-sedation is effective in utilizing diazepam and ketamine at very low but repetitive single dosages of 0.5–1 mg diazepam and 1 mg ketamine (both, 0.2–0.5 mg/kg). Hemodynamic reasons might also indicate the use of a 4-F open-end, inflated balloon catheter crossing the tricuspid valve for advancing a floppy wire through the duct in the DAO. Therefore, before a stent is delivered from the femoral vein and placed within the arterial duct, the hemodynamic stability has to be checked when a JR catheter is advanced in the PA. Borderline hemodynamic conditions, caused by persistent or artificial tricuspid and/or pulmonary valve regurgitation should decide for duct stenting by an arterial access (see Chap. 39).

29.9.3 Stent Placement

Following positioning of the stent-delivery system through the tricuspid valve, right ventricle, and pulmonary valve within the duct, careful releasing of the non-retrievable open-cell stent must be performed best guided by several markers as gastric tube and an MP in the DAO. Advancing of the delivery system needs to be performed without force during continuous invasively measured blood pressure.

29.10 Pitfalls (See Sect. 29.11)

1. *Morphology-dependent pitfalls:*
   Duct obstruction, duct aneurysm, aortic coarctation, tricuspid regurgitation

2. *Material-related pitfalls:*
   Stent expanding problems, stent-dependent obstructions, kinking; collapse (to less radial force); stent slipping caused by removal of the relatively stiff delivery system or balloon deflating weakness; open strut-related obstructions in term of placing a second stent; stent-related aortic isthmus obstruction

29.11 Complications

1. *Stent slipping, embolization*
2. *In-stent obstruction, insufficient stent expansion, secondary stent collapse*
3. *Retrograde aortic arch obstruction*
4. *Hemodynamic instability*

29.12 How to Manage Complications

To manage (1) stent slipping, embolization: snares and multiple stents of different lengths and diameter have to be in the catheter stock. Considering a stent diameter 1–2 mm bigger than the descending aorta, if, usually the stent migrates to the pulmonary artery side. If such a complication is observed during the release of the self-expanding Nitinol stent, the stent should be held with the relative ridge delivery system against the duct wall until a snare (5 or 10 mm) is prepared to catch the flexible stent from the femoral artery access. When the first stent is fixed by the snare and the stent is not migrated close to the pulmonary artery valve, but still with the distal part within the duct, the delivery system of the first stent can be removed, followed by a second stent placement, advanced from the femoral vein to the duct, both fixed by stent-in-stent placement in terms of telescopic stenting (Fig. 29.9).

To manage (2) immediate or short-term (in-) stent obstructions: utilizing SSF-DS™ means to place a self-expandable stent within a 1–2 mm smaller arterial duct lumen, in general without a significant stenosis. The open-cell device has the advantage of fixing

within a non-obstructed vessel. However, the relatively weak radial force of the very thin struts and the open-cell designed stent remains too weak to expand some obstructions. Therefore, in case of an insufficient stent expansion with observing a residual obstruction, re-dilation should be performed preferentially with a balloon also advanceable through a 4-F sheath; in most cases already a low pressure balloon with a diameter of 8 × 20 (30) mm (Tyshak Mini™) let expand the Nitinol stent, rarely high-pressure balloon (Sterling™) is necessary. Depending on the obstructed area, balloon catheters can be positioned from the venous or arterial access. Considering the high flow through the duct and the length of the stent-covered duct, re-dilation might be more effective utilizing a balloon with a length of 30 mm. Further, it has to be considered that a kinking within the nitinol stent or a placed second stent should not induce a secondary stent collapse during the immediate follow-up; rarely, a second balloon-expandable stent becomes necessary to solve the problem of an obstructed self-expandable stent.

To manage (3) retrograde aortic isthmus obstructions: we recommend proofing of the aortic isthmus before and after duct stenting, both by pressure measurements and by angiographic documentation. An acquired aortic coarctation is a serious complication. A low systolic blood pressure of less than 50 mmHg measured at the right arm, together with a high blood pressure gradient (>25 mmHg) between the noninvasively measured right arm blood pressure and the invasively measured blood pressure of the descending aorta, is one criterion of such a serious complication. The reasons for an obstruction could be a direct strut prolapse within an already slightly narrowed isthmus area, or an unfavorable strut position, which crossed an initial only mild coarctation. This complication can be minimized by placing a multipurpose catheter together with a coronary guidewire in the descending aorta. In case that over-stenting of the descending aortic arch is necessary, and a significant obstruction is detectable or the anatomy is unfavorable favoring a stenosis during the follow-up, stent placement should be performed in terms of a prophylactic or therapeutic tool. A coronary stent with a width of 4.5–5 mm

and a length of 8 or 9 mm can be usually used, but even a SinusRepo-DS™ closed cell device with a diameter of 5 or 6 × 9 mm (see Chap. 39).

To manage (4) any severe hemodynamic instability: pre-procedural assessment, which might include an advanced imaging, is most important to minimize intra-procedural instabilities. It has to be considered that duct stenting is not mandatory; only in prostaglandin-resistant duct obstructions, duct stenting is the most effective treatment. For cases presenting with severe hemodynamic instability despite an already performed bilateral PAB, duct stenting should be performed by femoral artery access. If possible, catheters from the venous access should be avoided; passing the tricuspid and pulmonary valves can force the hemodynamic instability or induce arrhythmias by touching a highly sensitive right ventricle (Chap. 39).

29.13 Post-procedural Care

Since we are using SSF-DS stents, prostaglandin infusion is not further stopped as previously reported when balloon-expandable stents were used. Further, we changed our management of prostaglandin infusion in terms that prostaglandin in a low dosage is continued for further 24–48 h in a dosage of 5 ng/kg/min together with heparin infusion (300 IU/kg/day). In the context of avoiding any duct constrictive or hyperproliferative measures, we also do not use cyclooxygenase inhibitors like acetylsalicylic acid (Aspirin®); routinely, we do also not administer any other antiaggregating drug; clopidogrel is used for anti-aggregation therapy, at a dose of 0.2 mg/kg per day, if two or three stents have been placed by the telescope technique within the duct, or when, in addition to the stented duct, a stent is placed within an aortic coarctation (see Chap. 39). A full stent covered duct without any kinking or slight obstruction is the most important preventive points avoiding follow-up problems. However, duct-related complications can develop anytime during the follow-up. Therefore, close follow-up observation needs to be performed not only

immediately during the few days after catheterization but depending on the aim for duct stenting by short- or mid-term controls.

In HLHS patients with a mandatory retrograde aortic flow perfusing coronary and the brain, clinical signs of heart failure (tachypnea) and blood pressure monitoring are essentials. It must be emphasized that systolic and diastolic, and not MEAN, blood pressures have to be measured and judged as single values, and in addition, the systolic pressure gradient between the right arm versus the leg that was not used for catheterization.

Echocardiography allows the assessment of early or late duct obstruction independently of an obstruction that is caused by in-stent proliferation or by constriction in an uncovered duct. A "pure" systolic velocity of less than 2.5 m/s across the stented duct is related to a less-compliant duct vessel but not to significant duct obstruction.

Discharge home is provided if the clinical, hemodynamic, echocardiography, and laboratory (brain natriuretic peptide [BNP] values) data obtained during several days after the transcatheter procedure are stable. A minimum post-discharge period of 10–14 days will probably be uneventful, and then the patient needs to be checked for clinical control as an out-clinic patient.

29.14 Follow-Up

Duct stenting is a palliative approach. Patients with completely duct-dependent systemic blood flow have a high mortality risk if there is any cause of duct obstruction. Therefore, close follow-up control is mandatory in all such patients until the next therapeutic step is performed.

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Part VI

Step-By-Step Procedures: Closing Or Creating A Defect

30 Step-by-Step Closure of Atrial Septal Defects (ASDs)

John Thomson

30.1 Introduction

Device closure of secundum atrial septal defects (ASDs) was initially described in 1974, but it was not until the Amplatzer septal occluder (ASO) (Fig. 30.1a) became available in the mid-1990s that it became a routine procedure. Since then, there has been significant progress in the ability of operators to tackle anatomically challenging defects, and transcatheter closure of ASDs is considered the procedure of choice for suitable defects in most countries.

There are broadly two types of occluder: self-centring (with a core) and the non-self-centring devices (with a thin central stalk). Self-centring devices are most commonly deployed due to their ability to deal with defects of most sizes.

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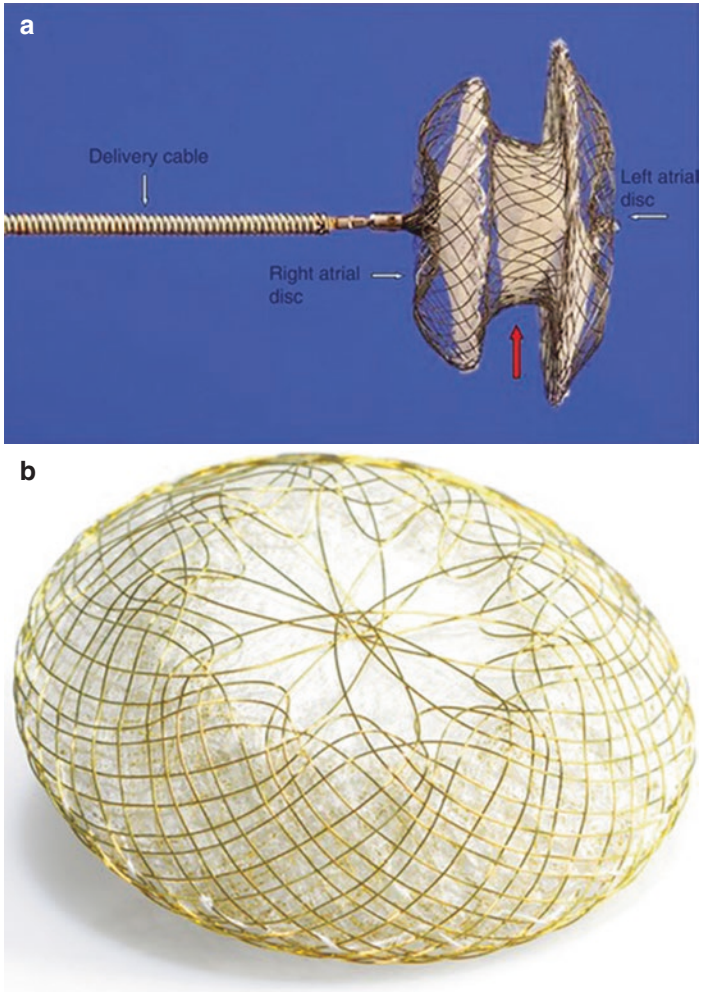

Fig. 30.1 (**a**) Amplatzer septal occluder (ASO). The red arrow points to the self-centring core of the device. (**b**) Occlutech Figulla occluder, left atrial aspect. (**c**) Occlutech Figulla occluder, articulation with the delivery cable. (**d**) Cardia Ultrasept occluder. Showing the left atrial disc and the central portion N.B. the right atrial disc is undeployed within the delivery sheath. (**e**) Gore septal occluder: side aspect. (**f**) Gore cardioform atrial septal occluder: side aspect. In contrast to the Gore septal occluder, (**e**) this has a central core

Fig. 30.1 (continued)

30.2 Anatomic Description

Deficiencies occur in a number of positions within the atrial septum, and an understanding of the anatomy is important when considering closure and the potential effects of a device within the heart (Fig. 30.2a). The atrial septum is not symmetrical. When

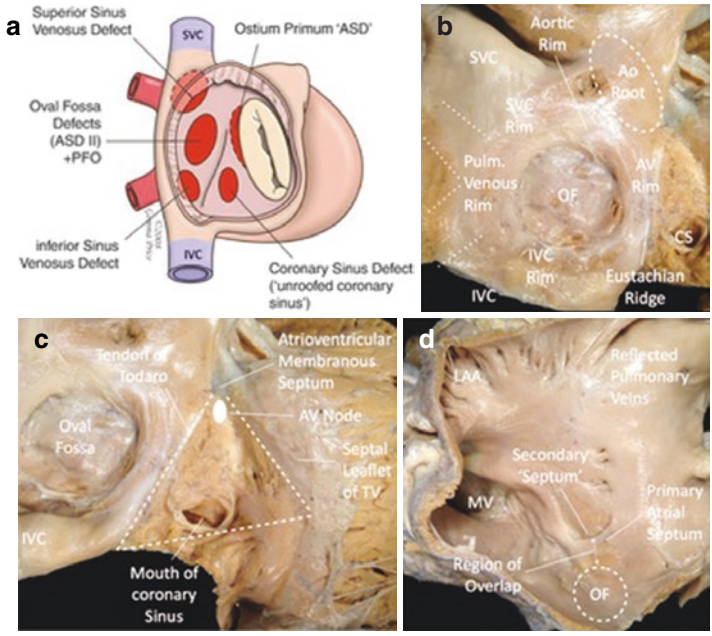

Fig. 30.2 (**a**) Schematic diagram of defects of the atrial septum, viewed through the right atrium (image courtesy of Dr Andrew Cook/Gemma Price). (**b**) Cadaveric specimen. View from the right atrium showing structures adjacent to the oval fossa (*OF*). *SVC* superior vena cava, *IVC* inferior vena cava, *CS* coronary sinus, *Ao* aorta (image courtesy of Dr Andrew Cook/Gemma Price). (**c**) Cadaveric specimen. View from the right atrium showing structures within the triangle of Koch. *TV* tricuspid valve (image courtesy of Dr Andrew Cook/Gemma Price). (**d**) Cadaveric specimen. View from the left atrium (image courtesy of Dr Andrew Cook/Gemma Price) *LAA* left atrial appendage, *MV* mitral valve

viewed from the right atrial side, a significant proportion of the atrial septum is made up of infoldings of the atrial wall, usually in association with other structures (Fig. 30.2b). From the right, postero-superiorly the septum consists of an infolding of the atrial wall between the superior caval vein and the insertion of the right pulmonary vein into the left atrium (as the heart sits within the chest). Anterior to this, the rim of the atrial septum continues behind the aortic root. Inferiorly, the rim of the atrial septum is in contact with the inferior caval vein and anterior to this margin is the interface with the coronary sinus, which in turn is separated

from the mouth of the inferior caval vein by the Eustachian ridge and valve (sinus septum). Deeper within the Eustachian ridge runs the tendon of Todaro, one of the three walls of the triangle of Koch, an area of the atrial septum containing several important structures including electrically active tissue (Fig. 30.2c).

When viewed through the left atrial wall, the anatomy is less complex. Only a small portion of the anterior region of the atrial wall is true septum, the rest representing overlap and fusion of the primary atrial septum (flap valve) with the anterior atrial wall (Fig. 30.2d).

The atrial septum is a curved structure and sits at an angle within the heart relative to the anteroposterior position of the thorax—this is a simple concept but one that poses challenges for the delivery of a device delivered through a sheath positioned along the natural line of the inferior vena cava. Defects in the secundum atrial septum are variable in both size and position. Although a proportion is truly central, eccentric holes with extension to any margin can occur. Secundum ASDs are not infrequently multiple and often sit within mobile or "aneurysmal" septal tissue. All of these anatomical variables must be defined prior to the delivery of a device.

30.3 Physiology

Assuming that an ASD is of significant size, the magnitude of the shunt is determined by the relative resistance to filling of the ventricles. Right ventricular resistance is usually less than the left, and therefore, the overall shunt is left to right. Flow across an ASD is phasic occurring predominantly in late ventricular systole and early diastole. Abnormalities of ventricular diastolic function in either ventricle (e.g. systemic hypertension leading to left ventricular hypertrophy) will affect the direction and magnitude of the atrial shunt and are part of the reason why atrial septal shunts increase in significance with age.

30.4 Clinical Scenarios: Natural History

The vast majority of infants and children with ASDs are asymptomatic, although many have a tendency to recurrent chest infections and respiratory symptoms. Very occasionally there will be an infant in whom an atrial shunt is responsible for failure to thrive.

During adult life, symptoms are progressive. Exercise intolerance is a common feature. Studies consistently show that with each passing decade, an increasing proportion of patients with ASDs display one or more of the characteristic sequelae of an important atrial shunt: pulmonary hypertension, atrial dysrhythmia or clinical right heart failure.

30.5 Indications for ASD Closure

Traditional indications for the closure of intracardiac shunts were based on invasive oximetry, with a shunt of >1.5:1 taken as significant. In the modern era, virtually all decisions on the significance of atrial shunts (and therefore the indications for closure) are made using non-invasive imaging *prior* to an attempt to close a defect.

There is an abundance of literature demonstrating that patients of any age with significant ASDs benefit from closure. In children, most units defer closure of ASDs until around the third birthday at the earliest unless the clinical situation is atypical, e.g. failure to thrive or there are particularly frequent chest infections.

Although in years gone by, there was an active debate about the need for ASD closure in older adults, the issue of benefit for these patients has been resolved by the publication of a number of studies. Established atrial dysrhythmia is rarely solved by closing an ASD in this age group, but shunt-related symptoms are improved, pulmonary hypertension is generally resolved, the right heart usually remodels to some degree, and clinical right heart failure, if present, is easier to treat medically.

Contraindications to ASD closure include defects that are anatomically unsuited to a device such as those that are greater than 40 mm in diameter or with inadequate margins. In these cases, surgery should be offered. Very rarely, there are patients who are affected by both primary pulmonary hypertension and a coexistent ASD. Often, these patients are younger women and almost always have clinical signs out of context with the ASD itself, e.g., cyanosis due to right-to-left shunting at atrial level. Closure of an ASD in this situation should be avoided.

Fig. 30.3 Occlutech flow regulator. Note the altered device weave leaving a permanent central hole within the device

In rare cases, there may be an argument for partially closing an atrial shunt. Often these are complex patients following previous cardiac surgery or with cardiac failure. The Occlutech atrial flow regulator (AFR), a device with a defined central hole is an option in some of these patients (Fig. 30.3), alternatively on table modifications to standard occluders can be employed.

30.6 Treatment Options

If an ASD is significant, then the options for closure are either surgery or a transcatheter-delivered device. It is important that patients and families are thoroughly counselled about the pros and cons of both the procedures, so that an informed decision can be made.

30.7 Device Options

1. Currently available self-centring devices:
   Amplatzer (St Jude) septal occluder (http://health.sjm.com/amplatzer-septal-occluder): Original nitinol framed occluder available in core central diameters from 4 to 40 mm (Fig. 30.1a).

Occlutech Figulla occluder (http://www.occlutech.com): Nitinol occluder with titanium coating (antithrombotic) in core diameters from 4 to 40 mm. The left atrial disc does not have a screw or metallic protrusion (Fig. 30.1b), and the delivery system is innovative, allowing articulation of the device on the delivery wire to facilitate closure of larger or asymmetric defects (Fig. 30.1c).

Cera (Lifetech) occluder (http://www.lifetechmed.com): Titanium-coated nitinol occluder in 6–42 mm core sizes.

Cardia (http://www.cardiainc.com): The Ultrasept ASD occluder is constructed of nitinol/titanium wires covered with Ivalon sails with core sizes from 6 to 34 mm (Fig. 30.1c).

Gore cardioform ASD occluder (http://www.Goremedical.com): Interlocking nitinol wire frame with an ePTFE cover. A series of sizes allowing closure of defects up to 35 mm including those with deficient retro-aortic rims are available.

Device delivery systems: The majority of the devices described above have their own pre-curved delivery sheaths of varying internal diameter (depending on the size of device) to facilitate delivery of the device. They are generally loaded into a short tube before being introduced into the delivery sheath. Modern self-centring devices are generally retrievable and repositionable prior to final release from the delivery cable.

2. Non-self-centring devices:

Occlutech, Cera and St Jude (Amplatzer) all produce a variant of their self-centring nitinol mesh-based device with a thin central core designed for coverage of multiple defects. These devices give maximal coverage of the defect without the restriction of a central core. This facilitates coverage of multiple holes at the expense of stability (i.e., the potential for movement) within the septum. These devices are delivered in exactly the same manner and through the same delivery sheaths as their self-centring equivalents.

Cardia: Cardia produce an Ultrasept cribriform based on the same principle as the Cardia ASD occluder but without the central core.

Gore Septal Occluder (GSO) (http://www.goremedical.com/eu/septaloccludereu/): Made from five interlocking nitinol/platinum wires covered with an ePTFE shell. The GSO is a low-profile device (Fig. 30.1d). The device comes pre-attached to a delivery handle, and the delivery sheath is part of the pre-assembled system. Device deployment is by a simple movement of the delivery button, and the device can be retracted and deployed as many times as required prior to final locking and deployment.

30.8 Pre-procedural Imaging

Most units rely on transthoracic echocardiography (TTE) for initial assessment. Clear evidence of right heart dilation on TTE is usually a marker of a significant atrial shunt. In children, TTE can also reliably delineate the anatomy, the margins of the defect and the presence of associated abnormalities (e.g., anomalous pulmonary venous drainage or mitral valve disease). In adults, TTE sometimes does not provide the resolution to accurately define the anatomy and margins of the defect. In many cases of this sort, it is appropriate to move on to attempted closure with a careful trans-oesophageal echocardiography (TEE) assessment to check suitability for closure before catheterisation. In some cases, a pre-procedural MRI can be helpful. MRI has the additional advantage of providing accurate volumetric analysis and relative pulmonary/systemic flow ratios if there is any doubt about the indication for closure. It also provides clear imaging of the pulmonary veins. Against this, the spatial resolution of MRI means that imaging of the atrial septum itself is not always as accurate as with ultrasound.

An important part of the assessment of an adult patient with an ASD should be a full assessment of left ventricular function. Impaired systolic and diastolic functions can be masked by an atrial shunt and closure of the defect in such patients can (rarely) precipitate pulmonary oedema. Therefore, in older adults (>50 years), it is important to assess and if necessary treat coexistent coronary abnormalities/disease prior to ASD closure.

30.9 Techniques: Step by Step

1. Pre-procedure

   Ensure appropriate patient selection.

   Is there adequate pre-procedural imaging?

   Has all comorbidity been appropriately excluded and/or treated?

   Has the patient/family been adequately counselled and consent properly sought and obtained?

   Is there a catheterisation plan in place? What is the agreed minimum dataset to be collected?

   Is the correct equipment available?

   Have "safe catheterisation" checklists been completed with the team performing the procedure?

2. Imaging at the time of the procedure

   Either using TEE or intracardiac echo (ICE)

   Establish anatomy including:

   Size and numbers of defect(s)

   Margins (Fig. 30.4a, b)

   Other important structures including anomalous pulmonary venous drainage

3. Catheterisation

   Initial placement of a 6-F venous catheter in the femoral vein using the Seldinger technique either using palpation/landmarks or ultrasound guidance.

   Systemic heparinisation (100 IU/kg)

   Diagnostic catheter study performed (agreed local unit minimum dataset obtained)

   Atrial septum crossed; catheter positioned in left-sided pulmonary vein

   Placement of a stiff exchange length wire

   Sizing of the defect (see below)

   Insertion of delivery sheath via the guidewire

   Placement of the correct-sized device:

   Each device type has its own deployment characteristics which should be mastered. The Amplatzer (St Jude), Occlutech and Cera occluders are relatively easy to deliver

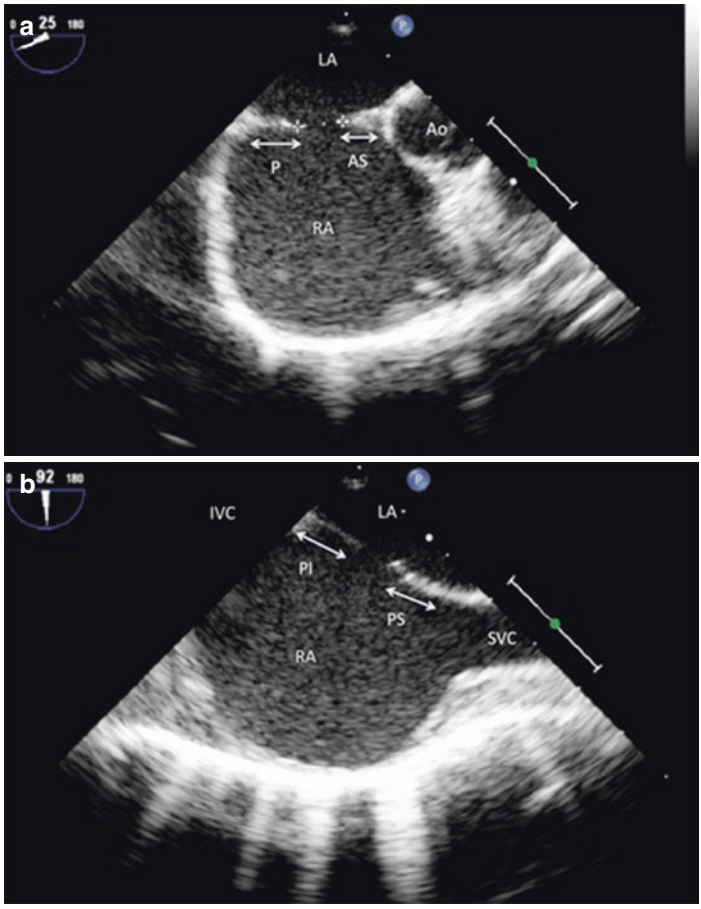

Fig. 30.4 (**a**) Trans-oesophageal echocardiography, "30°" view. *RA* right atrium, *LA* left atrium, *Ao* aorta, *P* posterior atrial rim, *AS* anterior-superior atrial rim. (**b**) Trans-oesophageal echocardiography, bi-caval view. *PI* postero-inferior atrial rim, *PS* postero-superior rim. The arrows delineate the extent of the tissue margin around the defect

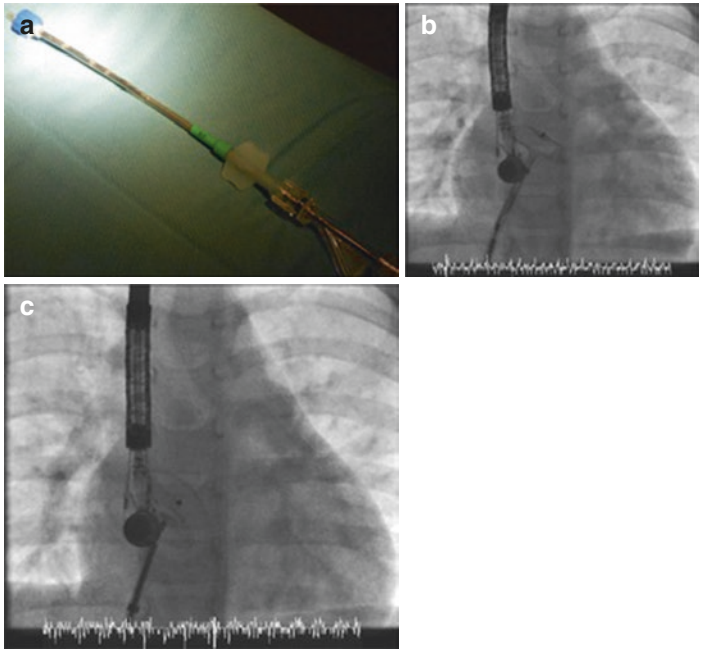

Fig. 30.5 (**a**) ASO loaded in delivery sheath. (**b**) Fluoroscopy. ASO, left atrial disc deployed. (**c**) Fluoroscopy. ASO, device fully deployed

into central, small- to medium (<20 mm)-sized defects. The chosen device is inserted into the delivery sheath using the proprietary loading tube (Fig. 30.5a) and thoroughly flushed with heparinised saline. Via the delivery sheath, the device is then advanced to the left atrium (Fig. 30.5b). Avoiding air ingress during this phase is important. The left atrial disc is reformed in the mid-left atrium by pushing the device forward and withdrawn onto the atrial septum using ultrasound guidance. Once the LA disc is against the left side of the septum, the core is developed, the right disc developed (Fig. 30.5c) (by pushing the device forward) and the occluder is thoroughly checked on ultrasound using a systematic review of the rims and edges of the device. Interference with other structures should be ruled out. Once the correct position

is confirmed, a stability check can be performed by pushing and pulling the device. The device is then unscrewed or unlocked from the delivery cable and released.

30.10 Materials

30.10.1 Essential Equipment for ASD Closure

1. Catheters/sheaths/wires:
   (a) Short access sheaths in sizes up to 12 F
   (b) 5/6 F Multipurpose-type catheters
   (c) 0.035″ Super-stiff-type exchange wire with a floppy tip
   (d) Standard device delivery sheaths in all sizes
   (e) Hausdorf modification of the Cook Mullins delivery sheath for difficult defects
   (f) St Jude/AGA "Rescue sheath" to enable the creation of an exchange system (by screwing together two wires to create a double-length cable) in the event of an emergency
   (g) Cook Flexor (or other braided type) sheaths (10–12 F) in case of the need to retrieve an embolised device
   (h) Goose-neck snares
2. Devices: A full range of occluders (4–40 mm in increments for the ASO)

30.11 Tips and Tricks

1. Sizing: For single ASDs many operators prefer to simply measure the defect using either TEE or ICE in two perpendicular planes, taking the largest diameter on colour Doppler flow and selecting the next device size up to avoid significant oversizing (Fig. 30.6). If balloon sizing is utilised, it is usually performed using the static balloon technique. The fluoroscopic angle should be adjusted to ensure that the balloon is truly perpendicular to the atrial septum and not foreshortened. Twenty-five percent contrast solution is used for gentle inflation. Care must

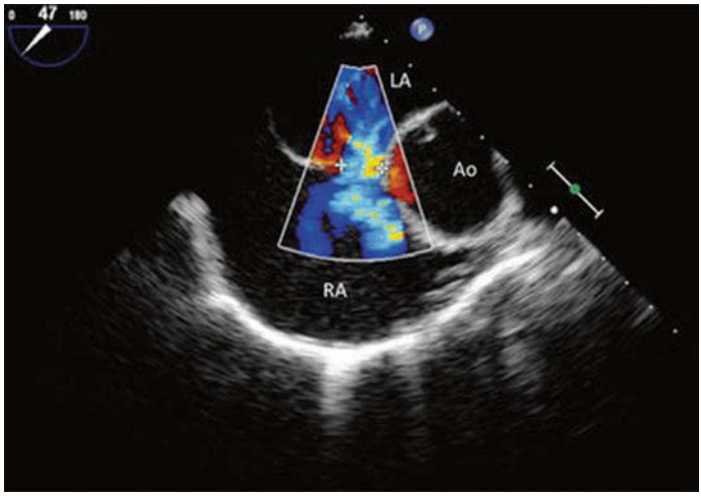

Fig. 30.6 Trans-oesophageal echocardiography, colour Doppler: markers showing sizing using colour Doppler. *RA* right atrium, *LA* left atrium, *AO* aorta

be taken not to "balloon dilate" and stretch the septum (some operators use a pressure monitoring device during inflation as a safety measure) as this can lead to systematic oversizing. Most device manufacturer instructions for use (IFU) now recommend the use of ultrasound imaging during balloon inflation using the point at which colour flow is abolished (the so-called stop-flow technique) for definitive sizing. Whilst the balloon is inflated, there should be a systematic evaluation of the atrial septum for additional defects.

2. Occluded femoral veins: Even using steerable sheaths, it is difficult to deliver an occlusion device using the jugular venous approach due to the angle of the atrial septum. If femoral venous access is not possible, then it is my preferred method to use a trans-hepatic approach.
3. Large defects: A significant proportion of defects are neither small nor central, and these are much more of a challenge for the delivery of a device. Deficiencies of the antero-superior septum are very common in defects larger than 20 mm. The

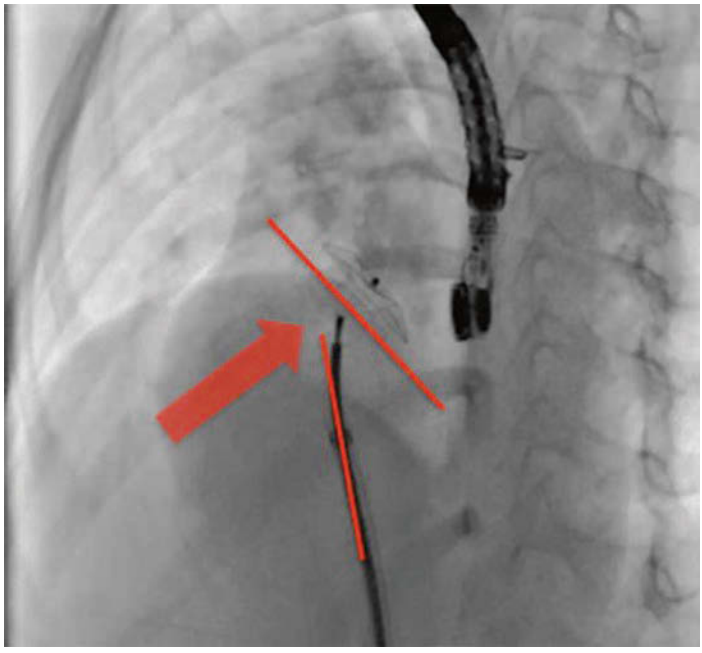

Fig. 30.7 Fluoroscopy: the angle between the septum and the delivery cable (*lines*)

usual approach from the inferior vena cava means that the Amplatzer septal occluder approaches the atrial septum at an angle (Fig. 30.7). In larger defects, it can be difficult to prevent the antero-superior rim of the device from pulling through from the LA to the RA before the core of the device can be developed. There are techniques to try and address this (discussed below), and there are devices, e.g. the Occlutech occluder (Fig. 30.8), which have a delivery cable which makes the angle between the device and the septum more favourable and delivery a little easier.

4. Technical modifications to close difficult defects: There are a number of tricks used by experience operators to place large occluders in challenging holes, generally these apply to the deployment of Amplatzer or "similar" types of occluder:

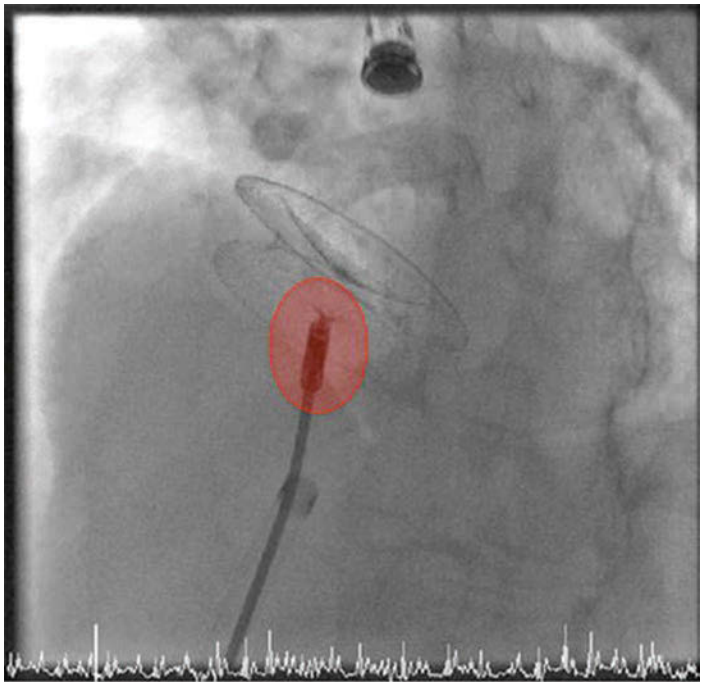

Fig. 30.8 Occlutech occluder. The flexible connection between the delivery cable and the device is *circled*

(a) Deployment manoeuvres: In some instances, a difficult ASD can be closed by changing the orientation of the left atrial disc of the device within the left atrium or altering the deployment sequence. Most operators will initially try to deliver the central core of the device slightly within the left atrium before bringing the device back towards septum in order for the stented portion of the device to offer support and prevent prolapse. If this fails, the left atrial disc of the device can be rotated towards the roof of the left atrium in an attempt to alter the delivery angle and give the device a chance to "sit" properly in the septum before prolapse through to the right atrium occurs. Another common technique is to deliver the left atrial disc within

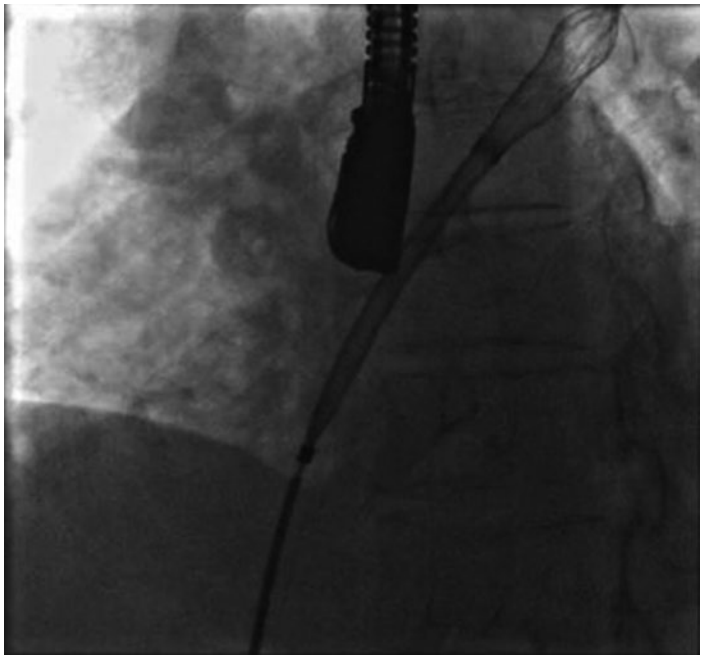

Fig. 30.9 Delivery of the left disc within the left upper pulmonary vein

the left or right upper lobe pulmonary vein in order to create tension on the system and allow the right atrial disc to fully appose to the septum whilst the left atrial disc remains within the pulmonary vein in an oval configuration (Fig. 30.9). With gradual removal of the sheath and increasing torque on the system, the left atrial disc will then pull back from the vein and engage the septum.

(b) Modifications to the device delivery sheath: These include the commercially available Hausdorf sheath modification (essentially a Cook Check-Flo sheath with a 3D curve to direct the device in a posterior direction) (Fig. 30.10), the steerable St Jude Agilis sheath (only available up to 8 F internal diameter and therefore limited in terms of the sizes of device it will accommodate) and the "home-made"

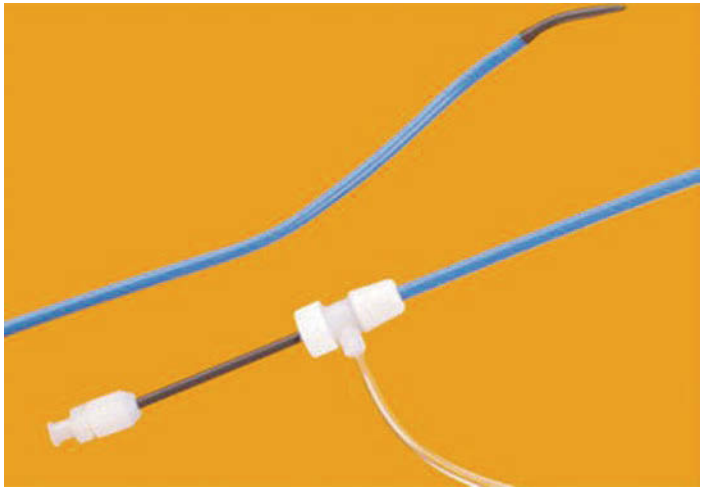

Fig. 30.10 Hausdorf delivery sheath. Note the "three-dimensional" curve which alters the approach of the device to the septum

creation of a bevelled edge by cutting the end of the standard sheath to allow the device to exit and reform at an altered angle.

(c) Balloon-assisted closure: A useful technique to assist in the closure of difficult ASDs is the use of a balloon to support the device during deployment (Fig. 30.11). An additional femoral venous access point is required, and through this, the balloon is positioned across the atrial septum. The left atrial disc of the device is then deployed within the left atrium and withdrawn onto the balloon which acts as a support for the device before the right atrial disc is extruded and the balloon deflated and removed.

(d) Other techniques: The use of a small snare threaded over the delivery cable to hold onto the device screw such that the main guidewire can be released, thereby removing the tension from the system and allowing reorientation of the device relative to the septum whilst still allowing retrieval is potentially helpful in very difficult ASDs (Fig. 30.12a, b).

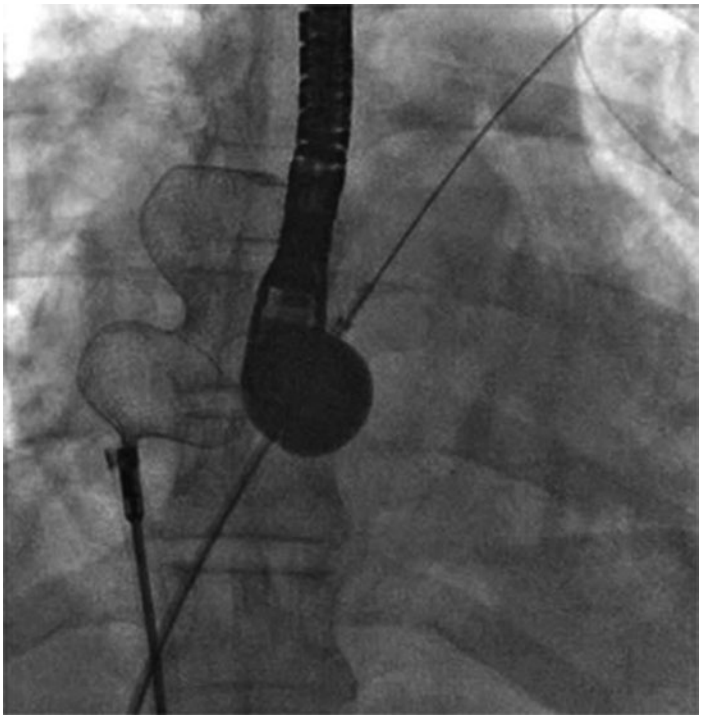

Fig. 30.11 Balloon-assisted technique

5. Multiple defects: Multiple ASDs vary from distinct defects within a relatively firm atrial septum to multi-fenestrated holes within a mobile structure. Sometimes multiple defects can be tackled with a single occluder, assuming that the edge of the defects is less than a few millimetre away from each other and that one of the defects is sufficiently small. Alternatively, if the tissue separating the defects is thin, then it can be possible to place an oversized occluder in the hope that this will break the tissue strand and effectively turn the defect into one large hole. If the defects are >5 mm apart, then usually two occluders are required. In this situation, both defects should be sized simultaneously with separate balloons and an occluder placed into the smaller hole first and then overlapped by the larger device. The smaller device is released first.

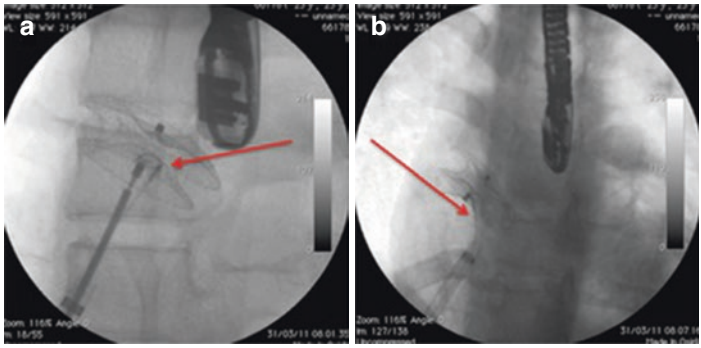

Fig. 30.12 (**a**, **b**) Snare (*arrowed*) around the screw of an ASO before (**a**) and after (**b**) release from the cable. (Images courtesy of Dr Gianfranco Butera)

30.11.1 Using Non-self-Centring Devices

Multi-fenestrated defects can be closed using a non-self-centring device (without a self-centring core). The key to this technique is to ensure that the catheter/wire is across the central defect, so that there is a uniform coverage across the rest of the atrial septum. It is important to check the rest of the atrial septum carefully once the device is positioned as sometimes occlusion of the major shunt "reveals" another defect that may require the insertion of an additional device.

The large degree of coverage relative to the small core means that these devices will easily cover multiple defects. They can also be deployed into smaller isolated defects and allow the operator to take advantage of devices with a low profile, e.g. the Gore septal occluder, which can be desirable, particularly in smaller children. In this situation, care must be taken to size the defect carefully and to "oversize" the device relative to the hole; e.g. for the Gore septal occluder, the current recommendation is to implant a device twice the diameter of the defect.

30.12 Pitfalls

30.12.1 Patient Selection

Pre-existing left ventricular dysfunction can be masked by the presence of an atrial septal defect. In this situation, closure of an ASD may precipitate pulmonary oedema. In elderly patients, particularly those with pre-existing coronary artery disease or systemic hypertension, a thorough clinical and echocardiographic assessment of left ventricular function should be made prior to device closure. Unfortunately, there are no large systematic studies to establish the margins of safety, but the available data suggests that a high LA pressure prior to occlusion of an ASD may be indicative of a latent left ventricular problem and that a significant rise in LA pressure with test balloon occlusion may predict problems after device closure. In patients with unfavourable haemodynamics or clinical/echocardiographic abnormalities, it is important to optimise treatment of co-morbidity, e.g. treatment of hypertension or coronary artery disease prior to considering ASD closure.

30.12.2 Younger Children

In small children, the capacity of the heart to accommodate an occluder is limited. Although there are published data demonstrating that ASDs can be closed percutaneously in children as small as 4 kg, the literature regarding the very young age range remains limited. In the early days of percutaneous ASD closure using the Amplatzer septal occluder (ASO), there was a "rule of thumb" that a device with a central core of no more than 1 mm per kg of body weight should be used in smaller children. Currently, many operators will routinely and successfully use larger devices that significantly break this rule, and the "safe" limits of percutaneous ASD closure in the very young and small remain unclear. The risk of electrical block is undoubtedly higher in smaller hearts, and retrieval of embolised devices in smaller patients can be a chal-

lenge; these issues should be borne in mind and discussed with parents prior to a procedure.

Although there will always be debate about the relative merits of surgical and transcatheter ASD closure in some patient groups between operators, those with limited experience are wise to remember that surgical closure of secundum ASDs remains safe and should not be discounted in this group as a reasonable alternative to a device.

30.13 Complications

Major procedure-related complications such as death and stroke are exceptionally rare. Device embolisation occurs in up to 1% of cases depending on the series. Acute cardiac perforation, usually as a result of catheter, wire or sheath damage to the free wall of the left atrium, is rare but can cause an important pericardial effusion or a laceration that may require surgery. Varying degrees of heart block can occur, particularly when relatively large devices are placed in smaller children. Paroxysmal atrial fibrillation occurs after device insertion in a proportion of older patients but is usually transient and usually responds to medical treatment or cardioversion. Migraine headaches can be transiently exacerbated by ASD closure in some patients.

Late cardiac erosion is feared, not only because it is potentially life threatening but because it is now apparent that it can occur many years after device placement. Patients should be informed about this rare complication prior to the procedure. Most reports relate to Amplatzer-type occluders but erosions are not limited to this occluder and have occurred with other devices. Perforations most commonly occur at the roof of the left atrium or between the aorta and right atrium (Fig. 30.13). Patients generally present with chest pain, dizziness and a pericardial effusion and require emergency cardiac surgery to explant the device and repair the perforation. Between 200,000 and 250,000, Amplatzer ASD devices have been shipped worldwide to date. On average, the US FDA has had reports of erosions in approximately ten cases per year. Risk factors for erosion are unclear, but speculation has centred on this

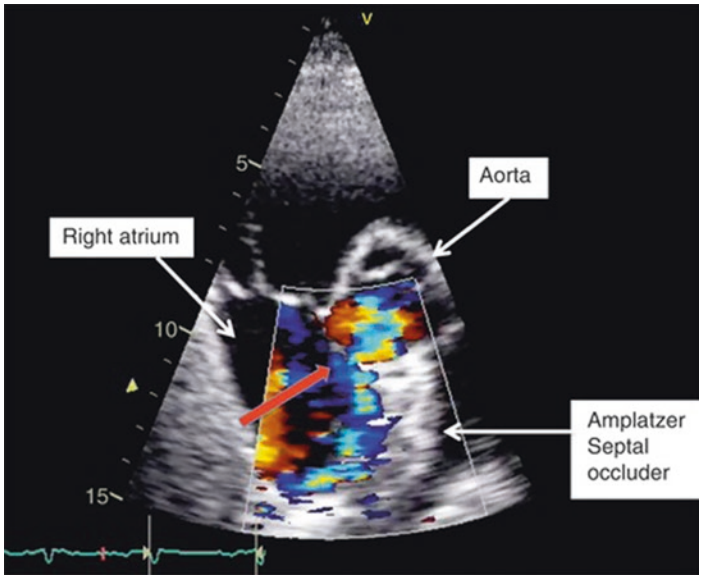

Fig. 30.13 Device-related perforation into the aorta (*arrowed*)

being more common with relatively oversized devices and in patients with absent retro-aortic rims.

30.13.1 Managing Complications

Embolised devices: It is inevitable that if an operator implants enough devices, then at some point he or she will have to deal with an embolised occluder. Those performing this procedure MUST be trained not only to retrieve a device but just as importantly to recognise when transcatheter retrieval is unsafe and emergency surgery required.

The degree of difficulty in device retrieval varies with device design. An embolised device within the aorta or the pulmonary artery can usually be safely retrieved via a catheter technique. Similarly an occluder free within the left or right atrium but away from AV valve or chordal tissue can be secured and retrieved. In

most cases, a device that is in contact with valvar tissue or chordal structures should be removed by a surgeon to avoid causing damage by dragging an occluder back into a sheath.

The most important tip when retrieving a device is to use a retrieval sheath that is sufficiently large. Unless there is a very good reason otherwise, I would currently use (at least) a 12-F Cook Flexor sheath, if necessary pre-closing an arterial access point prior to insertion with a Perclose suture. The armoured nature of this sheath means it is very difficult to buckle when applying traction.

The patient should be fully heparinised (100 IU/kg) and a catheter passed beyond the lost occluder before a wire is positioned and the long sheath advanced. A goose-neck snare is then used to snare the delivery screw. Considerable patience can be required to do this, and in some situations, e.g. when the device is in the aorta, a pigtail catheter can be required to turn the face of the device so that the screw is facing the snare. Once captured, the device is brought into the sheath. Commonly, the screw will hit the sheath sideways which can cause buckling. By careful manipulation and patience, the device will eventually come into the sheath.

30.14 Post-procedural Care

Standard care of the venous access site.

Post-procedural transthoracic echocardiography and ECG prior to discharge, focusing on device position, competence of AV valves and absence of pericardial effusion.

Six months of aspirin therapy.

30.15 Follow-Up

All patients: Review at 3/12 months.

30.15.1 Then.....

Children: Yearly follow-up until 3 years post device, then follow-up every 2 years whilst still growing.

Adults: Yearly follow-up for 3 years, then currently 3 yearly follow-ups. Currently there is debate about the need for review year a after implant but there is no doubt that although exceptionally rare late erosion does occur.

Further Reading

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Nassif M, Abdelghani M, Bouma BJ, et al. Historical developments of atrial septal defect closure devices: what we learn from the past. Exp Rev Med Devices. 2016;13(6):555–68.

Roos-Hesselink JW, Meijboom FJ, Spitaels SEC, et al. Excellent survival and low incidence of arrhythmias, stroke and heart failure long-term after surgical ASD closure at young age (a prospective follow-up study of 21–33 years). Eur Heart J. 2003;24:190–7.

31 Step-by-Step Device Closure of Persistent Foramen Ovale (PFO)

Michael Rahbek Schmidt
and Lars Søndergaard

31.1 Introduction

Persistent foramen ovale (PFO) is present in 20–30% of the background population, which indicates that the condition carries a relatively low risk for complications. Although a PFO provides the physiologic basis for a paradoxical embolism and thus is an intuitively likely prerequisite for so-called cryptogenic stroke, the causal connection has proved difficult to establish. Indeed, the presence of PFO in itself does not increase the risk for ischemic stroke in a general population [1], but in patients who already had a cryptogenic stroke, it increases risk for recurrent stroke.

Several clinical trials failed to prove a beneficial effect of device closure of PFO in this patient group, but with the publication of three large-scale clinical trials with a combined enrolment of more than 2300 patients younger than 60 years with cryptogenic stroke, the evidence for risk reduction by PFO closure become compelling [2–4]. This has only one changed guideline,

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but also the terminology to 'PFO-associated stroke'. However, the recurrent stroke rate in patients with PFO-associated stroke is low, at around 1–4% per year. It is, therefore, crucial that PFO closure can be undertaken safely, with a low risk of major complications and a high procedural efficacy. Importantly, although PFO closure reduces risk for recurrent stroke by approximately 50%, it does not fully eliminate the risk even in young patients with no known risk factors, possibly because of residual leaking after device closure or other risk factors.

As with ASD closures, wide availability of suitable devices for PFO closure began with launch of the Amplatzer PFO occluder. The majority of devices on the market, though constructed with slightly varying materials and engineering, are iterations of the nitinol-based non-self-centring Amplatzer device with a double umbrella connected by a central thin stalk (Fig. 31.1). An excep-

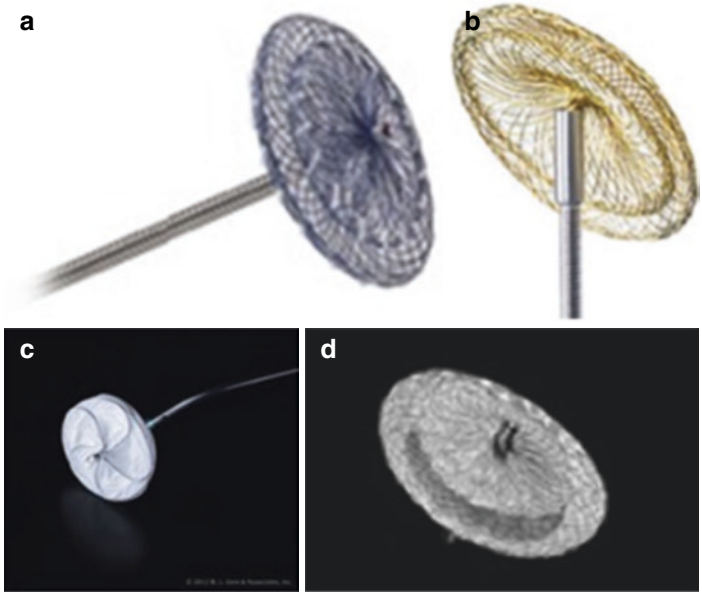

Fig. 31.1 PFO occluder devices. (**a**) Amplatzer PFO occluder, (**b**) Lifetech CeraFlex, (**c**) Gore Septal Occluder, (**d**) Cocoon PFO occluder

tion is the recently launched Noble Stitch® system, which leaves no device in the heart but closes the PFO channel with a double suture.

31.2 Anatomic Description

The foramen ovale is a foetal interatrial communication allowing the saturated blood from the umbilical veins to bypass the lungs and stream directly into, mainly, the upper body of the systemic circulation. In approximately ¼ of the population, foramen ovale remains patent due to failure of the primum and secundum septa to fuse (Fig. 31.2). The PFO is invariably located along the anterosuperior border of the septum primum next to the aortic root, but the width and length of the PFO channel are highly variable. Furthermore, some PFOs are anatomically more complex due to an atrial septal aneurysm (often defined as movement of aneurysmatic part of the septum primum of more than 10–15 mm during the cardiac cycle) or are held open by a fold in the atrial septum allowing for a continuous left-to-right as in ASDs [5]. Other ana-

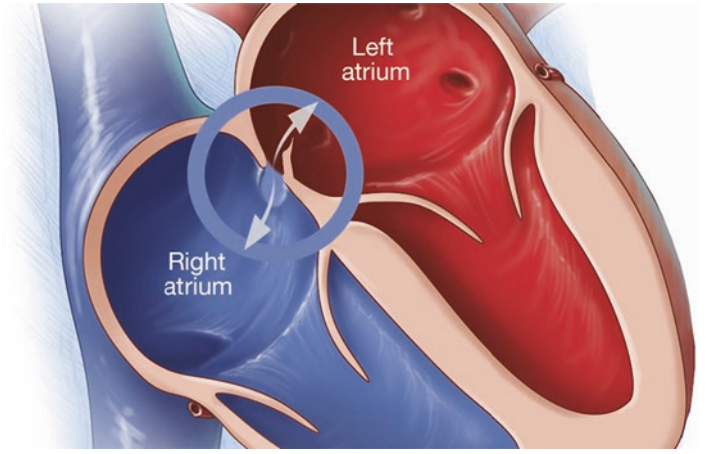

Fig. 31.2 PFO anatomy, on curtesy of Alma R. Stecher

tomic variations, which may complicate closure, include a Eustachian valve and Chiari network (see 'Pitfalls' below).

31.3 Physiology

Under normal physiological conditions, the slit-like opening (so-called 'flap valve') of the PFO is closed due to the higher blood pressure on the left atrial side. Thus, the PFO does not alter the haemodynamic condition of the normal heart, as there is no left-to-right shunt (Fig. 31.3). However, transient increases in right atrial (RA) blood pressure above the left atrial (LA) blood pressure (Fig. 31.3)—as during Valsalva manoeuvre or abdominal compression – leading to opening of the PFO, and right-to-left shunt may have dramatic consequences, as it allows systemic venous thromboembolisms to pass to the systemic arterial side. Certain conditions, such as acute pulmonary embolism, raises the RA blood pressure above the LA blood pressure for a longer time and, therefore, entails an increased risk of paradoxical embolism/stroke [6].

31.4 Clinical Implications of a PFO and Indications for Closure

As stated above, a PFO remains a clinically silent anatomical feature in the vast majority of the population. In the minority who experience clinical consequences, these fall into four categories.

31.4.1 PFO-Associated Stroke and Systemic Arterial Embolisation

PFO-associated stroke is the most prevalent clinical condition related to the presence of a PFO. For adult patients below 60 years with no or few competing risk factors, paradoxical embolisms through a PFO are believed to be the most frequent cause of stroke [8]. As these patients are often young, active, working and with

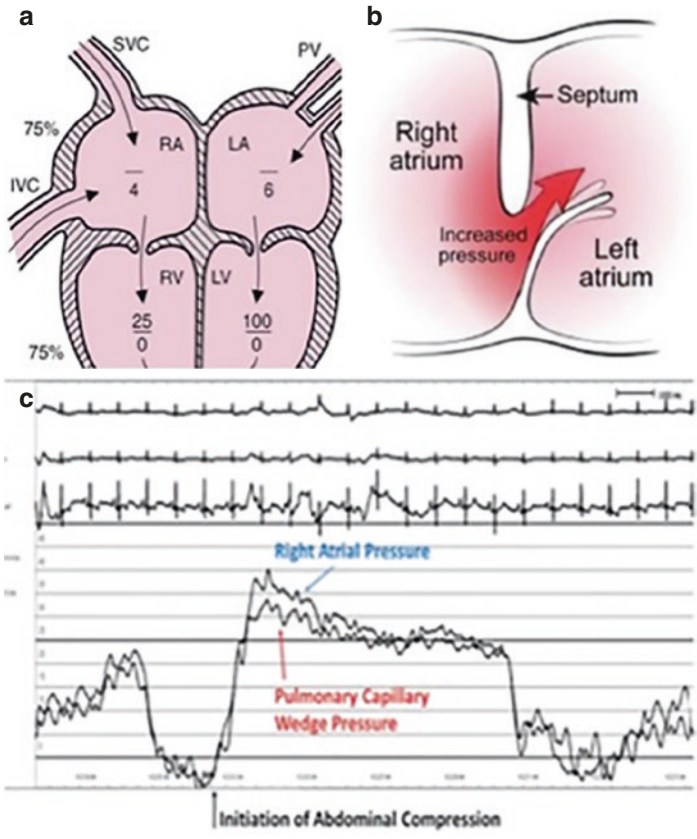

Fig. 31.3 Physiology of the PFO. (**a**) Pressures in the heart chambers during normal physiological conditions, (**b**) opening of the PFO during increased right atrial pressure, (**c**) changes in left and right atrial pressure during abdominal compression—from Beigel et al. [7] with permission

children, the consequences of a stroke are devastating. While the vast majority of clinical manifestations of paradoxical embolisms are strokes or TIAs, emboli may also travel to limbs, organs or coronary arteries [9].

As mentioned in the introduction, there is now substantial evidence supporting the beneficial effect of PFO closure in adult

patients below 60 years of age who already suffered an embolic stroke without other plausible explanations. This group of patients will most likely continue to comprise the vast majority of cases referred for PFO closure.

Importantly, other potential causes of embolic stroke should be thoroughly excluded. In particular, at least 48 h Holter monitoring (preferably longer) to exclude atrial fibrillation, carotid vascular ultrasound to exclude atherosclerotic causes and blood test to exclude coagulation disorders.

Furthermore, the likelihood of the PFO as the cause of stroke has been suggested assessed using the RoPE score calculator (Table 31.1).

While some types of anatomy may pose higher risk of paradoxical embolism (e.g. atrial septal aneurysm, larger defects, etc.), no types of PFO have been deemed risk-free. Therefore, any diagnosis of PFO in patients fulfilling the above criteria should lead to referral for PFO closure.

Indications for PFO closure after suspected paradoxical extracerebral embolization should be determined on a case-by-case basis. There is currently no clinical evidence proving the benefit of PFO closure in these cases, but clinical circumstances may support device closure.

Table 31.1 RoPE calculator. Maximum score 10, minimum score 0. A high score indicates high likelihood of PFO-related stroke

RoPE score calculator	
Characteristic	*Points*
No history of hypertension	1
No history of diabetes	1
No history of stroke or TIA	1
Non-smoker	1
Cortical infarct on imaging	1
Age	
18–29	5
30–39	4
40–49	3
50–59	2
60–69	1
>=70	0

31.4.2 Decompression Sickness

Diving sickness occurs when nitrogen accumulated in the body during scuba diving due to high ambient pressure is released as bubbles into the venous blood stream during the ascent to lower pressure. If the ascent is slow, the nitrogen bubbles are normally filtered in the lungs and cause no harm, but rapid ascent may overload the pulmonary filter and let the nitrogen bubbles pass through to the systemic arterial side causing tissue trauma and vascular occlusion, which frequently leads to neurological symptoms. If a PFO (or another potential right-to-left shunt) is present, bubble passage into the systemic arterial circulation may happen even during a slow ascent (Fig. 31.4). Accordingly, PFOs have been shown to be overrepresented among divers suffering from decompression illness [10].

Professional divers who experience neurological symptoms repeatedly during ascent to lower surround pressure despite strict adherence to decompression guidelines and with an identified PFO are often offered device closure. Though no randomised

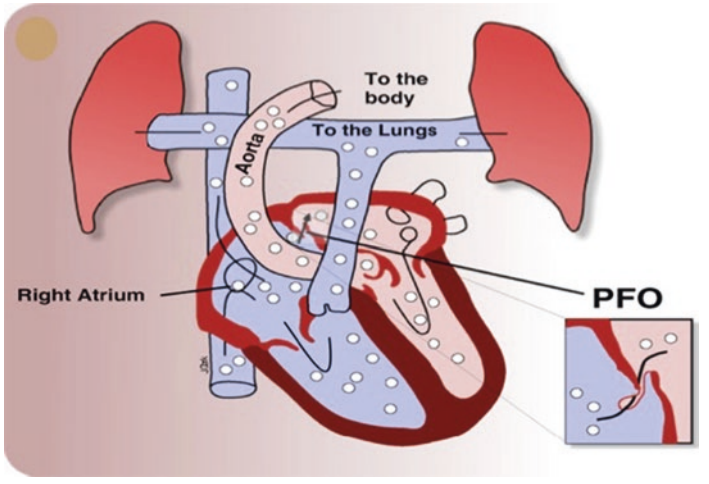

Fig. 31.4 Air embolism in diving sickness. Physiology of paradoxical air embolism in diving sickness—from Sykes and Clark [11]—with permission

clinical trial has been conducted, a significant amount of circumstantial and indirect evidence seems to support that device closure reduces recurrence of symptoms [10].

31.4.2.1 Migraine with Aura

The association between the presence of a PFO and migraine remains controversial. While several studies have established a disproportionally high prevalence of PFO among patients who suffer from migraine with aura [12], the only randomised clinical trial on the effect of PFO closure on migraine (MisT) [13] failed to show a beneficial effect of PFO closure. The MisT trial has been criticised for its design because the primary end point—complete cessation of headaches—may have been too ambitious. Indeed, a post hoc analysis indicated a reduction in the number of days with headache in the patients who underwent PFO closure. Similarly, it is frequently reported by patients who had their PFO closed on another indication (usually stroke) that pre-existing migraine symptoms decrease in frequency and severity [14], although some patients in contrast experience novel headache symptoms that were not present prior to PFO closure.

The evidence supporting PFO closure in patients suffering from migraine with aura is currently not considered sufficient for a generalised recommendation of PFO closure. While some interventions are performed on a compassionate individual basis, the indication is unlikely to enter the guidelines until more substantial clinical evidence is present.

31.4.3 Platypnea-Orthodeoxia

This rare syndrome is believed to occur due to either anatomic changes that produce a baffle that causes streaming through a PFO or to posture-dependent right-to-left pressure gradients. Platypnea-orthodeoxia is characterized by dyspnoea and hypoxaemia in the standing or sitting position, which disappears when lying down. Indication for closure is decided on a

patient-to-patient basis and requires thorough workup to exclude pulmonary hypertension/right heart failure, where the PFO may serve as right heart decompression, and closure may lead to clinical worsening.

31.5 Treatment and Device Options

While initial studies indicated similar efficacy of PFO closure and medical therapy in preventing recurrent strokes in patients with a PFO [15], the recent large-scale trials [2–4] have definitively established superiority of device closure.

There is currently no evidence supporting superiority of one type of device to another regarding complications or efficacy. However, operator preferences and PFO anatomy may infer specific devices, as the different devices demand slightly different implantation techniques, and the devices show somewhat different behaviour after implantation due to differences in stiffness and materials.

The suture-based Noble Stitch® system may be preferable in specific cases, but large-scale long-term data on efficacy and complication rates are still lacking.

31.6 Pre-Procedural Imaging and Diagnosis of a PFO

If the indication for PFO closure is embolic stroke, cerebral infarction should be confirmed my MRI. Similarly, for other indications, evidence of clinical significance of the PFO is mandated.

The PFO itself should be visualised by both transthoracic echo with bubble contrast with and without Valsalva manoeuvre and with transoesophageal echo, which should include detailed imaging of the atrial septum to exclude additional atrial septal defects (Fig. 31.5). It is important to visualise flow through the PFO channel, either by colour Doppler or bubble contrast.

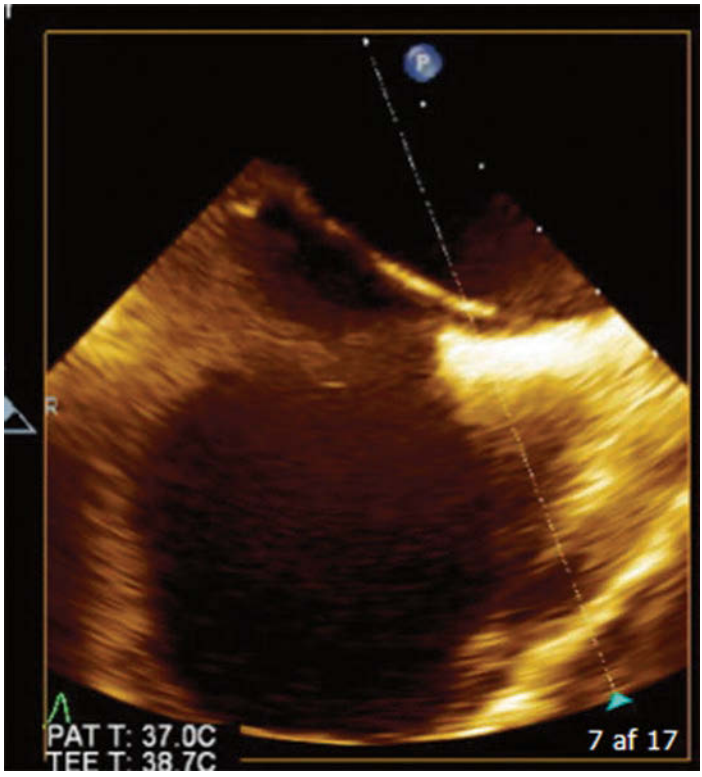

Fig. 31.5 TEE imaging of the atrial septum and the PFO

31.7 Technique Step-by-Step

The following steps describe the procedure used for deployment of double-disc devices.

1. Pre-procedure
   (a) Ensure appropriate patient selection and indication.
   (b) Ensure adequate pre-procedural imaging.
   (c) Have non-PFO-related causes been excluded.

2. Imaging at the time of the procedure (TOE or ICE).
   (a) Establish anatomy of PFO.
   (b) Exclude additional defects and anomalous pulmonary venous drainage.
3. Catheterisation and device deployment.
   (a) Initial placement of a 6F venous catheter in the femoral vein using the Seldinger technique either using palpation/landmarks or ultrasound guidance.
   (b) If ICE is used, insert an additional 6F venous sheath, which is subsequently upsized to a 10F long sheath (e.g. Arrow 30 cm). If the ICE imaging is performed by another operator, contralateral sheath position (left femoral vein) for ICE is usually most convenient.
   (c) Systemic heparinising (100 IU/kg).
   (d) The PFO is crossed using a diagnostic multipurpose catheter and a straight or J-tipped hydrophilic guidewire (e.g. Terumo).
   (e) Advance the catheter over the guidewire into the left upper pulmonary vein.
   (f) Exchange the wire for an exchange-length Amplatzer Super or Extra Stiff guidewire.
   (g) Remove sheath and catheter and introduce a sizing balloon (e.g. Amplatzer 18 mm) sheathless (or alternatively through an appropriately upsized sheath) into the PFO channel. Gently inflate (1:4 contrast to saline) when in position to interrogate size and shape of the PFO tunnel (Fig. 31.6). Be careful not to overinflate the balloon, which will transform the PFO tunnel into a circular defect.
   (h) Adjust fluoroscopy angle to avoid foreshortening and measure width and length of the PFO using markers on the balloon to calibrate distance measurement. A tunnel length of more than 15 mm bodes suboptimal device deployment and is a relative contraindication for device closure.
   (i) Choose the proper closure device. As a general rule, choose the smallest device with a size more than twice the width of the defect.

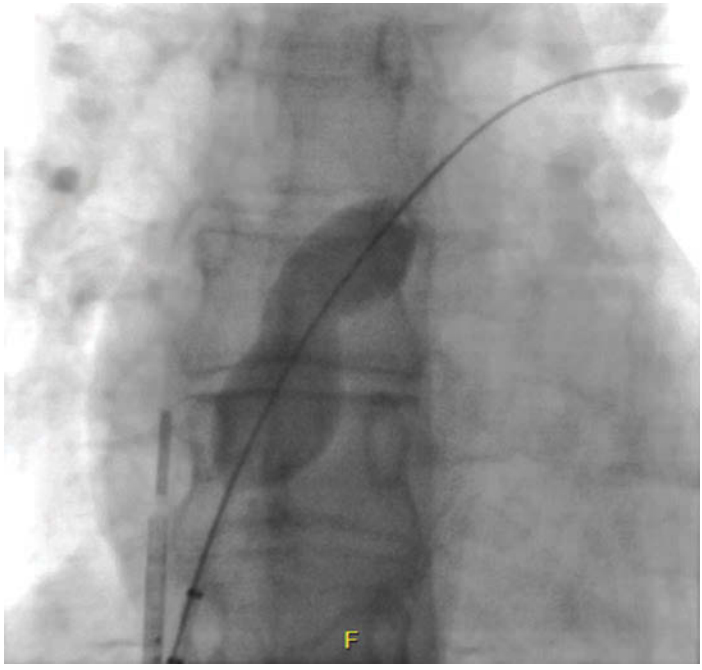

Fig. 31.6 Balloon sizing of PFO. Sizing balloon gently inflated in the PFO. Markers on the sizing balloon are used to calibrate the measurement

(j) Ensure guidewire is still in the correct position (Fig. 31.7a).
(k) Load the PFO closure device into the delivery system, which is flushed and de-aired with heparinised saline.
(l) Advance the saline-flushed delivery sheath into the left atrium and remove the guidewire (Fig. 31.7b).
(m) It is important to avoid air entry at the following stages. Remove the dilator from the delivery sheath while keeping it below the level of the heart. Thorough flushing and fluid-to-fluid techniques should be observed.
(n) While flushing the loading system, connect it to the sheath and advance the device into the delivery sheath.

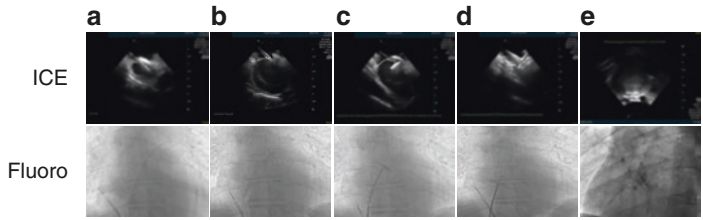

Fig. 31.7 Stepwise deployment of PFO device. (**a**) Wire through the PFO and positioned in left upper pulmonary vein. (**b**) Sheath through PFO. (**c**) Left atrial disc unfolded. (**d**) Left and right disc unfolded. (**e**) Device released

(o) Unfold left atrial disc and pull taut the atrial septum while observing the position on ICE or TOE (Fig. 31.7c).
(p) Develop the right atrial disc and push towards the septum (Fig. 31.7d).
(q) Gently test device position using push-pull.
(r) Confirm correct deployment with TOE/ICE.
(s) Unscrew/unlock the device from the delivery cable to release the device.
(t) Confirm correct deployment with TOE/ICE (Fig. 31.7e).
(u) Remove ICE and sheaths and ensure haemostasis by compression, perclose systems (e.g. ProGlide) or a 'figure-of-eight' suture.

31.8 Materials Required

1. Sheaths/catheters/wires.

(a) Standard short access sheaths up to 10F.
(b) 5F or 6F multipurpose catheters
(c) 0.035″ straight or J-tipped and Terumo wires
(d) 0.035″ Stiff wires in exchange length (>250 cm)
(e) Delivery sheaths for the available devices.
(f) Rescue sheaths and gooseneck snares for the rare complication of device embolization or need for device retrieval (see ASD chapter).

2. Devices: A minimum of two sizes of PFO closure device are recommended.
3. Intracardiac or transoesophageal echocardiography equipment.

31.9 Tips and Tricks

In most cases, the device implantation is:

1. When using ICE, place ICE in position in the RA and visualise the PFO prior to wire passing.
2. When wire passage of the PFO is difficult, try 'hooking' the defect with a standard diagnostic right coronary or internal mammary catheter.
3. Go for wire positioning in the LUPV—when difficult, enter the left atrium with the catheter, turn the counter clockwise and pull back while probing with the wire.
4. Refrain from unfolding left disc in the pulmonary vein.
5. Additional Defects: Small defects/holes in the PFO tunnel are often closed after device deployment and will not need selective closure. More distant or larger defects need separate closure—please see ASD chapter.

31.10 Pitfalls

Though procedural difficulties and complications are rare, these are important to know and handle. While the PFO itself shows little deviation in anatomy, the surround structure may vary significantly and confuse the operator. Broadly, three types of anatomical features may complicate the usually simple defect visualisation and device deployment.

1. Additional Atrial Septal Defects: Larger ASDs are usually diagnosed during workup, but smaller holes may be missed. Aneurysm atrial septum is not rarely associated with multiple defects. In particular, when these are present near or in the

PFO channel, the guidewire may cross the hole rather than pass through the channel, which will lead to incorrect device deployment and frequently incomplete closure of the PFO. In contrast, proper deployment of the PFO closure device, frequently also closes PFO-near defects as the discs are overlapping and covering.

2. Left Atrial Structures: Left atrial strings or septae (as in incomplete cor triatriatum) catch the left atrial disc during deployment leading to incorrect positioning of the device, and potentially, left heart embolization in the operator is not aware.
3. Right Atrial Structures: A large Eustachian valve or prominent Chiari network causes difficulties in the initial passing of the PFO because the catheter or wire has prevented its usual course towards the septum. During deployment, the right atrial disc may become entangled in strands from a Chiari network causing the disc to unfold incompletely or in distance to the PFO causing incomplete closure.

For all the above, thorough visualisation by ultrasound (ICE or TOE) and fluoroscopy is the key to avoid complications. For operators who rely on solely one imaging modality (TOE or fluoroscopy), any irregularity during deployment (unusual wire behaviour, distance between discs, etc.) should prompt additional imaging.

The riskiest pitfall in PFO closure is, however, not related to the procedure itself but rather to inadequate workup prior to the procedure. Overlooking paroxysmal atrial fibrillation or significant cerebrovascular atherosclerosis may have devastating consequences for the patient.

31.11 Post-Procedural Care

Post-procedural transthoracic echocardiography is encouraged to confirm correct device position and exclude pericardial effusion. Usually, the patient can be discharged the same or following day.

Post-procedural antiplatelet therapy is mandated for at least 6 months and for patients with a previous stroke and, importantly,

was prescribed life-long in the trials, which provided evidence for device closure in PFO-associated stroke.

Device closure of PFO may cause new-onset atrial fibrillation in the early post-procedural period. Since this implies a risk of stroke, appropriate monitoring for atrial fibrillation must be performed, and new arrhythmia addressed with oral anti-coagulation and/or conversion to sinus rhythm.

31.12 Follow-Up

A 6–12 months follow-up including thorough history (in particular neurological symptoms or palpitations) and a bubble contrast transthoracic echocardiography is mandated. History of arrhythmia symptoms should be further investigated by 2- or 7-day Holter monitoring. Neurological symptoms should be assessed by a neurologist. For patients who suffered from migraine prior to the PFO closure, it is common to experience changes in or cessation of migraine symptoms.

Significant residual shunting on bubble contrast echo (often described as more than 25 bubbles in the left atrium in a still frame), TOE, should be performed to visualise device position and exclude additional defects.

31.13 Further Reading and Future Developments

Although the current evidence for PFO closure is compelling, some questions remain [16]. The indications for device closure of a PFO are currently strict and limited to a very specific population of patients of 18–60 years of age with cryptogenic stroke and no competing potential causes. However, expansion of indication to older patient groups may be justified although evidence is currently lacking [17]. Similarly, conditions with elevated right atrial pressure, such as pulmonary embolism, may also become an indication for PFO closure [6, 18].

While the currently available device types are easy to deploy and show an acceptable efficacy, novel devices with more efficient closure of the PFO channel may be developed. Bioabsorbable devices are currently being developed for ASD closure and may also be relevant for PFO closure.

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Fontan Fenestration Closure

32

Derize E. Boshoff and Marc H. Gewillig

32.1 Introduction

Since Fontan and Baudet first described their technique for univentricular repair in 1971, numerous modifications have been described [1]. The technical approach to the Fontan operation itself has evolved into two major approaches: the lateral tunnel technique and the extracardiac technique. The risk of death is greatest in the immediate postoperative period, often in the setting of a low cardiac output state. Some of the complications that contribute to the early mortality may be transient or reversible, i.e. elevated pulmonary vascular resistance and ventricular dysfunction, or treatable in the case of residual distal pulmonary artery distortion. The concept of a fenestration between the systemic venous and the pulmonary venous pathways was introduced in 1971 [2], when the first patient of the atriopulmonary connections

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series received a 6-mm fenestration to serve as a "pop-off" valve, allowing decompression of the systemic venous pathway into the left atrium. Right-to-left shunt at the atrial level tends to improve cardiac output at the expense of systemic oxygen desaturation. Fenestration has been shown to improve early outcomes, including a decreased duration and quantity of chest tube drainage, a shorter duration of mechanical ventilation and a shorter postoperative hospitalization [3].

32.2 Timing of Closure

Spontaneous fenestration closure is well recognized, but persistent patency may warrant closure to improve arterial oxygen saturations and prevent cerebrovascular accidents due to paradoxical thromboembolism. Whether and when to intentionally close a fenestration remains debatable: depending on institutional protocol, management may vary from active fenestration closure at predetermined intervals to a "hands-off" approach, allowing the fenestration to follow its "natural history". In our institution, the total cavopulmonary connection (TCPC) by interposition of a 16–20-mm extracardiac Gore-Tex graft (WL Gore and Associates, Flagstaff, AZ) is currently the operation of choice, and a fenestration is created virtually in all patients. Despite all controversies surrounding routine fenestration closure, it seems reasonable to close fenestrations in patients with favourable haemodynamic assessment and clinically significant desaturation, based on the secondary effects of cyanosis.

Timing of fenestration closure remains debatable, but current recommendation is to postpone fenestration closure to at least 6 months after completion after the Fontan circulation if O_2 saturations are <90 % and test occlusion is tolerated.

32.3 Patient Selection

Patients with resting oxygen saturation (SaO_2) of less than 92 % or significant desaturation on exertion should be further assessed. Defining the "ideal" patient for fenestration closure is debatable,

but factors that can be considered more favourable are the following:

- Uncomplicated postoperative course after bidirectional Glenn shunt and Fontan procedure (absence of prolonged pleural effusions or chylothorax and discontinuation of diuretics within weeks after surgery)
- No clinical evidence of low cardiac output or systemic congestion
- Exclusion of a high-velocity shunt through the fenestration on echocardiography
- Unobstructed Fontan connections and low pulmonary vascular resistance (PVR)
- Good ventricular function and absence of significant valve regurgitation
- Unobstructed systemic outflow and pulmonary venous return
- Normal AV conduction on ECG and absence of significant arrhythmias

32.4 Evaluation Before Catheterization

Most information can be obtained by clinical assessment (including ECG and exercise testing, if appropriate) and transthoracic echocardiography (TTE). However, TTE may fail to detect some thrombi, therefore transoesophageal echocardiography (TEE) may be necessary in some patients. In case of suspected pulmonary venous obstruction or pulmonary artery distortion, computed tomography or magnetic resonance imaging should be performed prior to catheterisation. These imaging techniques can also give additional information about the presence of venovenous or aortopulmonary collaterals.

32.5 Catheterization Procedure

- Catheterization is performed with intubation and general anaesthesia in room air. Antibiotic prophylaxis and heparin (100 IU/kg IV, maximum 5,000 IU) should be administered routinely.
- After obtaining femoral venous and arterial access, a complete haemodynamic assessment should be performed, documenting saturations and pressures throughout the Fontan pathway and systemic circulation.
- Angiography should then be performed in the superior and inferior caval veins and pulmonary arteries to visualize the Fontan connections, surgical fenestration (Fig. 32.1), additional interatrial leaks and possible venous collaterals.

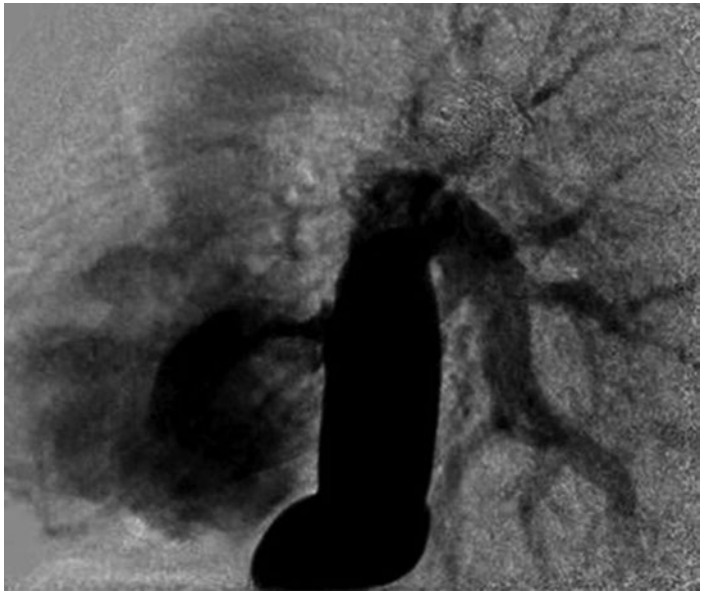

Fig. 32.1 Lateral view of contrast injection in inferior caval vein: a 20 mm conduit is mounted between the inferior caval vein and pulmonary artery; a 4.5 mm fenestration allows right-to-left shunt into the left atrium

- Selective injection in the innominate vein and right hepatic vein is indicated to exclude venovenous connections. An aortogram should be performed to exclude significant aortopulmonary collateral arteries.
- Anatomical abnormalities amendable to interventional treatment should be addressed first: balloon dilation and/or stenting of obstructed Fontan connections or stenosed/hypoplastic pulmonary arteries, occlusion of significant collaterals if appropriate and treatment of systemic obstruction (i.e. recoarctation) by balloon dilation and/or stenting.

32.6 Haemodynamic Assessment and Test Occlusion of Fenestration

- Identifying "favourable" haemodynamics for fenestration closure is ill defined [4].
- Measurement of PVR in Fontan circulation is fraught with difficulties due to inability in accounting for collateral circulation, possibility of pulmonary arteriovenous malformation, low cardiac output state, presence of systemic venous obstruction, unequal distribution of lung flow and possibility of pulmonary venous obstruction. All these factors multiply the error in accurate assessment of PVR.
- Test occlusion of the fenestration is used during catheterization to identify patients presumably unsuitable for fenestration closure, by quantifying changes in the systemic or mean venous pressure and systemic saturation. Whether temporary test occlusion in a sedated and intubated patient is a reliable surrogate for predicting physiology in the awake and spontaneously breathing Fontan patient is debatable, but is certainly recommended in case of unfavourable baseline haemodynamics and in high-risk patients.
  - Test occlusion can be performed using a 7-F balloon-tipped, multi-lumen catheter (Swan-Ganz catheter). The balloon catheter is passed over a wire into the systemic atrium; the balloon is inflated using 1 cc diluted contrast and pulled

back against the atrial wall/fenestration to allow for temporary occlusion (at least for 15 min).
- Alternatively, a small compliant balloon (typically a 6–8-mm Tyshak balloon (depending on fenestration size)) can be inflated within the fenestration itself, using the femoral sheath for pressure and saturation measurement.
- Complete occlusion should be confirmed by angiogram (through the proximal port of the balloon-tipped catheter or the femoral sheath) (Fig. 32.2).
- Measurements should be repeated, documenting VCI mean pressure and saturation and aortic pressure and saturation.
- Fenestration occlusion can probably be undertaken safely in patients with a systemic venous pressure of <18 mmHg dur-

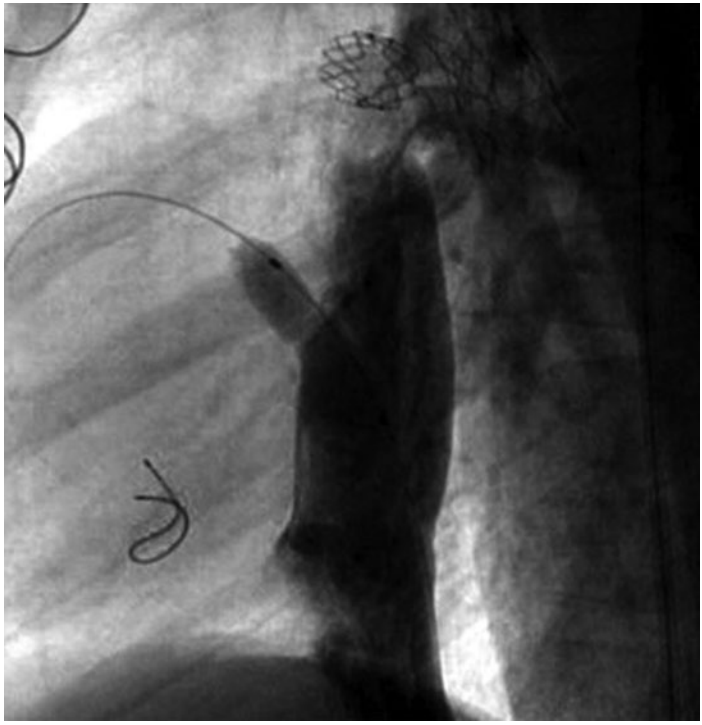

Fig. 32.2 Low-pressure balloon occlusion of 4.5 mm fenestration with a 6 mm Tyshak balloon; contrast injection through venous sheath confirms total occlusion

ing test occlusion or in the absence of a significant (>4 mmHg) increase in mean systemic venous pressure or reduction in mixed venous saturation of >10 %.
– We would strongly discourage fenestration closure in patients with systemic venous pressure of ≥20 mmHg.

32.7 Choice of Device

- When planning transcatheter closure, various factors should be considered, including the size and location of the fenestration, its geometry, the distance between the atrial chamber and the internal edge of the conduit and the possibility of placing a long sheath in the systemic atrium. Patient size and weight should also be taken into consideration.
- The ideal device must not only provide complete occlusion with reliable stability but also have a low profile without distorting the anatomy or obstructing flow within the Fontan conduit/baffle.
- Along with the diversity of methods to complete the Fontan operation, a number of techniques have been utilized to create fenestrations, depending on the type of Fontan operation (extracardiac conduit versus lateral intra-atrial tunnel) and institutional preference. For the lateral tunnel type of Fontan, a coronary punch is used to create a fenestration in the Gore-Tex baffle. In case of the extracardiac conduit, a fenestration can again be created by placing a coronary punch in the Gore-Tex conduit, after which the atriotomy resulting from detaching the inferior vena cava from the right atrium is sewn to the Gore-Tex graft as a circle of about 2.5 cm, with the fenestration in the centre of the circle. This prevents the adjacent atrial wall from impacting the size of the fenestration. In some centres, the fenestration is made using a short (5–7-mm) polytetrafluoroethylene (PTFE) shunt between the extracardiac conduit and the systemic atrium to decrease unexpected spontaneous closure. Kreutzer et al. described a novel method to create a fenestrated extracardiac Fontan conduit by means of a pericardial

tube anastomosed end to end with the inferior inlet of the right atrium [5].

- Fenestration size is usually between 3.5 and 5 mm, depending on the type of fenestration (punch hole vs. short PTFE shunt) and patient characteristics (risk stratification).
- In addition to intentional fenestrations, significant Fontan baffle leaks exist in up to 15 % of patients with a lateral tunnel-type Fontan. The baffle leaks are mostly located at the base of the right atrial appendage (RAA) at the suture line excluding the superior vena cava flow from the RAA. This suture line seems particularly susceptible to tiny leaks being left postoperatively due to the difficulty in tightly joining a smooth patch material to the corrugated surface created by the pectinate muscles. The increased venous pressure within the baffle can enlarge these channels creating a clinically significant shunt over time. While the origin of the leaks may be similar, the anatomy of the fistulous tract may vary.
- Due to the varying location, size and type of fenestrations, several catheterization methods have been described for fenestration closure by multiple authors, including Gianturco coils, detachable coils, clamshell devices, CardioSEAL devices, Amplatzer septal occluders, Amplatzer duct occluders, Amplatzer vascular plugs, Helex septal occluder, Angel Wings devices, Gianturco-Grifka vascular occlusion devices and CARDIA™ PFO star device.
- The placement of clips at the time of surgery to mark the location of the fenestration or to narrow the mid-portion of a tube graft for better anchoring of coils or devices has facilitated closure at the time of catheterization.
- Over the past two decades, the following devices have been used in our unit for closure of fenestrations and baffle leaks: Rashkind device, CardioSEAL, Amplatzer ASD occluder, Amplatzer VSD occluder and PFO star type device. In search of an ideal device, we modified a 15-mm PFO star (FFD15, CARDIA™, Burnsville, MN) by removal of the left disc to reduce thrombogenicity in the left atrium, increase the amount and length of the LA legs from 2 by 15 mm to 3 by 20 mm to prevent dislodgement and later adding a pivot between the left

and right umbrella [6]. We considered this device "ideal" because of its low profile, minimal fabric and metal, good closure rate and non-thrombogenicity. However, introducer sheaths are much larger than needed with the newer devices and although the loading mechanism has been simplified, there remains a learning curve. Currently, the Amplatzer duct occluder type II has become our device of choice for closing the typical punch-hole-type fenestration performed in our extracardiac Fontan conduits. This device has a high conformability and its dual articulating discs makes placement in the fenestration relatively easy. The fabric-free technology allows for delivery through a low-profile 4-F catheter while maintaining a high rate of occlusion without being bulky and potentially obstructive.

32.8 Crossing and Outlining the Fenestration/ Baffle Leak

- The position of the fenestration and/or baffle leak should be delineated using angiography in different views.
- TEE can give additional information in patients with a baffle leak, or in cases where the size of the atrial chamber is small or a residual atrial septum may be problematic.
- Depending on the location and shape of the fenestration, it can be crossed by the aid of various preshaped or custom heat-shaped catheters such as the right Judkins catheter and a floppy exchange wire [i.e. 0.035-in Terumo guide wire or Woolley Hi-torque Floppy wire (Mallinckrodt, St. Louis, MO)].
- Once the wire is advanced, it can be exchanged for a straight catheter if necessary, facilitating placement of stiffer wires.
- Depending on the method of test occlusion, a 7-F balloon wedge catheter is passed over an exchange wire, or a small compliant balloon (i.e. Tyshak balloon) is passed over the appropriate wire (depending on balloon). Test occlusion is performed, and if necessary, balloon sizing can be performed if the punch-hole size of the fenestration is not known or in case of a baffle leak or fistulous connection.

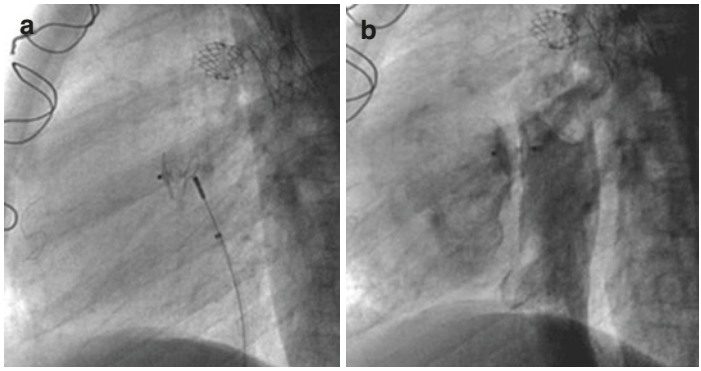

Fig. 32.3 (**a**) Deployment of Amplatzer duct occluder type II in fenestration, device still attached to delivery cable. (**b**) Cavogram after release of device: both disks are clearly at appropriate end of the fenestration; there is still some contrast like "smoke" through the device which will disappear within minutes after release

- After selecting the appropriate device, a long sheath with dilator or delivery system (depending on device) is passed across the defect over the exchange guide wire.
- The dilator and wire should be removed slowly, allowing for spontaneous backflow of blood through the sheath, followed by careful flushing to avoid air embolism.
- Loading and deployment of the device are performed in the usual way as described for the specific device. Prior and after release, the device position should be checked angiographically (Figs. 32.3a, b) and on TEE if necessary. Haemodynamic measurements and saturations should be repeated.

32.9 Alternatives to Device Closure

- Device closure necessitates introduction of a guide wire and a long sheath into the pulmonary atrium. Technical difficulties in closing fenestrations by different devices have been shown in TCPC patients with residual native atrial septum, forming an intermediate chamber on the pulmonary venous side of the

fenestration and additionally carrying the risk of systemic embolism in the case of difficult manipulations with wires and sheaths.

- In these situations the use of a covered Cheatham Platinum (CP) stent (NuMED, Hopkinton) could be a valuable option, at least in patients weighing more than 15 kg. A 12-F long sheath is advanced over a stiff guide wire across the TCPC conduit, positioning the tip of the wire in the superior caval vein. The stent is hand crimped onto a BIB balloon catheter (NuMED, Hopkinton), with a diameter equal to or 1–2 mm larger than the angiographic conduit diameter. Short procedural and fluoroscopy times required by this procedure are attractive, as well as the complete immediate fenestration closure. The technique also avoids protrusion of prosthetic material in the pulmonary atrium that could prompt apposition of thrombotic material and systemic embolism. Disadvantage of this technique is the relatively large sheath size needed for covered stent delivery.
- The combination of Fontan baffle stenosis "downstream" from the fenestration or baffle leak may significantly worsen right-to-left shunting especially during exercise. Device occlusion of fenestrations or leaks may additionally narrow the pathway in these patients and is therefore undesirable. Balloon expandable covered stents may be less desirable in this setting as there is often a significant size discrepancy between the stenotic area and the largest baffle diameter, which can potentially result in either incomplete closure of the baffle leak or an inadvertent baffle tear. Madan et al. recently described two patients with the combination of Fontan baffle stenosis and patent fenestration successfully treated with a Zenith abdominal aortic aneurysm endograft (Cook Medical) [7]. The Cook Zenith endograft is constructed using full-thickness woven polyester fabric sewn to a self-expanding stainless steel endoskeleton. This framework with fabric on the outside provides good graft to vessel wall apposition. The delivery system of the Cook Zenith stent offers an advantage over balloon expandable stents by enabling precise positioning and readjustment of the graft before final deployment. In addition, post-deployment, the self-expanding stent conforms to the vessel wall and selective

dilation of specific areas using different balloons can then be performed. This is advantageous in the Fontan patient where the baffle is not of uniform calibre in order to minimize residual leak. Due to the large delivery sheath size (16Fr), this technique should be reserved for older children or adults.

32.10 Closing the Stented Fenestration

- Spontaneous closure of a fenestration during the early postoperative period may lead to haemodynamic deterioration associated with elevated systemic venous pressures, low cardiac output, progressive oedema and effusions. The use of intravascular stents to reopen or create a fenestration in these unstable patients can be life-saving.
- Future reclosing of these stented fenestrations after patients have improved haemodynamically might pose some challenges. Figure 32.4a depicts such a stented fenestration that had to be created postoperatively in a patient due to prolonged chylothorax after early spontaneous closure of the fenestration. Ten months later, an Amplatzer duct occluder II was implanted within the stent, but 3 years later, saturations persisted below 88 % due to residual right-to-left shunting.

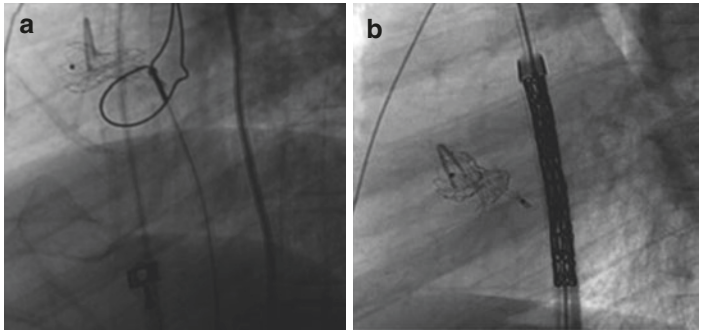

Fig. 32.4 Stented fenestration with previous attempt of closure with an Amplatzer duct occluder type II; now complete closure with a covered CP stent (see text)

Clinical and haemodynamic evaluations were favourable for complete fenestration closure, but technically the procedure proved to be challenging. The distal (conduit) part of the stent was snared from the femoral side and gradually pulled caudally against the conduit wall, to prevent sharp edges sticking into the conduit (Fig. 32.4a). The stent was then forced even more against the conduit from cranially to caudally by inflating a 20-mm Atlas balloon, also testing for the risk of balloon perforation due to residual sharp edges. Finally, a 45-mm covered CP stent was implanted using a 22-mm BIB (Fig. 32.4b), obtaining complete closure of the fenestration and a non-obstructive conduit.

32.11 Devices for Partial Occlusion

- In some patients with suboptimal Fontan physiology, the fenestration may be too large in the early postoperative period in a patient not yet stable enough for complete closure of the fenestration. The possibility to partially close such a fenestration could be an attractive option in this setting.
- A customized fenestrated atrial septal occluder device has been used in few patients; however, the incidence of spontaneous closure of the fenestration in the immediate follow-up period was high. We described a partial occluder, the 115S PFO star (CARDIA ™), designed by removing two opposite quadrants from the right atrial disc. These devices can also be manually tailored in the catheterization laboratory by removing one or more quadrants of the polyvinyl alcohol foam on the proximal disc (depending on the magnitude of residual shunt required). In the 18 patients in which the partial occluder was implanted, mild to moderate residual shunting remained in all but two after 1 month. Six months after device implantation, residual shunting was still documented by echocardiography in 12 of these patients (saturations 90 %±3 %). Closure of these shunts should be technically feasible using coils or a covered stent when indicated.

32.12 Follow-Up After Fenestration Closure

- Patients should be routinely evaluated (clinically and echocardiographically) 24 h, 1 month and 6 months after the intervention with specific attention to clinical signs of venous congestion or low output and evidence of thrombus or residual shunt on TTE. Complete closure is defined as improved saturations clinically and the absence of clinically significant shunt on colour.
- Transthoracic echocardiography may fail to detect some thrombi, and although routine transoesophageal echocardiography is more invasive, it should probably be considered in certain high-risk patients.
- The optimal anticoagulation regimen after Fontan completion is unknown. Previous reports have shown an incidence of 20–23 % of thrombus formation in the extracardiac conduit if anticoagulants were not given. Patients with a persistent right-to-left shunt and a tendency to form venous thrombi may be at increased risk for paradoxical embolic events and device occlusion of the fenestration may decrease the risks of systemic thromboembolisation. Treatment protocol before and after fenestration closure differs depending on institutional protocol and risk stratification in individual patients.
- In our institution all patients with a fenestrated Fontan circulation are treated with acetylsalicylic acid 1–2 mg/kg/day orally in combination with clopidogrel 0.2 mg/kg/day orally; the clopidogrel is usually stopped 6 months after fenestration closure, except in patients with “unfavourable” haemodynamics. In the event of previous thrombosis or high-risk patients, lifelong treatment with Coumadin is used aiming for a target prothrombin time of 1.5–2.

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Ventricular Septal Defects

33

Massimo Chessa, Gianfranco Butera, and Angelo Fabio d'Aiello

33.1 Clinical Indications

The defects that may be suitable for percutaneous closure are located within the muscular septum (muscular ventricular septal defects-MVSD) or in the perimembranous septum (perimembranous ventricular septal defects-PMVSD) with or without aneurysm, and they can be native residual post-surgery.

Surgical repair is currently the only option for doubly committed or supracristal defects, for perimembranous defects associated with prolapse of aortic valve and aortic regurgitation, and for any defect associated with malalignment of the muscular outlet septum, or straddling and overriding atrioventricular valves.

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Large defects give signs and symptoms of cardiac failure in early infancy, and they have to be treated surgically in the first months of life.

Clinical indications for the closure of ventricular septal defects (VSD) are:

- Symptoms of heart failure
- Signs of left heart volume overload with an echocardiographic evidence of a significant left-to-right shunt through the defect.

A shunt is considered significant when the following are found:

1. Left atrial enlargement, defined as a left atrial-to-aortic ratio >1.5
2. Left ventricular enlargement (left ventricular overload), defined as a left ventricular end-diastolic diameter >+2 standard deviation (SD) above the mean for the patient's age

Patients with left ventricular volume overload and haemodynamically significant shunts (Qp: Qs ≥1.5:1) should undergo VSD closure if pulmonary artery (PA) systolic pressure is less than 50% systemic and pulmonary vascular resistance is less than one-third systemic.

Closure may be needed in order to prevent pulmonary arterial hypertension, ventricular dilation, arrhythmias, aortic regurgitation and development of double-chambered right ventricle. In specific cases, small defects, with neither symptoms of cardiac failure nor overload, may need closure if an episode of infective endocarditis was experienced [1–6].

33.2 Patient Selection

– Absence of active infection; if a source of potential infection is founded, treat it before catheterization.
– Complete and deep analysis of previous medical history, cardiac catheterization and surgeries if the VSD is a residual post-surgical defect.

– Check personally the TTE before start the procedure.
– Take into consideration the possibility to treat associated anomalies if they are present (pulmonary valve or branch stenosis, atrial septal defect, etc.).
– Take personally the informed consent for all the planning procedures.

33.2.1 Technical and Equipment Issues

33.2.1.1 Device for MVSD

The Amplatzer MVSD occluder (Amplatzer muscular VSD Occluder, AGA Medical Corporation, St. Jude-Abbott, MN, USA) (Fig. 33.1) is a self-expandable device made of nitinol wires (thickness 0.004–0.005 in.), consisting of two flat discs having a diameter 8 mm larger than a central connecting waist (7 mm long). The diameter of the waist determines the size of the device, and it is available in sizes from 4 to 18 mm. Three Dacron polyester patches are sewn with polyester thread into both discs and the connecting waist. The device is secured to a delivery cable and is inserted into a delivery sheath ranging from 6 to 9 French in size [1, 2].

33.2.1.2 Device for PMVSD

The Amplatzer MVSD occluder (Amplatzer muscular VSD Occluder, AGA Medical Corporation, St. Jude-Abbott, MN, USA) (Fig. 33.1) could be used. Recently, the KONAR-MF™ (multifunctional) VSD device (Lifetech, China) has received CE marking approval for VSD closure (Fig. 33.2). The occluder is a self-expandable, double-disc device made from a nitinol wire mesh. The two discs are linked together by a cone-shaped waist. The waist of the four largest models (9–7, 10–8, 12–10, 14–12) is sewn with PTFE membranes securely using nylon threads in order to increase its occlusion ability and reduce the residual shunts, while the four smallest models (5–3, 6–4, 7–5, 8–6) have no membrane in it. A double-sided screw is available for retrograde and ante-grade approach. The device requires delivery sheaths from 4 to 7 French. It can be used for both perimembranous and MVSDs closure.

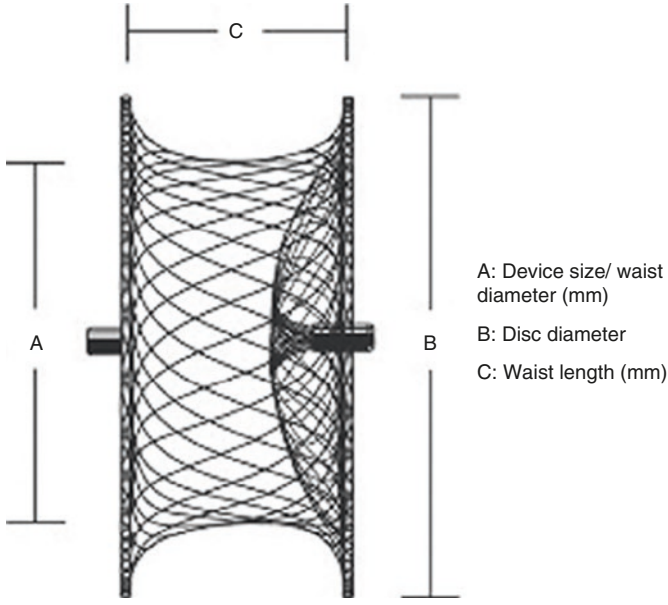

Fig. 33.1 The Amplatzer Muscular VSD Occluder, AGA Medical Corporation, St. Jude-Abbott, MN. *A* device size/waist diameter (mm), *B* disc diameter (mm), *C* waist length (mm)

Fig. 33.2 KONAR-MF™ (multifunctional) VSD device (Lifetech, China)

Procedure

Preparation

- General anaesthesia and orotracheal intubation.
- Biplane catheterization laboratory preferred.
- Patient position with arms lifted up, behind patient's neck (attention to brachial plexus overstretching).
- Patient is fully monitored including an arterial line for continuous arterial pressure monitoring, two peripheral venous lines or a central venous line, vesical catheter for diuresis evaluation.
- A transoesophageal echocardiography 2D or 3D (TEE) must be used to monitoring the procedure.
- Full heparinization with Heparin IV 100 UI/kg. Check hourly the activated clotting time >250 s. In case, add heparin IV during the procedure. Usually, it is not needed.
- Antibiotics IV: usually a cephalosporin.
- The procedure has to be considered as a surgical intervention. Therefore, special care has to be paid to strict asepsis. Special attention has to be given to operators' scrubbing and patient's preparation (including careful depilation). The personnel involved have to wear masks and hats.

Access Site

- A femoral vein (FV) access is used, to approach the closure of a PMVSD, and an internal jugular vein (IJV) access can be used for MVSD closure.
- An arteriovenous circuit must be created (IJV-femoral artery for MVSD and FV-femoral artery for PMVSD closure).
- Both sides for vascular femoral access are prepared.

Catheterization and Haemodynamic Evaluation for MVSD Closure

Left ventricular angiographies are obtained in axial projections for best evaluation of VSD size and position, in addition to TEE views. Left ventriculography in the hepatoclavicular projection (35° left anterior oblique/35° cranial) is performed to imaging mid-muscular, apical posterior defects. Anterior defects are better seen in 60° left anterior oblique/20° cranial (Fig. 33.3).

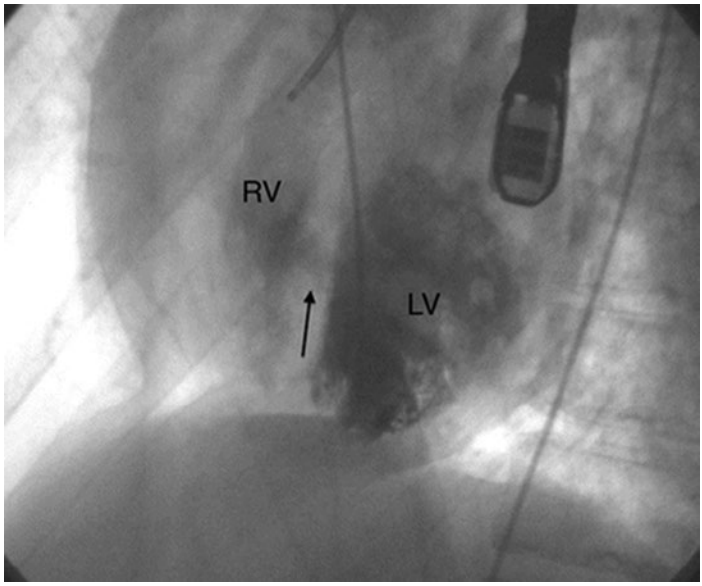

Fig. 33.3 Left ventricular angiogram: *LV* left ventricle, *RV* right ventricle, *Arrow* mid-muscular VSD

The VSD is crossed from the left side, by using a Right Judkins or Right Amplatz catheter and a soft glide wire (0.035 in., J tip, Terumo); the wire is advanced to pulmonary artery, where it is snared with a GooseNeck snare (Microvena Corporation, 20–25 mm in adults, 10–15 mm in children) (Fig. 33.4), and exteriorized out of right IJV or FV establishing an arteriovenous circuit.

Over the circuit, an appropriate size delivery sheath is advanced from the vein all the way until the tip of the sheath is in the ascending aorta. The dilator is withdrawn, and the sheath is pulled back in the left ventricle.

When the tip of the sheath is placed in the mid cavity of the left ventricle, the dilator and the wire are gently removed; a left ventriculogram is usually repeated, to confirm the position of the long sheath and also obtain additional information of the position and the size of the VSD.

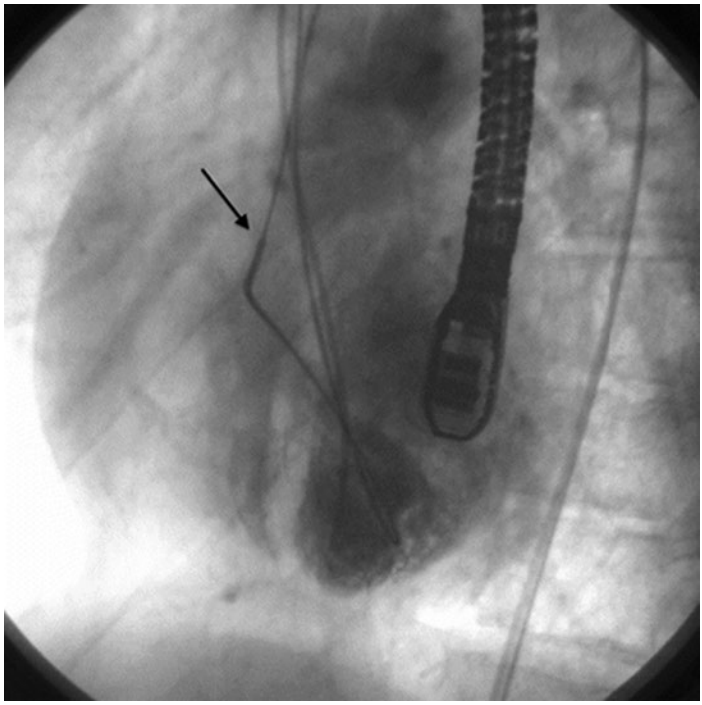

Fig. 33.4 Left ventricular angiogram: the *arrow* shows the arteriovenous circuit (femoral artery-IJV)

According to both angiographic and echocardiographic information, a VSD occluder 1–2 mm larger than the maximum size of the defect is chosen.

The device is attached to the delivery cable, loaded into the plastic loader, introduced and advanced into the sheath.

The left disc is deployed in the left ventricular cavity, making sure it is not impinged in mitral valve apparatus, then the entire system is withdrawn towards the septum (Fig. 33.5a), the central waist and the proximal disc are deployed; a test angiogram is done to verify the correct position of the device (Fig. 33.5b).

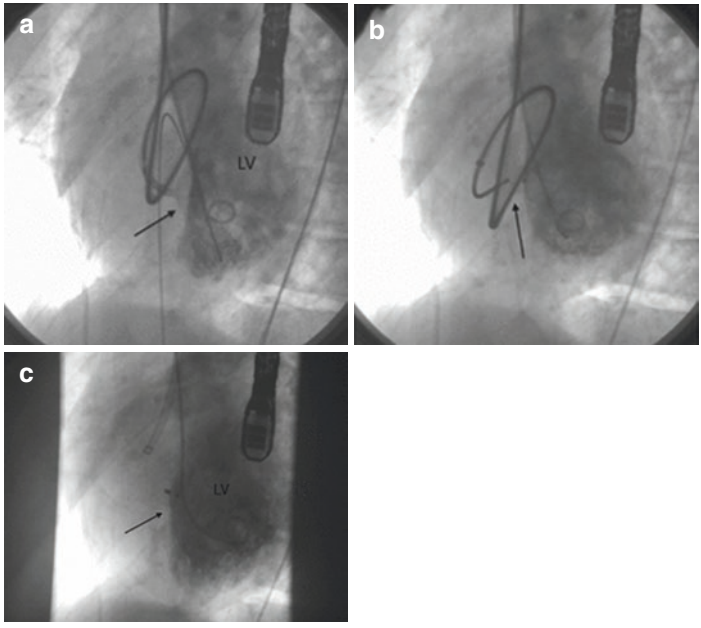

Fig. 33.5 Left ventricular angiogram: (**a**) The system is withdrawn towards the septum (*arrow*), (**b**) the central waist and the proximal disc are deployed (*arrow*), (**c**) a test angiogram is done to verify the correct position of the device (*arrow*). *LV* left ventricle

Echocardiographic views are also very important to confirm the position of the two discs on left and right side of the septum, respectively, and the central waist within the muscular septum.

The device is then released.

A final angiogram is performed approximately 10–15 min afterwards, to assess the position of the device and the possible residual shunt (Fig. 33.5c).

Patients receive acetylsalicylic acid (3–5 mg/kg/daily maximum 300 mg/daily) for 6 months and are asked to follow strictly endocarditis prophylaxis.

A similar approach may be used to close multiple muscular VSDs.

33.2.2 Alternative Techniques

33.2.2.1 Retrograde Approach

This approach can be used in adults and older children in whom a 7–8 French arterial introducer can be used safely.

The VSD is crossed from the left ventricle with the help of a soft 0.035 in. J-Tipped Terumo 260 cm exchange wire introduced through a Right Judkin or Right Amplatz coronary artery catheter.

The wire is then advanced in the pulmonary artery. The catheter is exchanged with an 80 cm delivery sheath (AGA medical or Lifetech SteerEase introducer) over the wire to the right ventricle apex. Wire and dilator are removed slowly in order to avoid air suctioning.

The chosen device is prepared and advanced into the long sheath (Fig. 33.6a). The distal disc is opened in the RV apex paying attention to the ventricular wall and tricuspid wall (Fig. 33.6b).

The whole system is then pulled back to approximate the interventricular septum. The sheath is further withdrawn to open the proximal disc onto the left ventricular surface of the interventricular septum (Fig. 33.6c).

Left ventricular angiograms and echocardiographic evaluations are performed to confirm the position of the device and the absence of complications (Fig. 33.6d).

The device is unscrewed from the delivery cable, and angiograms are performed in the ascending aorta and left ventricle to confirm the final position of the device, to search for residual shunt and to check aortic valve function.

Hybrid Approach [7, 8]

A hybrid approach has been developed to overcome the risks of the two procedures (percutaneous closure may be hazardous due to vascular access and haemodynamic tolerance of the procedure, and a surgical approach needs extracorporeal circulation and may be associated to significant morbidity and mortality, in particular, in case of apical defects) in smaller infants (less than 6 kg) [8].

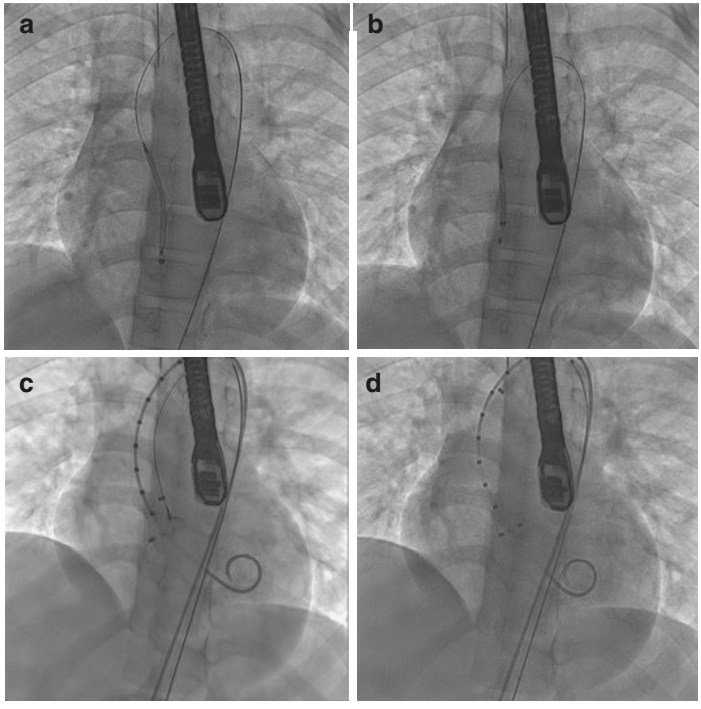

Fig. 33.6 Retrograde approach for high MVSD or PMVSD: (**a**) The delivery sheath is advanced over the wire in the right ventricle apex; the chosen device is prepared and advanced into the long sheath. (**b**) The distal disc is opened in the RV apex paying attention to the ventricular wall and tricuspid wall. (**c**) The whole system is then pulled back to approximate the interventricular septum. The sheath is further withdrawn to open the proximal disc onto the left ventricular surface of the interventricular septum. (**d**) Left ventricular angiograms and echocardiographic evaluations are performed to confirm the position of the device and the absence of complications

The chest and the pericardium are opened, under TEE control, an 18-gaude needle is used to puncture the right ventricle free wall.

A 5-0 polypropylene purse-string suture is placed around the puncture site.

The needle is introduced into the right ventricular cavity pointing towards the VSD.

A 0.0252 short guidewire is passed through the needle and the VSD in the left ventricle.

Over the wire, a short sheath is advanced to the left ventricle cavity.

A proper-size VSD device is delivered using TEE monitoring.

Catheterization and Haemodynamic Evaluation for PMVSD Closure

Angiographies are performed using 60° left atrial oblique plus 20° cranial view (Fig. 33.7).

An angiogram of the ascending aorta is also performed in 50° left atrial oblique view to check for aortic insufficiency.

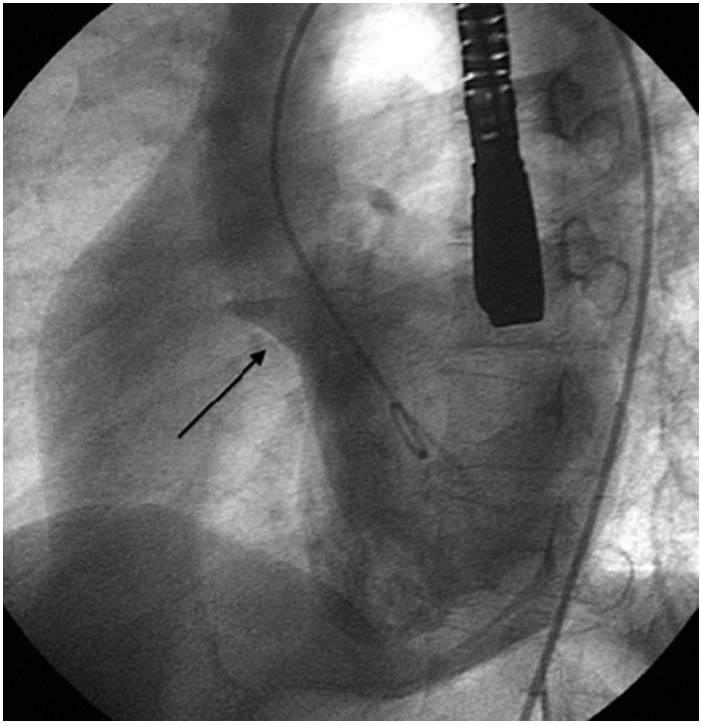

Fig. 33.7 Left ventricular angiogram: the *black arrow* shows the PMVSD

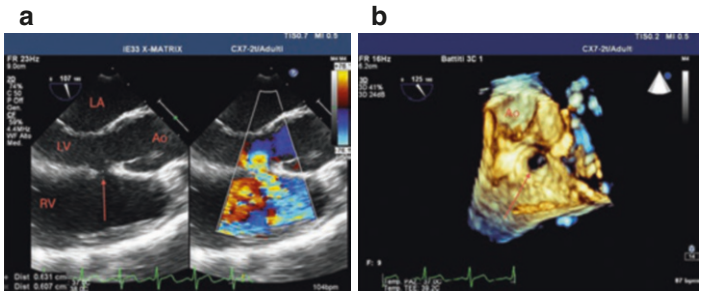

Fig. 33.8 (**a**) 2D-TEE long-axis view: *arrow*, PMVSD; *RV* right ventricle, *LV* left ventricle, *LA* left atrium, *Ao* aorta. (**b**) 3D-TEE: *arrow*, PMVSD; *Ao* aorta

The size of the defect and its relationship to the aorta are confirmed by TEE (Fig. 33.8).

The defect is crossed from the left ventricle by using a Right Judkins or a Right Amplatz catheter and a Terumo wire. The catheter is advanced to the pulmonary arteries or the superior or inferior caval vein.

The Terumo wire is then replaced by the soft exchange noodle wire (a dedicated 300 cm exchange guidewire, AGA Medical Corporation, Golden Valley, MN), snared with a GooseNeck snare and exteriorized from the FV (arteriovenous circuit) (Fig. 33.9a–b).

The delivery system is advanced over the wire up to ascending aorta.

Use a "kissing" technique if some resistance are encountered: both the tip of the sheath and of the arterial catheter over the wire must be in contact and pushed-pulled together.

When the long sheath is in ascending aorta, hold the guidewire circuit, withdraw the dilator of approximately 10 cm, withdraw slowly the sheath and advance the arterial catheter; make a loop of the wire and push it into the left ventricular apex.

The sheath is then advanced over the wire until it reaches the apex of the left ventricle, and the wire is gently removed (Fig. 33.10).

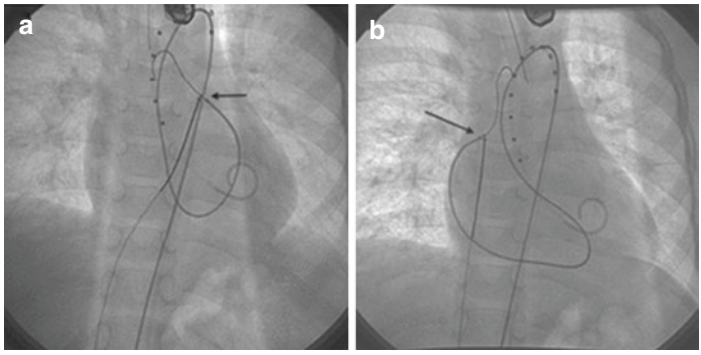

Fig. 33.9 The wire is snared with a GooseNeck Snare in the left pulmonary artery (**a**) or in the superior caval vein (*arrow*) (**b**) and exteriorized from the femoral vein (*arrow*) (arteriovenous circuit)

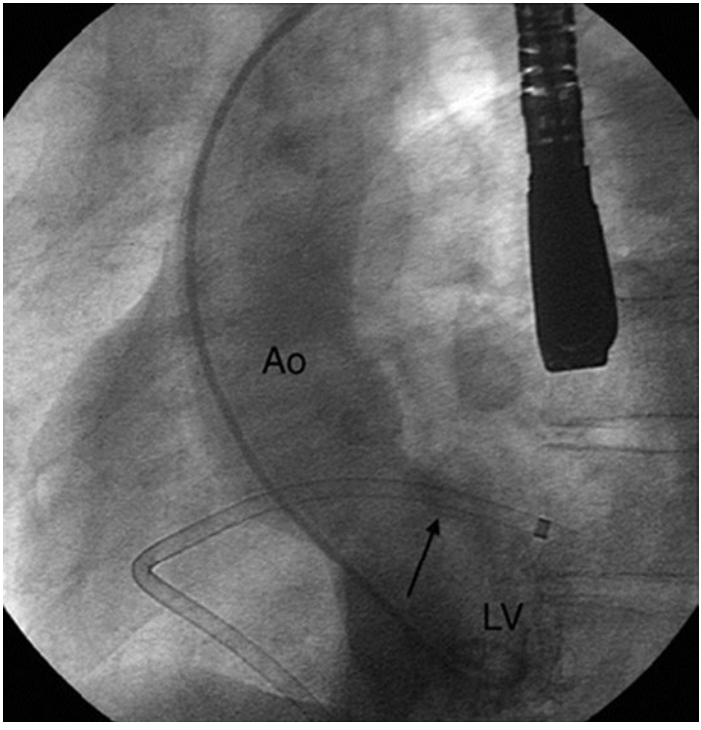

Fig. 33.10 The long sheath (*arrow*) into the left ventricle (*LV*), *Ao* aorta

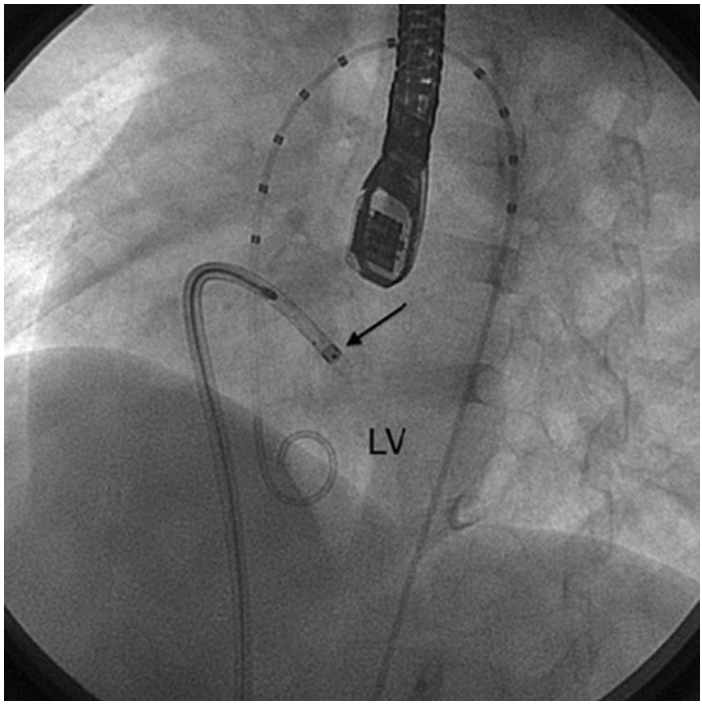

Fig. 33.11 The device is up to the tip (*arrow*) of the long sheath, and the entire system is withdrawn to the left ventricular outflow tract. *LV* left ventricle

The device, having been sized at equal to or 1 mm larger than the size of the defect, is secured on the delivery cable, and the flat part of the micro-screw is aligned with the flat part of the capsule of the pusher catheter.

The device is advanced up to the tip of the sheath, and the entire system is withdrawn to the left ventricular outflow tract (Fig. 33.11).

When the left disc is deployed, an echocardiographic monitoring is of paramount importance to confirm normal function of both mitral and aortic valve.

The platinum marker of the distal disc should point downwards.

The proximal disc is then deployed on the right side of the septum, and angiographic testing is done before releasing the device (Fig. 33.12a–c).

When it is difficult to achieve the position of the braided sheath towards the left ventricular apex, the sheath can be left in the ascending aorta. Before advancing the device, a coronary guidewire 0.014 in. could be advanced in ascending aorta for backup (Fig. 33.13).

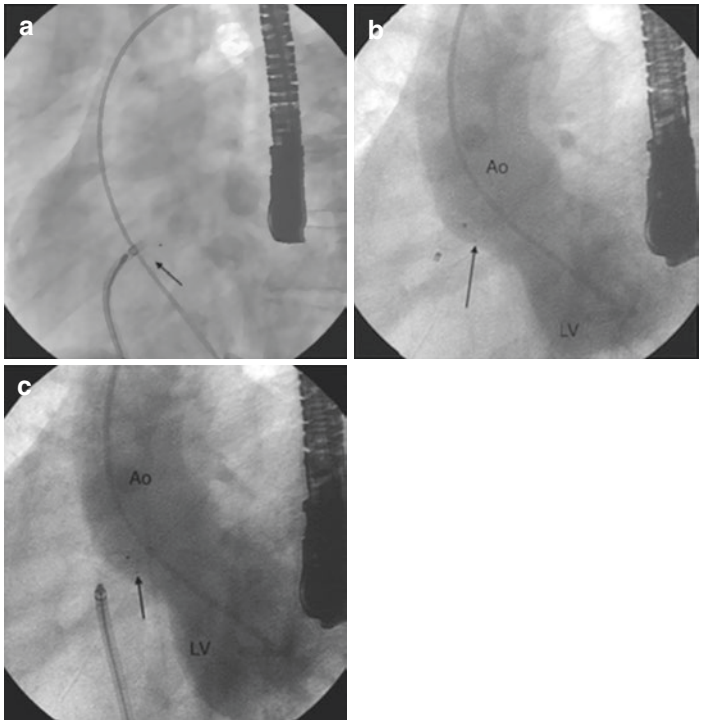

Fig. 33.12 Left ventricular angiogram: (**a**) The left disc is open and withdrawn to the interventricular septum. (**b**) The proximal disc is then deployed on the right side of the septum, and angiographic testing is done before releasing the device. The *arrow* shows the platinum marker of the distal disc pointing downwards. (**c**) The final angiography shows the complete closure of the defect. *Ao* aorta, *LV* left ventricle

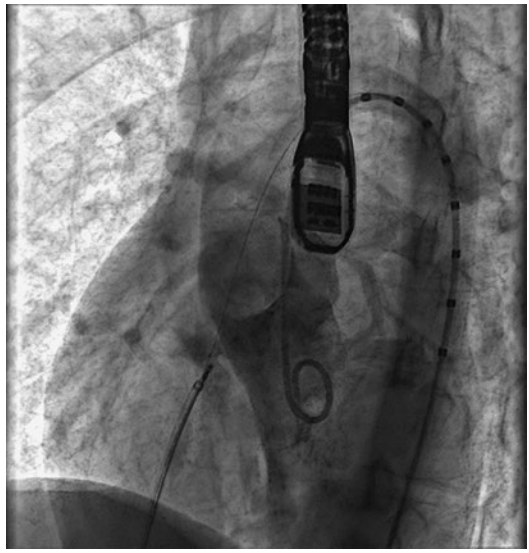

Fig. 33.13 Test angiogram is done before release to verify the correct position of the device. The coronary guide is positioned in ascending aorta during deployment

The left ventricular disc opened under the aortic valve while coming with the sheath from the aorta. Then the right ventricular disc is opened by advancing the delivery cable (Fig. 33.14a–b).

After 10–15 min, the left ventricular angiogram and aortogram are repeated to assess possible residual shunting or aortic regurgitation (Fig. 33.15). Throughout the procedure, the electrocardiogram is carefully screened in order to assess the occurrence of abnormalities of atrioventricular conduction or tachyarrhythmias.

The most common morphological variation is the presence of an aneurysm of the ventricular septum (Fig. 33.16).

Better try to close the true anatomical hole with the more appropriate device (muscular for perimembranous AGA device).

If the redundant tissue of the aneurysm is relatively small, the device could cover the hole along with the aneurysm (Fig. 33.17).

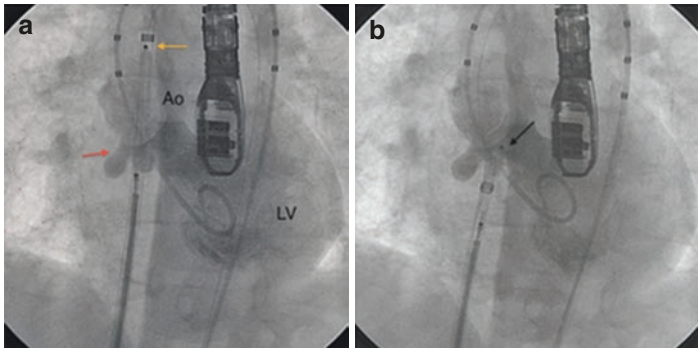

Fig. 33.14 Left ventricular angiogram: (**a**) The sheath can be left in the ascending aorta (*yellow arrow*). The *red arrow* shows the aneurysm of the PMVSD. (**b**) The left ventricular disc opened under the aortic valve while coming with the sheath from the aorta (*arrow*). *Ao* aorta, *LV* left ventricle

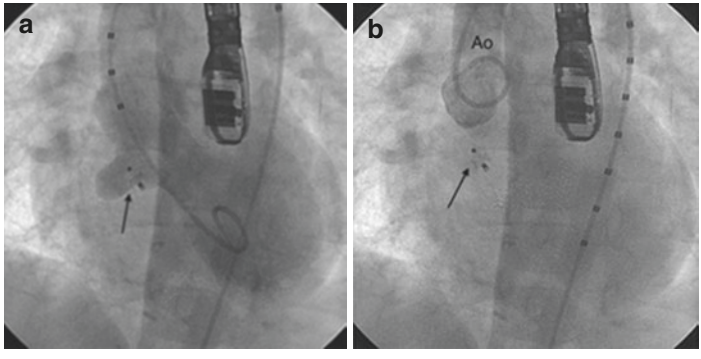

Fig. 33.15 The left ventricular angiogram (**a**) and aortogram (**b**) are repeated at the end of the procedure to assess possible residual shunting or aortic regurgitation. The *arrows* show the device setting into the aneurysm. *Ao* aorta

In case of very large aneurysms, the device may be implanted within the aneurysm itself, with the aim of closing the true anatomical hole, and not to place the device at the "entrance" on the left ventricular side, avoiding insertion of a dangerously oversized device.

In case of conic shape of the aneurysm, different devices (PDA I AGA Medical Corporation, St. Jude, MN, USA, or a Nit-Occlud®

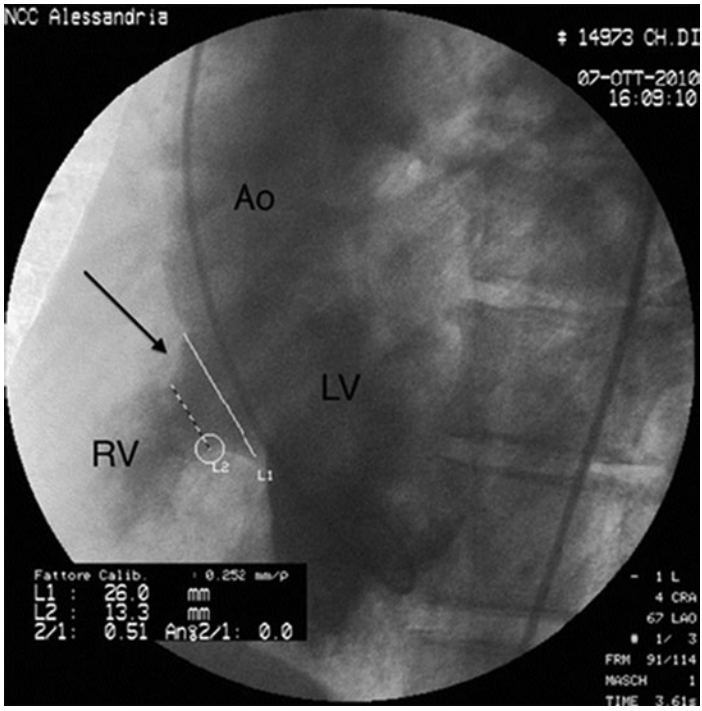

Fig. 33.16 Left ventricular angiogram showing a PMVSD with an aneurysm (*arrow*). *Ao* aorta, *LV* left ventricle, *RV* right ventricle

Lê VSD pfm medical ag.Köln, Germany, Konar-MF™ Lifetech, China) may be taken into consideration.

Specific Technical Aspects for Post-Surgical Residual VSD [9]

Balloon Sizing of the Defect Due to the varied anatomy of the substrate (presence of patches and patch leaks), sizing at the TEE and angio can be more difficult. Balloon occlusion of the shunt and assessment with TEE and angiography provides significantly better understanding of the shunt size and site.

Aortic Retrograde Approach The majority of these VSDs are located in the muscular septum, with a potential risk of the sheath passing through or under a trabeculation of the RV.

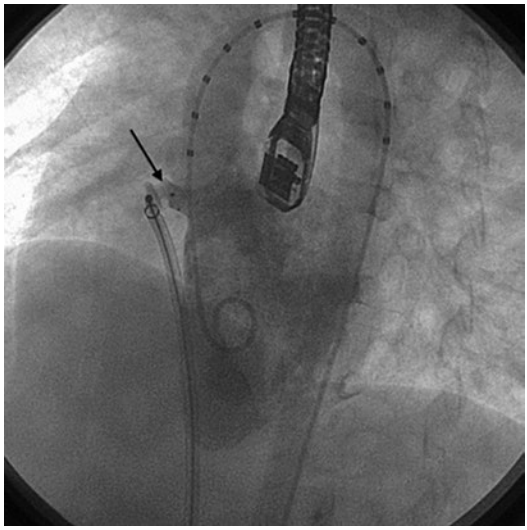

Fig. 33.17 Left ventricular angiogram. The *black arrow* shows the device deployed into the aneurysm

The standard anterograde approach may be more difficult (presence of the surgical patch, less space in the sub-aortic region to deploy the LV disc and increased risk of complications).

A retrograde approach may overcome these issues.

33.3 Complications [10–12]

Complications in closure are reported in 1, 3 up to 5%.

Major procedure-related complications include:

- Embolization of the device (likely to be related to the learning curve or lack of experience by the operator. An underestimated size device is usually implanted). The device can be retrieved, and a second device can be implanted.
- Cardiac perforation (be careful placing and moving guide-wires, delivery sheath and the device).
- Stroke (frequently related to air embolism).

- Deaths (rare).
- Haemolysis, frequently transient.
- Aortic regurgitation (related to PMVSD closure).
- Disturbances of conduction (related to PMVSD closure). Complete heart block (CAVB) is the most warried complication in children (not in adult patients). It may occur acutely, (transiently, during the procedure or permanent) or months after the procedure but permanently. Implantation of a pacemaker may be required.
- The exact mechanism of CAVB remains unclear (inflammatory reaction, formation of scarring in the conduction system, impingement against the vascular conduction system supply). To reduce the risk of CAVB, avoid to overestimate the device size more than 1 mm.

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34 Patent Ductus Arteriosus Closure

Ahmed Mohammed Alkamali
and Ahmed Adel Hassan

Isolated patent ductus arteriosus (PDA) in a full-term infant is one of the common congenital heart diseases. Its incidence ranges from 5% to 10% of all congenital heart diseases.

34.1 Patent Ductus Arteriosus Morphology

- The ductus arteriosus originates in the distal aortic arch just beyond and opposite to the left subclavian artery as a cone-shaped tube connected to the origin of the left pulmonary artery.
- It may have different sizes and shapes. PDA has been classified angiographically by Krichenko et al. into five types (Fig. 34.1). The most common is type A.
- Different approaches and devices may be needed in different morphologies.

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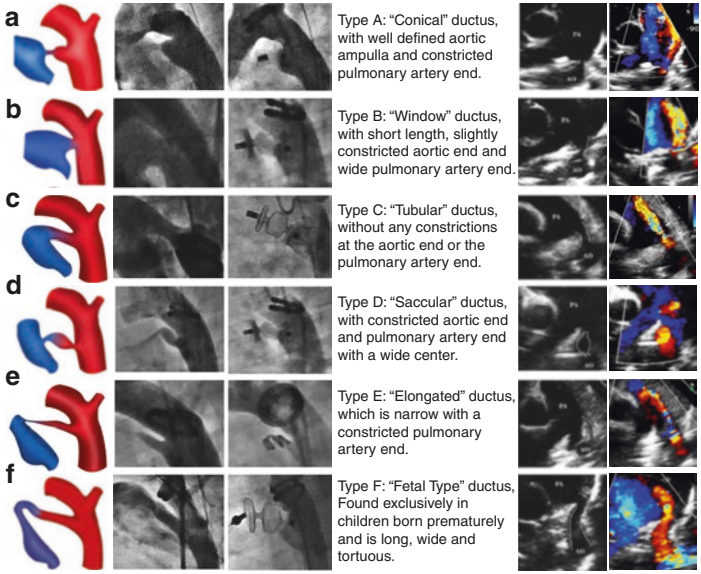

Fig. 34.1 Krichenko classification of PDA morphologies. To the left of the descriptive text is a figure of the different PDA types with their companion lateral aortograms before and after transcatheter device closure. To the right of the text is the concomitant 2D and color. (**f**) Fetal type (PDA morphology of premature children that did not fit the Krichenko et al. classification was grouped as Type F)

34.2 Indications of Closure and Patient Selection

- Clinical scenarios may range from patients with heart failure to asymptomatic subjects. Clinical findings of continuous murmur with bounding pulses and tachycardia indicate significant shunt.
- Symptomatic infants who do not respond rapidly to medical anticongestive treatment should undergo PDA closure. Failure to thrive with recurrent lower respiratory tract infections and exertional dyspnea is common in a large PDA with ventricular overload.

- Premature neonates commonly have PDA causing morbidity. The standard way of closing PDA in this group is either pharmacological or surgical. There is a growing body of evidence that transcatheter closure of hemodynamically significant PDA in prematures contributes to improvement of respiratory status sooner than surgical closure.
- Decision for closure of significant PDA in preterm is a discussion between neonatologist and cardiologist. Challenges include other comorbidities, duct morphology, and hemodynamic significance assessment.

1. In symptomatic infants weighing less than 5 kg, surgical closure of the PDA is recommended. In fact, the use of ordinary devices or coils in small infants with large PDA may have a high incidence of complications. A large device can protrude into the aortic arch causing obstruction. Embolization may occur. The use of large delivery system may induce arrhythmias and can stretch intracardiac structures like the tricuspid valve with severe iatrogenic regurgitation. Amplatzer Duct Occluder II AS (Amplatzer Piccolo™ Occluder) (Fig. 34.7) is a self-expanding nitinol mesh occlusion device with central waist designed to fill the ductus, been used in PDA of premature babies with promising result and less complications.

- In asymptomatic infants with volume-overloaded left heart, PDA closure can be delayed till the weight is above 6–7 kg.
- The indication for closure in patients with silent PDA remains controversial. In these patients, the main argument to close the PDA is the prevention of endarteritis.
- In adults with PDA, it is possible to have a complete spectrum of disease ranging from small asymptomatic PDA to cases of chronic volume-loaded left heart or to Eisenmenger syndrome.
- In patients with markedly increased PVR and those with fixed pulmonary hypertension emphasized by right-to-left shunt (Eisenmenger syndrome), PDA closure is contraindi-

cated. Pulmonary hypertension in this form progresses independently from shunt's closure.

- In patients with moderately increased PVR (>4 Wood units/m^2, ratio PVR/SVR > 0.35), the decision is difficult and is based on the pulmonary response to vasoreactivity testing.
- Echocardiography is the golden tool to select the cases for transcatheter closure.

34.3 Devices

34.3.1 Coil

- In this chapter, we will describe how to use single Flipper Cook coil to close a small PDA. Other PDA coils (e.g., PFM coil) and the use of multiple coils to close large PDAs are also possible.
- Flipper Cook coil comes in different diameters and loop number. They range from 3 mm diameter by three loops up to 8 mm diameter by five loops. The label in coil package will have two numbers (e.g., IMWCE-3-PDA4). The first is the diameter of the loop and second is the number of loops.
- The coil is loaded in clear cartridge with wide end at thread of coil side. The delivery system consists of a coil delivery wire (0.035″ thick) with a straightening mandril inside and the wire thread on other side (Figs. 34.2 and 34.3).
- The Flipper Cook coil is made from 0.035″ wire and can go easily through 4-F end hole, the only multipurpose catheter that accommodates 0.038″ wire.

34.3.2 Devices

2. The ADO I device design has a mushroom shape with a low profile and consists of a flat retention disk and a cylindrical main body, into which polyester fibers are sewn. A steel sleeve with a female thread is welded into the marker band at pulmo-

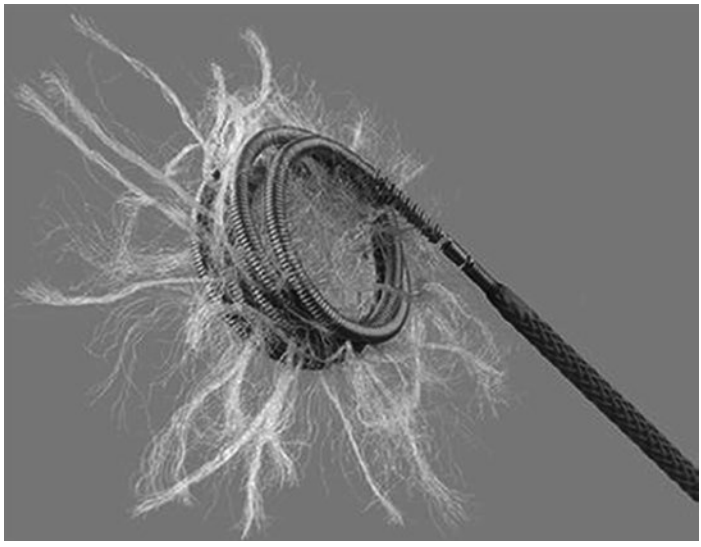

Fig. 34.2 Cook coil

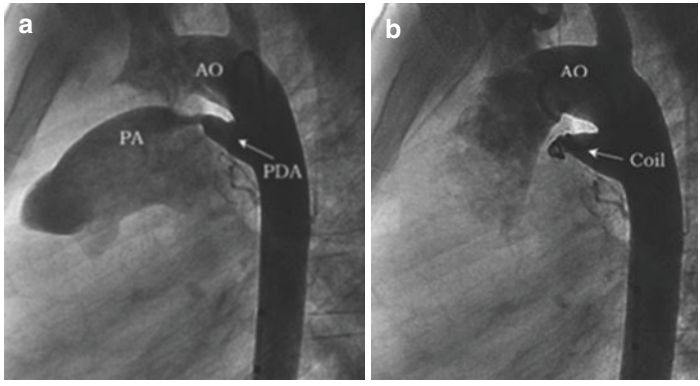

Fig. 34.3 Angiography in lateral view showing a small PDA (**a**) before and (**b**) after closure with detachable Cook coil

nary end. The retention disk, placed distally at the aortic end, is 4 mm larger than the main body, which itself has a conical structure.

- The standard device sizes are 5/4, 6/4, 8/6, 10/8, 12/10, 14/12, and 16/14 mm, respectively.
- The first number denotes the diameter of the larger distal (aortic) end of the device at the retention disk, whereas the second number, which is always 2 mm smaller, denotes the diameter of the proximal (pulmonary) end, where the stainless steel sleeve for screwing onto the cable is located. The smallest first two sizes are 7 mm in length and the remainders are 8 mm.
- The delivery system consists of a delivery cable, a Mullins-type sheath, a loader, and a pin vise.
- The required delivery sheath sizes from 5 to 8 F.
- The size of device chosen is generally such that the diameter of the pulmonary end of the device is at least 2 mm larger than the narrowest diameter of the duct. For example, if the narrowest PDA diameter is 4.8 mm, a 10/8 mm device should be selected.
- In adults with large PDA, it is recommended to oversize the device 4 or 6 mm more than the narrowest diameters (Figs. 34.4 and 34.5).

3. The use of ADO II and ADO II additional size has proven feasible and effective in providing rapid occlusion of PDAs with a diameter ≥2.0 mm and different morphologies.

- The ADO II is a self-expanding nitinol mesh device. Each occluder is made of a multilayered, flexible, and fine nitinol wire mesh shaped into a cylindrical waist with retention disks on either end to secure it in the PDA. It has a "fabric-free" technology, which allows for a very low profile of the device and delivery system. The central waist is designed to fill the defect, and the two retention disks are designed to be deployed on the arterial and venous sides of the defect.

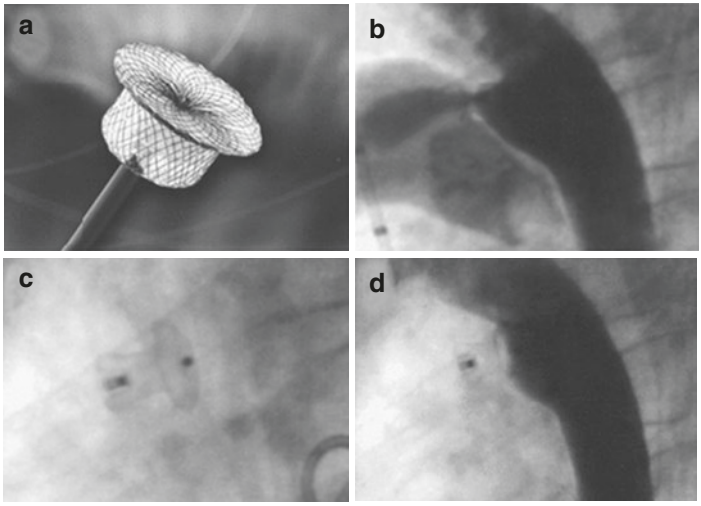

Fig. 34.4 (**a**) Amplatzer ADO I, (**b**) angiographic lateral view showing PDA, (**c**) ADO I in position, (**d**) aortography in lateral view showing complete PDA closure

- This design allows deployment from both the arterial and venous sides (Figs. 34.6 and 34.7).
- These devices are available in 4 or 6 mm lengths, with waist sizes of 3, 4, 5, and 6 mm in both lengths. Each disk diameter is 6 mm greater than the waist size.
- The Amplatzer Duct Occluder II (Fig. 34.6) can treat all types of PDAs in the Krichenko classification up to 5.5 mm in diameter.
- The "window-type" PDA is the only type that is unsuitable for closure with the ADO II. It is also contraindicated in PDAs measuring >12 mm in length and >5.5 mm in diameter on angiography.
- The device has a screw attachment for a delivery wire and radiopaque markers. The recommended delivery sheath is 4- and 5-Fr low-profile TorqVue® LP (AGA Medical) braided and tapered. It has a flexible distal catheter segment that allows for easy approachability. The wire for device

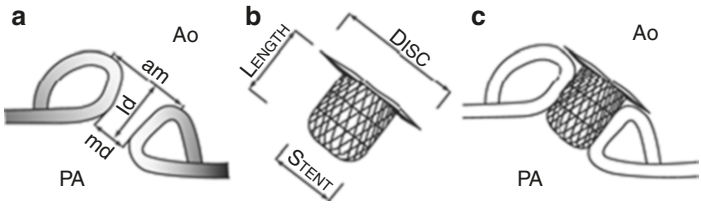

Fig. 34.5 Schematic view of duct measurements and ADO I occluding the PDA. *Ao* Aorta, *PA* Pulmonary artery, *am* aortic side diameter, *md* pulmonary artery side diameter, *Id* device length

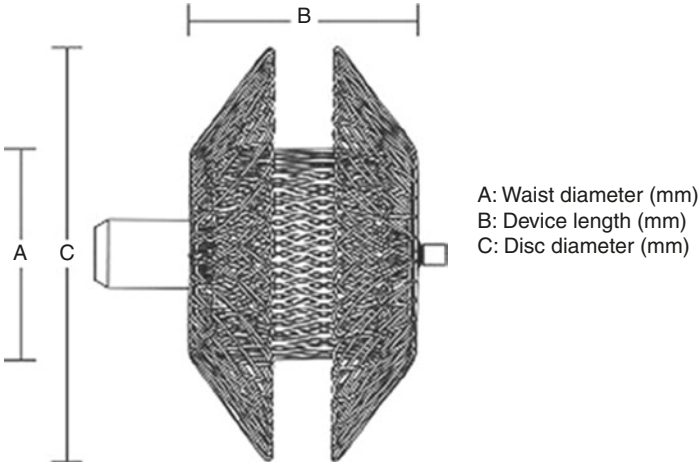

Fig. 34.6 Amplatzer Duct Occluder II. *A* Waist diameter (mm), *B* device length (mm), *C* disk diameter (mm)

positioning and deployment is braided with a flexible nitinol tip. The device can be deployed, recaptured, and redeployed for precise and secure placement.

- Usually, select ADO II 1–2 mm larger than the narrowest waist of the duct. Regarding the length, a 4 mm long device is used for PDAs ≤5 mm long, and a 6 mm length device for PDAs ≥5 mm long.
- The main advantage of ADO II as low-profile device with small caliper delivery sheath makes it feasible to be used in small-weight infants.

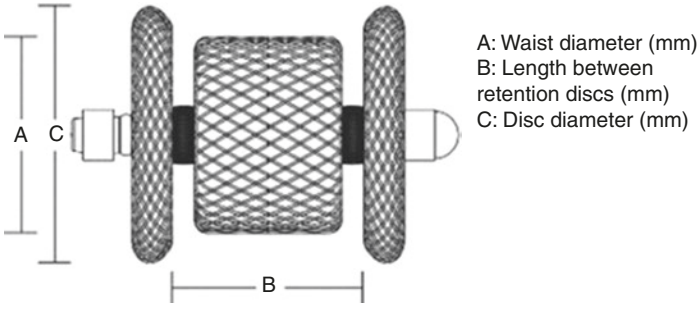

Fig. 34.7 Amplatzer Duct Occluder II AS (Amplatzer Piccolo™ Occluder). *A* Waist diameter (mm), *B* length between retention disks (mm), *C* disk diameter (mm)

4. The Amplatzer Duct Occluder II AS (Amplatzer Piccolo™ Occluder) (Fig. 34.7) is a self-expanding nitinol mesh occlusion device with central waist designed to fill the ductus. Having flat retention disks deployed at pulmonary and aortic ends of the duct with symmetric design allow for venous or aortic approach.

    - The trail of ADO II AS showed good outcome in closing ducts up to 4 mm and best outcome if tubular and in small or ex-premature babies as it goes through 4-Fr delivery system, designed to accommodate all sizes available for Amplatzer Duct Occluder II AS (Amplatzer Piccolo™ Occluder). The delivery cable has a flexible distal tip that allows device to take the final shape before deployment.
    - Only approved device in the USA now is Amplatzer Duct Occluder II AS (Amplatzer Piccolo™ Occluder); however, other devices are used off-label as Amplatzer Vascular Plug II (AVP II) and Medtronic Microvascular plug.
    - Amplatzer Duct Occluder II AS (Amplatzer Piccolo™ Occluder) device has the advantage of being softer and has variable lengths that allow intraductal placement to avoid protrusion problem in pulmonary artery or aorta. It comes with length between retention discs of 2, 4, and 6 mm and disc diameter of 4, 5.25, and 6.5 mm and waist diameter of 3, 4, and 5 mm.

5. For the less common ductal morphology, where the PDA is long and tubular and has no definite constriction at the pulmonary end, the relatively short ADO with only one retention disk may not be the most suitable device. The same limitation may apply in typical conical-shaped but large PDAs in adult patients because of its relative length. In both situations, the Amplatzer Muscular VSD Occluder is a more suitable device. For the short window-type PDA with no ampulla, the atrial septal occlude is more suitable.

34.4 Step-by-Step Procedure

- Cardiac catheterization is preferably performed under general anesthesia, especially in infants and in the presence of other comorbidities. Some centers chose to intervene with conscious or deep sedation without intubation. In some cases, the outcome of this procedure is not predictable, and the conscious sedation may end in a fully ventilated case under general anesthesia.
- In case of preterm PDA closure, it is advised to keep the catheterization lab warmer than usual and use fluid wormer along with other warming options and continuous temperature monitoring of the baby as they are prone to hypothermia.
- Prepare both groins and use a 4-F sheath for the artery and 5 F for the vein. In case of small PDA, only femoral artery access is enough.
- It is advised it use vascular ultrasound probe for access as proven to produce less vascular injury, especially in the prematures.
- Heparin and antibiotics are given according to the catheterization laboratory protocol.
  - If high PA pressures are suspected, right heart hemodynamic evaluation is obtained.
  - If the echo shows clearly restrictive left-to-right shunt with high pressure gradient, it is possible to avoid right heart catheterization and perform aortic hemodynamic study and aortography.

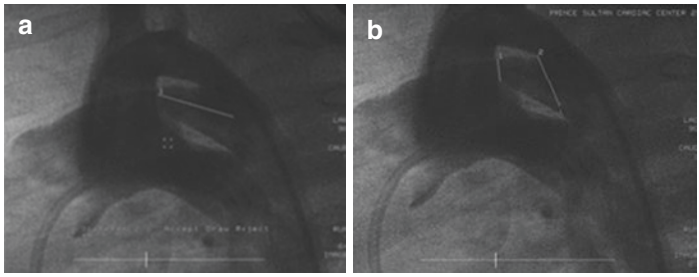

Fig. 34.8 PDA measurements: (**a**) pulmonary end, (**b**) length

- Use NIH or multipurpose catheter for the right side and pigtail for the left-side study.
- Place the pigtail catheter just above the PDA ampulla and perform test angiogram to fill the catheter with contrast and recognize the position of PDA.
- Use the lateral projection as the main projection to measure the size and shape of the PDA. In biplane projection, keep the other at straight PA or with RAO 20–30°.
- In case of large PDA, give up to 2 cc/kg contrast over 1–2 s to delineate the PDA morphology.
- In case of small tubular (type C) or elongated (type E) small PDA, it is possible to engage the ampulla with a multipurpose catheter and perform a hand injection (5–10 cc).
- The device is chosen according to PDA morphology and narrowest diameter (Fig. 34.8).
- In general, the commonest type A PDA with narrowest diameter below 2.5–3 mm can be closed with detachable Cook coil, and the one with narrowest diameter more than 2.5–3 mm can be closed with ADO device.
- Some centers use successfully multiple coils to close the large PDA, taking in consideration that the coil almost costs 10% of ADO device.
- In type B or large tubular PDA, the following measures have to be taken carefully: the narrowest diameter, the total length, and the ampulla diameter.

- In many cases, an Amplatzer Muscular VSD Occluder or ASD Occluder PDA has been used to close type B.
- In general, in type A or E PDA with narrowest diameter >2.5–3 mm, a ductal occluder device is chosen.

34.5 Coil Occlusion of Patent Ductus Arteriosus

- For small PDA with narrowest diameter less than 2.5–3 mm, Flipper detachable Cook coil can be used safely and easily (Figs. 34.2 and 34.3).
- Coil diameters have to be greater than or equal to twice the smallest diameter of the duct, and the ampulla on the aortic aspect of the duct should be large enough to accommodate the coil(s).
- A conical- or funnel-shaped ampulla is best suited for coil occlusion because it allows the coil loops to pack themselves without protrusion into the aorta. Fortunately, the vast majority of ducts have this shape.
- Tubular ducts have a relatively small diameter at the aortic end. This may prevent some of the coil loops from entering the ampulla as the coils are pulled toward the pulmonary arterial end.
- After hemodynamic study and accurate measure of all three diameters of the PDA (total length, ampulla diameter, and narrowest diameter) (Fig. 34.8), select the coil accordingly.
- Usually, Cook coil can be deployed from arterial or venous side. In selected cases with small PDA (<2 mm), PDA can be closed easily and safely from the arterial side without need for femoral vein access, thus avoiding crossing through intracardiac cavities.
- Cross the PDA from arterial side with the 4- or 5-F delivery catheter to main pulmonary artery directly or over a guidewire, just above the pulmonary valve.
- Prepare the selected coil and screw the thread of delivery wire to the thread of coil. Usually, this is the critical part that you should be sure that the mandril enters the thread of the coil.

You need good lighting and prefer to have a white background of the coil to get a clear view (use the white paper of coil package). Screw the delivery wire clockwise and observe the thickening of coil thread with the wire thread. Don't screw tight till the end and keep 2–3 screws free.

- Push the mandril carefully and proximal to the delivery wire preventing any kink of the soft mandril. Push till you feel resistance and the tip of coil moved indicating that the mandril reaches to the end of coil.
- Introduce the straight end of the coil cartridge at the end of the catheter tightly and push gently the delivery wire in the catheter. Under lateral projection fluoroscopy guide, push the coil to the tip of delivery catheter in main pulmonary artery. When the tip of coil is out, withdraw the mandril till the coil completely free from mandril. You will notice the tip of coil will bend slightly.
- Keep the catheter in the middle of MPA away from the pulmonary valve and PDA. With reference to the previous aortogram, push the delivery wire till you get 3/4–1 full loop in MPA. Hold the delivery wire and the catheter together and pull it back till the coil loop is stuck in the pulmonary side of the PDA.
- Keep the delivery wire fixed under gentle tension and milk out the delivery catheter till you free the whole PDA coils in the aorta. Immediately push the delivery wire gently with clockwise rotation to pack the PDA coils in the PDA ampulla.
- The coil can be gently and carefully manipulated to get proper shape and position in the ampulla.
- Release the coil carefully by rotating the delivery wire counterclockwise with pin vise. Don't pull or push the wire during the release.
- Mostly, the coil thread protrudes in the aorta but is usually attached to the wall of aorta. This is generally acceptable.
- After 5–10 min of releasing the coil, do aortogram for any residual leak or malposition of the coil. It is not possible to perform aortogram before release of the coil if occlusion was done from the arterial side.
- The same procedure can be done from the venous side with main numbers of loops in the aortic side and one loop in the

pulmonary side. Before releasing the coil, you can do an aortogram.

34.6 Device Occlusion of Patent Ductus Arteriosus

- After selecting the proper device and before preparing it, cross the PDA from the femoral vein up to the descending aorta with the multipurpose catheter. Usually, to cross the PDA, a standard straight tip guidewire or a floppy hydrophilic wire is needed.
- Advance the multipurpose catheter down to abdominal aorta and exchange the guidewire for an exchange 0.035 stiff wire that is placed down toward the contralateral iliac or femoral artery. Always keep the pigtail at the arterial side in the descending aorta for hemodynamic monitoring and to perform aortographies during and after deployment of the device.
- Prepare the delivery system by flushing the dilator and long sheath. The multipurpose catheter is exchanged for the delivery sheath and dilator over the 0.035″ exchange guidewire. While crossing the curve of the RVOT-PDA-descending aorta, keep the exchange wire straight and stable down to the iliac artery to facilitate smooth progression of the delivery system. The dilator is then removed, leaving the sheath in the abdominal aorta, just below the diaphragm level. At removal of the dilator, allow some backflow bleeding, de-air the delivery sheath, and then flush it gently.
- Open the proper device and merge it in pure saline. Insert the cable in the short sheath (loading pod). The device is screwed clockwise onto the tip of the delivery cable. When it is not possible to screw anymore, turning counterclockwise, a "click" can be felt. The side port allows easy flushing of the loaded device within the sheath. The loading pod is introduced into the delivery sheath, and the cable is then pushed to advance the

device. To prevent inadvertent unscrewing, rotation of the cable should be avoided when the device is being advanced.

- Keep a reference of aortogram at lateral projection with clear PDA site and morphology during the introduction and the deployment of the device.
- Under fluoroscopy, the device is advanced by pushing the delivery cable until it reaches the tip of the delivery sheath in the thoracic aorta. The sheath is gently withdrawn to deploy the retention disk only, following which the cable and delivery sheath are pulled as one unit under lateral fluoroscopy until the retention disk is against the ductal ampulla. This can be observed by fluoroscopy using the tracheal air column as landmark from the diagnostic aortogram previously done. Furthermore, a tugging sensation in synchrony with the aortic pulsation can be felt.
- It's preferable to push the pigtail carefully to the device side and perform an aortography to delineate relation of retention disk to the ampulla, especially if dealing with an adult/infant case or tubular PDA or type E.
- Once the position has been confirmed based on the location of the narrowest diameter in relation to the tracheal air column, the cylindrical portion of the device is deployed by retracting the delivery sheath while applying slight tension on the cable. This is for ADO I-type devices.
- Avoid pushing or pulling the cable while retracting the delivery sheath. After deployment of second retention disk, some operators wiggle gently the cable to test device stability.
- Before detaching the device, perform another aortogram to verify correct positioning of the device. This is evident by the retention disk being well apposed to the ampulla and a slight waist seen in the middle portion of the device induced by constriction at the narrowest part of the PDA.
- If the position is satisfactory, unscrew the cable by rotating counterclockwise under fluoroscopy. Keep the delivery sheath

near the tip of the cable to prevent any traumatic jump of the cable tip at the time of release.
- After 5–10 min, do other aortogram to confirm position and observe any significant residual leak. It's acceptable to see residual through the device as the process of clotting will take some time. If there is a jet around the device, consider recapturing the device and change it to a bigger size.

In the case of use of ADO II and Amplatzer Duct Occluder II AS (Amplatzer Piccolo™ Occluder), the procedure of device delivery is very similar to ADO I.

With small infants, it is recommended to deliver the device using an anterograde transvenous approach and to avoid arterial access whenever possible. It is recommended not to deliver the device in small infants (≤2 kg) using the retrograde approach since small infants are at an increased risk for arterial injury.

- With the device still attached to the cable, a descending aortogram in the lateral projection to confirm the position.
- Once proper device position is confirmed, the device is released by anticlockwise rotation of the delivery cable. A repeat descending aortogram 10 min after the release to check for residual shunts can be done (Fig. 34.9).

34.7 Hints and Pitfalls in Coil Implantation

- Retrieval of an embolized coil can be performed by using a 10 mm gooseneck snare and a 4–5-Fr snare catheter.
- Sometimes, it can be very difficult or impossible to retrieve the coil. If there is no problem with the pulmonary flow, the coil can be leaved.

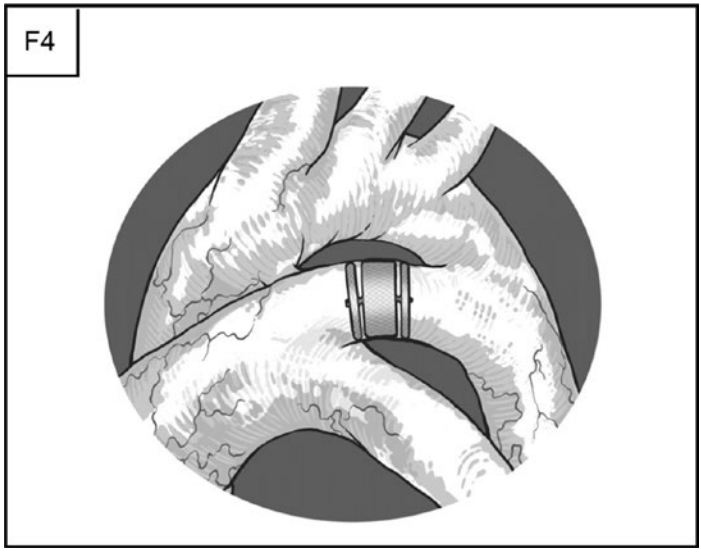

Fig. 34.9 An example of the correct shape for a deployed occlude. The discs should not protrude or bulge into the surrounding vessels

34.8 Hints and Pitfalls in Amplatzer Device Implantation

- Kinking of the long sheath in the RVOT in infants <10 kg may occur.
- Window-like defects can be treated by using an Amplatzer ASD Occluder.
- PDA with pulmonary hypertension can be treated by using an Amplatzer Muscular VSD Occluder in order to have a good support from both sides (pulmonary and arterial side) in cases of isosystemic pulmonary pressures. This device can be used in older children because the proximal disk can give stenosis on the LPA in smaller babies.

- It was reported in the literature the added value of combined use of fluoroscopy and echocardiography guidance.
- In small babies, keep BP cuff in the lower limb and palpate the femoral pulses post device placement to assess for aortic obstruction.
- In prematures, utilize the esophageal temperature probe as a landmark for aortic isthmus.

34.9 Possible Complications

- Device Embolization: Retrieval of an ADO device can be performed by using gooseneck snare and a long sheath appropriate for the device's size.
- Hemolysis.
- Left pulmonary branch obstruction.
- Aortic isthmus obstruction.
- Be careful when closing large ducts in small babies!
- Reported complications for premature PDA closure (Post PDA closure syndrome, LPA stenosis, aortic stenosis, tricuspid or pulmonary valves injury, femoral artery thrombosis, hemolysis, embolization, IVC dissection reported as well).

Further Reading

Amplatzer Piccolo™ Occluder, Instructions for Use.

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35 Percutaneous Closure of PDA in Premature Babies

Carles Bautista-Rodriguez, Helene Bouvaist, Tosin Otunla, Alban-Elouen Baruteau, and Alain Fraisse

35.1 Background

Patent ductus arteriosus (PDA) is the most common cardiac diagnosis in premature newborns. A persistent PDA, defined as failure of the ductus to close within 72 h postnatally, is seen in around 50% of extremely premature newborns [1]. The presence of a haemody-

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namically significant PDA increases the risk of chronic respiratory disease, prolonged assisted ventilation, necrotizing enterocolitis, pulmonary haemorrhage, bronchopulmonary dysplasia, intraventricular haemorrhage, renal impairment and death [2–10].

Despite this, the treatment strategy remains controversial [2, 11, 12]. Treatments with cyclooxygenase inhibitors or paracetamol have been used to promote closure when a PDA was found to be haemodynamically significant. However, significant adverse effects are reported, such as renal function impairment, bleeding, developmental delay and motor impairment [13, 14]. Medical treatment fails in 20–30% of the cases [15, 16].

Surgical ligation of the PDA is often the alternative to failed pharmacological treatment, and it is usually performed through left thoracotomy. Thirty days mortality rate is 5–8% [17, 18]. It has been associated to haemodynamic instability (post-ligation syndrome), phrenic nerve and vocal cord palsy, pneumothorax, bleeding and infection [19–21]. Evidence also suggest that it may also worsen long-term outcomes, including increasing the risk for bronchopulmonary dysplasia, retinopathy of prematurity and neurosensory impairment [22–24].

Conservative management consisting of fluid restriction, diuretics and positive end-expiratory pressure is still advocated in many neonatal units because of the lack of scientific evidence regarding PDA closure in this population [25–28]. Although this approach has been associated with increased mortality and morbidity [8, 9, 11, 26, 27, 29], other studies conclude that conservative management is a reasonable option as compared to pharmacologic or surgical treatment [28, 30–32].

In the most recent years, several cohort studies reported preliminary experience with transcatheter technique, using various devices for PDA closure in premature babies [33–44]. Comparison with surgical ligation revealed a positive impact on post-procedure pulmonary outcome [38, 39]. The procedure has been well described in preterm infants, including some patients as small as 640 g [39]. Following the results of an ongoing prospective trial using the ADO II AS [40], this device received FDA and CE approval for transcatheter closure of PDA in premature babies weighting more than 700 g and over 3 days old [41]. This has been renamed as Amplatzer Piccolo Occluder® (APO).

35.2 Patient Selection and Organisation of the Procedure

Preterm infants are always referred by neonatologist for percutaneous closure, usually after failure of medical therapy and/or failure to wean from the ventilator. Pre-procedure transthoracic echocardiography confirms the haemodynamic significance (left cavities dilation, functional mitral/aortic regurgitation and ductal morphology), and transcatheter closure is usually performed in catheterisation laboratory. In some centres with portable fluoroscopy equipment, the procedure can be performed at the bedside, especially in unstable patients. Transcatheter closure at the bedside under echocardiography guidance-only has also been reported [33] but is almost never performed because of potentially severe, life-threatening complications.

35.3 Available Devices

Over the last few years, different devices have been used to close PDAs in premature babies (Fig. 35.1). PDA coil occlusion was initially achieved in selected low-weight infants with symptomatic PDA [34, 42]. More recently, the Amplatzer Vascular Plug II (AVP II) was successfully implanted with good results [43, 45]. Unfortunately, this device is often not suitable for short ducts in

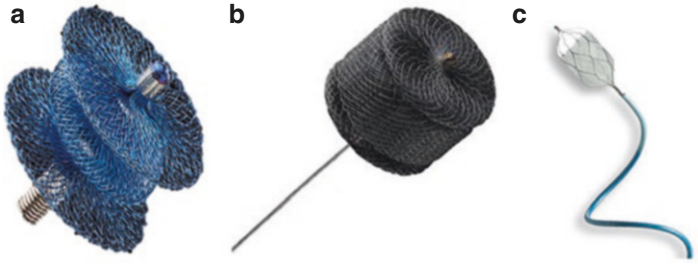

Fig. 35.1 Devices used for percutaneous PDA closure in premature babies. (**a**) ADO II AS/Piccolo (Abbot). (**b**) Amplatzer vascular plug II (Abbot). (**c**) MVP microvascular plug (Medtronic)

extremely low-weight infants. In such cases the central disk, which is the same diameter than the proximal and distal disk, can stretch and make the whole device too long, increasing the risk of LPA and/or aortic stenosis. Conversely, in very large ducts (>4 mm diameter) that are not suitable for the largest APO devices, the 8 mm AVP II is sometimes the only alternative. The Amplatzer Vascular Plug IV has also been used, but this length is also a limit in the smallest babies [44]. Similarly, the retention disks of the Amplatzer duct occluder II are too large for infants weighing <1500 g. The Medtronic Micro Vascular Plug® (MVP), initially designed for occlusion of abnormal blood vessels, has shown excellent results for PDA closure in premature babies [36]. It is made of a nitinol framework covered partially by a polytetrafluoroethylene (PTFE) membrane at the proximal portion. It is delivered through a microcatheter with two sizes of 5.3 mm and 6.5 mm. The APO is currently the only dedicated device for this procedure. It is a self-expanding occlusion device with a central waist and retention disc on both ends, delivered through a 4-French delivery catheter. The device is intended to be used in ducts with <4 mm diameter and with a minimum length of 3 mm [41].

35.4 Description of the Procedure

Ultrasound-guided access of the femoral vein increases the likelihood of successful access and reduces the risk of local complications, especially inadvertent puncture of the femoral artery. Femoral artery access is not recommended and even contraindicated in smaller patients (<1500 g) due to a high risk for vessel occlusion and potential ischemia of the lower limb [46]. Aortic angiogram before and after device deployment is unnecessary when appropriate echocardiographic guidance can be performed. A 4-French sheath is usually inserted in the femoral vein.

Heparin administration remains controversial [47].

Whereas some operators give no heparin, others would give 50–100 units/kg of unfractionated heparin bolus once access has been achieved. Prophylactic antibiotics are administered.

Under fluoroscopic guidance, a 3.3-French Mongoose catheter (PediaVascular, Chagrin Falls, Ohio, USA) or 3.0-French multipurpose BALT catheter (Montmorency, France) is advanced over a wire (a 0.018 hydrophilic wire (Terumo®) or a 0.014 soft coronary wire) through the inferior vena cava towards the right atrium and right ventricle. Baseline haemodynamic measurements are usually not collected. The PDA is crossed and the catheter is positioned in the descending aorta. Sathanandam et al. have reported a similar technique using a 4-french-angled glide catheter (Terumo, Japan) and a 0.035″ Wholey wire (Medtronic, Minneapolis, MN, USA) to cross the PDA ante-gradely into the descending aorta [48]. Other teams have used 4-French Swan-Ganz catheter instead [45].

If a 3F catheter is used to cross the PDA, then a 0.021 Fixed-Core Guide Wire (Cook Medical®, USA) is advanced in the descending aorta and the 3F catheter is directly exchanged with the 4Fr TorqVue® delivery sheath. The tip of the TorqVue® sheath is placed in the descending aorta, slightly lower than the PDA. A hand angiogram of 1.5–2 cc is performed in RAO and lateral projection. Other teams perform the angiogram with the same catheter used to cross the PDA [48]. Also, fluoroscopic guidance only is used in some centres [40, 42]. In our experience, most of the patients had type C (tubular) and F (foetal) PDA [43] (Figs. 35.2 and 35.3).

The appropriate device is selected based on echocardiographic and angiographic measurements (Fig. 35.4). As a rule, the shortest APO device is used to avoid protrusion of the device in aorta/pulmonary artery branches. In almost all cases, only the 2 and 4 mm length devices are used.

The device is positioned under fluoroscopy guidance with the goal to implant the entire length of the device into the PDA. Successful positioning is defined by complete occlusion of the duct and the absence of aortic or LPA obstruction by echocardiography.

Following release of the device, echocardiographic evaluation is repeated, paying also particular attention to the tricuspid valve function. Follow-up echocardiography is performed within 24 h after procedure.

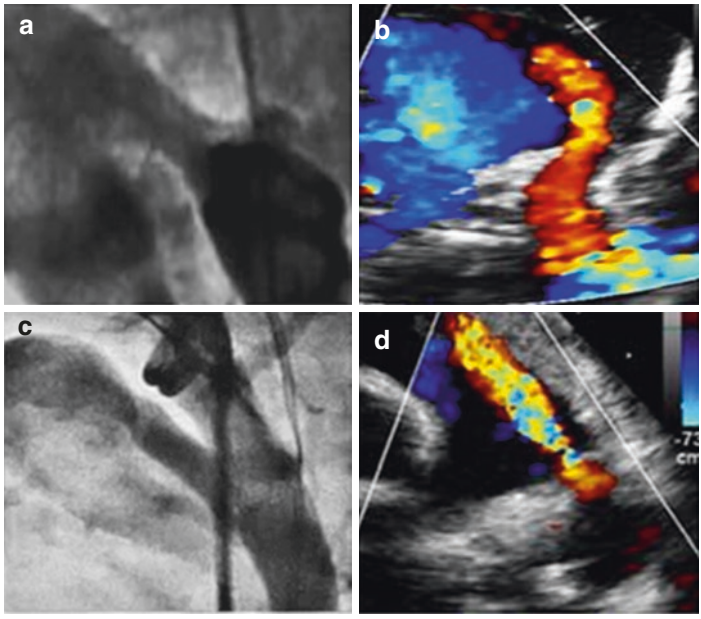

Fig. 35.2 Most common ductal morphology in premature babies undergoing percutaneous closure. Panels (**a**, **b**) showing type "C" tubular ductus without any constrictions at the aortic end or the pulmonary artery end. Panels (**c**, **d**) showing a "foetal-type" ductus, which is typically long, wide and tortuous

35.5 Complications

One of the most frequent complications is LPA obstruction due to protrusion of the device. In a 1 kg baby, the LPA diameter is approximately 3 mm, whereas the APO retention discs for the 4 and 5 mm devices are 5.25 and 6.5 mm, respectively. Despite the goal is to implant the whole device inside the duct itself, protrusion of the proximal disk may occur, especially in patients with a large duct and restriction at the pulmonary end. In cases with severe LPA stenosis (max velocity >3 m/s on CW Doppler interrogation) diagnosed before the device is released, it is recommended to reposition it or to change it for a shorter and/or a

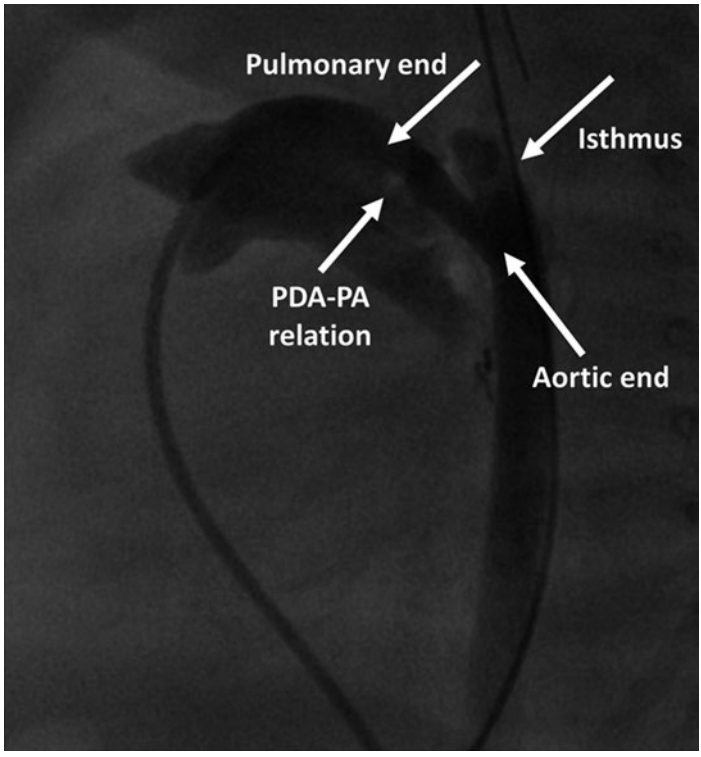

Fig. 35.3 Imaging of the duct in premature patients undergoing percutaneous closure. Angiogram performed through a catheter via RV—MPA—PDA—descending aorta. Angiogram is useful to delineate both PDA's aortic and pulmonary ends as well as to assess its relationship with pulmonary arteries and rule out aortic coarctation

smaller one. Significant LPA stenosis has been described with AVP II requiring LPA stenting [49]. When the LPA stenosis is mild, the device can sometimes be released as this will resolve in most of the cases during follow-up [38]. Finally, LPA stenosis seems less frequent with the Medtronic vascular plug because of the circular shape of the device (Fig. 35.5).

Coarctation of the aorta related to the device is an uncommon complication. It is diagnosed with TTE after device implantation

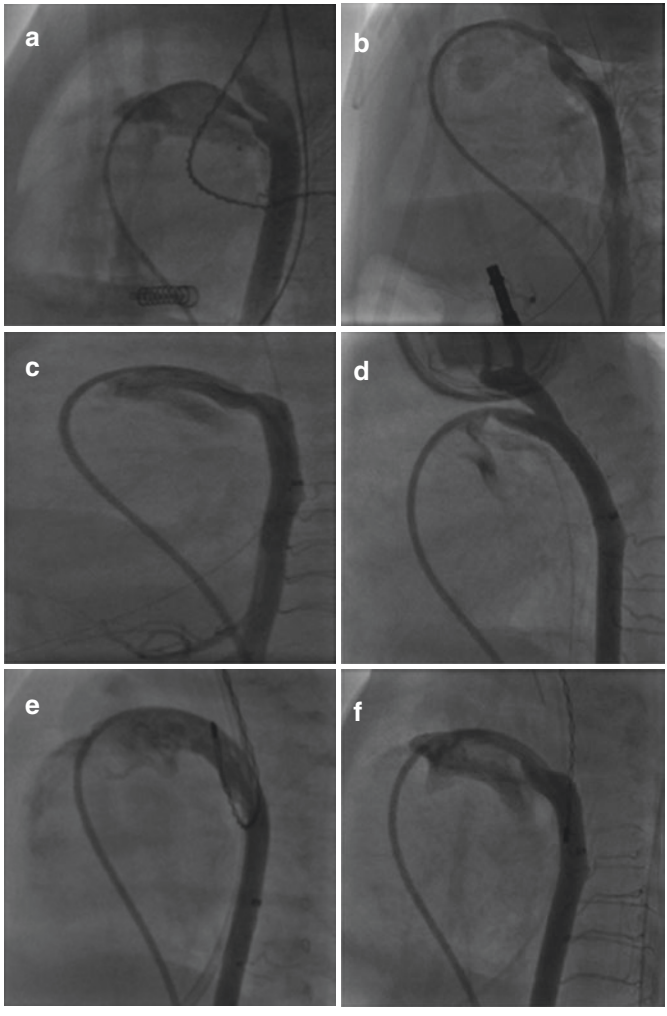

Fig. 35.4 PDA morphology and size are extremely variable in premature babies. Angiogram helps to delineate duct features and choose the most appropriate device. (**a**) Relatively long duct with mid-course constriction. (**b**) Long and tortuous duct with pulmonary end constriction. (**c**) Long and tortuous duct with widely open pulmonary and aortic ends. (**d**) Short duct with pulmonary end constriction and LPA origin stenosis. (**e**) Extremely large and dilated duct (note that its diameter is bigger than descending aorta). (**f**) Long and tortuous duct with variable diameter along its course

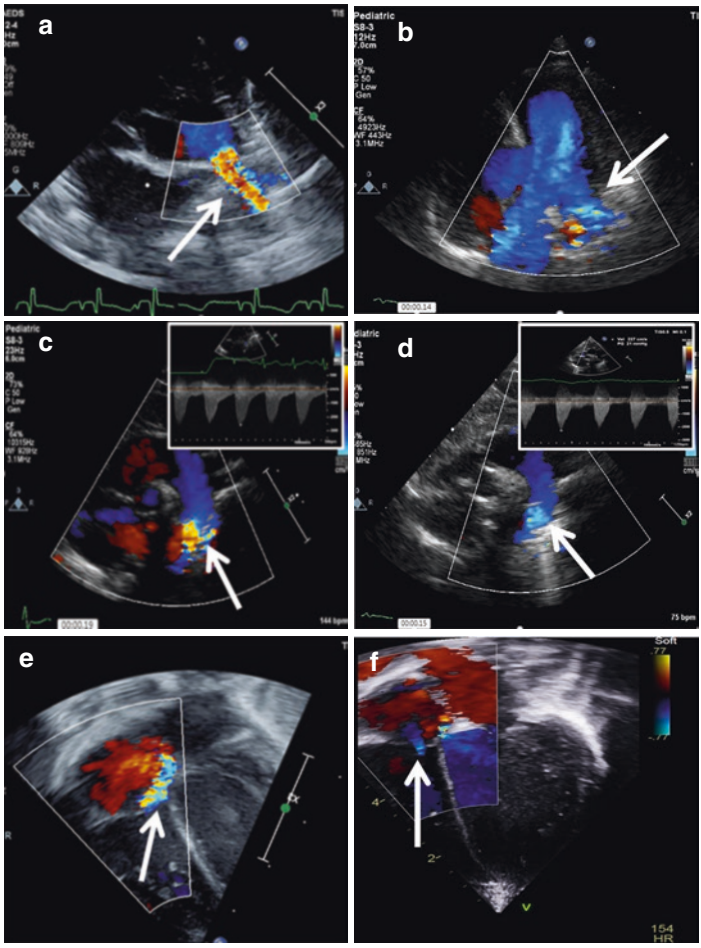

Fig. 35.5 Complications encountered after PDA device closure. Panel (**a**) shows LPA origin stenosis following percutaneous closure, which is fully resolved 5 months after follow-up (**b**). (**c**) Mild aortic coarctation after device deployment, which is fully resolved after 11 months of follow-up. (**d**) Tricuspid valve trauma noted after successful PDA device closure. Tricuspid regurgitation is trivial 12 months after the procedure

and device recapturing, and repositioning is required in such cases. In our experience, no clinical coarctation was encountered, and echocardiographic signs of coarctation disappeared few months after with growth of the aorta. However, at least one patient requested delayed surgical repair of a coarctation induced by protrusion of the device (A Baruteau, unpublished data).

Device embolisation has also been reported during, immediately after and up to 9 days after the procedure [37, 38, 50]. This seems to rather happen in patients over 1.2 kg with large PDA. The device can be retrieved percutaneously in most cases through a 4- or 5-French sheath.

Tricuspid valve traumatism may happen and is often due to chordae rupture when the tricuspid valve is crossed. Use of balloon catheter and soft wires may minimise such risk. In our experience, this has resulted in only mild tricuspid regurgitation that remained stable over time. However, long-term outcome remains unknown.

Rupture and/or IVC dissection has been reported as a fatal complication [43]. We believe that the position of the wire during the femoral vein access should be checked by fluoroscopy before advancing any sheath to minimise this potentially life-threatening complication.

Morville et al. reported fatal cardiac perforation in a 680 g patient. During the procedure, the right ventricle was perforated by a 4F catheter over a 0.018 Terumo wire, creating a hemopericardium [51].

Device-induced haemolysis has not been reported with the APO to our knowledge. Residual shunt after PDA closure usually disappears within 24 h [38, 48, 49, 51].

Low-cardiac output syndrome or heart failure (also known as post-ligation syndrome) following percutaneous closure has not been reported.

35.6 Conclusion

Percutaneous PDA closure in premature babies is safe and effective with excellent short-term outcome. The current technique and available devices apply for most of the population and may

become the first-line strategy as an alternative to surgical ligation and even to medical therapy. Further experience is needed, and multicentre studies and registries are urgently needed to better clarify the results and the outcome as well as the timing for this procedure.

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36 Catheter Closure of Coronary Artery Fistula

Kothandam Sivakumar, Ajit Mullasari, and Bharat Dalvi

36.1 Anatomic Description and Physiopathology

Coronary artery fistula (CAF) is a direct communication between one or more coronary arteries and a cardiac chamber or a great vessel bypassing the capillary network. The true incidence of CAF is unknown since most are silent and therefore undetected. The incidence of CAF is 0.3–0.8 % in patients undergoing diagnostic cardiac catheterization. Most fistulae arise from the right coronary artery (RCA), followed by the left anterior descending (LAD) and the left circumflex artery (LCx) in that order. Rarely, fistulae may arise from more than one coronary artery. Over 90 % of the CAFs drain in the right heart chambers.

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36.2 Pathophysiology

1. Shunt through the fistula: Magnitude of the shunt is determined by the size of the communicating orifice and the pressure difference between the site of origin and insertion. Therefore, those with nonrestrictive communication draining into low-pressure right atrium (RA) or the superior vena cava (SVC) will have a large shunt resulting in heart failure. On the other hand, long and tortuous fistulae with small communicating orifice draining in a high-pressure left ventricle (LV) will result in a small shunt with patients remaining asymptomatic.
2. Size and tortuosity of feeding artery: The fistula may arise from the proximal main coronary artery or one of its branches. The more proximal the origin, the more dilated it tends to be. Some of the feeders enlarge very rapidly and become aneurysmal, resulting in cardiomegaly on chest X-ray due to stretch of the pericardium over the fistula.
3. Secondary effects: Some fistulae may steal blood from the neighboring myocardium and cause coronary ischemia, while the others may compress soft cardiac structures and produce arrhythmias. Large fistulae may rarely obstruct systemic or pulmonary veins.
4. Natural history during adulthood: With the onset of atherosclerosis or due to thrombus formation within the dilated fistulous tract with distal embolization, some adults may present with angina, myocardial infarction, or a sudden cardiac death. Rupture of aneurysmal fistula and infective endarteritis have been uncommonly reported. Very rarely, spontaneous thrombosis of a slow-flowing fistula may result in its natural closure.

36.3 Clinical Scenarios

A few clinical case studies are presented to highlight the varying clinical presentation of patients with CAF in different age group and discuss the various indications for interventional treatment.

Case Study 1

A CAF arising from the left main coronary artery (LMCA) and draining into the RA was identified on a routine fetal echo performed at fifth month of gestation. During the entire pregnancy, there was no evidence of ventricular dysfunction or hydrops fetalis. Postnatally, this large fistula caused features of heart failure due to the large left-to-right shunt. The fistula was closed with two coils at 4 months of age, when the child weighed 4 kg (Fig. 36.1).

Case Study 2

A 4-month-old asymptomatic infant was incidentally found to have a murmur. His echo revealed a CAF from RCA to RA. On follow-up, there was a progressive enlargement of the fistula from 5 mm at 4 months to 11 mm at 1 year of age. The rapid enlargement of the feeding artery on echocardiography indicated closure of fistula despite absence of symptoms (Fig. 36.2).

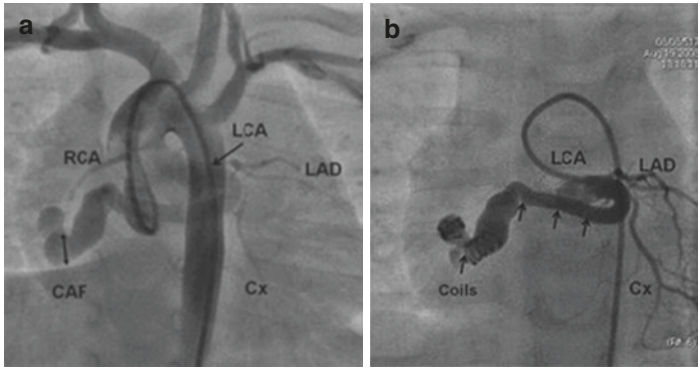

Fig. 36.1 (**a**) Aortic root angiogram in anteroposterior view shows tortuous coronary artery fistula (CAF) (*arrow*) arising from the left coronary artery (LCA) entering right atrium (RA). Normal branching of the LCA into the left anterior descending (LAD) and left circumflex (Cx) is well seen. Proximal portion of the right coronary artery (RCA) is also opacified. (**b**) It was closed with two Gianturco coils (*arrow*)

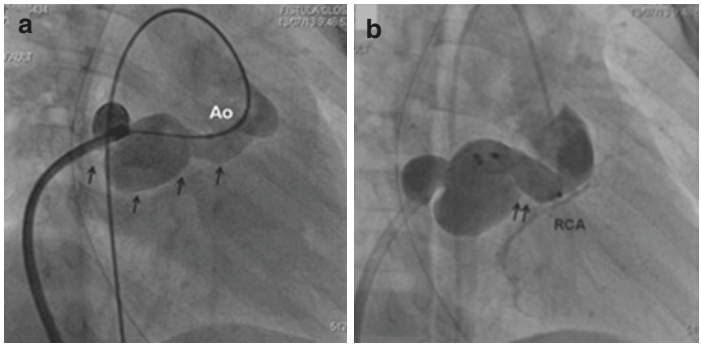

Fig. 36.2 (**a**) Angiogram through venous sheath after AV loop formation in right anterior oblique projection shows a large fistula (arrows) from the proximal part of right coronary artery (RCA) coursing posteriorly to enter the right atrium. Faint opacification of the aortic root (Ao) is also seen. The RCA branches are not seen due to high flows through the fistula. (**b**) After closure with a 14–12 duct occluder (arrows), RCA is seen well

Case Study 3

Angiogram of a 15-day-old presenting with heart failure showed a large fistula from LCA to the RA coursing posterior to the aortic root. Surgery was performed through midline sternotomy, and a large coronary fistula from the left coronary sinus behind the aortic root was identified in the transverse sinus of the heart. The fistula was clipped in its course without the use of cardiopulmonary bypass. Disappearance of the thrill was taken as confirmation of closure of the fistula and sternotomy was closed. The neonate remained ventilator dependent due to significant residual flow through the fistula. Lung infections complicated the course of the child further. Transcatheter closure of the residual shunt through the fistula was prompted by persistent heart failure, growth failure, and recurrent pneumonia warranting ventilatory support (Fig. 36.3).

Case Study 4

A 7-year-old asymptomatic child was followed up for CAF from the RCA to the right ventricle (RV). Oximetry showed Qp/Qs of 1.7:1. Magnitude of the left-to-right shunt prompted transcatheter closure of the fistula (Fig. 36.4).

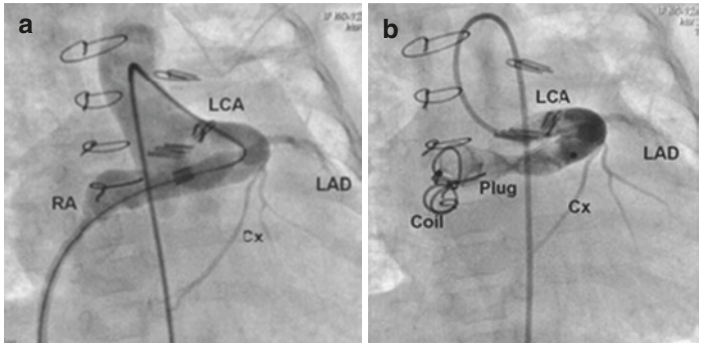

Fig. 36.3 (**a**) Angiogram through venous sheath after arteriovenous (A-V) loop formation in a patient with a significant residual flow from left coronary artery (LCA) to right atrial (RA) fistula after surgical ligation shows the long fistulous tract and normally branching LCA into the left anterior descending (LAD) and the left circumflex (Cx) arteries. Sternal wires and multiple clips placed to close the fistula surgically are seen. (**b**) Repeat angiogram after placement of Amplatzer vascular plug II through the venous sheath and an additional coil at the most distal end, shows a complete closure of the fistula with better visualization of the branches of the LCA

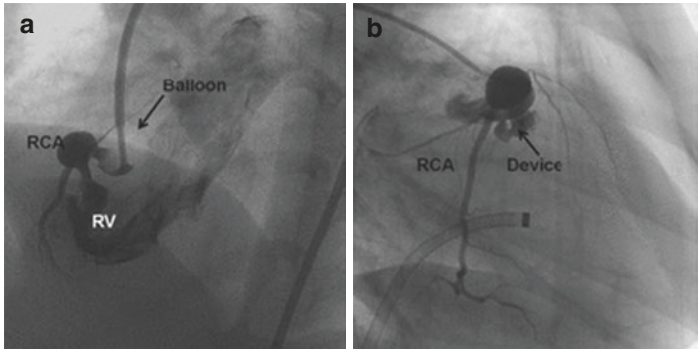

Fig. 36.4 (**a**) Aortogram in left anterior oblique view with cranial angulation using a balloon tipped catheter shows large fistula from proximal right coronary artery (RCA) to the right ventricle (RV). The branches of the RCA distal to the fistula are seen due to proximal balloon occlusion. (**b**) After closure with a duct occluder, angiogram in right anterior oblique view shows better opacification of the RCA with branches

Case Study 5

A 21-year-old asymptomatic man was identified to have a large fistula from the LCx to the ostium of coronary sinus on a preemployment medical examination. Angiogram showed a fistula arising from a markedly dilated LCx, measuring 22 mm, draining into the ostium of the coronary sinus, with a 1.9:1 shunt and mildly elevated pulmonary artery and LV filling pressures (Fig. 36.5).

Case Study 6

An 8-year-old asymptomatic child with RCA to the RV fistula had entire RCA aneurysmally dilated and measuring 12 mm from its aortic origin to the crux of the heart where the posterior descending interventricular branch (PDA) was given off. The fistula terminated immediately before the origin of the PDA. Even though the shunt was only 1.4:1, transcatheter closure was indicated by a threat of rupture of the aneurysmal RCA, which measured 12 mm (Fig. 36.6).

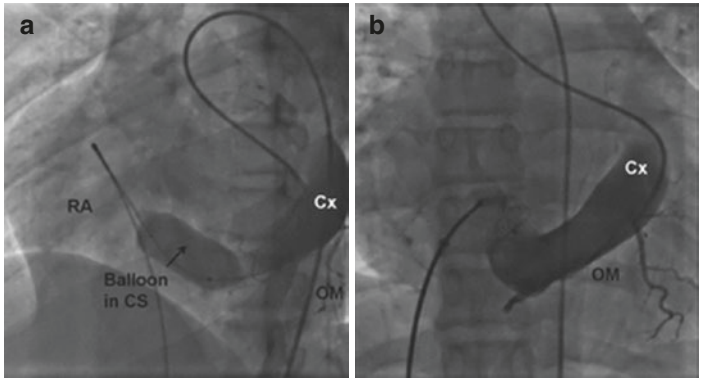

Fig. 36.5 (**a**) Selective angiogram in the left circumflex artery (Cx) after occluding the fistula opening into the right atrium (RA) by a 25 mm Tyshak II valvuloplasty balloon shows the obtuse marginal (OM) branches of the Cx. (**b**) After the distal end of fistula was closed with a large duct occluder, the contrast is seen to opacify the Cx and OM branches more intensely

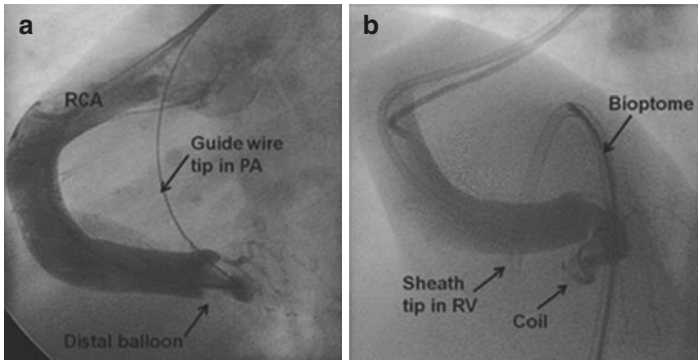

Fig. 36.6 (**a**) Selective right coronary artery (*RCA*) angiogram in left anterior oblique view shows a large RCA to right ventricle (*RV*) fistula. Through a second arterial access a guidewire was advanced through the fistula into the pulmonary artery and a distal balloon occlusion was done. The posterior descending and posterolateral branches of the RCA are better visualized only after the distal balloon occlusion. (**b**) The distal end of the fistula was closed from the venous side with bioptome assisted delivery of two intertwined embolization coils

Case Study 7

A 40-year-old man was diagnosed to have a small fistula from the atrial branch of the left circumflex artery to the right atrium during device closure of secundum atrial septal defect. After 6 years, he developed effort angina with reversible perfusion defect on myocardial perfusion sestamibi nuclear scan. The effort angina and nuclear perfusion defect were a result of myocardial steal through the fistula (Fig. 36.7).

Case Study 8

A 3-year-old asymptomatic young boy was diagnosed to have a fistula from a single left coronary artery to the right ventricle. The LAD continued beyond the apex in the posterior interventricular groove as the posterior descending artery (PDA) and subsequently coursed in the posterior right atrioventricular groove and terminated in a sac before entering the right ventricle. There was a large right-sided branch from the LAD that coursed in the right anterior interventricular groove which also terminated in the same sac

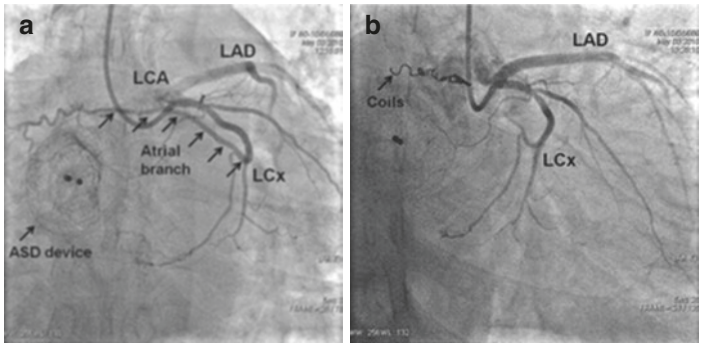

Fig. 36.7 (**a**) Left coronary artery (LCA) injection through right radial arterial access in right anterior oblique view shows a small fistula (*multiple arrows*) from the left circumflex artery (LCx) coursing posteriorly and terminating in the right atrium in a patient who had closure of atrial septal defect with an Amplatzer septal occluder (*single arrow*) earlier. The left anterior descending (LAD) artery is seen to be normal. (**b**) This fistula was closed with six micro coils (0.018" Hilal embolization coils, Cook medical) using a microcatheter through a left coronary artery guide catheter

before entering the right ventricle. The entire myocardial supply in the region of the right coronary artery in the right atrioventricular groove and the posterior interventricular groove was given off from the LAD (Fig. 36.8).

36.4 Indications

1. Heart failure and growth impairment
2. Clinical features of large left-to-right shunt
3. Enlarged heart on X-ray
4. Echocardiographic evidence of dilated left ventricle and diastolic flow reversal in aorta
5. Myocardial steal on stress ECG or myocardial perfusion scan
6. Aneurysmal fistula with risk of rupture or thrombosis
7. Progressive enlargement of fistula on serial follow-up
8. Coronary artery fistula in the setting of single coronary artery

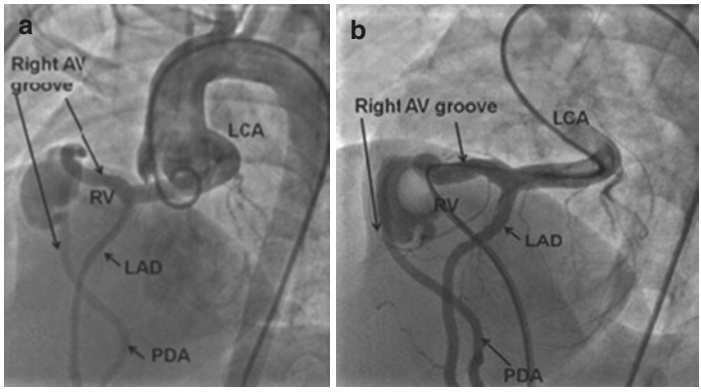

Fig. 36.8 (**a**) Aortic root angiogram in left anterior oblique projection shows absence of right coronary artery with faint opacification of the left coronary artery (LCA). A dilated left anterior descending (LAD) artery continues beyond the apex in the posterior interventricular groove as posterior descending artery (PDA) and then subsequently courses in the posterior right atrioventricular groove in region of RCA and drains finally into the right ventricle (RV). There is another anterior branch from the LAD that courses in the anterior right atrioventricular groove in the region of RCA and again enters into the fistulous sac. (**b**) In this high flow fistula, all the branches viz. left anterior descending (LAD), posterior descending (PDA) are better delineated by a selective LCA angiogram with distal occlusion done with a balloon wedge catheter from the right ventricle (RV)

36.5 Patient Selection

Age and weight: Although it can be performed at any age, it is safer in children weighing >5 kg when one needs to close the fistula from the venous end by creating an AV loop.

Symptomatic status: The patient has to be symptomatic to justify closure. In the absence of symptoms, there has to be an evidence of significant left-to-right shunt or presence of myocardial steal resulting in ischemia or any other feature (presence of aneurysmal sac, progressive enlargement of the feeding vessel, arrhythmias either at rest or exercise induced) which can result in a life-threatening complication.

Anatomy of the fistula: Side branch fistulae are safer to close because they rarely compromise blood flow through the parent coronary artery. On the other hand, distally draining fistula arising from one of the major coronary artery, if closed, has a very high risk of compromising the flow through the main coronary artery with resultant myocardial ischemia/infarction.

Multiple fistulae or a single fistula with multiple drainage sites is technically more challenging than a single fistula with one site of drainage.

36.6 Treatment Options

In asymptomatic young patients with a small left-to-right shunt, normal somatic growth, with incidental detection of CAF on echocardiogram and no progressive dilatation of the feeding vessel or the cardiac chambers can be followed up medically with yearly echocardiogram. Adults with small fistula identified incidentally on coronary angiogram with no evidence of myocardial steal can also be followed medically.

Surgical correction on cardiopulmonary bypass is indicated in young symptomatic infants or children with large complex fistula, multiple exits, and extremely sinusoidal tracts and in those who had failed transcatheter closure.

36.7 Preprocedural Imaging

Echocardiogram in young patients gives information about origin of fistula, course, and distal exit points. The quantity of shunt is assessed from left ventricular volumes and diastolic flow reversal in aortic arch.

Multidetector CT (MDCT) produces high-quality images especially in adults with ECG-gated image reconstruction algorithms. The high-resolution images from MDCT give an in-depth anatomical information about the fistula, presence of side branches proximal and distal to its drainage, and the size of the fistula at various sites (Fig. 36.9). This crucial information avoids surprises

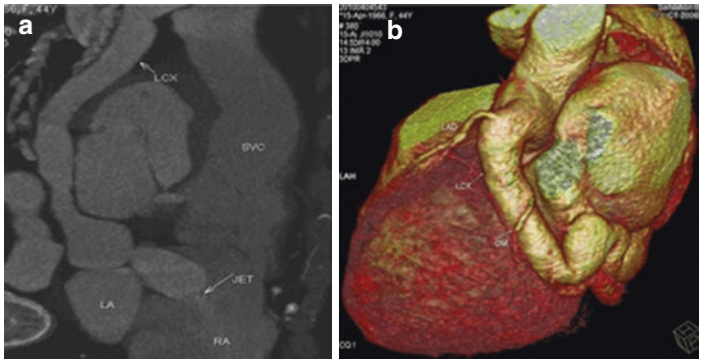

Fig. 36.9 (**a**) CT coronary angiogram showing a fistula arising from the left circumflex artery (LCx) coursing in front of the left atrium (LA) and draining into the RA just below the entry of superior vena cava (SVC). Severe restriction at the point of exit into the RA is producing a jet effect. It shows the entire fistulous tract which is dilated with two outpouchings in its course. One can appreciate the tortuosity of the tract and measure the dimensions at various sites including at the point of exit in the RA. (**b**) Three dimensional reconstruction shows the number of obtuse marginal branches arising from the dilated fistulous left circumflex artery (LCx) along the way before it opens in the RA. The left anterior descending artery (LAD) is normal in its course and caliber. These distal fistulae need to be closed at the point of exit to prevent ischemia in the region of the side branches

in the catheterization laboratory and reduces contrast volume, procedure time, and radiation dose. However, MDCT is challenging in young children due to faster heart rates, breathing, and movement artifacts and because of difficulty in tracking the contrast. It also has limitations in adults with cardiac arrhythmias.

36.8 Technique (Step by Step)

1. A resting, baseline 12-lead ECG is recorded for future comparisons. A single low-dose aspirin 5 mg/kg body weight is given on the morning of the procedure.
2. Anesthesia: General anesthesia is preferable in infants and children.

3. Vascular access: For a single, side branch fistula where a retrograde delivery of coils or device is contemplated, a single femoral arterial access is adequate. However, in adults with small fistula from proximal coronary artery to pulmonary artery amenable for closure with microcoils, radial artery access is chosen (Fig. 36.7). In cases where AV loop formation and transvenous delivery of the device are planned, it is necessary to have an additional venous access.
4. Heparin at 100 units/kg body weight is given to avoid catheter- or guidewire-induced thrombus formation and subsequent embolization into the dilated fistulous tract. Additional doses of heparin, if the procedure is prolonged, are given empirically or by assessment of activated clotting time.
5. Aortic root injection in left anterior oblique view (LAO 60° cranial 20°) as an initial projection shows both coronary arteries without overlap of branches and delineates the anatomy of the fistula.
6. Selective coronary angiogram in small infants with large run-off is better done with coronary guide catheters rather than the coronary diagnostic catheters. The coronary guide catheters have a lumen equal to or larger than 0.056″ and are capable of delivering more volume of contrast with hand injection. Since the affected coronary ostium is dilated and comes off from a dilated aortic sinus, a large guide catheter can be easily manipulated in aortic root to cannulate the coronary artery.
7. Selective angiogram helps in delineating the anatomy of the fistula better in terms of its origin, course, and site of drainage and in identifying single/multiple feeders to the fistula (Fig. 36.8).
8. In high-flow fistula, the coronary branches are delineated better by proximal or distal balloon occlusion angiogram (Figs. 36.4, 36.5, and 36.8).
9. Proximal occlusion is carried out with a balloon-tipped wedge catheter placed well within the dilated coronary ostium, preferably in the proximal coronary artery. Hand injection is made after temporary inflation of the balloon with care to avoid balloon rupture (Fig. 36.4).

10. Distal occlusion with balloon floatation wedge catheters (Figs. 36.5 and 36.8) or compliant occlusion balloons (Fig. 36.6) is carried out after forming an arteriovenous (AV) loop. The waist on the balloon gives additional information regarding the size of the distal exit. Another way of doing a distal balloon occlusion is to float a Berman angiographic catheter to the end of the fistula, inflate the balloon, and inject through the proximal holes.
11. Basic principle in choosing the site of closure is to occlude the fistula as proximally as possible to avoid a long cul-de-sac with large thrombus that may migrate proximally. So, in fistulae with no side branches, the occlusion is done very proximally (Fig. 36.2). However, in fistulae with multiple side branches up to its exit, the occlusion needs to be done at the exit point thereby protecting the flow in the side branches (Figs. 36.5 and 36.10).

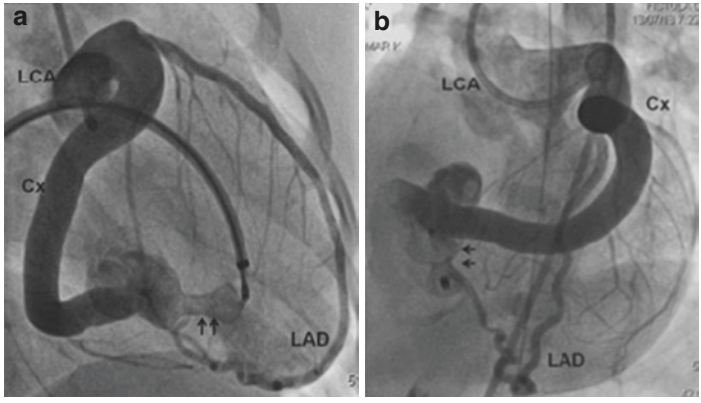

Fig. 36.10 Left coronary artery (LCA) injections in right anterior oblique projection (**a**) and left anterior oblique view (**b**) show a large feeder from the dilated left circumflex (Cx) artery and smaller additional feeder from the terminal part of the left anterior descending (LAD) artery entering the right ventricle. In this situation, occlusion of the most distal portion of the fistula sparing all the coronary branches is achieved by an Amplatzer duct occluder (*two arrows*)

12. Forming AV loop is mandatory to close a fistula distally near the exit point. This is done by advancing a guidewire from the aortic root catheter through the fistula into the cardiac chamber and snaring the guidewire from the venous end. A 0.035″ Glidewire (Terumo) is used to cross the fistula from the arterial side. If the feeding vessel is extremely tortuous, 0.014″ floppy-tip coronary guidewire (sometimes supported by microcatheters) can be used. Gooseneck snare (ev3 medical) is used to snare the guidewire. If the catheter can be pushed over the guidewire into the cardiac chamber, we exchange the Glidewire/coronary wire with a Noodle wire (St. Jude Medical) for forming the AV loop.
13. For a more proximal occlusion, an antegrade closure from an aortic end can be done with a standard-lumen coronary diagnostic catheter (with coils or AVP IV), large-lumen coronary guide catheter (for AVP I or II), or arterial long sheaths (for larger devices).
14. Prior to actual occlusion, it is good practice to occlude the fistula temporarily for 10–15 min and look for ECG changes suggestive of ischemia.
15. Aspirin is continued for at least 6 months. In addition, clopidogrel or warfarin is given if the fistula is closed distally and there is a slow flow in the fistulous tract after closure (Figs. 36.5 and 36.6).

36.9 Materials

Catheters: Judkins left and right diagnostic and guide coronary catheters in smaller curves (JL and JR2 and JL3), pigtail catheters, extra backup curve (EBU) guide catheters, and microcatheters (Cantata, Cook Medical, and Progreat, Terumo Corporation)

Guidewires: Exchange length hydrophilic 0.035″, 0.025″, and 0.018″ Glidewires (Terumo Corporation), 0.014″ floppy coronary guidewires, and Noodlewire

Occluders: MReye embolization coils, Flipper or Detach controlled release coils, Hilal and Nester 0.018″ Dacron fibered micro

platinum coils (Cook Medical), Amplatzer duct occluders I and II, and Amplatzer vascular plugs I–IV (St. Jude Medical)

Long sheaths: Flexor sheaths and Mullins sheaths (Cook Medical), and TorqVue Delivery System (St. Jude Medical)

Occlusion balloons: 6–8 French balloon floatation wedge catheters (Arrow Medical), Berman angiographic catheters (Arrow Medical), Amplatzer sizing balloon (St. Jude Medical), and Tyshak II balloons (NuMED Corporation)

Snare and other retrieval devices: Gooseneck snare (eV3 medical) and bioptome (Cook)

36.10 Tips and Tricks

1. AV loop formation: In younger patients with tortuous fistulae, 0.035″ guidewires may be stiffer and hence difficult to manipulate through the tract. In such instances, thinner 0.018″ or 0.025″ guidewires may be more easily advanced to form the AV loop. Microcatheters help in manipulating these guidewires along the tortuosities. Hypotension occurs during passage of rigid braided sheaths after AV loop formation; however, the blood pressure quickly recovers once the AV loop is broken.
2. Braided hydrophilic sheath (Flexor sheath, Cook Medical) is preferred to avoid kinks.
3. If a non-braided sheath is chosen in a tortuous fistula, the supporting guidewire is retained in place to prevent kinks and bends till the occluder is advanced through the sheath.
4. For antegrade closure from the aortic end, a diagnostic catheter is carefully advanced deep into the fistula to the selected occlusion site. Embolization coils and Amplatzer vascular plug IV can be delivered through 0.038″ lumen diagnostic catheters (Fig. 36.11). Coronary guide catheters are used for delivery of AVP I and II plugs.
5. Given the safety and efficacy of vascular plugs and nitinol occluder devices, coils are less commonly used in recent times.

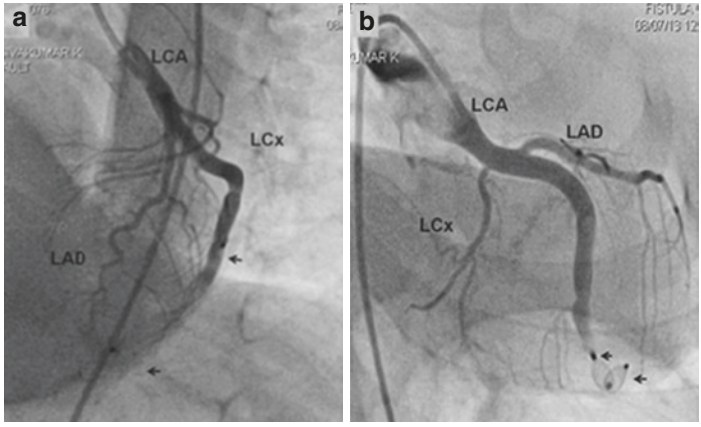

Fig. 36.11 (**a**) Left coronary artery (LCA) injection in left anterior oblique and (**b**) right anterior oblique views show a fistula from the ramus intermedius branch of the LCA arising between the left anterior descending (LAD) and the left circumflex (LCx) branches. This fistula was closed through a 0.038" lumen standard diagnostic multipurpose catheter advanced into the fistula with a very low profile Amplatzer vascular plug IV (*arrows*). The plug is placed more distally to allow flow into all its myocardial branches.

6. When multiple coils are chosen, they are intertwined together and delivered with the aid of bioptome in the desired location (Fig. 36.6). Bioptome helps in controlling the coil delivery.

36.11 Pitfalls and Complications

Minor complications include vascular access complications such as pulse loss and local hematoma, transient hypotension due to rigid guidewires and sheaths, transient arrhythmias, ST-T wave changes on the ECG, contrast allergy and contrast-induced nephropathy, and minor elevations of cardiac enzyme or troponin I levels after the procedure.

Major complications include death, myocardial infarction, left ventricular dysfunction, occlusion of coronary artery branches, marked elevations of cardiac enzymes or troponins, coronary dissection, myocardial stunning, and coronary air embolism.

36.12 How to Prevent and Manage Complications

1. Thrombus formation: Adequate heparinization with monitoring of ACT is ideal to prevent this complication. Long sheaths and large-lumen guide catheters with higher propensity to form thrombus should be flushed frequently. If, however, a thrombus is noted in the coronary arteries, the patient must be given additional dose of heparin. In addition, platelet glycoprotein IIb/IIIa receptor antagonist abciximab may be given if there is large thrombus. If there is an ST segment elevation and evidence of myocardial infarction, thrombolysis using lytic drugs is to be considered. Very rarely, if thrombotic burden is too large and myocardial ischemia has resulted in hemodynamic compromise, one may have to resort to mechanical means such as thrombosuction.
2. Air embolism: Aspiration of blood before flushing, tapping the hub during aspiration, and letting the sheath to back bleed are measures to prevent air embolism. Supportive care with fluid infusion, inotropic support, and injection of atropine to combat bradycardia may rarely be required if there is hemodynamic compromise.
3. Coronary artery dissection: Use of soft-tip guidewires, monitoring for constant free movement of guidewire tip, and avoiding use of undue force in pushing catheters prevent this complication. If the dissection is non-flow-limiting, it may be treated conservatively; otherwise, the dissection flap needs to be tacked up against the wall with the help of a stent.
4. Cardiac arrhythmias: They can be prevented by avoiding catheter wedging before coronary injections and gentle manipulation of guidewires and catheters. Most arrhythmias are transient. If there is a hemodynamic compromise, one may have to resort to cardioversion.

36.13 Post-procedural Care

Aspirin is continued for at least 6 months after the procedure. In selected patients with very large aneurysmal fistulous tracts, if they are a part of the main coronary artery, it may be continued indefinitely. In such patients, additional agents like warfarin or clopidogrel are also added.

36.14 Follow-Up

All patients should be followed up indefinitely at 6–12 monthly intervals. An ECG is recorded during each visit to look for changes of ischemia. In older children and adults, a computerized stress test is recommended every year to confirm the absence of exercise-induced ischemia. An echocardiography is done for assessing global and regional wall motion of the left ventricle, residual flows through the fistula, remodeling of the coronary arteries, and reduction in size of the cardiac chambers. In patients with very large fistulous tracts, a repeat coronary angiogram is recommended after 1 year of the procedure to study the remodeling of the tract, level of thrombus propagation, and flow in the branches of the affected coronary artery.

Further Reading

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2. Armsby LR, Keane JF, Sherwood MC, Forbess JM, Perry SB, Lock JE. Management of coronary artery fistulae. Patient selection and results of transcatheter closure. J Am Coll Cardiol. 2002;39:1026–32.
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37

Vessel Embolization: Transcatheter Embolization of Pulmonary Arteriovenous Malformations and Aortopulmonary Collateral Arteries

Liang Tang, Zhen-fei Fang, and Sheng-hua Zhou

37.1 Transcatheter Embolization of Pulmonary Arteriovenous Malformations

37.1.1 Anatomic Description and Physiopathology

Pulmonary arteriovenous malformations (PAVMs) are direct high-flow, low-resistance fistulous communications between the pulmonary arteries and veins, bypassing the normal pulmonary capillary bed and resulting in an intrapulmonary right-to-left shunt.

Most PAVMs are congenital, with 80–95% cases are associated with hereditary hemorrhagic telangiectasia.

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Acquired PAVMs are even less frequent, occurring after heart surgery (Fontan or Glenn procedure), trauma, and pulmonary operations [1].

Most PAVMs are identified in the lower lobes, with the left lower lobe being the most common location. PAVMs are usually classified into single or multiple types. Approximately, 80% of PAVMs are simple in which the feeding arteries arise from one or more branches of a single segmental pulmonary artery.

The majorities of the rest are complex PAVMs, which have multiple segmental feeding arteries from more than one pulmonary segment. A smaller percentage of PAVMs are diffuse in which there is disseminated involvement of multiple pulmonary segments. Diffuse lesions are difficult to treat with transcatheter embolotherapy and are often referred for lung transplant.

PAVMs can be further characterized according to their radiological appearance. The fistula-type PAVM has a feeding artery directly connected to a draining vein, with a venous sac.

Less commonly, PAVMs are plexiform with a multiseptated aneurysm or a cluster of vascular channels [2].

37.1.2 Clinical Manifestation

Clinical manifestation of patients with PAVM varies depending on the size, number, and flow through the PAVM.

Most PAVMs, especially if they are small ones, can be clinically silent for a long time. However, large or diffuse malformations can cause a wide spectrum of clinical manifestations including exertional dyspnea, fatigability, cyanosis, and neurologic disorders (infarcts, transient ischemic attack, and brain abscesses) secondary to paradoxical embolism as well as life-threatening hemoptysis due to sac rupture.

Because the pulmonary capillary bed is bypassed, the blood flowing through a PAVM is not oxygenated and directly drained into the pulmonary veins, resulting in systemic hypoxemia.

In addition, the absence of the normal filtering of the pulmonary capillary allows particulate matter (air bubbles or clots) to enter into the systemic circulation leading to serious neurologic complications.

37.1.3 Indications and Patient Selection

Transcatheter embolization of PAVMs is indicated for patients who have evidence of significant systemic hypoxemia or for patients at risk for or who have a documented history of a paradoxical embolic event and also for the prevention of pulmonary hemorrhage [1].

The current guidelines recommend transcatheter embolization of PAVMs for all symptomatic patients and for asymptomatic patients with discrete lesions with feeding arteries greater than 3 mm in diameter [3].

For patients with complex or diffuse PAVMs that cannot be safely or completely occluded, a reduction in the volume of right-to-left shunting by partial or staged transcatheter closure may also be indicated in order to reduce cyanosis and alleviate symptoms.

37.1.4 Treatment Options

Therapeutic options for PAVMs include transcatheter embolization with coils or Amplatzer devices and surgical excision.

Currently, transcatheter embolization has become the mainstay of treatment for PAVMs. Surgical resection is currently rarely necessary and reserved for patients who are not candidates for embolization (e.g., in patients with diffuse lesions) or when embolization fails or is unavailable.

The primary aim of embolotherapy is to eliminate or reduce the right-to-left shunting to relieve desaturation symptoms, prevent pulmonary hemorrhage, and, most importantly, prevent neurologic complications associated with paradoxical embolism.

37.1.5 Pre-procedural Imaging

37.1.5.1 Contrast-Enhanced Echocardiography

Contrast-enhanced echocardiography is useful in the assessment of PAVM since it helps to distinguish between intracardiac and extracardiac shunts. Intracardiac shunts are characterized by the

visualization of bubbles in the left heart chambers within 1–2 cardiac cycles after appearing in the right atrium. In patients with PAVMs, this event occurs after a delay of 3–8 cardiac cycles. This delay is the time needed to traverse the pulmonary capillary beds and shunt back through the pulmonary veins to the left atrium.

37.1.5.2 Chest Computed Tomography

Multidetector CT (MDCT) has been established as the primary imaging modality in the detection of PAVM. CT angiography, especially with three-dimensional reconstruction, can provide important details to inform subsequent catheter-based treatment including the location, number, and size of the arterial feeding vessels and the presence of multiple, smaller malformations.

37.1.5.3 Contrast-Enhanced Magnetic Resonance Angiography (MRA)

Contrast-enhanced magnetic resonance angiography (MRA) has high sensitivity and specificity and should be considered in young patients, where radiation exposure will be of greater concern. It is potentially able to provide precise information on the number, location, and complexity of PAVMs.

37.1.6 Technique (Step-by-Step)

37.1.6.1 Diagnostic Pulmonary Angiography

Following femoral venous access is obtained, weight-adjusted unfractionated heparin (100 U/Kg) is given intravenously.

Routine right heart catheterization is performed to assess the pulmonary artery pressure.

The initial diagnostic pulmonary angiogram is usually performed in the anteroposterior (AP) projection and ipsilateral 40° oblique (this projection places the heart over the injected lung and spreads the basal segments) using a 6-Fr pigtail catheter or other catheters.

Complete angiography in both lungs prior to any attempt at embolization is mandatory in order to identify all feeder vessels to a PAVM, their diameter, and length. This determines the occlusion strategy.

37.1.6.2 Occluding Materials

The choice of the occlusion device is primarily based on the anatomy morphology and size of the vessel as well as on the personal experience and preference of the interventionist. In general, PAVM with feeding artery diameters of less than 5 mm may be treated with coils, whereas those with diameters larger than 5 mm should be preferably treated with ADO or AVP [4].

Coils

Magnetic resonance-compatible steel or platinum pushable or detachable coils are used in the majority of cases. It is recommended to choose coils that are at least 20–30% larger than the vessel to be occluded. The main drawbacks of coil occlusion include the risk of embolization, the need for multiple coils, the potential for recanalization, and the resulting long procedure time without complete occlusion. Pictures regarding COOK coils are reported in the chapter on ductus arteriosus closure. Of note, the MicroPlex cosmos complex coils (MicroVention, Terumo, Japan) or Axium™ helical or 3D detachable coil system (ev3 Neurovascular, Irvine, CA, USA), which is primarily used for the embolization of intracranial aneurysms and arteriovenous malformations, can also be off-label used for occlusion of PAVMs. These kinds of coils as well as interlocking detachable coils (IDCs, Boston Scientific, MA, USA) are usually used for packing the venous sac.

ADO

Before the advent of AVP, the Amplatzer duct occluder (ADO) device is used for the occlusion of medium-sized to large PAVMs. Several reports in the literature have described successful transcatheter treatment of large PAVMs using the ADO device. However, their application is limited by the need for relatively large long sheaths or large guiding catheters. With the availability of various kinds of AVP, the ADO has been rarely used for PAVM occlusion. Pictures regarding ADO are reported in the chapter on ductus arteriosus closure.

AVP

The AVPs are particularly suitable for embolization of large high-flow feeding vessels. They are a woven nitinol wire cylinder that can be delivered via small catheters such as standard 5–8-Fr coronary guiding catheters (Fig. 37.1a, b). The recent developed AVP IV (Fig. 37.1c) can even be introduced through a diagnostic catheter. During embolization, at least a 30–50% oversizing of the device to the feeding vessel is recommended for the prevention of device migration and total occlusion. The only potential drawback to the AVP appears to be the relatively long length of the occluder that may limit its use if the target vessel is too short.

37.1.6.3 Techniques for Closing PAVM with Coils

To date, the most common approach to closing PAVM is embolization of the feeding artery using pushable fibered or detachable coils delivered via coaxial catheters.

Once a PAVM and its feeding arteries had been identified, selective catheterization of the target vessels is performed using a coaxial guide system with an outer 6-Fr 80 cm guide catheter and inner 5-Fr 100 cm end-hole coil delivery catheter (i.e., multipurpose catheter, Cook).

The added support provided by the guiding catheter prevents the inner-coil delivery catheter from backing out of the target vessel during embolization, thereby allowing the coils to be delivered more precisely and in a tighter mass.

The use of such a coaxial guide system also allows smaller coils to be positioned within a larger anchor or scaffold coil.

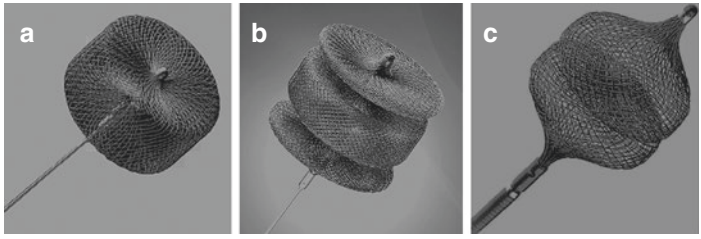

Fig. 37.1 Amplatzer plug I (**a**). Amplatzer plug II (**b**). Amplatzer plug IV (**c**)

Access to the middle and upper lobes can be challenging and is facilitated by the use of a 5-Fr Judkins left/right coronary catheter (cordis) or internal mammary catheter.

Once a feeding segmental artery is catheterized superselectively, the guiding catheter is advanced over the inner catheter to secure a stable position (i.e., placed in the parent segmental vessel), and the inner catheter is advanced into the vessel that feeds the malformation. The catheter tip is positioned beyond the origin of any proximal artery that supplies normal lung parenchyma.

Subsequently, hand-injected angiography (usually four frames) in multiple projections is performed to confirm position and define exact anatomy of the PAVM to determine site of implantation as well as size of the device to be used.

The basic principle for choosing the closure site is complete obliteration of the PAVM without compromising normal pulmonary parenchymal blood flow. Therefore, it is important to achieve as distal an embolization as possible in the feeding artery, preferably at the neck of the venous sac. This not only reduces the risk of occluding branches to normal adjacent lung, but may also reduce the likelihood of persistent perfusion of the venous sac by bronchial collaterals and of pulmonary artery recanalization. This is especially true when multiple PAVMs are present and multiple indiscriminate proximal occlusions could result in a significant reduction in pulmonary blood supply.

Coil size is an important consideration. Undersized coils are at risk to pass through the malformation and becoming an embolic agent, while oversized coils may be difficult to form a tight nest. Many interventionist empirically oversize the initial coil to the feeding vessel by at least 20%. After placement of the first coil, additional coils must be positioned until blood flow to the PAVM has ceased. In order to create a dense, cross-sectional occlusion for durable result, packing of subsequent smaller coils in the center of the first deployed coil is essential (Fig. 37.2).

The “anchor or side-branch technique” and “scaffold technique” have been documented to be very useful in achieving complete cross-sectional occlusion and avoiding paradoxical embolization of the coil via the PAVM [1].

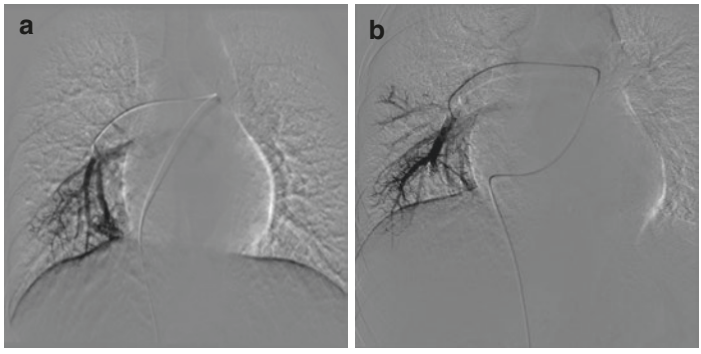

Fig. 37.2 (**a**) Pulmonary angiogram showing a PAVM of the right lower lobe. (**b**) Dense packing of two 6 mm COOK coils producing complete occlusion

The "anchor technique" is characterized by the first 2 cm of the coil and is purposely anchored in a side branch close to the aneurysmal sac, and the remainder of the coil positioned in the feeding artery and additional coils are densely packed so that cross-sectional occlusion is obtained. By securing the tip in a side branch, the risk of coil dislodgment is minimized.

The so-called scaffold technique is mainly used for high-flow vessels or when there is no anchoring vessel available. Initially, a high-radial force, fibered coil with a diameter 2 mm larger than the feeding artery is placed to create a scaffold. Then several small diameter high-radial force coils are placed as well into the endoskeleton, followed by several softer coils, until cross-sectional occlusion is obtained.

Packing of the aneurysm sac has been proposed as an alternative to feeding artery embolization when the feeding artery is too short to avoid sacrifice of large normal pulmonary artery branches or when the artery is a high-flow type with a higher risk of paradoxical embolization of coil.

Following the feeding artery that is catheterized superselectively with a 6-Fr guiding catheter, a coaxial microcatheter (i.e., a 2/2.6-Fr Excelsior microcatheter (Boston Scientific, MA, USA), 2.1/1.7-Fr Echelon™-10 microcatheter (ev3 Neurovascular, CA, USA), 2.4/1.7-Fr Headway microcatheter (MicroVention,

Terumo, Japan), etc.) is advanced coaxially through the catheter into the aneurysmal sac. After confirmation of the right position of microcatheter, venous sac embolization (VSE) with detachable coils can be performed. Several microcoils such as MicroPlex cosmos complex coils or Axium detachable coil system and IDCs are densely filled within the venous sac until a large matrix is established (Fig. 37.3). Sometimes, these detachable coils were

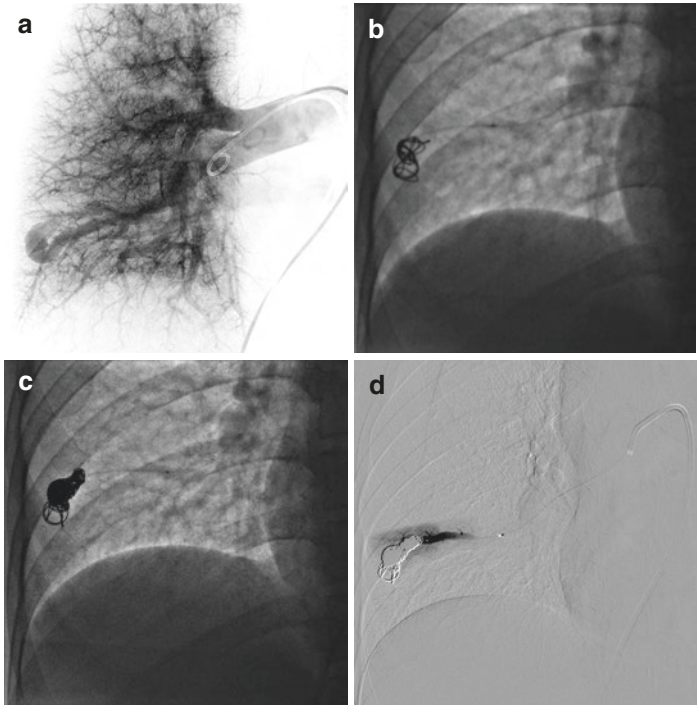

Fig. 37.3 A 49-year-old asymptomatic woman with simple-type PAVM treated by venous sac embolization. (**a**) Right pulmonary artery angiogram shows large basal PAVM of simple type with an aneurysmal venous sac. (**b**) Following an Echelon™-10 microcatheter placed in the aneurysmal sac, Axium™ helical detachable coils were sequentially deployed into the sac. (**c**) The sac was densely filled with 12 Axium helix coils, of which the largest one is 10 mm in maximum diameter. (**d**) Selective arteriogram a few minutes later demonstrates complete packing of the sac

used to form an initial wire mesh framework to prevent coil migration of the subsequent pushable small coils.

37.1.6.4 Techniques for Closing PAVM with Amplatzer Devices

For patients with PAVMs of large feeding artery or high-flow pattern, occlusion with coils is technically demanding and has a high risk of coil migration through the sac into the systemic circulation.

Alternatively, the recently developed Amplatzer vascular plug (AVP) appears to be an effective tool for embolization of PAVMs, particularly in patients with large outflow or short feeding arteries in whom embolization using coils entails a great risk of paradoxical embolization. At present, AVPs are the most commonly used devices for large PAVM occlusion.

The AVP is made from densely woven nitinol mesh wires that can be delivered via small catheters such as standard 5–8-Fr coronary guiding catheters and can be repositioned multiple times prior to its final release.

Following pulmonary artery pressure recording and angiography, the feeding artery is selectively cannulated using an appropriate-sized guiding catheter (5-Fr guiding catheter for AVPs 4–8 mm in diameter, 6-Fr for AVPs 10–12 mm in diameter, and 8-Fr for AVPs 14–16 mm in diameter).

Once a suitable position has been achieved as distally as possible within the feeding vessel and beyond any branches to normal lung, the AVP is then delivered to the target area.

The diameter of the AVP is selected to be at least 30–50% larger than the diameter of the feeding artery, according to the manufacturer's recommendation. Satisfactory positioning of the AVP is confirmed by repeat arteriography via the guiding catheter before its final detachment. If suboptimally positioned, the AVP is resheathed and redeployed in a more appropriate site. Since the AVP does not cause instantaneous thrombosis and in high-flow situations thrombosis typically takes up to 15 min, control angiography should be performed for at least 15 min after deployment of the occluder (Fig. 37.4).

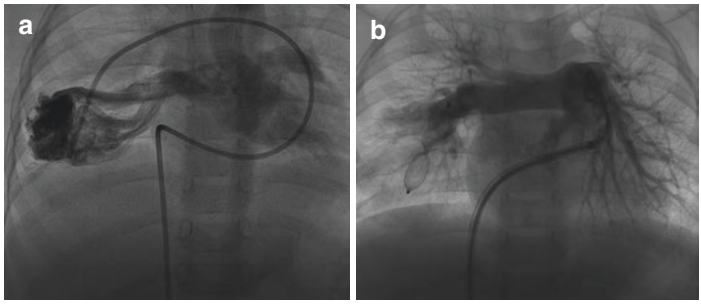

Fig. 37.4 Occlusion of a large right lower lobe PAVM with an AVP II. (**a**) Selective right lower lobe pulmonary angiogram demonstrates a large PAVM, which had a feeding vessel measuring 9 mm in diameter. (**b**) Control angiogram 10 min later after releasing a 14 mm diameter, AVP II demonstrates its optimal positioning at the neck of the PAVM and completes vessel occlusion with preservation of normal pulmonary artery branches. (This figure is provided courtesy of Prof. Yu-mei Xie, Department of Pediatric Cardiology, Guangdong Provincial People's Hospital, China)

The ADO can also be an alternative for closing large PAVM (Figs. 37.5 and 37.6). After catheterization of the feeding vessel, a long delivery sheath is introduced over a stiff exchange wire and placed in the feeding artery as close to the malformation as possible. The size of the ADO selected for embolization should be 2–4 mm larger than the caliber of the feeding vessel at site of implantation.

37.1.6.5 Techniques for Closing PAVM Using Venous Sac Embolization (VSE) in Combination with Feeding Vessel Embolization (FVE)

The combined treatment approach including the VSE using the detachable coils, followed by occlusion of the large feeding arteries using the AVP devices, has also been documented to be a highly efficient method for the treatment of the complex PAVMs with large outflow vessels or short feeding arteries. The combination of these two occlusion techniques enhances the ability of clot formation in the PAVM. The decrease of blood inflow into the aneurysmal sac due to the application of the AVP in the feeding

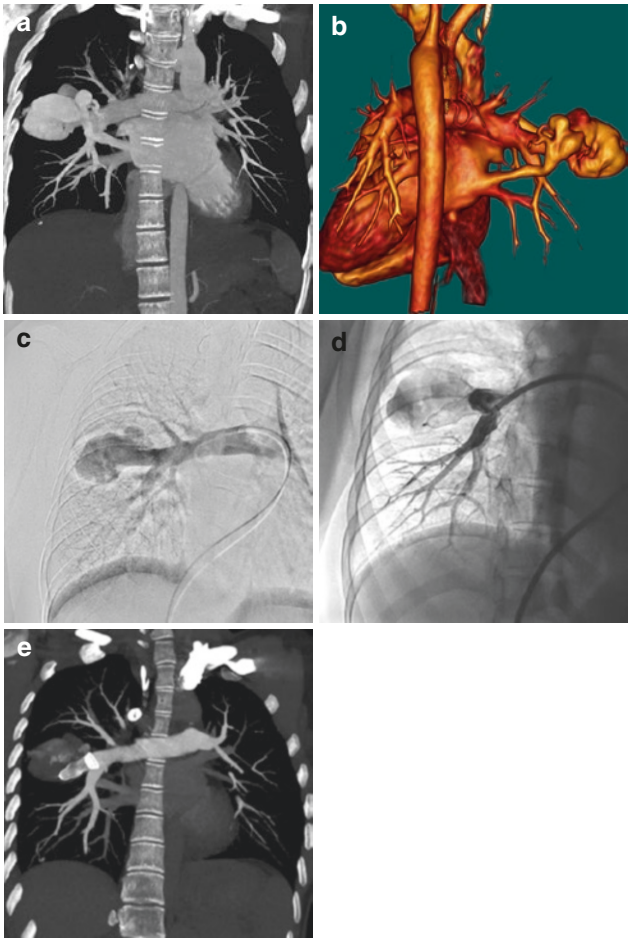

Fig. 37.5 A female patient had embolization of a right middle lobe PAVM with an ADO. (**a**) Initial three-dimensional reconstructed CT scan of the chest and (**b**) 3D volume-rendered CTA images showing a very large PAVM in the right middle lobe. (**c**) Right pulmonary artery angiogram demonstrating a large PAVM, with a feeding vessel measuring approximately 12 mm in diameter at its point of communication with the aneurysmal venous sac. (**d**) Angiogram after positioning a 16–18 mm ADO demonstrates its optimal positioning at the neck of the PAVM, and the PAVM was successfully occluded. (**e**) A follow-up CTA 1 month after embolization demonstrates the PAVM remains occluded. (This figure is provided courtesy of Prof. Yuan Feng, Department of Cardiology, West China Hospital, China)

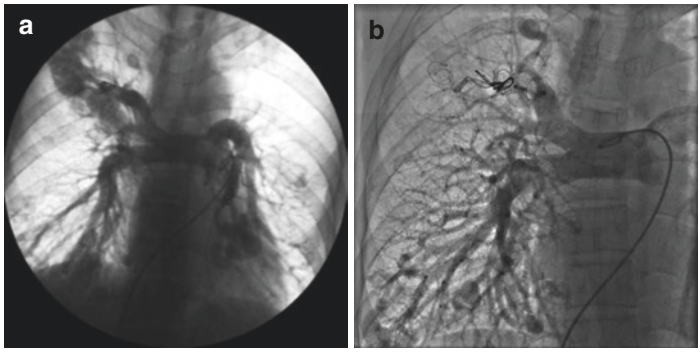

Fig. 37.6 A 29-year-old man with multiple PAVMs was referred for repeated PAVM occlusion. (**a**) Pulmonary angiogram revealed recanalization of previously embolized PAVMs. (**b**) PAVMs in the right upper and lower lobe were completely occluded with ADO

vessels increases the blood clotting ability and improves the occlusion of the aneurysmal sac. The steps of dense packing of venous sac using detachable coils combined with FVE using the AVP devices have been described above (Figs. 37.7 and 37.8).

After embolization of the feeding artery or arteries by any of the above methods, repeated segmental and lobar angiography should be performed to assess for complete occlusion and any accessory feeding vessels that may have been missed on the initial planning run and might also require embolization.

37.1.7 Expected Results

1. Pulmonary angiogram postembolization confirmed complete occlusion of the PAVMs.
2. A significant improvement in systemic arterial oxygen saturation and sustained relief of clinical symptoms attributed to the PAVMs on follow-up are obtained after embolization.
3. Contrast-enhanced CT at follow-up showed that the PAVMs remained occluded, with a significant shrinkage of the vein sac or complete resolution of the malformations.

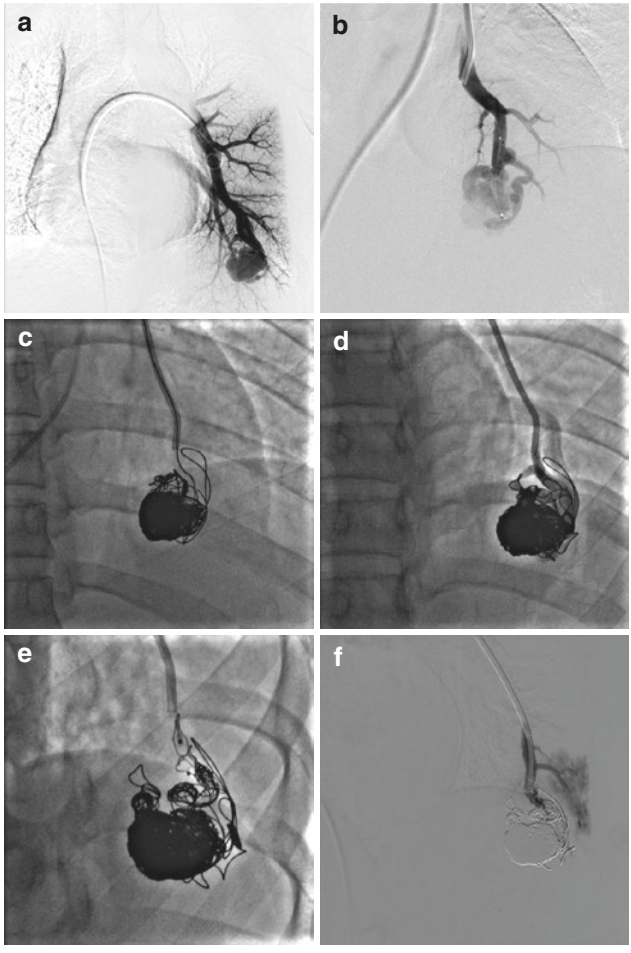

Fig. 37.7 A 25-year-old woman with hereditary hemorrhagic telangiectasia (HHT), large PAVM, and history of transient ischemic attack treated by VSE combined with FVE. (**a**) Left pulmonary arteriogram shows a complex-type PAVM with three feeding artery and a venous sac. (**b**) A 2.4/1.7 Fr Headway microcatheter was advanced coaxially through the 6-Fr MPD guiding catheter into the sac. (**c**) Twelve MicroPlex cosmos coils are densely filled within the venous sac. (**d**) There is still flow through the malformation following coil embolization of the aneurysmal sac. (**e**) A 6 mm AVP I was deployed at the neck of the PAVM. (**f**) Selective arteriogram a few minutes later demonstrates complete vessel occlusion with preservation of normal proximal pulmonary artery branches

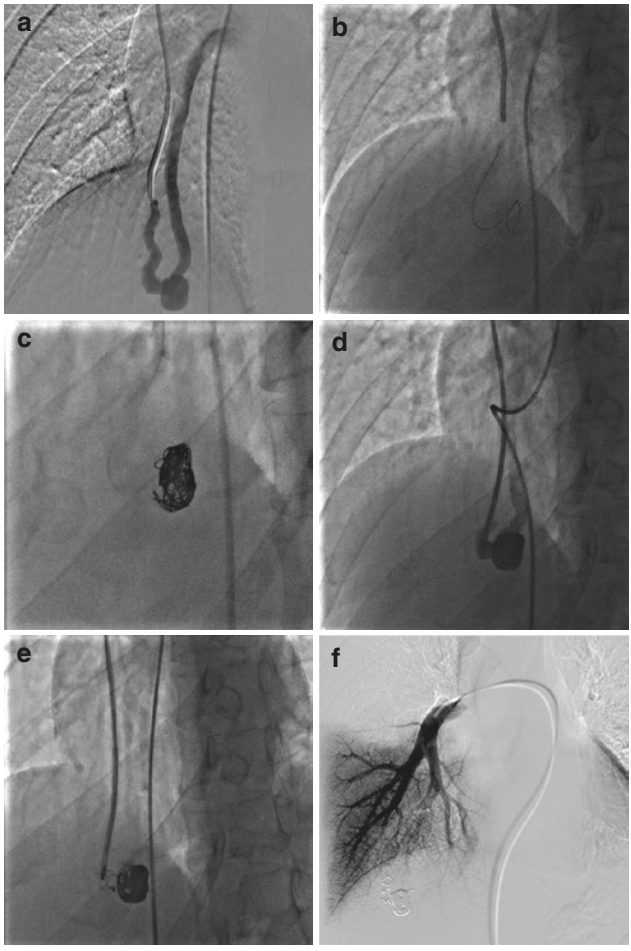

Fig. 37.8 A 35-year-old man with large bilateral PAVMs, and history of surgical resection of left lower lobe PAVM, was treated by VSE combined with FVE. (**a**) Subselective right pulmonary angiogram demonstrates a simple-type large PAVM with 7 mm feeding vessel, an aneurysmal venous sac. (**b**) A coaxial 2.4/1.7 Fr Headway microcatheter was placed in the aneurysmal sac. (**c**) The venous sac was densely packed with eight MicroPlex cosmos coils and one MWCE-35-5/6 mm coil. (**d**) Control angiogram performed 5 min later showing significant residual shunt. (**e**) A 10 mm AVP I was deployed at the neck of the PAVM. (**f**) Angiogram performed a few minutes later demonstrates complete vessel occlusion

37.1.8 Complications and Its Management

To date, there is no mortality occurred during the procedure and long-term follow-up. The complications of transcatheter embolization of PAVMs documented in the literature have been infrequent and are listed as follows.

37.1.8.1 Device Embolization

Device migration with paradoxical embolization is one of the most severe complications and occurs in 0.7–3% of treated patients, especially in cases of high-flow malformations with large outflow vessels. Paradoxical coil embolism into the cerebral artery, left popliteal artery, and left carotid artery has been reported. The choice of appropriate-sized coils is crucial to minimize the risk of coil embolization. The "anchoring" and "scaffolding" techniques are also frequently applied to overcome the problem of coil migration. In PAVMs patients with big-sized feeding arteries (>5 mm), coils probably should not be the first choice for embolization, and AVP alone or in combination with coils might be a better primary option for embolization in these patients.

37.1.8.2 Pulmonary Infarction

Pulmonary infarction has been observed in about 3% of patients. It usually occurs when the embolization causes occlusion of normal branches secondary to overly proximal positioning of embolization materials. To minimize this event, the embolization materials should be placed as close to the PAVM and as distal to normal side branches as possible.

37.1.8.3 Air Embolization

Air embolization is not infrequently encountered during the procedure. This usually occurs when a catheter or wire is withdrawn rapidly out of the sheath. When blood cannot replace the space previously occupied by the retrieved catheter, air will be suck into the delivery sheath. Air accidentally enters into the coronary arteries causing acute chest pain, bradycardia, and temporary ECG

changes. This usually resolves within l5min. A continuous flushing of catheters, observation for back-bleeding, and removal of catheters or wires "underwater" can largely prevent this complication.

37.1.8.4 Pleurisy

Pleurisy is the most frequent complication of embolization occurring in approximately 15–31% of patients. Delayed pleurisy (4–6 weeks after the procedure) with fever and infiltrates has been reported mainly with larger PAVMs. It is thought that this is due to delayed thrombosis of the aneurysmal sac and is usually self-limited and responsive to nonsteroidal anti-inflammatory drugs.

37.1.8.5 PAVM Recurrence

Recurrence of PAVMs can occur in 15% of cases, but is not considered a real complication or failure of the treatment, since it can result from recanalization of previously occluded PAVMs, collateral reperfusion about the embolized site from adjacent small pulmonary branches that have grown or developed over time, collateral flow from bronchial or other systemic arteries distal to the occlusion site, or missed accessory pathways. Risk factors for recanalization have been established, including large feeder vessel, poor coil packing, use of an insufficient number of coils or coil elongation (use of oversized coils), complex PAVMs, and embolization more than 10 mm from the venous sac. This complication can be reduced by good closure technique and by selecting appropriate occluder for large PAVM.

37.1.9 Post-procedural Care and Follow-Up

Most patients can be discharged on the next day following the procedure. For patients with large feeding arteries and received large occluder, daily oral aspirin (5 mg/kg/day) is recommended for 6 months to prevent thromboembolic complications. Prophylactic antibiotics are not routinely recommended for all treated patients. In patients with incomplete occlusion with residual shunting, physicians should be aware of the risk of mechanical

hemolytic anemia. Care should also be taken to early detect femoral thrombosis and local hematoma at puncture site.

Long-term follow-up of treated patients with imaging modality and clinical and physiologic evaluation should be performed in order to document recanalization of embolized PAVMs early, as well as to detect growth or enlargement of the untreated small lesions. It is recommended that a combination of clinical evaluation, physiologic testing, and contrast-enhanced CT scan is the best algorithm of follow-up.

37.2 Transcatheter Embolization of Aortopulmonary Collateral Arteries

37.2.1 Anatomic Description and Physiopathology

Aortopulmonary collateral arteries (APCs) can be detected in association with various congenital heart diseases (CHDs), from simple malformations to complex cyanotic CHDs such as tetralogy of Fallot, pulmonary atresia, and single ventricle with pulmonary stenosis, resulting in varying degrees of left-to-right shunting.

APCs may be masked by another predominant cardiac lesion and are not discovered until after surgical repair of the major lesions. They are often found in cyanotic CHDs patients who have undergone a Glenn shunt procedure or Fontan repair.

The APCs typically originate from the anterior wall of the descending thoracic aorta at the level of the carina. However, they can also arise from the lower descending thoracic and abdominal aorta or innominate arteries.

They frequently run a retroesophageal course. Occasionally, the collateral arteries may arise from the coronary artery.

In patients with cyanotic CHD and reduced pulmonary blood flow, the additional pulmonary blood flow provided by APCs can relieve systemic hypoxemia prior to surgical correction. However, APCs' flow can result in significant volume overloading of the left ventricle, compete with and limit blood flow via the pulmonary arteries, and increase pulmonary arterial pressure and vascular resistance during the postoperative period of corrective surgery.

37.2.2 Clinical Manifestation

Small APCs are usually clinically silent, but large or multiple APCs can result in pulmonary overperfusion and symptomatic cardiac volume overload manifested as exertional dyspnea, recurrent pleural effusion, protein-losing enteropathy, frequent lower airway infection, and hemoptysis.

37.2.3 Indications and Patient Selection

37.2.3.1 Indications

The decision to occlude APCs primarily depend on the degree of left-to-right shunting and degree of dual supply between the aortopulmonary collateral and the native pulmonary artery to the segmental branches and pulmonary vascular bed. Transcatheter occlusion of APCs is indicated for the treatment of aortopulmonary collateral vessels with documented large left-to-right shunting in biventricular or single-ventricle physiology that results in congestive heart failure, pulmonary overcirculation, and respiratory compromise, or development of pleural effusion or protein-losing enteropathy [3].

37.2.3.2 Relative Indications

1. Transcatheter occlusion of APCs may be considered in the presence of moderate-sized collaterals found in asymptomatic single-ventricle patients undergoing routine pre-Glenn or pre-Fontan cardiac catheterization.
2. Transcatheter occlusion of APCs may be considered in patients with pulmonary atresia and aortopulmonary collaterals that have adequate dual supply from native pulmonary arteries.

37.2.3.3 Contraindications

1. Transcatheter occlusion is not recommended for the presence of APCs of any size in biventricle or single-ventricle patients who have significant cyanosis due to decreased pulmonary flow.

2. Transcatheter occlusion is not recommended for patients in whom the responsible collateral arteries directly supply a large area of pulmonary parenchyma, when embolization could result in infarction of the lung parenchyma [3].

37.2.4 Treatment Options

Therapeutic options for APCs include transcatheter embolization and surgical ligation. Surgical ligation can be technically challenging because of the identification, and dissection of the APCs can be very difficult, especially when they are transdiaphragmatic, and the operative field can be flooding by APCs supplying. Transcatheter occlusion is currently the preferred method for the management of APCs. The primary goal of embolotherapy is to control excessive flow of blood to the lungs.

37.2.5 Pre-procedural Imaging

Conventional angiography remains the gold standard for morphological assessment of the APCs. Noninvasive imaging modalities such as contrast-enhanced MRA and multidetector-row computed tomography (MDCT) with three-dimensional reconstruction are also useful for the assessment of APCs. Both of them can clearly identify the number, origins, course, and diameter of the APCs.

37.2.6 Technique (Step by Step)

37.2.6.1 Aortography and Pulmonary Angiography

Access is obtained in both the femoral artery and vein. Systemic anticoagulation (heparin 100 U/Kg) is provided, and the activated clotting time is maintained within the therapeutic range. A standard right heart catheterization is performed to assess the degree of shunting and evaluate the pulmonary artery pressure. A diagnostic aortogram and pulmonary angiogram is performed with a 5-Fr pigtail catheter. The goal is to identify the anatomic

characteristics of the APCs including the number, origin, course, diameter, and flow distribution pattern of the APCs and the presence or absence of native pulmonary arterial supply in the region of the "target" vessel. Since there is considerable anatomic variation among APCs in their origins (it can arise anywhere along the aorta or its major side branches), course, and branching patterns, selective angiography at multiple sites is required to fully assess for APCs. In some circumstances, it may also be necessary to perform selective angiography of the right or left subclavian artery, and even the coronary arteries, to fully disclose the collateral arteries.

37.2.6.2 Occlusion Techniques and Devices

Currently, transcatheter occlusion of APCs is performed most commonly with detachable or undetachable coils and AVP. Device selection is made according to the angiographic features of the target vessels.

Following diagnostic aortogram, the target collateral arteries are selectively engaged using a coaxial guide system with an outer 6-Fr Judkins right guiding catheter (Cordis, USA) or Cobra catheter (Terumo, Japan) and inner 5-Fr multipurpose catheter (COOK Corp., USA). The use of coaxial catheters allows for deep coil delivery and reduces the risk of proximal coil malposition. Once a suitable position has been achieved as deeply as possible within the target vessel, the appropriate-sized coils are then delivered to the target vessel. If undetachable coils are used for occlusion, the target vessels are selectively catheterized with a 5-Fr Judkins right guide catheter or Cobra catheter through which a microcatheter is introduced. The use of a coaxial microcatheter avoids the risk that a catheter may be dislocated by tension during the advancement of microcoils and the subsequent problem of coil deployment in an inappropriate systemic artery. With desired catheter position obtained, microcoils are delivered into the target vessel by saline flush.

The AVP is particularly suited for embolization of large, short, high-flow, or tortuous collateral arteries, where coil migration is possible or multiple coils may be needed. An appropriate-sized guiding catheter or long sheath is advanced over a

hydrophilic-coated guidewire into the collateral artery as deeply as possible. The AVP is then advanced via the guiding catheter or sheath into the vessel. Hand-injected angiogram is performed 15 min later to confirm coils or AVP position within collateral arteries.

It is important to recognize that APCs may have multiple sources of arterial supply, and occlusion devices should be delivered as selectively and as deeply into the target vessel as possible to block all potential arterial supply to the final pulmonary exit point.

Herein, a few clinical cases are briefly presented to illustrate the transcatheter occlusion techniques:

Case 1 A 1-year-old boy had successfully undergone surgical correction for double-outlet right ventricle and transesophageal echocardiography-guided device closure of muscular ventricular septal defect. At surgery, there was no evidence of increased pulmonary venous return. On postoperative day 3, attempts to wean the child from mechanical ventilation after surgery were unsuccessful. The patient developed progressive congestive heart failure and need for escalating doses of inotropic support after surgery. He was referred for transcatheter occlusion of multiple APCs (Fig. 37.9).

Case 2 A 6-month-old boy who had undergone surgical correction for pulmonary atresia and repair of atrial septal defect and ventricular septal defect was referred for emergent transcatheter occlusion of APCs due to a progressive low-output syndrome and the need for escalating doses of inotropic support after operation (Fig. 37.10). Transcatheter embolization of the APC resulted in prompt improvement in patient's conditions and rapid weaning from mechanical ventilation support.

Case 3 A 4-month-old boy had undergone a successful surgical correction for T.O.F. However, the patient was failure to wean from mechanical ventilation support. The postoperative period was further complicated by airway infections and dependence on inotropic support due to postoperative left ventricular dysfunc-

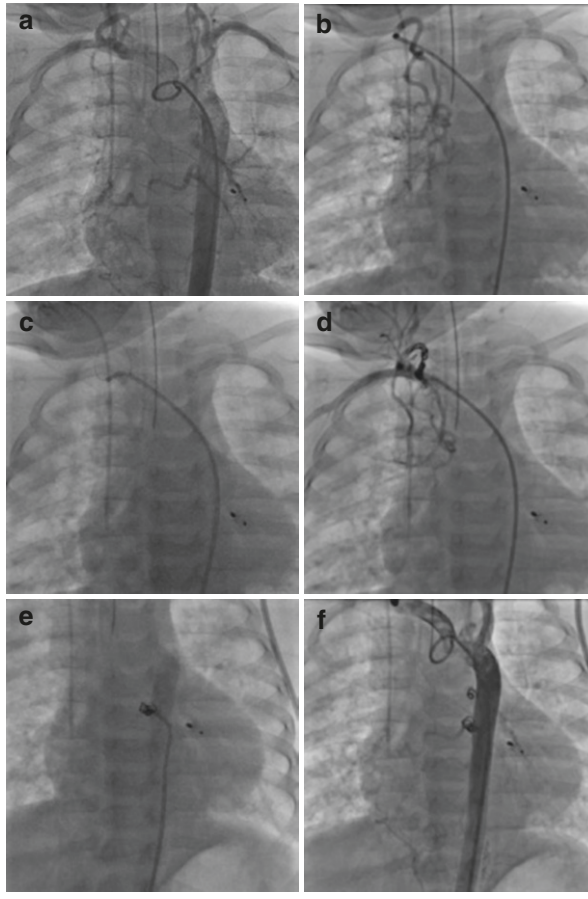

Fig. 37.9 (**a**) Ascending aortogram showing multiple APCs arising from the right subclavian artery and descending aorta. (**b**) The APC artery arising from right subclavian artery was selectively engaged by a 5-Fr Cobra catheter (Terumo, Tokyo, Japan) through which a 0.014 in. guidewire was advanced into the target vessel distally. (**c**) Over the guidewire, a 1.9-Fr, 130 cm Instant-pass microcatheter (APT Medical, Hunan, China) was introduced into the target vessel as deep as possible. (**d**) Subsequently, three MWCE-18S-5/2 mm tornado coils (COOK Medical, IN, USA) were advanced via the microcatheter by the 0.014 in. guidewire into the target vessel. The coils were packed densely at its narrowest segment. Following placement of coils, selective right subclavian artery angiogram demonstrates blood flow of the collateral artery was significantly decreased. (**e**, **f**) The remaining two APCs arising from the descending aorta were successfully treated in the similar fashion with two and one MWCE-18S-5/2 mm tornado coil, respectively

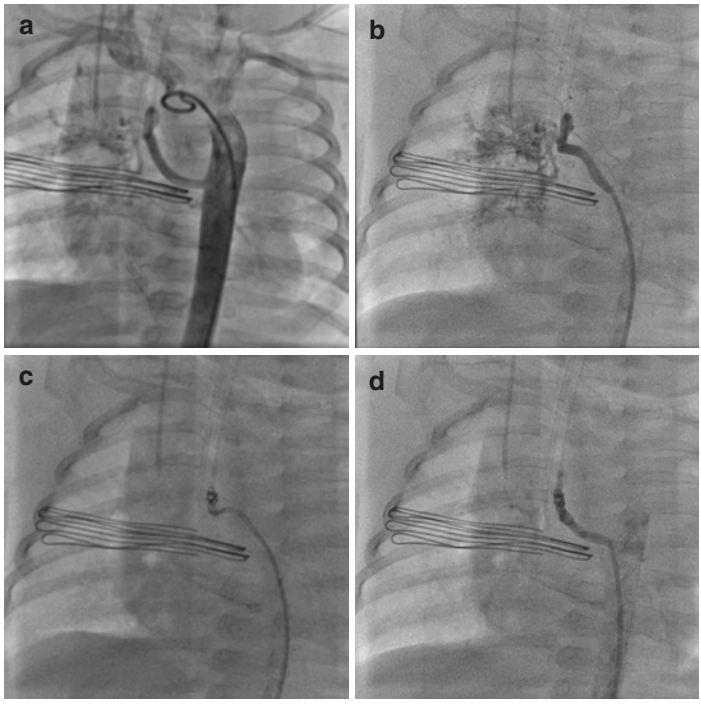

Fig. 37.10 (**a**) An angiogram in the aorta arch demonstrating a large APC artery arising from the descending aorta, which mainly supplying the right lung. (**b**) The APC artery was selectively engaged with a 5-Fr Judkins right diagnostic catheter (Cordis, FL, USA). The catheter was placed as distally as possible within the target vessel. (**c**) Two coils (MWCE-35-5/3 mm, MWCE-35-3/4 mm coils, COOK Medical, IN, USA) were sequentially advanced via the catheter by the 0.035 in. hydrophilic wire (Terumo, Tokyo, Japan) and placed at the narrowest part of the collateral artery. (**d**) Following placement of coils, selective angiogram demonstrating satisfactory occlusion of this collateral artery

tion. He was referred for transcatheter occlusion of APCs (Fig. 37.11).

Case 4 A 2-month-old boy diagnosed with ventricular septal defect and pulmonary atresia was referred for transcatheter occlusion of APCs prior to surgical correction, to avoid excessive pul-

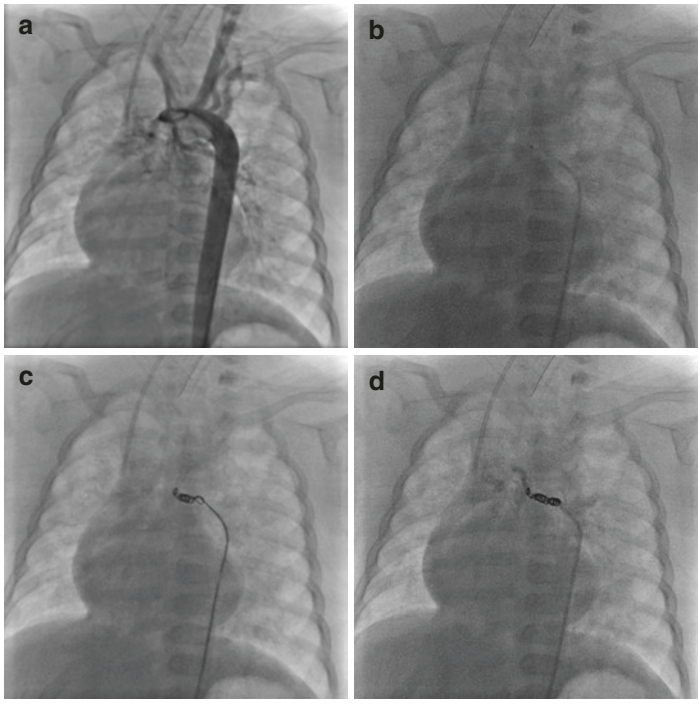

Fig. 37.11 (**a**) Angiogram in the aorta arch demonstrating a large APC artery arising from the descending aorta, which supplying bilateral lungs. (**b**) The target collateral artery was cannulated selectively using a coaxial guide system with an outer 5-Fr Cobra catheter (Terumo, Tokyo, Japan) and inner Instantpass microcatheter (APT Medical, Hunan, China). (**c**) Then, four tornado coils (MWCE-18S-4/2 mm, 18S-3/2 mm coils, COOK Medical, IN, USA) were sequentially advanced via the microcatheter by the 0.014 in. guidewire and placed at the narrowest segment of the target vessel with dense coil packing. (**d**) Selective angiogram after deployment of coils demonstrating complete occlusion of the collateral artery

monary blood return during the corrective surgery. Transcatheter embolization of a large APC using the AVP in combination with coils (Fig. 37.12). Considering the patient's condition could not tolerate long operation time, the planned procedure for occlusion of the remaining APC artery was aborted.

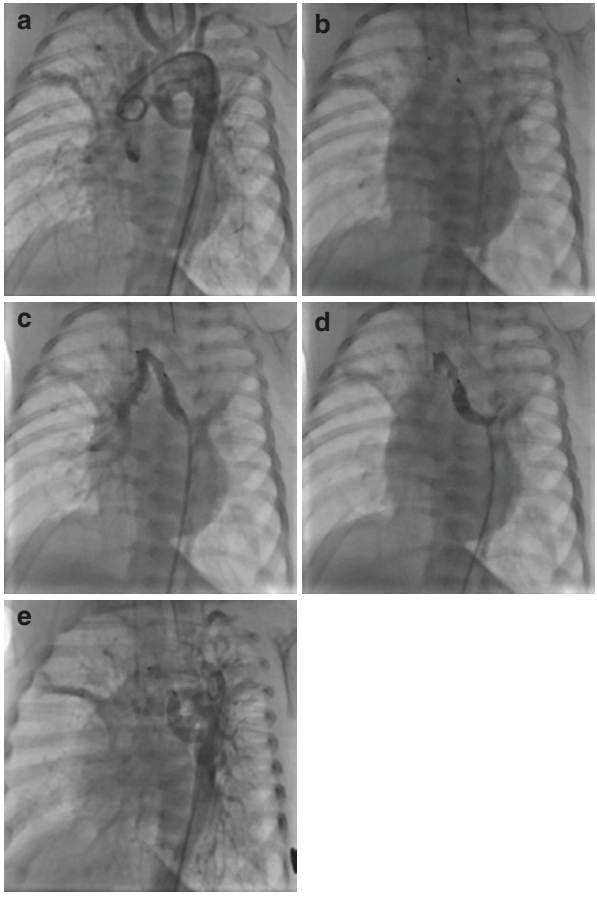

Fig. 37.12 (**a**) An angiogram in the ascending aorta demonstrating the presence of multiple APCs arising from the descending aorta, which supplying bilateral lungs. The larger one measuring approximately 4 mm in diameter at its narrowest. (**b**) The larger collateral vessel was selectively engaged with a 5-Fr Judkins right 4.0 guiding catheter (Cordis, FL, USA), and an 8 mm AVP (AGA Medical, MN, USA) was deployed at its narrowest position. (**c**) Control angiogram was performed 5 min later after the AVP deployment, demonstrating significant residual shunt. (**d**) Therefore, an addition MWCE-18S-8/5 mm Tornado coil (COOK Medical, IN, USA) was implanted, resulting in immediate, complete occlusion of this vessel. (**e**) Repeat descending aortogram performed 10 min later, demonstrating no residual shunting across this collateral vessel

37.2.7 Expected Results

1. Cardiac catheterization postembolization confirmed complete occlusion of the APCs with the pulmonary arterial pressure and oxygen saturation decreased, while the systemic pressure elevated to the normal level.
2. Significant improvement or resolution of symptoms attributed to the APCs at physician follow-up is obtained after embolization.

37.2.8 Complications and Its Management

37.2.8.1 Device Embolization

Device embolization into an important systemic artery occurs in about l% of embolization attempts, mainly with coils. It usually occurs when a coil can't be fully accommodated by the target vessel that led to coil bouncing out of the collateral during or after implantation. In order to avoid such a complication, the selected coils should be of appropriate size, and it should be placed as deeply into the target vessel as possible. When coil embolization occurs, retrieval of the coil with a snare may be considered.

37.2.8.2 Pulmonary Infarction

The complication of pulmonary infarction occurs when the APCs constitute the sole supply to the affected lung or the responsible collateral arteries supply directly a large area of pulmonary parenchyma. Prior to embolization, a careful analysis has to be made based on the collateral circulation to ensure that the collateral arteries targeted for embolization are not the sole source of flow of blood to a parenchymal segment.

37.2.8.3 Hemolysis

Hemolysis has been rarely reported with embolization of APCs. This rare complication occurs if there is significant residual shunting across the occluder. Once it occurs, the patient should be monitored and treated medically. If hemolysis is so significant that

medical treatments are not effective, the residual shunt should be eliminated by further embolization, and surgical removal is an alternative option.

37.2.9 Post-procedural Care and Follow-Up

Most patients can be discharged in a few days following the procedure. A pre-discharge imaging study including echocardiography, chest X-ray should be performed to assess the cardiac function and occluder position. Long-term follow-up of treated patients with imaging modality and clinical evaluation is recommended.

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38 Closure of Residual Postsurgical Defects

Gerrit Kaleschke
and Helmut Baumgartner

38.1 Anatomic Description and Physiopathology

Residual leaks occur in a wide anatomic variety after repair of heart defects and may not rarely be underdiagnosed, especially after complex surgery.

Residual defects after *surgical ASD closure* (direct suture or patch) are rare and have been reported in 2–7% of patients. Defects may occur at any site but are most likely posterior-inferior, where surgical closure is more demanding. Compared to native ASD, residual defects are more rigid. Again, inferior leaks are frequently complex and complicated by deficient rims making them not suitable for interventional closure. "Simple" residual ASDs result—when large enough (greater than 5–10 mm)—in significant left-to-right shunt and right ventricular volume overload. Inferior residual defects may cause right-to-left shunt in the

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presence of normal hemodynamics due to anatomic features. These again are in general not suitable for interventional closure.

Residual VSDs have been observed in up to 25% of patients and may be caused by ruptured sutures, patch dislodgement, or primarily incomplete closure of the defect(s). Many of these defects are restrictive, cause only insignificant shunt, and do not require treatment. The rarely encountered larger defects will cause significant left-to-right shunt with left ventricular volume overload and eventually pulmonary hypertension up to the development of Eisenmenger physiology if large enough and untreated for a long time. Recurrent surgical therapy may be associated with increased risk and is not always successful. In general, residual defects along the margin of patches used to close VSDs in the membranous or outflow portions of the septum are suitable for transcatheter closure by their size, shape, and location. However, proximity to heart valves has to be assessed with care and may cause unsuitability for catheter intervention. Residual muscular VSDs may be complex in shape and more difficult to cross, but the availability of a wide variety of devices may frequently facilitate effective interventional closure. This is also true for patients with *muscular defects* after myectomy in HOCM or after aortic valve surgery (Fig. 38.1). Another example of a young patient after surgical resection of a subvalvular aortic stenosis and mechanical aortic valve replacement is shown in Fig. 38.2.

Baffle leaks may be present after repair of *anomalous pulmonary venous drainage* and atrial switch operation. While obstruction of systemic or pulmonary venous return after the *Mustard or Senning operation* has been reported in up to 16% of patients, *baffle leaks* in this setting were found in approximately 10%. Both lesions may be present in combination [1]. Shunt direction and shunt portion depend on defect size and hemodynamics determined by ventricular filling characteristics and associated pathologies. Additional obstructions have particular impact and may cause right-to-left shunt. These patients are at risk for *paradoxical embolic events* or may be cyanotic and present with secondary *erythrocytosis*, but the latter findings are rare. Given the increased risk of thromboembolism after permanent pacemaker implantation, every patient after atrial switch operation should meticu-

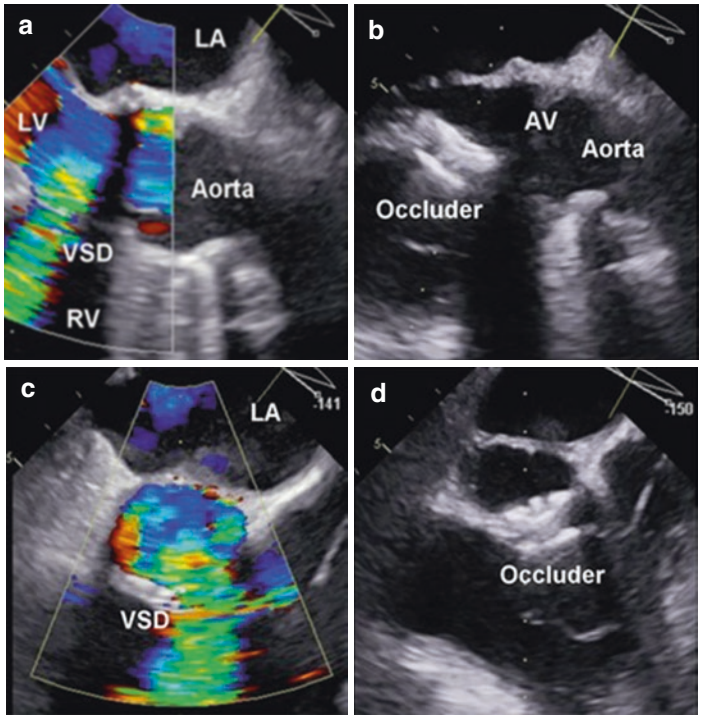

Fig. 38.1 Example of postsurgical muscular VSD closure. Transesophageal echocardiography of a postsurgical muscular VSD in a 73-year-old man after recurrent aortic valve surgery and myectomy (Qp/Qs ratio 2, evidence of pulmonary hypertension, (**a**) color Doppler long axis and (**c**) short-axis view visualizing the defect). A maximum defect diameter of 7 mm was measured by echo and angiography, and direct retrograde closure could be achieved with an Amplatzer™ Muscular VSD Occluder 10 mm without residual shunting (**b**: long-axis view, **d**: short-axis view with implanted occluder). *LA* left atrium, *LV* left ventricle, *VSD* ventricular septal defect, *RV* right ventricle, *AV* aortic valve

lously be examined with echocardiography and catheterization because smaller leakages may easily be missed. Defects can virtually occur at all sites of central venous return [2] but are more often located at superior or inferior caval connection [3]. They can, however, also be found in the central part of the baffle, where

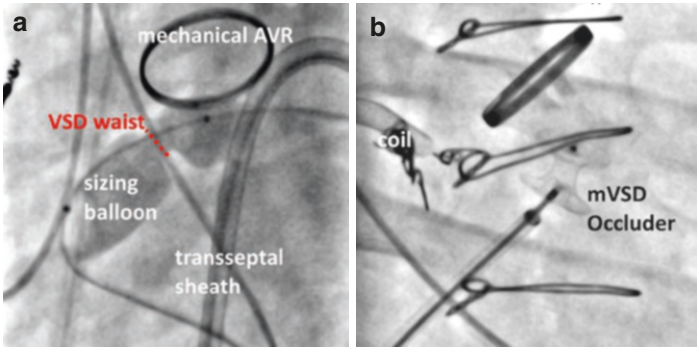

Fig. 38.2 Pat. with mechanical aortic valve and two VSDs after correction of an aortic and subaortic stenosis with relevant left-to-right shunt (Qp:Qs 3.1). After transseptal puncture, a steerable sheath, a judkins right coronary catheter and a Terumo wire was used to probe the VSD from left ventricular side. An arteriovenous loop was established and a soft balloon (Tyshak II) used for sizing of the defect (**a**). Due to shadowing induced by the mechanical aortic valve, ultrasound could not be used for proper sizing. The smallest diameter was 5 mm, VSD closure was performed with an Amplatzer™ 6 mm *muscular VSD occluder* from the right ventricular side (**b**). The second VSD was closed in an additional session. Note the proximity of the mechanical aortic valve, which was not compromised by the occluder

posteriorly the pulmonary veins may be accessible and anteriorly the adjacency to the AV valves has to be considered. Reoperation is associated with higher risk and mortality; hence, interventional therapy advanced to primary treatment option in most cases. *Residual patent ductus arteriosus* (*PDAs*) after surgical closure carry the risk of endocarditis and hemolysis as well as hemodynamic sequela (volume overload, pulmonary hypertension).

38.2 Clinical Scenario

Residual defects may be diagnosed in asymptomatic patients during routine follow-up visits. Patients present themselves with symptoms such as reduced exercise capacity, shortness of breath, and arrhythmias in case of significant left-to-right shunt with ventricular volume overload and eventually pulmonary hypertension. Symptoms at late stages may include signs of right heart failure.

Patients with right-to-left shunt may present with cyanosis and/or paradoxical embolism. Brain infarcts and abscesses should trigger the search for such residual defects.

38.3 Indications and Patient Selection for Defect Closure

- Patients with symptoms related to residual shunt.
- Asymptomatic patients with significant left-to-right shunt defined by signs of volume overload with enlargement of the ventricles (LV enlargement in defects on ventricular level, RV enlargement in defects on atrial level) or shunt ratio (Qp/Qs) >1.5.
- Asymptomatic patients with elevated pulmonary pressure (see Chap. 34 for specific considerations when severe pulmonary hypertension precludes defect closure).
- Otherwise unexplained stroke or other systemic embolism, likely due to paradoxical embolism.
- Cyanosis not caused by pulmonary hypertension (residual ASDs with specific anatomic features causing right-to-left shunt, baffle leaks in combination with baffle obstruction).
- Baffle leaks in patients with indication for *pacemaker implantation.*

38.4 Treatment Options

For most residual leaks after surgical ASD or VSD closure, self-centering double-disk devices or their derivatives (e.g., Amplatzer™, St. Jude Medical Inc. MN, USA) are suitable. For residual ASDs, ASD occluders will be the devices of choice, while VSD and PDA occluders as well as vascular plugs may be chosen for residual VSDs depending on the specific anatomy. In some cases with long or tortuous tunnels or aneurysm formation after VSD closure, nitinol spiral systems may have an advantage over the more rigid meshed nitinol devices (e.g., Nit-Occlud®, pfm medical ag Köln,

Germany). Multiple baffle leaks or leaks with *concomitant obstructive lesions* can be treated with balloon-mounted covered stents such as covered CP Stents™ (NuMed Inc. NY, USA) or self-expanding covered stents (e.g. Gore® Excluder ®).

38.5 Pre-procedural Imaging

Most appropriate information can be gained from *transesophageal echocardiography*. In residual ASDs native size of the defects, rims, and proximity to atrial wall, veins, and valves can easily be assessed. Residual VSDs should be addressed in terms of tunnel configuration (e.g., funnel-shaped), maximum diameter on left/right ventricular side, distance to the valves, and accessibility of the defect from right/left ventricle. Due to the limitations of TOE in posterior/inferior defects of the atrial septum, intracardiac echocardiography is a valuable addition for detection of residual defects (see also chapter "intracardiac echocardiography").

The arcuated course of baffles accounts for difficulties in uncovering and defining the location of leaks, which can also be missed by angiography or MRI. Color Doppler may detect even small defects and is more sensitive than angiography. 3D echocardiography may help to understand the orientation of the defect because this can be—as mentioned before—very variable. In some patients, bubble studies help to understand the course of shunt defects and shunt direction. MRI and MSCT may be particularly helpful for the evaluation of baffle anatomy and venous connections. MRI allows calculation of ventricular volume overload and shunt flow, but also multiplanar reconstruction if a 3D volume acquisition can be performed.

38.6 Technique (Step-by-Step) and Materials

Setting If prior diagnostics or pathophysiology proposes a *TEE-guided procedure* (especially in complex anatomic situations), then general anesthesia or deep sedation is recommended in most cases. Furthermore, complex defect closure with multiple

puncture sites can be time-consuming and exhausting for the awake patient. Biplane fluoroscopy reduces the amount of contrast medium and facilitates orientation on surrounding anatomic structures (e.g., ribs, vertebra). In selected cases, intracardiac echocardiography may replace the transesophageal approach, which supersedes general anesthesia/sedation.

Medication If device implantation is planned, pretreatment with aspirin and prophylactic administration of antibiotics (e.g., cefazolin) are generally recommended. Furthermore, heparin is administered (70–100 U/kg, ACT 200–250 s) during the procedure.

Access and Crossing the Defect Vascular access is mostly obtained from femoral arteries/veins but also jugular veins.

Postsurgical ASD and VSD closure follows the same principles as they are also described in the corresponding chapters for native defects. This includes for VSD closure arteriovenous loops whenever needed, and direct retrograde approach from the aorta is not feasible (depends on occluder type and size, sheath length, and accessibility of the defect). Although superior baffle leaks may be easily approached by *jugular access*, the angle of attack to the defect sometimes requires access from the femoral vein. Baffle leak crossing can be managed with a right coronary Judkins catheter whenever a rectangular approach is needed; if the defect is positioned more in line with caval veins, a multipurpose catheter is preferable. A hydrophilic j-tipped guidewire facilitates probing the leaks. In order to achieve a stable wire position that can be maintained during balloon testing of the defect and advancing the delivery sheath, the exchange to a stiff guidewire (e.g., Amplatz Extra Stiff, Cook Medical, IN, USA) is recommended. When balloon sizing is neccessary, the correct guidewire profile (e.g. 0.025" or 0.035") matching to the balloon has to be considered. Some smaller sized semi-compliant balloons can only be used with 0.025" wires (e.g. Tyshak®, NuMED Inc. USA).

Defect Sizing, Positioning, and Prevention of Complications Balloon sizing of residual leaks provides information on stretchability and reveals the shape and diameter of the defects more accurately than echocardiographic measurements alone and should include

biplane view if possible because tears of the suture lines may result in slit-shaped (and not circular) defects. In such defects delivering a self-centering device with a circular waist can result in a "mushroomed" conformation of the occluder, an Amplatzer™ cribriform septal or PFO-occluder may be a better choice. Furthermore, the radial strength of the device can lead to further disruption of the sutures with subsequent occluder dislodgment after implantation. In circular defects, the waist of the sizing balloon can directly obtain the optimal occluder size (e.g., Amplatzer™ septal occluder, which was also mostly used for baffle leak closure). Oversizing should be avoided in this context. Before releasing, the device potential complications have to be excluded. Obstruction of the systemic or *pulmonary venous return* must be avoided by optimal device sizing and positioning. This has to be checked meticulously before device release by echocardiography and angiography. Increase in wedge pressure (compared to the contralateral side) should be ruled out (Fig. 38.3). When covered stents are used for the treatment of leaks (especially in combination with baffle stenosis), balloon interrogation of the area with special regard to potential obstruction of the pulmonary venous return should be considered. Interference of the device with surrounding structures, mainly valves, must be excluded. Stent positioning should be guided by biplane angiography and adoption to fluoroscopic landmarks that have been acquired during diagnostic part of the procedure.

Multiple Devices If there are two or more adjacent leaks that necessitate the implantation of two devices, the smaller occluder should be placed first and initially being screwed upon the delivery cable, until the second occluder is in place. By doing so, correction of position remains possible and optimal overlap of the disks can be achieved.

The same principles also apply for the intervention of postsurgical ASD and VSD. In the latter, device type and size selection is even more difficult, despite the diversity of the available products. Defect size should be determined by angiography (LAO cranial projection), by contrast injection in the LVOT using a pigtail catheter, and/or by injection via the delivery sheath already in place. If

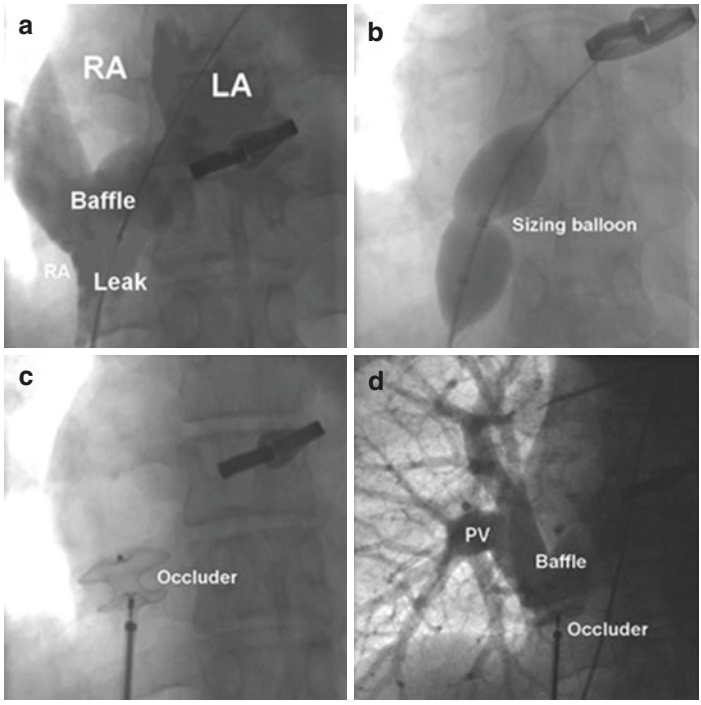

Fig. 38.3 Example of baffle leak closure. A 45-year-old woman with correction of scimitar syndrome at the age of 10 years, recurrent embolic events (TIA, myocardial infarction without evidence of coronary artery disease, meningitis). Diagnosis of baffle leak based on contrast echo was confirmed during catheterization (**a**), balloon sizing of the leak (**b**), effective closure with Amplatzer™ 11 mm ASD device (**c**), pulmonary venous angiography rules out obstruction (**d**), no residual shunting and no further clinical events. *LA* left atrium, *RA* right atrium, *PV* pulmonary vein

the defect is stretched by the delivery sheath, additional angiography is useful to reassess the size. 3D TEE helps to depict the shape and orifice proximity to surrounding structures.

In circumscription postsurgical muscular VSDs with distance to the aortic valve, the Amplatzer™ Muscular VSD Occluder provides good closure rates and should be sized 2–3 mm larger than the defect. Perimembranous defects are located in the left

ventricular outflow tract and entail proximity to the aortic valve; furthermore, closure comprises the risk of total heart block. Attempts to develop a modified device (lower clamp force, softer outer waist, larger wing span to enhance stabilization of the occlude) have not yet led to marked release. Device size should be 1–2 mm larger than the defect, but orientation of the eccentric device can be cumbersome. Aneurysmatic defects are challenging, and different orifice diameters on left/right ventricular side hamper distinct choice of device size and may lead to squeezed conformation with *outflow tract obstruction* or *valve dysfunction*. When closing perimembranous VSDs using an Amplatzer ™ Duct Occluder ADO-I [4], it has to be considered that the cable is screwed to the right side of the occluder. In most cases, probing the VSD from the left side is easier, which means that creating an arteriovenous loop is necessary (device size +2 mm of smallest defect diameter). After introducing the sheath from venous side, it has to be positioned through the defect in the ascending aorta. While gently pulling back to the defect, one has to take care not to damage the aortic valve after partial release of the device. Sequential aortogram and ventriculogram before releasing help to rule out relevant residual shunt and device malpositioning.

Beside the Amplatzer™ VSD devices, the Duct Occluder 2 (ADO-II) is a well described and frequently used (off-label) option due to its softness and flexibility with low risk of heart block. It can also be used in postsurgical Gerbode defects (between LVOT and RA), but there is a limitation in the available sizes (not usable in defects greater than 6 mm or large aneurysm). The ADO-II can be delivered from both arterial and venous side (mostly after retrograde crossing from the arterial side), where one disk is positioned on the right side and central waist/second disk of the left side of the defect. Device waist size is usually 1 mm greater than smallest defect diameter [5].

Residual PDA closure follows mainly the same principles of a native PDA intervention (addressed in the chapter “patent ductus arteriosus closure”), but coils are more frequently used in this situation.

38.7 Expected Results

Data about results and closure rates of interventional therapy in postsurgical ASD or VSD defects are sparse, and retrospective analysis of patient cohorts included native defects and/or referred to outdated occluder types. Otherwise, there are only case reports and small series. Overall, procedural success is high with an acceptable rate of complications. This also applies for interventional baffle leak closure, where atypical occluder positions are common. Small postinterventional leaks may disappear over time. In conclusion, device therapy is, therefore, well accepted as treatment of choice to overcome the disadvantages of reoperations.

38.8 Complications and How to Manage

Occluder therapy always implies the risk of laceration of surrounding structures. If the margin of the defects is close to free wall of the atria, sufficient distance of the disks must be ensured. The same is true for stents with low flexibility and sharp edges. If device occlusion is considered adequate, *rims* to the tricuspid valve and pulmonary and caval veins must be reassured to avoid the complication of obstruction and valve malfunction. Hemolysis may occur after device implantation, mostly after incomplete VSD closure. In severe cases, the device has to be removed surgically with concomitant closure of the defect. The primary goal to avoid hemolysis must be a complete as possible shunt closure. ECG telemetry in patients at risk for AV block (mainly VSD closure) is mandatory, especially when big devices or models prone to induce AV block (pmVSD) were used. Device embolization may eventually happen, which is the rationale to balloon-size the defects before closure whenever possible. Snaring of the device is feasible in most cases either from venous side if the device embolized to the pulmonary artery or otherwise from the arterial side. Pericardial effusion may occur after every complex cardiac intervention and should always be ruled out by echocardiography during or after the procedure.

38.9 Post-procedural Care and Follow-Up

Routine echocardiography is able to document correct positioning of the devices in most cases and should be performed directly after the procedure, being repeated within the first 2 days after the procedure and at follow-up visits. Pericardial effusion must be ruled out. Response of the ventricles and pulmonary circulation to defect closure can be evaluated. *Contrast echocardiography* is helpful for the detection of residual shunts and should be repeated during follow-up if incomplete closure is documented. Especially after VSD closure, ECG must be used to detect potential alterations of the conduction system. Atrial arrhythmias can occur after ASD and baffle leak closure. In the rare case of atrial fibrillation, anticoagulation and further antiarrhythmic treatment may be required. After implantation of occluders or stents, antithrombotic therapy with ASA is recommended for at least 6 months; we mostly add Clopidogrel for 3 months whenever the implanted material is exposed to systemic bloodstream.

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39 ASD Closure in Special Situations: Elderly, PA-IVS

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Transcatheter closure is nowadays considered as the first-choice treatment of atrial septal defect (ASD). However, indication, technique, and results of this approach are still challenging and under debate in particular settings, as in elderly or in patients with pulmonary atresia with intact ventricular septum (PA-IVS) previously submitted to right ventricular decompression.

39.1 ASD Closure in Elderly

Over time, ASD tends to progressively increase its size and hemodynamic burden due to the pathophysiologic changes that ensue from aging. Progressive enlargement of ASD from pediatric age to adulthood could be hypothesized by analyzing the mean size of the closing devices used in pediatric patients vs. adult ones. In addition, the physiologic decrease of left ventricular (LV) compli-

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ance due to aging and/or associated chronic diseases, such as aortic valve sclerosis, essential arterial hypertension, coronary artery disease, tends to increase the left-to-right atrial pressure gradient and hence the total atrial shunt. Overall, these anatomic and pathophysiologic changes result in always greater increase of pulmonary over-circulation and progressive LV "deconditioning" caused by a preload decrease. These changes tend to increase the risk of atrial arrhythmias and cause a progressive decrease of the systemic cardiac output, eventually resulting in worsening of effort tolerance of this subset of patients. Thus, treatment of this malformation should always be indicated regardless of age, provided its hemodynamic feasibility.

Nowadays, transcatheter approach is worldwide deemed the first-line treatment of this cardiac malformation, and in addition, it allows to "test" the pathophysiologic consequences of defect closure by temporary balloon test occlusion (Fig. 39.1). In fact,

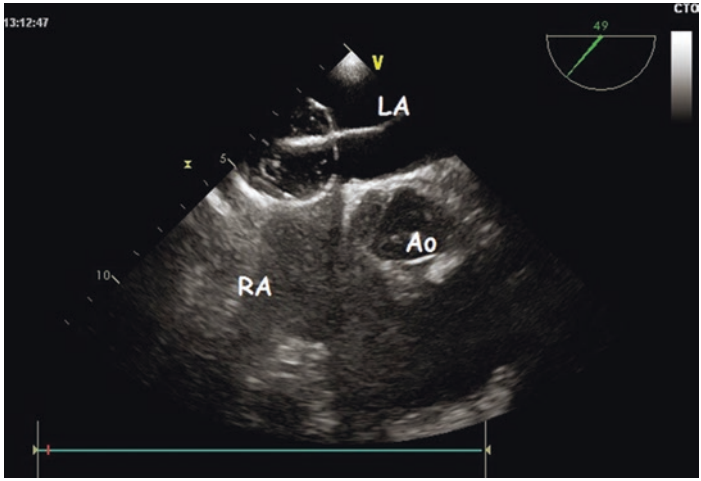

Fig. 39.1 Temporary ASD closure in elderly patient to test the hemodynamic changes resulting from sudden left-to-right shunt disappearance. The occluding balloon is inflated into the right atrium and carefully advanced to the atrial septum. In this setting, the temporary balloon occlusion should be performed using the "dynamic" technique inflating the occluding balloon (Equalizer Balloon Occlusion Catheter, Boston Scientific, Natick, Massachusetts, USA) in the right atrium so less interfering with the volume and compliance of the left heart chambers. *Ao* aorta, *LA* left atrium, *RA* right atrium

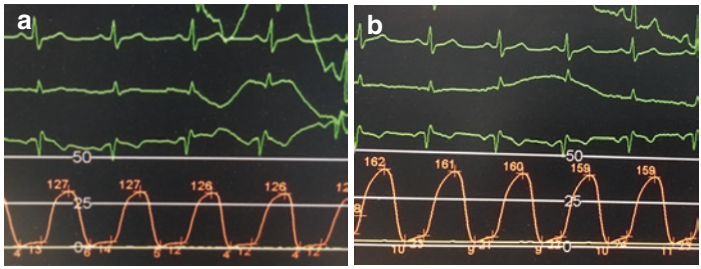

Fig. 39.2 Left ventricular (LV) pressure tracing before (**a**) and during (**b**) temporary dynamic ASD balloon occlusion performed from the right atrium showing a significant increase of both proto- and end-diastolic pressure. These changes are almost universally recorded in elderly patients during test occlusion. However, in the vast majority of cases, there is a tendency to reduce or normalize following a few minutes of occlusion. Conversely, if the LV end-diastolic pressure persistently increases during balloon occlusion or does not show any trend toward the baseline values, a further test of ASD closure should be performed following a trial of diuretic and ino-lusitropic drugs

this maneuver makes possible to temporarily abolish the atrial shunt, so testing the physiopathologic consequences of hemodynamic "normalization" in terms of left and right ventricle preload changes. The temporary closure of ASD causes a sudden decrease of RV preload, and hence pulmonary blood flow, and a sudden increase of left chamber preload. These latter changes may be poorly tolerated by an under-trained LV and may cause a significant rise of the left atrial mean pressure and LV end-diastolic pressure (Fig. 39.2) that may clinically appear as pulmonary congestion or frank edema.

At the temporary balloon test occlusion, relative contraindications for definitive shunt closure are the persistency for more than 15 min of unfavorable hemodynamic data as:

- Increase of LV end-diastolic pressure (>20 mmHg and/or increase >50% compared to baseline).
- Decrease of systemic arterial pressure as higher as 20% with respect to baseline values.
- Appearance of pulmonary edema signs (need for increase of postexpiratory peak pressure during mechanical ventilation or breath fatigue in awake patients).

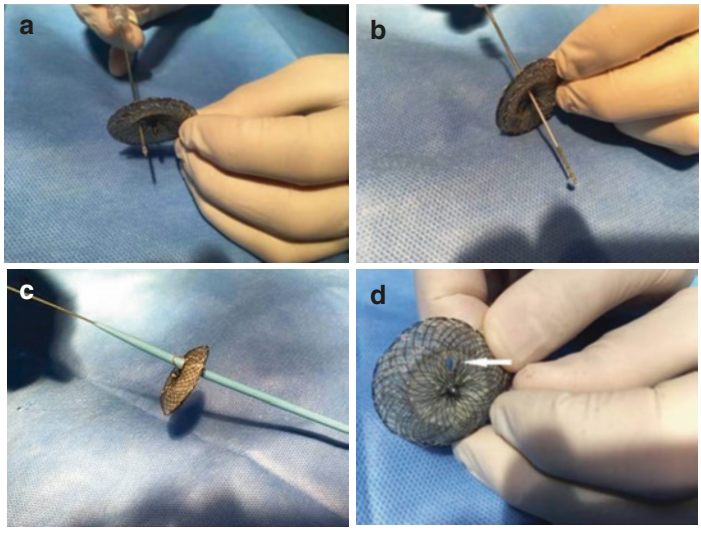

Fig. 39.3 Handmade fenestrated ASD-occluding device. (**a**) The device is perforated close to the central hub using an 18 G Seldinger needle. (**b**) Then, the stiff end of a 0.035″ standard guidewire is passed through the needle. (**c**) Finally, the dilator of a standard 10 Fr introducer is firmly advanced over the wire. (**d**) As a final result, a 3–4 mm large hole (*arrow*) is created. This maneuver may be repeated several times if multiple holes have to be made

This subset of patients may benefit of:

- Partial ASD closure with a fenestrated device (Fig. 39.3). Fenestration may be obtained by perforating a self-centered occluding device using a Seldinger technique with 10–12 Fr femoral sheath, so creating a 3–4 mm hole within the device. If needed, the hole can be further increased "in vitro" by using a 6–8 mm peripheral angioplasty balloon, or multiple fenestrations may be sequentially made. The fate of these fenestrations is usually their spontaneous closure within a few months.
- Pharmacologic trial with intravenous anticongestive drugs (diuretics and ACE inhibitors) for 3–5 days or with oral drugs (diuretics and ACE inhibitors) for 3 months, followed by further reevaluation of the hemodynamic data during

ASD balloon occlusion. Also in this setting, an improved but persistent borderline hemodynamic profile should advise for partial ASD closure with fenestrated devices.

Technical steps of transcatheter ASD closure in elderly are not significantly different from what widely described in younger patients. However, based on the previous pathophysiologic considerations, the procedure demands some additional tips:

- Adequacy of the respiratory pattern, obtained either with a comfortable position in the awake patient or optimizing the mechanical ventilation in anesthetized ones.
- Systemic artery and left ventricle pressures recording by small-size catheters both at baseline and during the temporary occlusion test.
- Right atrial, pulmonary artery, and pulmonary capillary wedge pressures as well as systemic pressure recording at the end of a quiet respiratory cycle. Superior vena cava, inferior vena cava, pulmonary artery, and femoral artery spO2 measurements in triplicate.
- Evaluation of pulmonary venous saturation through the ASD.
- Evaluation of potential coronary artery pathophysiologic abnormalities by coronary angiography and/or intracoronary pressure and/or flow recordings since any significant LV preload increase resulting from atrial shunt closure might potentially unmask subclinical, borderline coronary artery stenoses.
- Temporary balloon-occlusion test for 15 min mimicking the device deployment. In patients with borderline coronary artery stenoses, balloon-occlusion is maintained for a longer time, looking for ischemic EKG changes or regional systolic/diastolic LV abnormalities.
- Careful evaluation of LV end-diastolic pressure and systemic arterial pressure before, during, and after balloon testing.
- Pulmonary artery pressure evaluation during and after balloon testing in subjects with high baseline values.
- ASD closure in elderly patients is performed following a routinely, well-described technique.

39.2 ASD Closure in Pulmonary Atresia with Intact Ventricular Septum

ASD or patent foramen ovale (PFO) is almost invariably present in the setting of pulmonary atresia with intact ventricular septum (PA-IVS) submitted to right chamber decompression by percutaneous or surgical valvotomy. Right ventricular (RV) hypoplasia and/or abnormal compliance almost always condition the long-term pathophysiology of this malformation, resulting in bidirectional atrial shunt (Fig. 39.4) either at baseline or during effort as a safety valve to unload right chambers and/or to increase systemic ventricle output. Thus, ASD closure may be hemodynamically dangerous and clinically poorly tolerated in this subset of patients, in that it causes the increase of the systemic venous pressure (particularly harmful at hepatic and renal level) and a decrease of the systemic output due to under-filling of the LV. However, ASD closure should be always attempted in patients with PA-IVS submitted to RV decompression, in order to avoid the right-to-left shunt caused by borderline right chamber size and/or compliance resulting systemic hypoxia at rest or during effort as well as potential paradoxical embolism.

Indication to ASD closure derives from baseline clinical and instrumental findings, as well as uneventful balloon occlusion test of the septal defect.

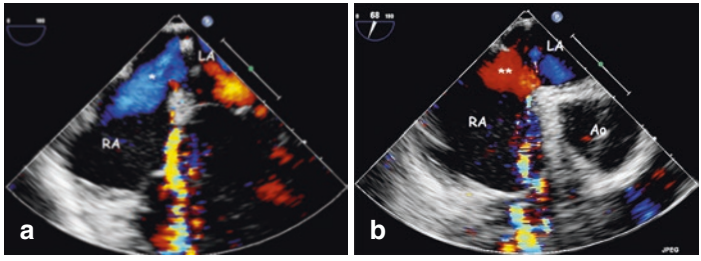

Fig. 39.4 ASD closure in a patient with pulmonary atresia and intact ventricular septum (PA-IVS) submitted to right ventricular (RV) decompression. Bidirectional left-to-right (**a**) and right-to-left (**b**) atrial shunt as imaged at TEE color-Doppler analysis. *Ao* aorta, *LA* left atrium, *RA* right atrium

Anatomic and/or functional findings suggestive of RV unsuitability for biventricular physiology, such as tricuspid valve hypoplasia (z-score < 3), RV hypoplasia (bipartite morphology or severe apical hypertrophy), significant and almost exclusive right-to-left atrial shunt, should be considered as absolute contraindications to ASD closure. However, patients with mild-to-moderate systemic desaturation at rest (>85%) and/or bidirectional atrial shunt at low velocity at Doppler examination should be considered for potential ASD closure. In this setting, the temporary balloon occlusion test of the PFO/ASD could give the physician more information about the final hemodynamic picture.

Mandatory technical steps of the temporary transcatheter ASD closure in these patients are:

- Complete right and left heart catheterization.
- Transesophageal echocardiographic and fluoroscopic monitoring.
- Balloon occlusion test performed from left atrium (Fig. 39.5) in order to avoid any interference with the right chambers vol-

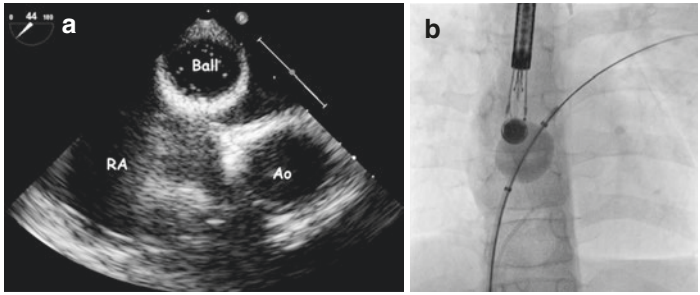

Fig. 39.5 Temporary ASD closure using an occluding balloon to test the hemodynamic changes resulting from the sudden shunt disappearance, as imaged at TEE (**a**) and fluoroscopy (**b**). Also in this setting, the "dynamic" occlusion test should be preferable in order not to interfere with the right atrium. Thus, the occluding balloon is inflated into the left atrium and carefully pulled back toward the atrial septum in order to reduce the volumetric impact into the right atrium. Noteworthy, a multipurpose catheter is left inside the right atrium in order to record any potential pressure change at the time of shunt disappearance. *Ao* aorta, *Ball* occluding balloon, *LA* left atrium, *PA-IVS* pulmonary atresia with intact ventricular septum, *RA* right atrium, *RV* right ventricle

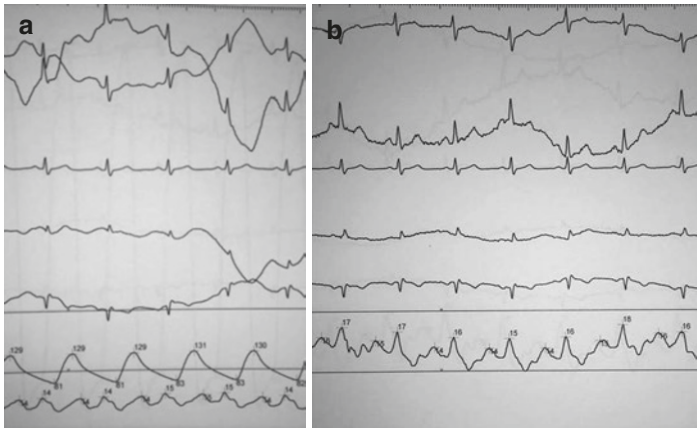

Fig. 39.6 Right atrium pressure tracing before (**a**) and during (**b**) the temporary "dynamic" ASD occlusion. Noteworthy, during the ASD closure, no significant change in RA pressure as a result of abolition of the right-to-left shunt is recorded. However, a significant and persistent increase of the RA pressure and/or a decrease of the mean systemic pressure would preclude the ASD closure. *LA* left atrium; *RA* right atrium

ume and compliance. The occluding balloon should be inflated for at least 15 min in order to evaluate any "subacute" hemodynamic modification. Due to this shrewdness, the dynamic balloon testing is preferable to the static one.

– During the test occlusion, the following hemodynamic parameters should be monitored: right atrial pressure (Fig. 39.6), systemic arterial pressure, and systemic oxygen saturation. Ideally, the mean right atrial pressure should not increase more than 20%, the mean systemic arterial pressure should not decrease more than 20% as compared to the baseline values. In addition, the arterial oxygen saturation should increase over 95%, and no significant changes should be recorded at blood gas analysis. In the case of borderline changes, a short-term course of diuretics may be given, in order to proceed to a second attempt of closure after some few days. In alternative, the ASD closure may be performed and followed by a short-term

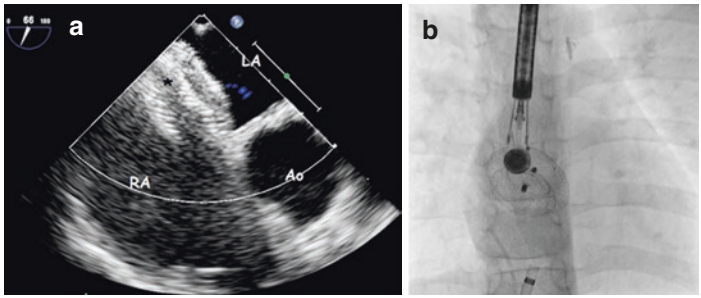

Fig. 39.7 ASD closure using an Amplatzer Septal Occluder device as imaged at TEE (**a**) and fluoroscopy (**b**)

(3–6 months) trial of diuretic therapy. Finally, the device fenestration could be considered as a safety solution in some borderline patients despite the diuretic therapy.

- ASD/PFO closure in PA-IVS is performed following a routinely, well-described technique (Fig. 39.7).

40 Creating an Interatrial Communication

Derize E. Boshoff, Gianfranco Butera, and Marc H. Gewillig

The presence of an interatrial shunt may be important to augment cardiac output in obstructive lesions of the right side of the heart, to enhance mixing in patients with transposition of the great vessels, to off-load the right side of the heart in pulmonary vascular obstructive physiology, to relieve left atrial hypertension in left-sided obstructive lesions, and to decompress the right atrium in postoperative right ventricular failure. With the use of extracorporeal membrane oxygenation for circulatory support, an interatrial communication is necessary to relieve left atrial hypertension from the nonejecting left heart, and in those children with a failing Fontan circulation, an adequate interatrial communication may lessen systemic venous hypertension, improve systemic perfu-

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sion, and perhaps relieve sequelae such as protein-losing enteropathy. Rashkind balloon atrial septostomy, described in 1966 in patients with transposition of the great arteries, was the first percutaneous atrial septostomy [1]. Few transcatheter techniques have been developed over the years to create or enlarge an interatrial communication. These include balloon atrial septostomy, blade atrial septostomy, static balloon dilation of the septum, and radio frequency (RF) perforation or transseptal puncture of the septum, followed by one of the above procedures. These techniques provide a temporary solution; for longer-lasting mixing/relief of obstruction, stent implantation in the septum or perforated devices may provide a more durable solution.

40.1 Anatomy of the Oval Fossa and Surrounding Structures

The key to a successful atrial septostomy is knowledge and understanding of the anatomy of the fossa ovalis and the surrounding landmarks.

The interatrial septum is bounded posteriorly by a fold of pericardium between the atria, superiorly by the vena cava superior, anterosuperiorly by the noncoronary sinus cusp of the aortic valve, anteriorly by the septal tricuspid annulus, anteroinferiorly by the coronary sinus ostium, and inferiorly by the vena cava inferior. The interatrial portion is relatively small, and its most prominent feature is the fossa ovalis, comprising an average of 28% of the total septal area. During fetal and neonatal life, the valve of the fossa ovalis is a paper-thin, delicate, and translucent membrane. With increasing age, however, the valve becomes thicker, tougher, and more opaque, due to deposition of collagen and elastin. Because of hemodynamic streaming within the right atrium during fetal life, the poorly oxygenated blood from the vena cava superior is directed toward the tricuspid valve, while well-oxygenated placental blood from the vena cava inferior is directed via the Eustachian valve toward the fossa ovalis and into the left atrium. As a result of this orientation of the venae cavae, transseptal access is much easier via the vena cava inferior, in contrast

to right ventricular biopsies, which can be more readily performed via the vena cava superior. Occlusion of the femoral veins may, therefore, be considered as an anatomical limitation for transseptal access. The umbilical vein is a good alternative in the newborn baby, but if obstructed, the transhepatic access can be used safely even in small infants. The advantages of this route include a better angle to access the atrial septum and the possibility of using larger sheaths in small children without vascular damage.

40.2 Catheterization Procedure: General Principles

The procedure is typically performed under general anesthesia. The exception to this rule is routine atrial septostomy in newborn babies performed in the neonatal intensive care unit under echocardiographic guidance. Prophylactic antibiotics and heparin sulfate (100 units/kg) should be given intravenously. In patients who require mechanical or RF transseptal access, heparin should be given only after entering the left atrium. The creation of an atrial communication should include surgical and circulatory support backup. The surgical backup does not necessarily mean a standby operating room but rather the availability of surgeons and anesthesiologists who can manage neonates and infants in case of complications from the procedure.

40.3 Imaging Techniques

40.3.1 Fluoroscopy

Biplane fluoroscopy is preferred for performing an atrial septostomy, with the only exception being routine atrial septostomy in newborn babies with simple transposition of the great arteries.

It is of particular importance in small patients or conversely in larger patients with either a very large or very small atrium, a large dilated aortic root, no vena cava inferior access to the atrial septum, or any abnormal cardiac chamber or great vessel posi-

tional abnormalities. Several different angiographic projections have been described to best visualize the interatrial septum during transseptal procedure (discussed in detail in Chap. 15).

40.3.2 Echocardiography

Echocardiographic imaging has greatly improved the success and safety of transseptal interventions.

Transthoracic echocardiography (TTE) may permit visualization of the interatrial septum and the adjacent structures, but its role in guiding complex transseptal catheterization (i.e., stent implantation) is limited due to poor image quality, difficulty to identify the fossa ovalis correctly, and disruption of the sterile field.

The fossa ovalis can be accurately located using intracardiac echocardiography (ICE), but it has limitations as sheath size, additional puncture in the femoral vein, possible longer procedural time, and significantly higher costs.

Transesophageal echocardiography (TEE) is probably the modality of choice in addition to fluoroscopy, particularly when visualizing a specific area of the fossa ovalis to be punctured, the thickness of the septum at that point, and the degree of anterior-posterior direction of the intended puncture and certainly in the case of complex (congenital) anatomy when stent implantation is performed. In small infants, the use of the higher profile biplane or multiplane pediatric TEE probes may cause airway and even left atrial compression, which may distort the underlying septal anatomy and limit even further the already restricted space in the left atrium for transseptal procedures.

Limited reports suggest that the 8-French AcuNav (ACUSON Acunav, Siemens Medical Solutions, USA) probe can be used transesophageally in small infants. Although the AcuNav is a monoplane probe and does not have an attached thermistor, the quality of the pictures seems to be sufficient, and thermal damage in the esophagus did not seem to be an issue in these limited reports.

40.4 Balloon Atrial Septostomy

Balloon atrial septostomy (BAS) should be available in every institution that cares for infants with congenital heart disease.

Because of septal thickening with age, it is usually not consistently effective beyond the neonatal period. Emergency BAS is performed in any infant with simple transposition of the great arteries who exhibits evidence of acidosis as a result of inadequate interatrial mixing.

It is also indicated in all infants with simple transposition of the great arteries who are younger than 1 month of age with a restrictive interatrial communication and not otherwise scheduled for immediate surgery.

It may also be indicated for palliation in neonates with other congenital heart lesions in whom all systemic, pulmonary, or mixed venous blood must traverse through a restrictive interatrial communication to return to the circulation.

40.4.1 Balloon Catheters

BAS catheters are available from various manufacturers and in different designs. Currently, there are four different catheters that can be used for this purpose:

1. *The Miller-Edwards Catheter (Edwards Lifesciences)*
   This is a single-lumen catheter with a 5-Fr shaft but requires a 7-Fr sheath. It has a 35° hockey stick angle 2 cm from the tip, which allows easy entry into the LA. The fairly compliant latex balloon is capable of accepting 4–5 ml of fluid. At that volume, the diameter of the balloon sphere is 17–18 mm. Due to the relatively high compliance, large balloon inflations are often required to successfully perform a septostomy, which is a considerable disadvantage (especially in small infants <3 kg, or a small LA). Although a favorite balloon for many interventionists, the balloon was recalled in march 2019 because of

deflation, fragmentation, and detachment issues; it is unlikely it will be reintroduced.

2. *The Rashkind Balloon Catheter (USCI-CR Bard)*
   This septostomy catheter has a recessed, low-profile balloon and can be introduced through a 6-Fr sheath. The balloon accepts 1.5 ml of contrast to give a balloon diameter of 12–13 mm. Larger volumes will only elongate the balloon without *increasing* the diameter.
3. *The Fogarty (Paul) Balloon Catheter (Edwards Lifesciences)*
   Introduced via a 6-Fr sheath.
4. *The NuMED Z-5 Atrioseptostomy Catheter (NuMED)*
   This is the only catheter with an end hole that enables the operator to advance it over a guidewire and to confirm position by injecting contrast in the left atrium. It is available with balloon sizes of 1 ml (9 mm diameter) and 2 ml (13.5 mm diameter) and can be passed over a 0.014/0.018-in. wire (5-Fr/6-Fr sheaths). The noncompliant nature of the Z-5 septostomy catheter and its relatively small size offer distinct advantages when performing BAS in patients with a small left atrial size (i.e., HLHS). Of note is that a radiopaque marker is located in the midportion of the balloon. However, the wrapped balloon will extend a fair amount beyond the end of the catheter shaft, and one has to be very careful when advancing this balloon to avoid pushing the fairly stiff tip against the left atrial wall or appendage, which can easily induce atrial tachycardia [2].

40.4.2 Procedure

Access is obtained from either the umbilical vein or the femoral vein by use of an appropriate-sized sheath (5–7 French, depending on the type of septostomy catheter to be used). If the procedure is done in the catheterization laboratory and the baby is stable, routine hemodynamic assessment may be performed, followed by the septostomy. When using the umbilical venous approach, the progress of the catheter through the ductus venosus can be monitored either by fluoroscopy (in which case the catheter passes from the right of the midline superiorly toward the right

atrium in the AP projection and from front to back in the lateral projection) or by cross-sectional echocardiography. It may sometimes be difficult to pass the catheter into the VCI due to stenosis or closure of the ductus venosus. In this case, a 0.018″ guidewire and 4-French end-hole catheter combination can be introduced into the umbilical vein and then manipulated into the right atrium. Thereafter, an appropriate-sized sheath can be used to introduce the septostomy catheter. When using a sheath in the umbilical vein, it must be kept in mind that the tip of the sheath is often inside the RA and may impede withdrawal of the inflated balloon across the septum if not withdrawn into the ductus venosus before performing the septostomy. Once the balloon is positioned in the left atrium and the position is confirmed (by fluoroscopy and/or echocardiography), the balloon is inflated with the appropriate volume of saline/contrast mixture (80/20%) while holding the balloon against the atrial septum (to prevent passage across the mitral valve).

The stopcock is closed, and the balloon advanced 1–2 mm of the atrial septum and then jerked/pulled briskly to the right atrial/vena cava inferior junction. The balloon is subsequently advanced promptly to the mid right atrium and deflated as quickly as possible. The balloon must be watched on fluoroscopy or echocardiography during inflation: if it does not retain a perfectly circular shape even at its highest inflation volume, it is probably not free in the atrium and must be deflated and repositioned. Care must be exercised as to how vigorously the balloon is pulled into the inferior vena cava. This process is repeated at least once until there is no resistance to passage of the full balloon across the defect. A gradient across the septum may be measured, and if still significant, a BAS may be repeated as above. Alternatively, echocardiography along with Doppler assessment of the residual gradient may be used to determine the adequacy of the septostomy.

40.4.3 Tips for Crossing the "Difficult" Septum

A variety of techniques can be used to advance the septostomy catheter across the interatrial septum. Direct advancement of the

preshaped catheter is successful in most cases. In some patients, advancing a sheath across the interatrial septum may facilitate passage of the septostomy catheter. The Cordis 6-FR BRITE TIP sheath (Cordis Corp., Miami, FL) has a sufficiently smooth transition to pass over a 0.018-in. guidewire into the left atrium. It is important though to pull back the sheath sufficiently into the vena cava inferior, prior to performing the BAS. If all techniques fail, advancing a low-profile balloon, such as the NuMED Tyshak Mini (NuMED, Hopkinton, NY), across the interatrial septum may allow predilation of the interatrial communication to subsequently allow passage of the septostomy catheter.

40.4.4 Intact Interatrial Septum

Perforation of the interatrial septum may be required whenever the interatrial septum is intact or the existing interatrial communications are unsuitable for BAS (superior or inferior location). The use of the standard Brockenbrough needle for transseptal puncture in patients with complex anatomy or a small left atrium (HLHS and variants) may be rather cumbersome, with the potential risk of atrial perforation. The Nykanen RF perforation wire and the 180 cm 0.035-in. outer diameter coaxial injectable catheter (both Baylis Medical Corporation, Montreal, CA) can be controlled and appropriately directed using a Judkins right coronary catheter. This is particularly beneficial in patients with a small left atrium or unusual anatomy.

40.5 Blade Atrial Septostomy

In infants older than 1 month of age, and certainly in older children, the atrial septum is usually too tough or thick for a simple BAS to tear the septum. The indications for blade atrial septostomy are the same as considered for a balloon septostomy or for surgical atrial septostomy that otherwise would be needed in the older infant. Blade septostomy catheters (Cook, Bloomington, IN) are available with three blade lengths: 1.0, 1.34, and 2.0 cm.

The two smaller blades (the PBS 100 and 200) are available on a 6-French catheter, and the 2.0 blade (the PBS 300) is on an 8-French catheter. Both blade catheter sizes require a sheath one size larger than the catheter for smooth introduction. The blade is controlled by a wire that has a moveable "handle": if the wire is fully retracted so that the blade is inside the catheter shaft, the handle may be locked against the hub, preventing inadvertent blade protrusion. A side port is available to flush the catheter with saline (or contrast); the direction of the port (off the side of the catheter) is roughly the same as the direction of the curve and of the blade when it is protruded. The blade should always be tested outside the patient to be sure it opens and closes fully without resistance. The blade catheter is advanced through the previously placed long Mullins sheath into the left atrium, and the sheath is then withdrawn well into the vena cava inferior. The blade is then opened carefully in the left atrium while it is continuously observed on fluoroscopy (and ideally also under TEE guidance). The tip is directed anteriorly and either to the patient's right or left side. In contrast to the balloon septostomy, the blade catheter is withdrawn slowly in a controlled maneuver. Resistance may be quite considerable, so bracing one's hands against the patient's leg (and pulling with the fingers) during the maneuver may prevent sudden retraction of the open blade down the vena cava inferior. If the septum proves too rigid to cross with a fully opened blade, the opening angle should be adjusted to 45–60° before pulling across and then repeated with a fully opened blade. Once the blade has crossed the septum, the catheter should be slightly advanced and the wire withdrawn to retract the blade back inside the catheter. The blading is repeated at least four times while changing the angle of extension of the blade as necessary and changing the blade direction from side to side until there is no further resistance to withdrawal of the fully opened blade catheter. The blade septostomy is followed by a balloon septostomy (standard BAS or static balloon dilation). Blade septostomy should probably be avoided though in patients with complex anatomy and small left atrial size, and the combination of cutting balloon septostomy and static balloon septostomy is likely a safer alternative in patients with a small left atrial size.

40.6 Cutting Balloon Septostomy

With the availability of larger cutting balloons of ≤8 mm in diameter (Boston Scientific, Boston, MA), the combination of static cutting balloon septoplasty, followed by the use of larger diameter static balloons or standard balloon atrial septostomy, has become a valuable alternative to blade atrial septostomy in patients with a thickened interatrial septum. It is suggested that the microsurgical blades of the cutting balloon allow controlled tearing of the septal wall rather than stretching of the thickened interatrial septum, as seen with static balloon dilation alone. Rotation of the cutting balloon, followed by repeat inflations may tear the interatrial septum in different locations and improve the response to static balloon septoplasty. The smaller the preexisting septal defect, the higher the likelihood that the use of a cutting balloon will achieve an adequate result. If the existing interatrial communication is "stretchable" (i.e., floppy valve), cutting balloon dilation will be inefficient. In this situation, it may be better to perform a transseptal puncture and start with a new diminutive opening to obtain a better result with cutting balloon septoplasty. The cutting balloon catheter (typically 4–8 mm) is advanced through a 6- or 7-French (short/long) sheath over a 0.014-in. coronary angioplasty wire or a 0.018-in. guidewire (Roadrunner, Cook) positioned in the left upper pulmonary vein or alternatively curled in the body of the left atrium.

40.7 Static Balloon Septostomy

This modality can be used primarily or after blade septostomy or cutting balloon septoplasty. The balloon dilation is performed with a high-pressure balloon (Fig. 40.1). Balloon diameter will depend on patient/atrial size and underlying cardiac anomaly.

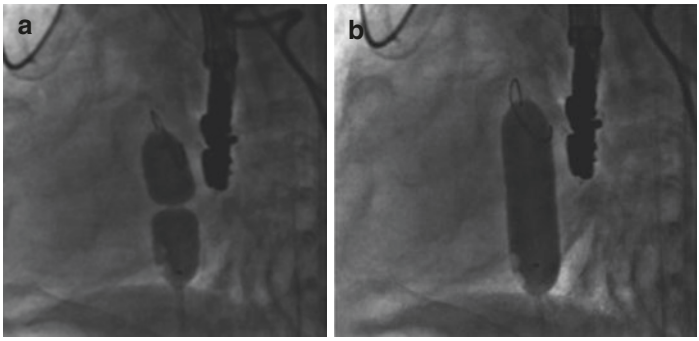

Fig. 40.1 Static balloon dilation of restrictive atrial communication: (**a**) 12 mm balloon inflated at mild pressure delineating the small atrial communication, (**b**) full inflation stretches-tears the septum

40.8 Stent Implantation in Congenital Heart Defects: Nonrestrictive Technique

A restrictive interatrial communication in patients with univentricular anatomy significantly affects surgical outcomes. In patients with univentricular hearts, wide-open atrial communication leads to lower pulmonary artery pressure, which is one of the most important factors influencing the success of bidirectional Glenn and Fontan operations. In some patients, recurrence of restricted interatrial communication can be observed despite initially successful interventional or surgical creation of unrestrictive interatrial communication. Atrial stent septostomy can provide a reliable long-lasting restrictive or nonrestrictive interatrial communication.

40.8.1 Procedure

After access has been obtained in the left atrium, a wire should be positioned in a pulmonary vein and an appropriate-sized long sheath advanced over the wire with the tip across the atrial septum. Premounted balloon-expandable stents are preferable in this

setting although self-expandable stents have been used with success in some patients. The stent diameter will depend on the age and size of the patient and the type of congenital anomaly (especially atrial size), aiming to provide an unrestrictive and potentially durable flow through the interatrial septum for several months. One of the crucial facts is to avoid implanting too long stents due to the risk of atrial erosion, thrombus formation, and obstruction of the pulmonary veins. The stent should be long enough though to allow adequate stabilization within the interatrial septum, minimizing the risks of embolization due to movement during inflation or due to foreshortening after expansion. We advise using the technique of sequential stent flaring to facilitate accurate stent positioning. The stent is advanced through the long sheath into the left atrium. Half of the stent is exposed by pulling back the sheath and the balloon is inflated in the left atrium, expanding the distal half of the stent. Pressure in the balloon is maintained using a stopcock. Next, the entire system is firmly pulled back against the atrial septum. The pressure in the delivery balloon is slightly released, allowing the right atrial portion of the stent to be unsheathed. The balloon is then fully inflated, opening the proximal portion of the stent. The deflated balloon should be removed carefully out of the stent into the long sheath, avoiding stent dislodgment. We advise against crossing the newly implanted stent with a catheter unless certainty of adequate fixation. Gradients and flow across the interatrial septum should be assessed using TTE or TEE (Fig. 40.2). In contrast to conventional balloon atrial septostomy, stent implantation requires antiaggregation treatment (acetylsalicylic acid 2–5 mg/kg/day) to prevent thrombus formation.

40.9 Stenting of the Interatrial Septum: Restrictive Technique

40.9.1 Pulmonary Arterial Hypertension

Atrial septostomy for severe pulmonary arterial hypertension (PAH) improves cardiac index and functional class by the creation

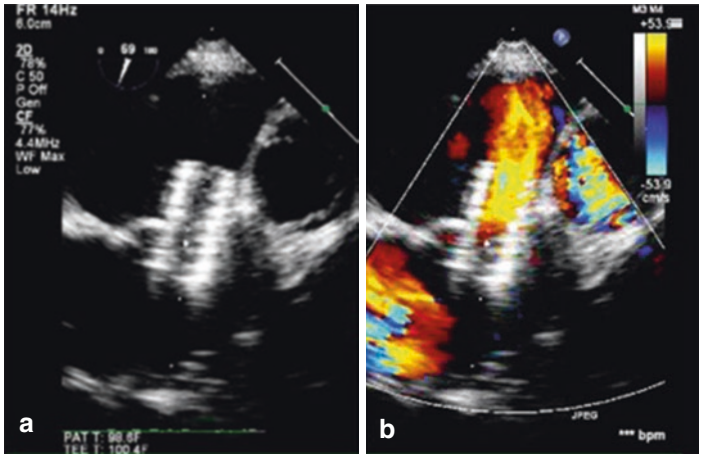

Fig. 40.2 TEE of stent across atrial septum: (**a**) a Genesis 1910 stent is nicely positioned across the atrial septum, (**b**) color flow mapping shows right-to-left shunt across the stent

of a right-to-left atrial shunt and may even improve the survival in some patients. The presence of this iatrogenic shunt decompresses the failing right ventricle and improves left ventricular preload and thus cardiac index. Early series reported a high mortality, largely caused by difficulty in achieving accurate control of the size of the atrial shunt. Improvements in patient selection and septostomy techniques (i.e., sequential balloon dilation) have increased the safety of the procedure; however, a high spontaneous closure rate is observed after balloon dilation, necessitating repeated procedures in an already critically ill patient group. The recent evidence-based treatment guidelines for PAH list the indication for the atrial septostomy procedure as Class 1C, generally limited to specialized centers and reserved for patients with recurrent syncope and those who are refractory to, or intolerant of, medical therapy or as a bridge to transplantation. In contrast with balloon septostomy, restrictive stenting of the interatrial septum with the use of a diabolo-shaped (bow tie or dog bone stent) allows for a predictable and long-lasting interatrial shunt in these patients. The fenestration technique currently used in our unit has

been adapted from [3], who described a small mixed series of primary PAH patients and patients with a failing Fontan circulation. A venous sheath up to 12-Fr is placed into the right femoral vein, followed by a puncture of the interatrial septum with a Brockenbrough needle.

40.9.1.1 Stent Preparation

A loop of 3–5 mm diameter is created using a set of temporary epicardial pacing wires. The needle ends are removed, including the distal 5 cm length of isolative coating, allowing making a low-profiled tight knot with bare metal wire. The two wires are tied together to provide a length of about 90 cm, allowing the wire to leave the sheath at the operators end. Using the bare end of the wire, a secure double knot is formed over a 10–14-French dilator. The resultant loop is then placed over the midportion of a standard 15 mm valvuloplasty balloon catheter. A standard stent (PALMAZ GENESIS stent 1910, Cordis Corporation, Miami Lakes, FL) is gently dilated with the help of the tapered end of the 10–14-French dilator. The stent is then mounted on the valvuloplasty balloon, taking care that the loop created from the pacing wire is placed accurately in the center of the balloon and the stent (Fig. 40.3). The stent is then manually crimped and its stability tested.

40.9.1.2 Stent Deployment

The mounted stent is delivered through the long sheath, securing the end of the temporary pacing wire. The stent is then deployed using the technique of sequential stent flaring, as described above (Fig. 40.4). After the stent has been deployed in diabolo across the septum, the balloon (with the metal-knot wire) is removed. We gradually increase the size of the fenestration until arterial saturation has decreased down to 80–85%.

40.9.2 Fontan Circulation

Secondary fenestration of a failing Fontan circulation is a valuable technique to improve the hemodynamic condition of the patient. The fenestration is created to allow a restrictive

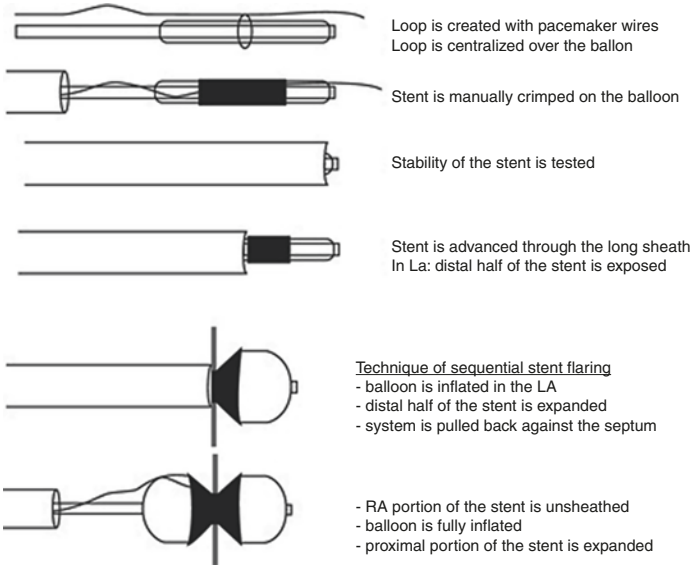

Fig. 40.3 Cartoon with various steps for a diabolo stent

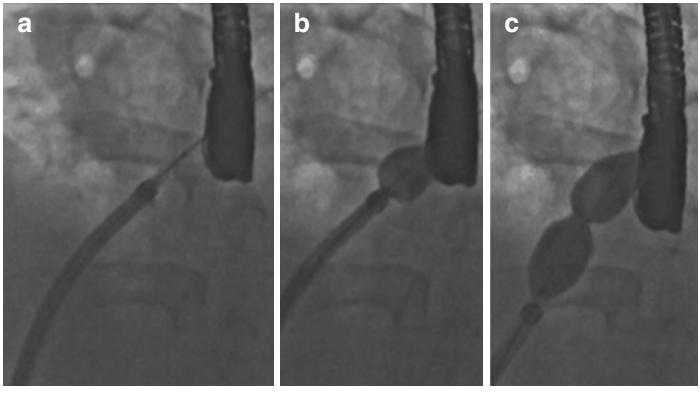

Fig. 40.4 Deployment of a 1910 Genesis stent: (**a**) 10F sheath into left atrium, stent is partially uncovered, (**b**) inflation of 15 mm balloon results in flaring of distal end of the stent, (**c**) after pulling whole system against the septum, the sheath is pulled back to uncover the RA end, and further inflation of the balloon results in diabolo shape of the stent across the atrial septum

right-to-left shunt, decreasing the systemic venous pressure and congestion, with increase in cardiac output, but at the expense of arterial desaturation. However, cyanosis is better tolerated than low cardiac output with congestion [4]. Fenestrating the extracardiac Fontan circuit may be more challenging, due to separation of the different wall layers during needle puncture and sheath placement [5]. The optimal perforation site in the extracardiac Fontan conduit is the point that has the most acute angle coming from the inferior (exceptionally the superior) caval vein and which is in contact with the atrium. Gore-Tex conduits are poor conductors and are, therefore, not vulnerable for RF perforation. Puncturing the conduit with a Brockenbrough needle may require considerable force and consequently adequate immobilization of the patient. Currently, we prefer puncture of the inferior caval vein just below the conduit. Once the Gore-Tex or caval wall is crossed, the point of the needle will be in the atrial wall. By giving a small contrast injection, the atrial wall can be tagged and some overflow may be observed in the pericardial space. The needle should then be further advanced until it pops through the atrial wall (position confirmed by small contrast flush). A 0.014-in. coronary wire is then advanced through the needle until well within the atrium (preferably the left upper pulmonary vein). Advancing the dilator and sheath over the Brockenbrough needle may again require considerable force and wringing of the sheath. It is sometimes necessary to predilate the Gore-Tex conduit with a 4 or 5 mm cutting balloon before the sheath can be advanced through the Gore-Tex. After crossing the detached pericardial space, the sheath should be advanced through the atrial wall until well within the atrium before the needle and dilator are withdrawn. The techniques for stent preparation and implantation are identical to the method described for restrictive stenting in patients with PAH. Sequential stent flaring allows for re-approximation of the different layers during stent deployment, creating a predictable restrictive right-to-left shunt.

40.10 Atrial Flow Reducer

The atrial flow regulator (AFR) (Occlutech, Istanbul, Turkey) (Fig. 40.5) is a self-expandable double-disc wire mesh device constructed from 0.004–0.0075-in. nitinol braided into two flat discs connected by a waist of 1–2 mm and central fenestration, which enables bidirectional flow. A welded ball connector located on the device's proximal disc serves as an adapter to connect the delivery system for deployment. After implantation, the AFR conforms completely to the atrial septum leaving an interatrial communication with a preselected fixed diameter. The device self-centers following deployment and is retrievable prior to release. The device is available in 4, 6, 8, and 10 mm fenestration diameters with a total device diameter from 16 to 23 mm delivered via 8–14-F sheath. The device exists also in different waist size ranging from 2 to 10 mm in order to accomodate different "septal" thickness. Its use is only possible as a compassionate procedure. Similarly to other procedure used to create an interatrial communication, a transseptal puncture is performed, fol-

Fig. 40.5 Atrial flow regulator. Left: "right" atrial view; Right: "left"atrial view. The fenestration is clearly delineated

lowed by static balloon dilatation. Then, the delivery sheath is introduced and the device is implanted similarly to standard double-disk ASD closure devices. Its use has been described in a patient with a failing Fontan with successful early and medium-term results [6].

AFR implantation has been also described for patients with symptomatic PAH, right ventricular failure, left atrial congestion, and symptomatic heart failure (HFpEF or HFrEF).

40.11 Complications

Complications of BAS include tears to the left atrium, pulmonary vein, and right atrium, as well as atrial dysrhythmias (usually transient). Atrial septal interventions in patients with HLHS can pose a considerable technical challenge, and procedure-related mortality has been reported to be as high as 15%. In patients with a thick interatrial septum, even a partially inflated balloon may not tear the interatrial septum, causing a shearing force on the pulmonary veins, leading to pulmonary vein avulsion and death in the treated patient. Complications inherent to atrial septal puncture are cardiac perforation and puncture of an inappropriate atrial septal site. Occasionally, the valve is extremely floppy so that when pushed with the tip of the catheter, it may even extend to the lateral wall of the LA, risking exit into the pericardial space when the "septum" is punctured. Prompt recognition and management of cardiac tamponade are essential to minimize the mortality in these patients.

40.12 Conclusions

Creation or enlargement of interatrial communications can be achieved using a variety of transcatheter techniques including transseptal needle puncture or RF perforation, balloon septostomy, blade septostomy, and stent implantation. The procedure can improve hemodynamics acutely in a variety of compromised circulations or provide effective palliation until definitive surgery can be attempted.

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41 Transcatheter Correction of the Superior Sinus Venosus ASD

Eric Rosenthal and Jan Hinnerk Hansen

41.1 Anatomic Description and Pathophysiology

The superior sinus venosus atrial septal defect (SVASD) is a rare interatrial communication, which only accounts for approximately 10% of all atrial septal defects. It is situated outside the fossa ovalis above its superior rim, immediately inferior to the junction of the superior vena cava (SVC) and the right atrium (RA). It is associated with partial anomalous pulmonary venous drainage of the right upper and/or right middle pulmonary veins (RUPV and RMPV). The defect results from a deficiency in the atrial wall that forms the posterior wall of the SVC and the anterior

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wall of the right- sided pulmonary veins. The RUPV and/or RMPVs are no longer committed to the left atrium (LA) and drain into the RA at the mouth of the SVC, which overrides the atrial septum. In addition, anomalous pulmonary vein branches may also drain into the SVC above the SVC–RA junction (Fig. 41.1).

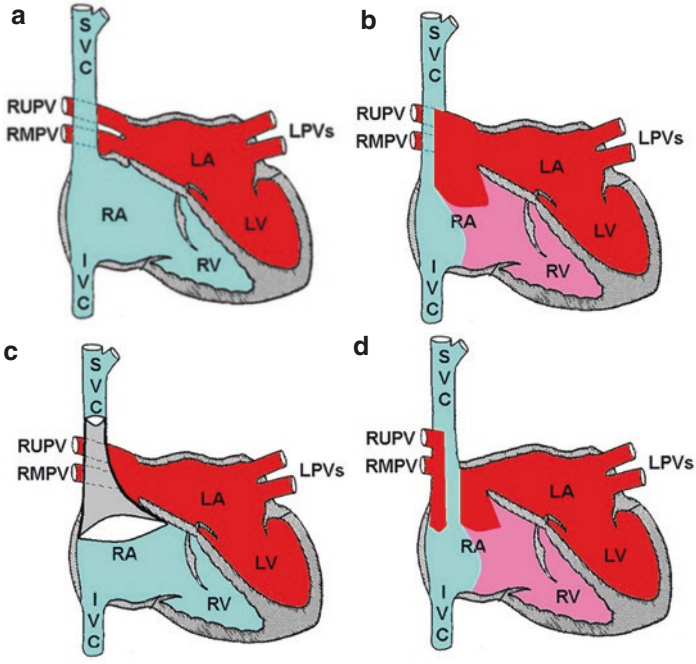

Fig. 41.1 Illustration of the concept: (**a**) In the normal heart, the right pulmonary veins (RUPV and RMPV) are seen to drain behind the SVC into the left atrium (LA). (**b**) In the commonest form of sinus venosus atrial septal defect with partial anomalous pulmonary venous return, the wall between the RUPV and RMPV and the SVC is absent and the RUPV and RMPV drain into the back of the SVC—RA junction with an attachment to the LA. (**c**) Stent correction is possible with the anatomy in (**b**) where the covered stent (in grey) closes the SVASD and the RUPV and RMPV now drain behind the stent into the LA. (**d**) The RUPV and RMPV drain directly into the SVC without continuity to the LA—while the RMPV might still be able to drain behind a stent into the LA, the RUPV will be occluded by the stent and this type is not usually suited to stent correction (Modified from JACC Cardiovasc Imaging 2021. https://doi.org/10.1016/j.jcmg.2020.11.010)

The pathophysiological consequences of the SVASD are determined by the defect size, pulmonary and systemic vascular resistance and right and left ventricular compliance. The resulting left-to-right shunt from the interatrial communication and the anomalous pulmonary venous return into the right atrium lead to right ventricular volume overload and right ventricular dilation. Increased pulmonary blood flow may result in pulmonary vascular changes and elevated pulmonary vascular resistance.

41.2 Clinical Effects

Due to its complex anatomy, the diagnosis of SVASD can be challenging and may even be delayed into adulthood. The defect is often an incidental finding. All patients with unexplained dilation of the right atrium or the right ventricle should undergo evaluation for intracardiac shunts. Symptoms related to intracardiac shunting become more frequent with increasing age and include dyspnoea with exertion, atrial arrhythmias and right heart failure. In addition, stroke can result from paradoxical thromboembolism.

41.3 Indications for Treatment

The indications for treatment are similar to those for secundum atrial septal defects. Treatment aims to attenuate symptoms and prevent right-sided heart dysfunction and atrial arrhythmias. Indications include:

- Right atrial and/or ventricular enlargement with or without symptoms.
- A significant left-to-right shunt with a pulmonary to systemic blood flow ratio greater than 1.5:1 (e.g. determined by cardiac magnetic resonance imaging).
- Paradoxical thromboembolism.
- Need for a transvenous pacemaker.

SVASD closure is contraindicated in irreversible pulmonary hypertension.

41.4 Treatment Options

Surgical correction is the standard of care and until recently was the only option. Surgical repair is by patch-closure of the interatrial communication. The patch is fashioned so that drainage of the pulmonary veins is to the left atrium. Redirection of the pulmonary veins sometimes requires enlargement of the interatrial communication to create an unobstructed pulmonary venous pathway and/or patch augmentation of the SVC to avoid pulmonary vein or SVC stenosis. The Warden procedure —which involves division of the SVC, translocation of the cephalic end to the right atrial appendage and baffling of the caudal end to the LA —is usually reserved for SVASDs with anomalous pulmonary veins draining remotely into the mid or upper SVC.

In general, SVASDs were not considered amenable to transcatheter correction. A transcatheter approach using a covered stent deployed in the SVC–RA junction was first described in 2013 and 2014 [1, 2] and has been adapted and modified by us [3]. The covered stent replaces the deficient posterior wall of the SVC separating the SVC and RA from the RUPV and LA, so that the SVASD is closed and the anomalous pulmonary venous drainage is redirected to the LA behind the stent (Fig. 41.1c). This technique may be considered as an attractive alternative to surgical treatment in adult patients. For children, not fully grown to adult size, surgical correction is currently the usual option.

41.5 Pre-procedural Imaging

An in-depth understanding of the anatomy is essential to assess the likelihood of successful occlusion of the interatrial communication without compromising pulmonary venous return. A prerequisite for simultaneously closing the interatrial communication and redirecting pulmonary venous flow to the left atrium is that the posterior wall of the anomalous pulmonary vein is either in direct continuity with the left atrium or another pulmonary vein with continuity to the left atrium.

Suitability for percutaneous treatment can be assessed with multiple modalities and should routinely include multiplanar and 3D reconstruction of cardiac MRI or CT imaging. Cross-sectional imaging can delineate the anatomy of the pulmonary venous return, and its anatomical relationship to the SVASD. The number and size of anomalous pulmonary veins and their distance from the SVC–RA junction can be assessed (Fig. 41.2).

Stent implantation can be simulated ex vivo with printed or virtual 3D models. Cardiac CT or MRI datasets can be used for 3D model segmentation and reconstruction [3]. Patient - specific 3D models printed in a hollow fashion with soft transparent material allow simulation of the procedure and direct visual inspection of the results (Fig. 41.3a, b). Virtual 3D models facilitate assessment of suitability by allowing virtual implantation of different pre-designed stent prototypes. Stent shape, size, length and choice of sizing balloon can be planned. After virtual stent placement, potential areas of residual shunt or obstruction of the pulmonary venous pathway can be assessed by direct visualisation or 3D cropping (Fig. 41.3c). With these techniques, the ability to predict suitability for transcatheter correction has improved, and standalone diagnostic catheterisation has become redundant in the vast majority of cases.

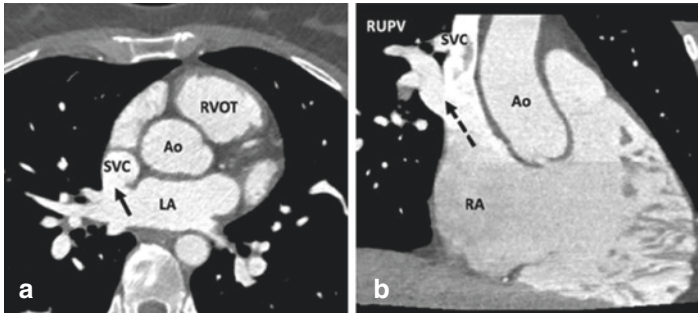

Fig. 41.2 Cardiac CT, axial (**a**) and modified frontal stack (**b**) showing the sinus venosus defect (solid arrow) at the posterior wall of the superior vena cava (SVC) and the anterior wall of the right- sided pulmonary veins. The right upper pulmonary vein (RUPV) drains into the SVC–RA junction (dotted arrow)

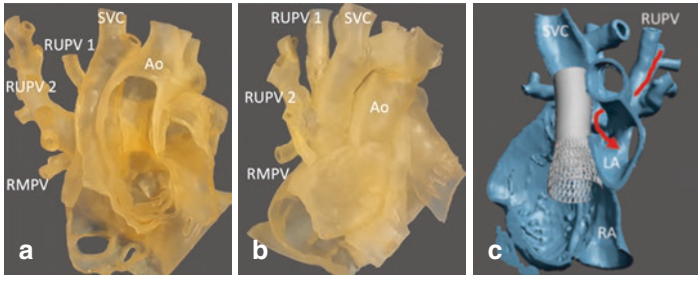

Fig. 41.3 Pre-procedure evaluation with printed and virtual 3D models In (**a**) a small RUPV1 is draining remotely into the high SVC and large RUPV2 at the SVC–RA junction potentially redirectable with a stent. The patient proceeded to stent implantation leaving RUPV1 draining into the SVC. In (**b**) the large RUPV1 is unlikely to be redirected with a stent while it is amenable to surgical redirection. The patient proceeded to surgical repair. (**c**) Covered stent generated by computer-assisted design placed into a patient-specific heart model. The sagittal section of the left atrium and the bicaval view are shown. The view is en face to the sinus venosus defect. The anomalous RUPV is potentially redirected towards the left atrium behind the stent (red arrow). The stent has a flared segment in the lower third to ensure occlusion of the interatrial communication. The patient proceeded to stent implantation. (Reproduced with permission from J Am Coll Cardiol 2020; 75: 1266–1280)

41.6 Patient Selection

Factors precluding transcatheter correction include large pulmonary veins draining exclusively into the SVC well above the right atrial junction that are amenable to surgical redirection (Fig. 41.3b). Smaller RUPV branches that are too small and too far from the SVC to merit surgical redirection are not considered to be a contraindication for percutaneous treatment — they are left draining into the SVC above the stent (Fig. 41.3a). The potential for pulmonary venous compression with stenting is another concern and may rule out transcatheter correction. Large defects with extension into the secundum atrial septum may not be closable with a covered stent alone as the stents used only dilate up to 35 mm — indeed there are isolated reports of simultaneous covered stent and atrial septal

defect device implantation to close such defects [1]. Surgical correction is also preferred if concomitant intra-cardiac procedures are required.

41.7 Technique (Step-by-Step)

Transcatheter correction of SVASD is not an established routine procedure. The proposed step-by-step closure technique is based on a single-centre experience [3].

1. General Aspects.
   (a) Transcatheter SVASD closure is performed under general anaesthesia.
   (b) Prophylactic intravenous antibiotics are administered before vascular access.
   (c) Heparin is given to maintain an activated clotting time of over 250 s after transseptal puncture.
   (d) Radial artery canula avoids the need for femoral artery cannulation.
   (e) Transoesophageal echocardiographic guidance is used throughout the procedure to guide transseptal puncture, stent positioning and deployment, to assess pulmonary venous return and to check for a residual shunt.
   (f) Biplane fluoroscopy and angiography facilitate the procedure. We use the AP and left lateral or 110° LAO projections. In a single-plane laboratory, the AP and 45° LAO projections may be more convenient.
2. *Vascular Access and Preparatory Steps:*
   (a) Right internal jugular vein (RIJV) is used for SVC angiography during balloon testing and stent deployment. It is the exit site for the femoral to jugular venous guide wire circuit over which sizing balloons, delivery sheath, stent/s and post deployment balloon/s are passed.
   (b) Right femoral vein (RFV) is used to advance the guide wire for snaring from the RIJV and subsequent balloon interrogation of the defect, sheath placement, stent

deployment and post stent balloon dilation. A vascular closure device anticipating the large bore sheath (18 –20F) can be placed.

(c) Left femoral vein (LFV) is used for transseptal puncture to access the LA with an 8F SRO sheath for constant LA pressure measurement. Through the sheath, a pigtail catheter is advanced into the RUPV for continuous pressure measurement (simultaneous with the LA) and pulmonary vein angiography. When necessary a balloon can be passed into the pulmonary vein orifice to protect it during stent deployment.

(d) A veno-venous guidewire circuit with an Amplatzer 0.035″ guide wire is established from the RFV to the RIJV to facilitate balloon test occlusion and stent implantation using standard snares. Tension on the guidewire exiting the RIJV sheath is used to control the position of the balloon during balloon test occlusion and stent placement.

3. *Balloon Interrogation of the Defect:*

(a) Test occlusion of the defect is performed before stent implantation to confirm defect occlusion without pulmonary vein obstruction.

(b) The balloon is inflated in the anticipated stent landing zone at the SVC–RA junction, until TOE confirms elimination of the interatrial shunt. An SVC angiogram from the RIJV sheath is obtained to document SVC flow occlusion and balloon apposition to the SVC walls. During balloon inflation, pulmonary venous return is assessed by Doppler echocardiography, angiography from the RUPV catheter and simultaneous RUPV and LA pressure measurements (Fig. 41.4).

(c) Compliant sizing balloons (e.g. 24 mm or 34 mm Amplatzer sizing balloon, St. Jude Medical, St. Paul, MN, USA) may be overinflated at the defect level. This can lead to bulging of the balloon through the defect causing pulmonary venous obstruction.

(d) Non-compliant balloons (e.g. 6 cm AltoSa-XL PTA balloon, AndraTec GmbH, Koblenz, Germany) should be used if there is apparent pulmonary vein occlusion when

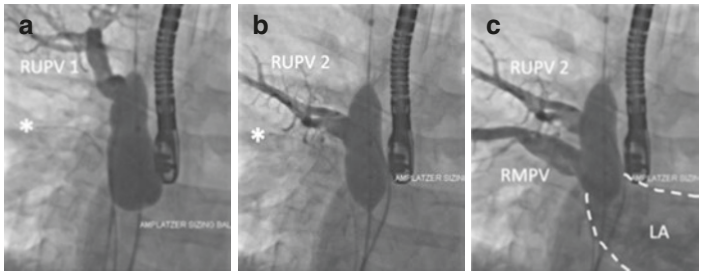

Fig. 41.4 Balloon testing with Amplatzer sizing balloon and selective pulmonary vein angiography. Two right- sided upper pulmonary veins (RUPV1 and RUPV2) are occluded with the balloon inflated in the SVC–RA junction (**a**, **b**), while the right middle pulmonary vein (RMPV, * tip of the catheter in RMPV) is diverted into the left atrium (**c**). The patient was deemed unsuitable for a percutaneous approach and underwent surgical repair. (Reproduced with permission from J Am Coll Cardiol 2020; 75: 1266–1280)

using compliant balloons. The initial balloon size should be 2–4 mm larger than the SVC diameter. Non-compliant balloons, however, may not fully occlude the defect and judgement is needed to determine that the residual leak will be abolished or small enough to ignore after flaring of the RA portion of the stent (Fig. 41.5).

4. *Stent Choice:*
   (a) Measurements obtained during balloon test occlusion are used to choose the stent length and the appropriate balloon diameter for implantation. Custom-made 7 cm or 8 cm long 10-zig covered Cheatham-Platinum (CP) stents are preferred in our unit. Significant shortening of the stent with deployment has to be anticipated particularly at the RA end. The stent length should be selected based on having at least 2 cm of the unexpanded stent in the SVC to prevent stent migration and 2 cm protruding into the RA below the inferior margin of the defect initially.
   (b) The 6- cm long stents should be reserved for cases with an additional pulmonary vein draining into the SVC that cannot be redirected to the LA. To avoid occlusion of the pulmonary vein branch, the cranial end of the covered stent is

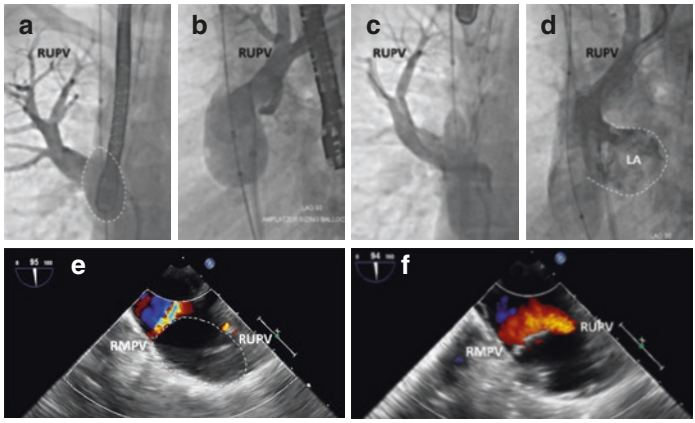

Fig. 41.5 Balloon test occlusion with Amplatzer sizing balloon (**a**, **b**, **e**) and AltoSa-XL balloon (**c**, **d**, **f**) in the same patient. Inflation of the Amplatzer sizing balloon resulted in obstruction of the RUPV (**a**, **b**). TOE (high oesophageal bicaval view with clockwise rotation) confirmed RUPV obstruction and also showed RMPV compression (**e**). During repeat balloon testing with the AltoSa-XL balloon, pulmonary venography confirmed RUPV drainage to the LA (**c**, **d**) with a tiny residual shunt into the right atrium. The TOE revealed no compression to the RMPV but accelerated flow in the RUPV—suspicious for potential obstruction with stenting (**f**). (Reproduced with permission from J Am Coll Cardiol 2020; 75: 1266–1280)

placed just below its orifice allowing continued drainage into the SVC. Additional anchoring with a bare stent then needs to be considered due to the short stent segment apposed to the SVC.

(c) The stent is mounted on a custom-made Balloon-in-Balloon (BiB) catheter. The minimum diameter of the balloon should exceed the SVC diameter by 3 –4 mm, and the balloon length is as long as the stent length.

5. *Stent Implantation:*
   (a) The RFV sheath is upsized to an adequately sized long delivery sheath (e.g. 85 cm 18F Check-Flo Performer, Cook Medical, Bloomington, IN, USA or 65 cm 20F DrySeal, Gore, Flagstaff, AZ, USA) which is placed across the SVC–RA junction.

(b) The stent is positioned using simultaneous TOE imaging and angiography from the RIJV and RUPV. The stent should be long enough that the caudal end of the stent extends 2 cm below the lower edge of the defect on TOE and the cranial end sufficiently high in the SVC to allow secure anchoring (Fig. 41.6).

(c) The stent is deployed by sequential inflation of the inner and the outer balloons of the BiB catheter. Due to the different calibre of the SVC and the RA, there is a tendency for the balloon to milk downwards and change the position of the stent. This can be avoided by tension on the wire externalised from the RIJV while fixing the wire at the RFV onto the balloon catheter.

(d) To ensure complete coverage of the interatrial communication, the RA portion of the stent is flared with the outer balloon of the BiB used for stent deployment, which does not necessarily achieve its maximum diameter in the SVC. If additional flaring is required, a Coda Balloon (Cook Medical, Bloomington, IN, USA) is used. Flaring of the RA portion needs to be performed with small increments under TOE guidance while monitoring the RUPV and LA pressure as overinflation may cause compression and stenosis of the pulmonary venous pathway behind the stent (Fig. 41.6).

6. *End of Procedure.*

(a) A TOE assessment and final angiograms from the SVC and RUPV, as well as pressure measurements in the RUPV, LA and RA, complete the procedure. Due to the increased left ventricular preload, a significant rise in RUPV and LA pressure is common and flow across the transseptal puncture will be present.

(b) If paradoxical embolism was the indication for treatment, complete abolition of interatrial shunting needs to be achieved. Device closure of additional shunts (e.g. PFO) should be performed.

(c) The large bore access in the RFV can be closed with an appropriate vascular closure device or with a subcutaneous Z-suture/figure-of-eight suture.

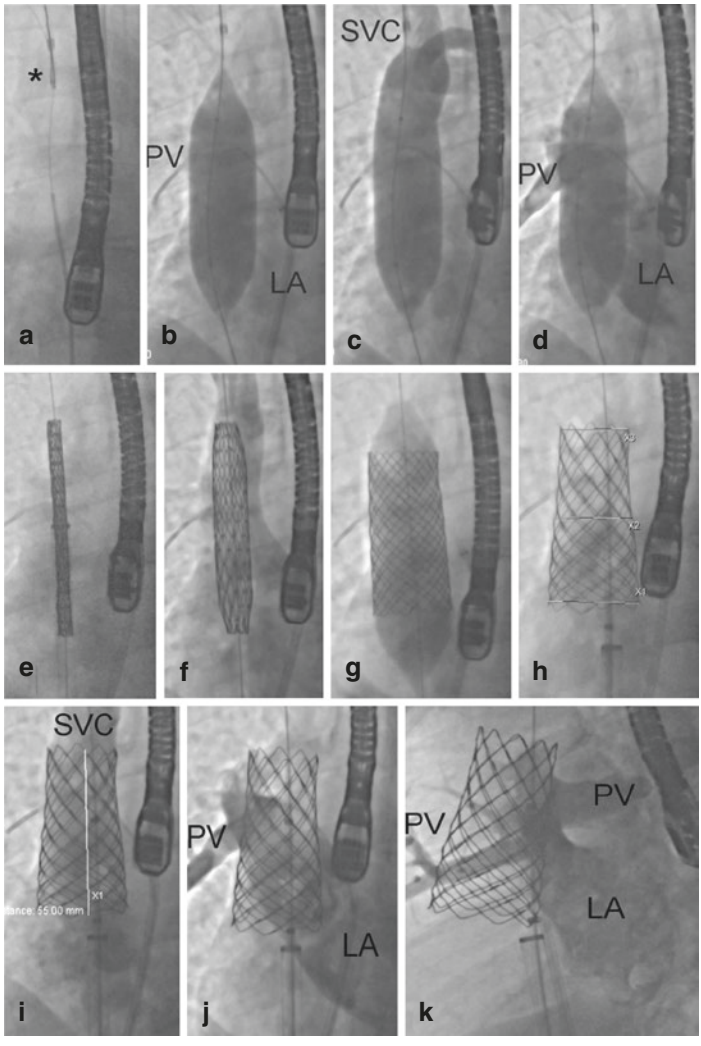

Fig. 41.6 Stent deployment steps: (**a**) Guide wire from femoral vein snared from jugular. (**b**) 30 mm non-compliant balloon inflated with SVC occlusion in (**c**) and patency of pulmonary veins to left atrium in (**d**). In (**e**) the 8 cm stent is exposed from the long sheath, in (**f**) the inner balloon and in (**g**) the outer balloon of the BiB are inflated. After further inflation with a Coda Balloon in (**h**), the stent has shortened to 5.5 cm (**i**). Pulmonary flow to the left atrium is unobstructed in (**j**) AP projection and (**k**) lateral projection with no leak into the right atrium

41.8 Materials

- 6 –8F vascular access sheaths
- Multipurpose and pigtail catheters.
- Vascular closure device or Z-suture.
- 260 cm 0.035″ Amplatz Extra-stiff wire, (Cook Medical, Bloomington, IN, USA)
- Brockenbrough transseptal needle (St. Jude Medical, St. Paul, MN).
- 8F SRO sheath (St. Jude Medical, St. Paul, MN, USA)
- Snaring catheters.
- 24 mm or 34 mm Amplatzer sizing balloons (St. Jude Medical, St. Paul, MN, USA)
- 6 cm AltoSa-XL PTA Balloons, diameter 16 – 30 mm (AndraTec GmbH, Koblenz, Germany)
- 85- cm 18F Check-Flo Performer (Cook Medical, Bloomington, IN, USA) or 65 cm 20F DrySeal sheath (Gore, Flagstaff, AZ, USA)
- 10-zig covered CP stent, length 6 – 8 cm (NuMED, Hopkinton, NY, USA)
- Balloon-in-Balloon catheter, length 6 – 8 cm, outer balloon diameters between 18 and 34 mm (NuMED, Hopkinton, NY, USA).
- 8-zig bare metal CP stent, length 2.8 – 3.9 cm (NuMED, Hopkinton, NY, USA) and appropriate Balloon-in-Balloon catheters
- Coda Balloon (Cook Medical, Bloomington, IN, USA).
- Atlas PTA Balloons 12 – 20 mm diameter (Bard Peripheral Vascular, Tempe, AZ, USA).

41.9 Tips and Tricks

41.9.1 Balloon Testing

- Distention of the native biological tissues is difficult to predict accurately in the printed or virtual 3D models. In vivo simulation by balloon inflation in the SVC–RA junction prior to stent implantation is important to evaluate the potential for

pulmonary venous obstruction after stent placement, as well as guiding the choice of balloon size for mounting the stent.

- The choice of the balloon for testing is important. Compliant sizing balloons can simulate flaring of the inferior part of the stent at the SVC–RA junction to achieve complete occlusion of the defect. If these eliminate the shunt without causing pulmonary vein obstruction then stent implantation can follow immediately. They may, however, also be overinflated leading to bulging of the balloon into the defect causing pulmonary venous obstruction. In these cases, balloon testing should be repeated using a non-compliant balloon 2 –4 mm larger than the SVC to re-evaluate the pulmonary venous drainage, accepting the likelihood of a residual shunt on imaging as the fixed diameter balloon will not fully simulate flaring of the stent at the SVC–RA junction (Fig. 41.5).

41.10 Monitoring and Protection of Pulmonary Venous Return

- To avoid pulmonary venous compression, pulmonary venous and left atrial pressure should be monitored continuously and flow into the left atrium should be assessed by TOE during balloon testing and stent deployment and flaring.
- To avoid interference with stent placement and the theoretical risk of catheter trapping, the RUPV is entered from the left atrium after transseptal puncture. Others have placed the catheter directly into the pulmonary vein from the transvenous route through the sinus venosus defect for balloon testing. There is no further monitoring if the catheter is removed before stent deployment or a risk of catheter trapping and stent displacement if the catheter is removed past the newly deployed stent. Alternatively, a retrograde arterial approach is possible, but may be cumbersome —and we have abandoned this approach.
- Accessing the RUPV from the left atrium facilitates protection of the pulmonary venous pathway during stent deployment and flaring (Fig. 41.7). If obstruction of the pulmonary venous return behind the stent appears likely, a high-pressure

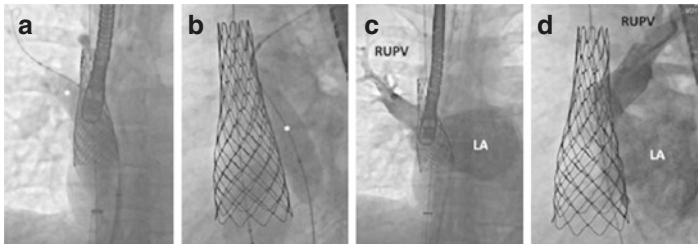

Fig. 41.7 Protection of the right upper pulmonary vein (RUPV) with an Atlas Gold balloon (*) during stent deployment and flaring preventing protrusion of the stent into the pulmonary venous pathway in same patient as in Fig. 41.5 (**a**, **b**). Pulmonary vein angiography confirms unobstructed flow into the left atrium (**c**, **d**). (Reproduced with permission from J Am Coll Cardiol 2020; 75: 1266–1280)

balloon (e.g. Atlas PTA Balloon, Bard Peripheral Vascular, Tempe, AZ, USA) at least 2 mm larger than the pulmonary vein can be placed in exchange for the angiographic catheter. The pulmonary vein balloon is inflated to at least six atmospheres while the SVC stent is deployed at a low pressure to allow "molding" of the SVC stent around the balloon in the pulmonary vein orifice. The stent balloon is deflated before deflating the pulmonary vein balloon. Subsequently further stent dilation and flaring at higher pressure during careful monitoring (with/without re-inflation of the pulmonary vein balloon) may be needed if this technique leaves a significant shunt. It is preferable to leave a tiny shunt than to risk pulmonary vein compromise as small shunts often seal over the next year.

- Pulmonary venous access from the left atrium also offers the potential for bail-out balloon dilation or even stenting in the event of unforeseen RUPV obstruction.

41.11 Choice of Stents, Positioning and Fixation

- The 10-zig covered CP stent can be inflated to a nominal diameter of 30 mm and up to 35 mm without tearing the covering, which may be needed at the RA portion to close the defect.

- The use of self-expanding stent grafts has also been reported infrequently. However, there is more control with balloon expandable stents, which do not have capacity for continued expansion, potentially impinging on the pulmonary vein orifice after deployment.
- The balloon-expandable stent is placed in an unobstructed SVC and flared in the right atrium. To prevent stent migration, the diameter of the stent should exceed the SVC diameter by 3 to 4 mm aiming for at least a 2 cm zone of apposition in the SVC.
- Despite this, stent migration may occur during the procedure. In those cases, it is usually possible to reposition the stent with the help of the delivery sheath and anchor it with an overlapping bare metal stent in the SVC. The anchoring stent is delivered through the RIJV sheath while the RFV delivery sheath secures the position of the covered stent (Fig. 41.8).
- The 6-cm 10-zig covered CP stent is currently the longest balloon-expandable stent with a CE mark. However, in many cases, the 6 cm stent will be too short to completely cover the defect in the right atrial portion and has a long enough zone of apposition in the SVC without using an additional anchoring stent. The custom-made 7- or 8- cm- long covered CP stents facilitate an adequate zone of apposition in the SVC and full coverage of the defect. The 6 cm stent can be used, when a small upper pulmonary vein that cannot be diverted to the LA is left above the covered stent. In those cases, a bare metal anchoring stent should be used if necessary, to allow continued drainage of this vein to the SVC.

41.12 Complications

Potential procedure- related complications are those as for any catheter procedure but stent embolization and pulmonary vein obstruction are specific to this procedure. Avoidance of femoral arterial access by using a radial artery cannula for hemodynamic

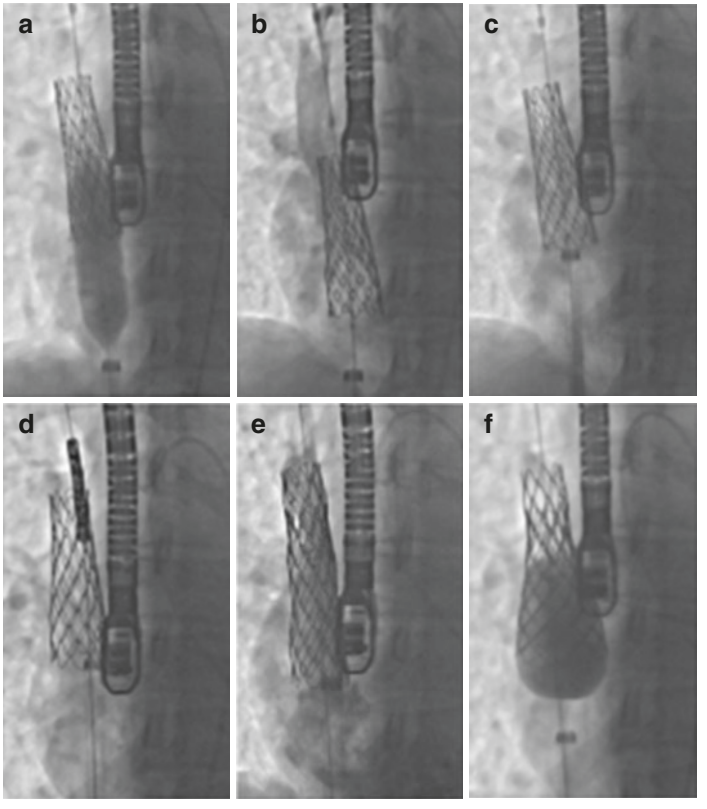

Fig. 41.8 Flaring of the inferior stent part (**a**) after deployment results in displacement inferiorly (**b**). The stent is re-advanced into position with the delivery sheath (**c**). An additional stent is passed from the jugular vein access to overlap the covered stent (**d**) and is deployed (**e**), allowing stable flaring of the covered stent (**f**). (Reproduced with permission from J Am Coll Cardiol 2020; 75: 1266–1280)

monitoring reduces vascular complications. Retrograde arterial access to the pulmonary vein can be time consuming with increased radiation exposure. Transseptal access may increase the risk of pericardial tamponade.

41.13 Post-procedural Care

- Dual antiplatelet therapy is recommended with clopidogrel 75 mg and aspirin 75 mg daily for 2 months followed by aspirin alone for a further 4 months.
- Patients with other indications for anticoagulation should remain anticoagulated (without additional antiplatelets) for at least 6 months.
- A PA and lateral chest x-ray and transthoracic echocardiogram are performed the day after the procedure.
- Clinical follow-up with transthoracic echocardiography and 12-lead ECG is recommended at 3 and 12 months after the procedure and at yearly intervals thereafter.
- A cardiac CT is performed after 2 to 3 months to confirm stent position and exclude pulmonary vein obstruction (Fig. 41.9).
- Right heart remodelling and quantification of any residual shunt is assessed with cardiac MRI after a year.

41.14 The Authors' Current Experience

- Between March 2016 and January 2020, we have successfully implanted stents in 29 adult patients.
- In an additional two patients who underwent balloon testing of the defect, there was a potential for pulmonary vein obstruction and we did not implant a stent —these patients underwent surgical repair.
- The stent embolised a few hours after the procedure in one patient. The stent was retrieved uneventfully during cardiac surgery to repair the defect.
- Follow up now extends to 4 years, and 16 patients have been followed up for more than a year. CT scans have confirmed the stent position and unobstructed pulmonary venous drainage. MRI scans have shown a reduction in RV size and no residual shunt in all except one patient with a residual leak. In this case, a 6 cm stent had been used and did not cover the caudal aspect of the defect fully.

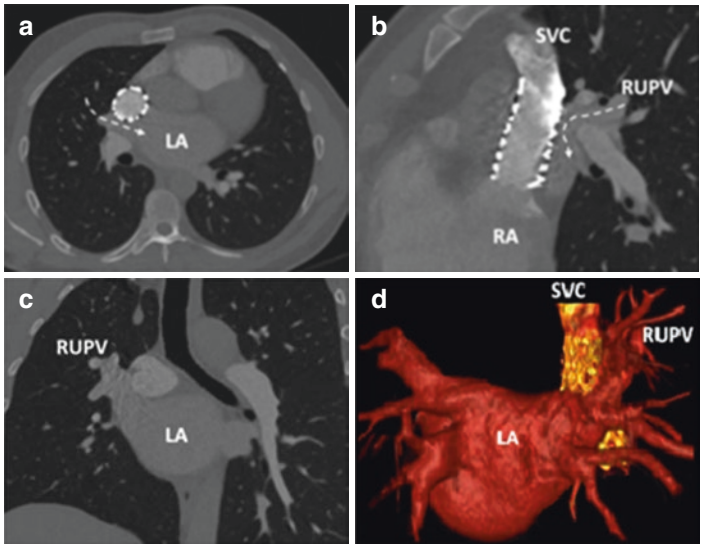

Fig. 41.9 Cardiac CT after SVC stent implantation. In the axial (**a**) and sagittal (**b**) views, the patent SVC stent and the unobstructed re-directed right upper pulmonary vein (RUPV, dotted line) draining into the left atrium (LA) posterior to the SVC stent are visualized. In the coronal view, the continuity of the RUPV to the LA is shown with the stent out of plane (**c**). The 3D volume rendered reconstruction (posterior coronal view) shows the RUPV draining into the LA behind the SVC stent. (Modified from J Am Coll Cardiol 2020; 75: 1266–1280)

41.15 Future Directions

This technique appears feasible in about 75 –80% of patients with this condition. We have not extended use into the paediatric age group but believe it would be appropriate in patients >45 kg who have undergone most of their growth. Others have used this in smaller children —but there is no follow- up information on the behaviour of the SVC and pulmonary veins during growth of the child.

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Part VII

Step-By-Step Procedures: Valve Implantation

42 Melody Valve Implantation in Pulmonary Position

Gianfranco Butera, Simon Thomas MacDonald, Massimo Chessa, and Philipp Bonhoeffer

42.1 Clinical Indications

Clinical indications for the treatment of pulmonary regurgitation and/or stenosis, whether surgical or percutaneous, are subject to ongoing discussions and there are no unifying guidelines [1, 2].

Despite of this, the consensus indications are:

1. RV systolic pressure >2/3 systemic with clinical symptoms or.
2. RV systolic pressure >3/4 systemic without clinical symptoms.

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3. Severe pulmonary valve regurgitation associated with one or more of the following.
   (a) RV dysfunction and/or dilatation (assessed on echocardiography and/or cardiovascular magnetic resonance imaging (cMRI)).
   (b) Decreased exercise capacity (peak VO_2 < 65–70% predicted for normal).
   (c) Arrhythmias: atrial or ventricular sustained arrhythmias.

42.2 Patient Selection

- Absence of active infection.
- Check and treat all sources of potential infection: ensure patient has had dental check.
- Complete a thorough analysis of previous medical history, cardiac catheterization and surgical interventions: in particular check previous surgical notes if available.
- Check renal and hepatic function, full blood count, CRP.
- Candidates for Melody valve have a conduit, bioprosthetic valve or an anatomical stenosis in the RVOT or main pulmonary artery [3–5].
- Check MR and/or CT. CT can give exact measurement of coronary position and assessment of calcification. MRI allows assessment of right ventricular size and volumes, shunts and differential flows.
- Check if associated anomalies are present: Pulmonary branch(es) stenosis, bifurcation stenosis, residual intracardiac defects, with coronary position also being at risk with potential compression during the procedure.
- The ideal candidate for Melody valve should have a dysfunctional previously implanted conduit ranging between 16 and 22 mm in diameter or previous bioprosthetic valve.

42.3 The Valve and the Delivery System

42.3.1 Device

The Melody trans-catheter pulmonary valve (Medtronic, US) is composed of a segment of bovine jugular vein with a thinned down wall and a central valve. The vein is sutured inside an expanded platinum-iridium stent with a length of 28 mm and a diameter of 18 mm that can be crimped to a size of 6 mm and re-expanded from 14 mm up to 22 mm (Fig. 42.1). The current stent design, which has an eight-crown zig pattern with six segments along its length, is reinforced at each strut insertion with gold weld. The venous segment is attached to the stent by continuous 5–0 polypropylene sutures around the entire circumference at the inflow and outflow portion, as well as discretely at each strut insertion. The suture is clear coloured for all the points except the outflow line, which is blue to signify the outflow end of the device. The venous segment is fixed in a buffered glutaraldehyde solution in a concentration low enough to preserve the flexibility of the

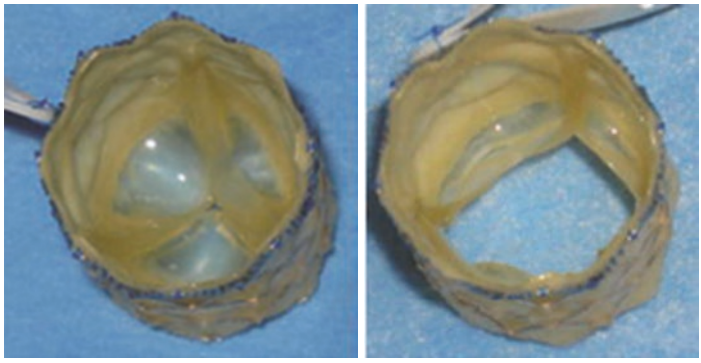

Fig. 42.1 Melody percutaneous pulmonary valve, in the close (left) and open (right) position, mounted on a platinum-iridium stent

venous valve leaflets. A final sterilization step is performed on the combined device using a sterilizing solution containing glutaraldehyde and isopropyl alcohol, in which it is then packaged.

42.3.2 The Delivery System

The delivery system, Ensemble, also manufactured by Medtronic MN, includes a balloon-in-balloon (BiB) design onto which the valved stent is front-loaded and crimped (Fig. 42.2). The system is available with three outer balloon diameters: 18, 20 and 22 mm. The tip of the system is blue to correspond with the blue outflow suture on the device to help with correct orientation. The body of the Ensemble system consists of a one-piece Teflon sheath containing a braided-wire reinforced elastomer lumen. The design minimizes the risk of kinking whilst optimizing flexibility and retaining the pushability required for the procedure. There is a retractable sheath which covers the stented valve during delivery

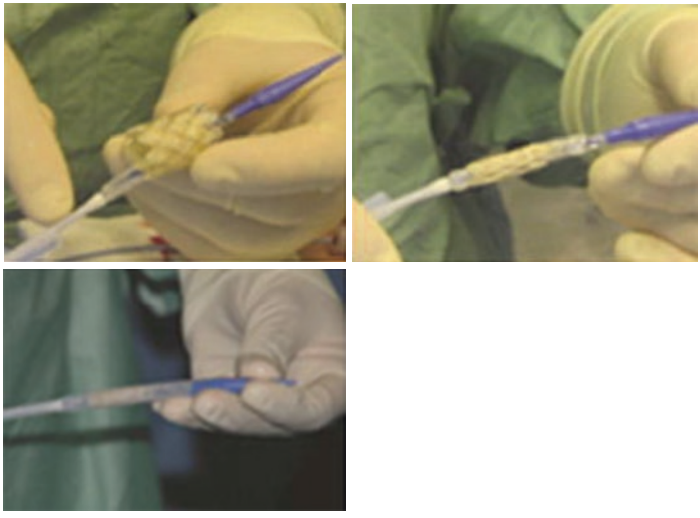

Fig. 42.2 Loading of the PPVI stent onto the delivery catheter

and is withdrawn just prior to deployment. Proximally, there are three ports, one for the guidewire (green), one to deploy the inner balloon (indigo) and one to deploy the outer balloon (orange).

42.4 Procedure

42.4.1 Preparation

- General anaesthesia and orotracheal intubation are generally preferred; deep sedation with propofol and local anaesthesia is also possible with appropriate staff and monitoring.
- Biplane catheterization laboratory preferred.
- Patient position with arms lifted up, behind patient's neck (attention to brachial plexus and overstretching).
- Patient is fully monitored including an arterial line for continuous arterial pressure monitoring, two peripheral venous lines or a central venous line, urinary catheter for urine output.
- Full heparinization with Heparin IV 100 UI/kg. Check every 30 min that the activated clotting time is >250 s. Further boluses of heparin iv may be needed during the procedure.
- Antibiotics IV: Usually a cephalosporin or flucloxacillin and gentamicin.
- The procedure has to be considered a surgical intervention. Special care has to be paid to strict aseptic technique. Attention has to be given to operators' scrubbing and patient's preparation (including careful depilation). The personnel involved have to wear masks and hats.
- Cross matching of blood. Provisionally up to 4 units is generally needed in case of vessel/conduit rupture.

42.5 Access Site

- Usually femoral venous access is used. Sometimes internal jugular access can be used.
- Both sides for vascular femoral access are prepared.

- A 12 Fr femoral sheath is placed: this avoids the need for changing the femoral sheath during diagnostic catheterization, balloon sizing, balloon testing and angiography with 6 Fr Multitrack catheter (NuMED Inc., Hopkinton, NY) or 9 Fr Mullins sheath (*Cook Europe, Bjaeverskov, Denmark).*
- Arterial access is obtained by using a 5Fr sheath.

42.6 Catheterization and Haemodynamic Evaluation

- Right heart catheterization is performed by standard techniques to assess pressure and saturations with a right coronary catheter, JR 4, or any other catheter with a curved tip.
- Routinely, pressure measurements are obtained in the right ventricle, pulmonary artery and aorta with additional measurements, for example, in the branch pulmonary arteries.
- A 0.035″super-stiff guidewire is then positioned in a distal branch pulmonary artery to provide an anchor over which the delivery system can be advanced. It is important to avoid as much as possible curves, to place the tip of the wire as distal as possible (ideally in the pulmonary arterial bed at the level of the diaphragm), to make sure the wire is not passing through the chordae of the tricuspid valve (check by using a multitrack catheter or a balloon tipped catheter with the balloon inflated (e.g. 7Fr Swan Ganz catheter that takes a 0.035″ wire).
- The preferred guidewire is usually a 0.035″ exchange wire (Amplatz Ultra-Stiff wire 260 cm long (*Boston Scientific Corp., Natick, Massachusetts*), Lunderquist 260 cm long (*Cook Europe, Denmark*) or Back-up Meier 300 cm long (*Boston Scientific Corp., Natick, Massachusetts*).
- Special care has to be paid to wire stability and position. This should not move throughout the procedure. If during any phase of the procedure the wire moves and the position is not satisfactory, it should be replaced in the ideal position again using a catheter. Do not push forward the stiff wire by itself due to the risk of pulmonary vascular bed injury.

- Angiography is performed using a 6 Fr Multi-Track catheter (NuMED Inc., Hopkinton, NY) or through a 9 Fr Mullins long-sheath with the tip placed just beyond the pulmonary valve. In the latter case, a pigtail with radiopaque markers is placed across the wire inside the Mullins sheath in order to allow for precise measurements (Fig. 42.3).
- Angiography is performed on the pulmonary artery trunk, right ventricle and ascending aorta (Fig. 42.3).

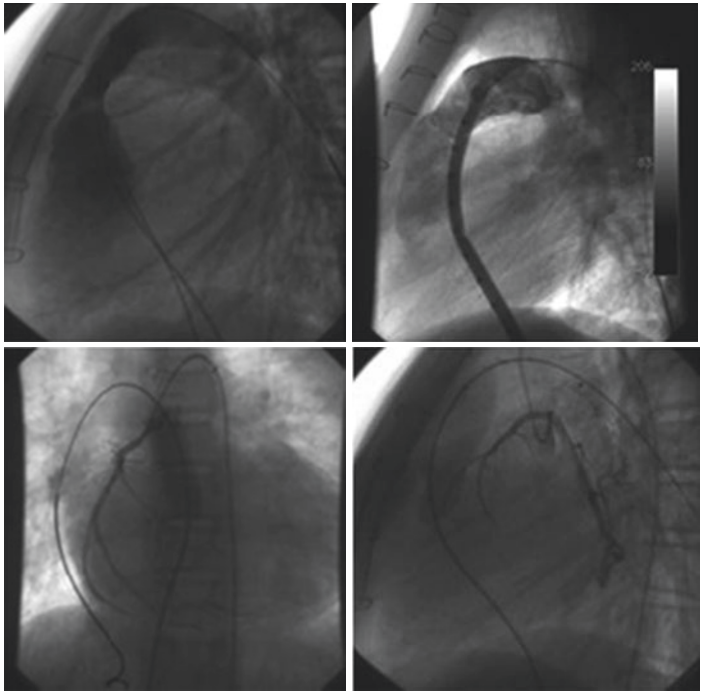

Fig. 42.3 Left upper: Angiography in lateral view showing severe stenosis and regurgitation at the level of the RVOT; right upper: Angiography in lateral view of the RVOT through the lateral port of the Mullins sheath after pre-dilatation of the RVOT; left and right bottom: balloon RVOT testing during simultaneous coronary angiography

- Usually, the following imaging planes are used:
  - *Lateral view*:
    Visualize anterior chest, landing zone and proximal end of pre-stented outflow tract.
    Useful to check coronary artery position during balloon testing.
  - *Anterio-posterior view with cranial angulation usually with left anterior oblique angulation on biplane:*
    Visualize relationship of pulmonary bifurcation to distal end of stent.
- If needed, selective coronary artery angiography is performed.
- Delineate landing zone and decide final target diameter to be achieved with Melody valve.

In case of doubt on the characteristics of the landing zone, it can be useful to do the following:

- Use a sizing balloon (e.g. 34 mm Amplatzer ASD sizing balloon (NuMED Inc., Hopkinton, NY) or 25 mm PTS-X balloon (NuMED Inc., Hopkinton, NY)). This gives a precise negative image of the RVOT and landing zone.
- Then use a low-pressure angioplasty balloon for example, a Z-Med II balloon (NuMED Inc., Hopkinton, NY) or Crystal balloons (Balt, Montmorency, France) to check RVOT/conduit tissue distensibility.
- As a further step, if tissue looks poorly distensible with low-pressure balloon, use a high-pressure balloon Mullins X-Ultra high-pressure balloon catheter (NuMED Inc., Hopkinton, NY), Atlas PTA balloon dilatation catheter (Bard Peripheral Vascular Inc., Temple, AZ, US), True balloon (Bard Peripheral Vascular Inc., Tempe, AZ, US).
- In this latter case, two scenarios should be considered:
  - Presence of a lesion with high risk of tear. This can be anticipated in cases with a heavily calcified lesion.
  - Potential for coronary compression.
- If there is a possibility that a coronary artery is at risk of compression from valve implantation, coronary angiography is performed with an angioplasty balloon inflated simultaneously

in the right ventricle to pulmonary artery conduit. If there is a high risk of coronary arterial compression, valve implantation should not be attempted and the patient should be referred for surgery.

  - Selective coronary angiography is preferred in multiple projections (Fig. 42.3).
    If coronary arteries are distant, low pressure will be enough.
    If coronary arteries are close, full inflation (with either low-pressure or high-pressure balloons) is indicated.
    Note: Care is required in case this causes a conduit tear.

- Control RVOT angiography is performed to rule out possible extravasation:
  - If a tear is confirmed then covered stents (CP covered stents (NuMED Inc., Hopkinton, NY)) should be implanted covering not only the conduit length but going 1–2 zigs below and above the conduit length.
  - Initially there is no need to fully open stent (especially if danger of coronary compression): appose stent to wall and flare ends against wall to seal tear.

42.7 Pre-Stenting

Exchange the 12 Fr sheath for an 18 Fr venous sheath in order to facilitate the exchange of balloons and long sheaths for pre-stenting (Fig. 42.4). A 65 cm 24 Fr Gore® DrySeal Flex Introducer Sheath (WL Gore and Associates, Flagstaff, AZ, US) can also be used.

42.8 Stent Choice

- Bare stent (CP stent (NuMED Inc., Hopkinton, NY), Intrastent (ev3, Inc., Plymouth, MN), AndraStent (Andramed, Reutlingen, DE).
  - When no extravasation is seen.
  - Full balloon expansion achieved by pre-dilatation.

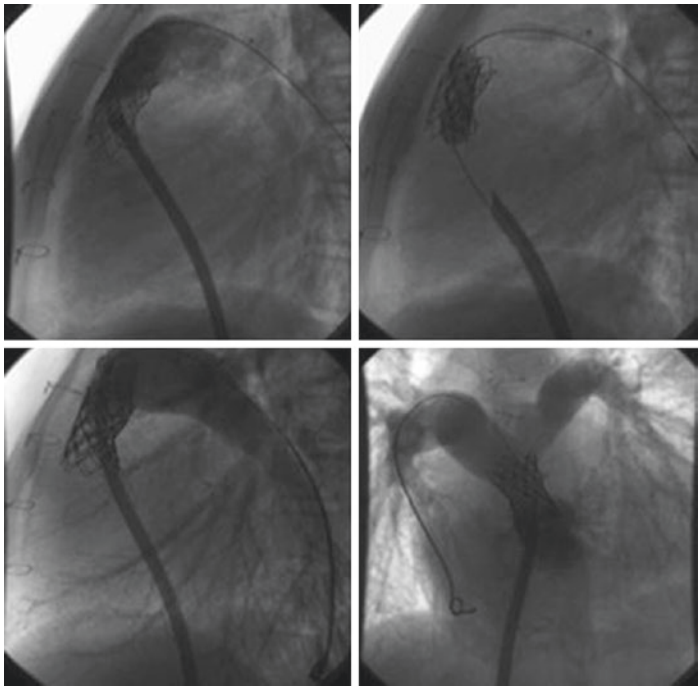

Fig. 42.4 Left upper: Angiography in lateral view after pre-stenting; right upper: Ensemble is in place and inner balloon is inflated; left bottom: Melody is correctly placed and angiography in the pulmonary trunk shows no pulmonary regurgitation; right bottom: Angiography in antero-posterior view with cranial and left anterior oblique angulation: normal flow in both pulmonary arteries

- Covered stent (CP covered stent (NuMED Inc., Hopkinton, NY).
  - When extravasation is seen.
  - Anticipated risk of conduit/outflow tract tear or fracture.

Length of the stent should be enough to cover the stenotic area and the entire length of the Melody valve.

42.9 Stent Implantation

- Use a 14 Fr Mullins long-sheath.
- If a bare stent is used and there is jailing of a PA, it may be possible to dilate cells with an Atlas ultra-high-pressure balloon >20 atmospheres to re-open the stent into the PA (it is usually preferred to use open-cell or a partially open celled stent such as the AndraStent).
- If a covered stent is used, special care should be observed to avoid jailing. Furthermore, as covered stent use has the aim of avoiding extravasation, it is important to cover not only the conduit length but to go with 1–2 zigs below and above the conduit. Finally, the ends of the covered stents have to be flared to get maximal apposition of stent against wall to seal any expected tear.
- During stent implantation, record on fluoroscopy both balloon inflation and deflation. In case of significant recoil or presence of compressing forces on the stent, implant a second or a third (or even more) stents until the area looks stable (this is in order to avoid the risk of Melody valve stent fracture during follow-up).
- In case of any doubt, repeat coronary angiogram before further dilations.
- Dilate stent until desired internal diameter is achieved.
- Re-check haemodynamics (RV and PA pressure and gradient).
- Repeat RVOT and PA angiography for extravasation.
  - Place additional covered stent(s) in the case of extravasation.

42.10 Melody Valve Implantation

- Prepare Melody valve and Ensemble.
- The valve is taken out of the packaging by using sterile forceps to avoid contamination.
- The valved stent is prepared in three sequential saline baths (5 min in each) to wash off the glutaraldehyde, in which it is stored.

- The label of the valve is removed.
- The size of the valved stent is reduced by crimping it to increasingly smaller sizes prior to front-loading onto the delivery system. It is recommended to use a 2.5 ml syringe for crimping to an intermediate size prior to the final crimping onto the balloon catheter.
- The blue stitching on the distal portion of the device is matched to the blue portion of the delivery system and verified by an independent observer to guarantee correct orientation of the valve.
- Further hand crimping of the device onto the balloon is performed, following which the sheath is advanced to cover the stent whilst a saline flush is administered via the side port to remove air bubbles from the system.
- Special care is paid during all these phases concerning strict asepsis.
- Predilate the groin with 22 and 24 Fr dilators.
- Insert the Ensemble in the groin and flush the haemostatic sleeve valve at the access site.
- Advance Melody to the preferred landing zone:
  - It is important to have a simultaneous view of right atrium, target zone and distal tip of wire. Choose the best view to have all at once. Usually the lateral or (sometimes better) the anteroposterior with cranial angulation view.
  - Push the Ensemble from the groin: usually, keeping the wire fixed, it advances quite easily. This is particularly the case when pre-stenting/dilatation has been performed.
  - Sometimes things are more difficult. The following tricks can be used:

    Ensemble dilator may encroach into the RVOT, because of the angle and of the characteristics of the Ensemble itself. In these cases, pushing on the Ensemble will pull back the wire.

    Push on Ensemble while applying gentle traction on the wire (beware of significant wire coming back). With this manoeuvre, usually the dilator comes away from the RVOT wall and allows advancement.

Push on the guidewire/delivery system in order to create a loop in the right atrium. This manoeuvre changes the angle and may help entering in the RVOT. Once the system has passed the target zone, straighten the wire in order to avoid any interference with tricuspid valve function.

With the right atrial loop in place, pull back the Ensemble/guidewire system. This may help jump the system into the landing zone.

Consider changing wire position into the other pulmonary artery.

Consider using internal jugular vein access.

A 65 cm Gore® DrySeal Flex Introducer Sheath or similar may reach beyond the RVOT in smaller patients and ease the Ensemble getting into position, the DrySeal sheath being pulled back to reveal the Ensemble in position.

- Uncover the Melody valve by pulling back the sheath to the double marker on the Ensemble system. Usually, there can be slight forward movement of the balloon/melody valve with this. Sometimes the operator cannot uncover the valve because of too much friction. In this case, the Ensemble can be advanced further into the PA, the system is straightened and the valve partially uncovered. Then the Melody/balloon system can be pulled back in the landing zone.
- Position and complete uncovering of the valve can be further checked by hand contrast injection through the side arm of the Ensemble.
- Partial deployment of the stent is achieved by hand inflation of the inner balloon. The Ensemble may move forward when the inner balloon is inflated (Fig. 42.4).
- After final confirmation of the position, the outer balloon is also hand inflated to complete deployment.
- The balloons are deflated (inner balloon first) and the delivery system withdrawn carefully and slowly.
- The Ensemble is exchanged for the 18 Fr femoral venous sheath.

- Pressure measurements are obtained to confirm the result.
- Angiography is performed (Fig. 42.4).
  - In the RVOT in order to show if any extravasation of contrast has occurred and should be performed in case of any hemodynamic instability.
  - In the main pulmonary artery to show competence of the valve and normal flow in the two pulmonary arteries (use the AP with cranial angulation ± LAO view).
- Post-dilatation of the valve may be needed in the presence of a residual gradient (>20 mmHg) and incomplete expansion of the valved stent.
  - Verify if the gradient is caused by the valve diameter (limited by the pre-existing conduit, which may be insufficient for patient size/body surface area) or another sub- or supravalvular structure.
  - Use an appropriately sized ultra-high–pressure balloon (e.g. Atlas Gold) with a maximum balloon size of 24 mm.
- Obtain pressure measurements to confirm the result and satisfactory gradient.
- Final angiography is performed.

42.10.1 Complications

A rate of 4–6% of serious complication has been reported [3, 6, 7].

- Device instability and stent migration/embolization: This is a very rare event. If it occurs, surgery is usually needed. It is possible to attempt to recapture and reposition any stent/valve if the guidewire is still correctly in place. In the case of distal migration, the stent/valve can be dilated in the branch pulmonary arteries, in proximal migration they can be deployed in the IVC with care (Fig. 42.7). The Melody valve acts as a covered stent and there is thus a risk of branch vessel occlusion, however. A vavuloplasty balloon can be used to destroy the valve as appropriate.

- Homograft rupture: It has been reported previously how to avoid this complication. If bleeding occurs (hemothorax), a chest tube is placed and auto-transfusion should be initiated as soon as possible to re-establish a sufficient circulation for further intervention or "wait and see" approach. Acute thoracotomy is not usually advised as decompression of the chest may exacerbate bleeding and lead to later difficulty in locating the source of bleeding. If it possible to identify the rupture point, covered stents can be implanted [8]. It may be possible to buy time by inflating a PTS-X balloon within the homograft as surgeons are called. In high-risk cases, prior femoral and venous access can allow insertion of large French ECMO sheaths to allow an ECMO circuit to be set up as surgical intervention is instigated.

Strategies to avoid RVOT conduit rupture include sequential balloon dilation, starting 2–3 mm $\geq$ the stenotic area, and increasing by around 2 to 3 mm with each balloon. Repeated conduit angiography between angioplasties is performed. If possible, avoid over-dilation of the conduit beyond the implanted diameter.

Once coronary artery anatomy assured, covered stent (s) can be implanted.

- Compression of the coronary artery: Careful evaluation of the implantation site, RVOT distensibility and of coronary artery anatomy are routinely performed prior to Melody valve implantation. This is usually done with cMRI or CT scanning study and by performing balloon testing of the RVOT simultaneously with coronary angiography. Sometimes this assessment has not been enough to avoid complication however.
- Injury to a distal branch pulmonary artery or tricuspid valve. Damage to distal pulmonary artery branches can be minimized by ensuring stable guidewire positioning at all times, and avoidance of damage of the tricuspid valve can be achieved by use of a balloon catheter for the initial manoeuvring of the catheter through the right heart. In case of damage to a

pulmonary branch, the approach is similar to that of conduit rupture (Fig. 42.5). Compliant balloon (e.g. PTS-X balloon) inflation in a branch PA will be better tolerated with ventilation and perfusion of the opposite lung buying time.

- Melody stent fractures (Fig. 42.6): These are a potential complication of all cardiovascular stent applications. In PPVI procedures, the prevalence of stent fractures can be as high as 21–40%, at up to 3 years follow-up [9, 10]. Implantation in a native RVOT, absence of RVOT calcification, and qualitative recoil of the valved stent just after implantation may be

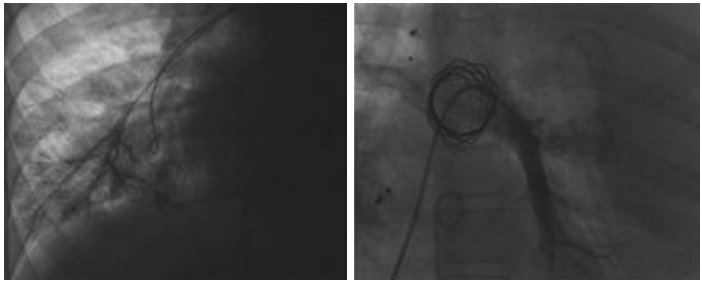

Fig. 42.5 Perforation of distal (right) or main (left) pulmonary artery

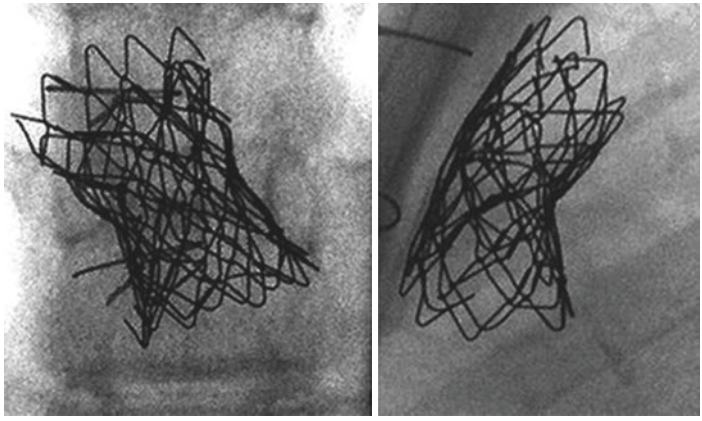

Fig. 42.6 Type II stent fractures in anteroposterior and lateral view

predictors of stent fracture. A classification that can guide management has been formulated by Nordmeyer, whereby type I fractures (no loss of stent integrity) can be managed conservatively, type II fractures (loss of stent integrity with echocardiographic signs of restenosis) should be considered for repeat PPVI (valve-in-valve procedure) or surgery, and type III fractures (separation of fragments/embolization) necessitate surgery [9].

Pre-stenting with a bare metal or covered stent reduces the risk of stent fractures. Pre-stenting has to be performed until recoil is eliminated (Fig. 42.7).

Serial radiographic and echocardiographic follow-up is mandatory to detect and monitor stent fractures and facilitate timely intervention. Fluoroscopy is a useful adjunct to assess fractures and stent stability. Repeat Melody implantation can be performed for stent fracture and residual stenosis. The procedure is feasible and has excellent and sustained haemodynamic results.

- Endocarditis has been documented on both the venous wall and the valve itself (annual risk of endocarditis approximately 2.4% per patient year) [6, 11] The Melody valve, being a bovine-based valve, may be more prone to endocarditis than compared to a homograft or Edwards percutaneous pulmonary valve [12, 13]. Age at time of implant and degree of residual RV-PA obstruction may be factors but care is required regarding overexpansion at time of implant and conduit rupture [14]. Long-life antibiotic prophylaxis is mandatory afterwards with subsequent invasive dental or other surgery. It may or may not result in device dysfunction, and surgical or medical management strategies should be employed accordingly [15].
- Vascular access complication: as can be found in other catheterisation procedures. Using ultrasound to obtain access in femoral vessels, particularly in patients with previous interventions via this route, is particularly useful. In acute femoral bleeding, obtaining access in the opposite femoral vessel and crossing over using a wire and compliant balloon (e.g. PTS-X)

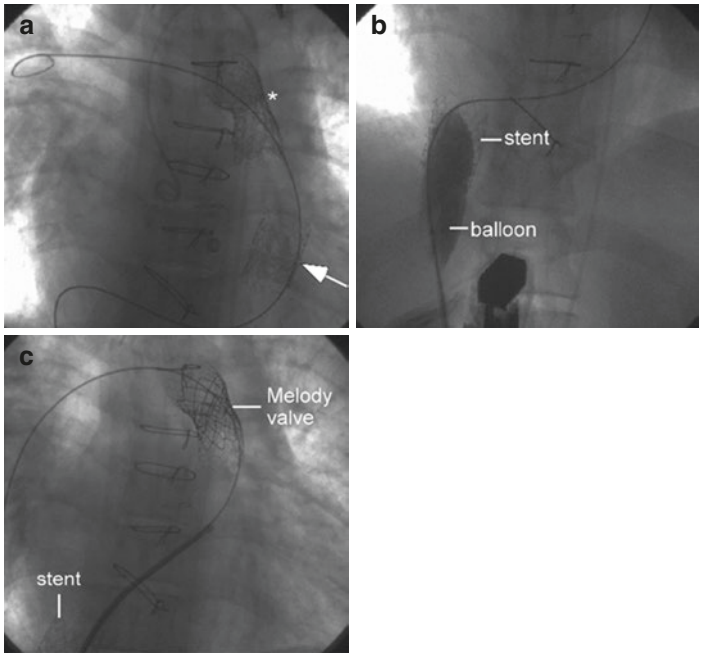

Fig. 42.7 Proximal stent migration, panels A-C. An AndraStent 39 mm XXL Cobalt Chromium stent (Andramed, Reutlingen, DE) has migrated proximally into the right ventricle during pre-stenting. Stenting the distal outflow tract end failed to capture it and prevent further proximal migration (*, **a**). Using a partially inflated balloon, it was manipulated into the IVC where it was finally deployed (**b**). A Melody valve was subsequently implanted in the correct position, severe pre-existing tricuspid regurgitation and annular dilatation facilitating embolised stent manipulation (**c**)

may stop flow temporarily, allowing haemostasis or buying time until surgical evaluation. It may also allow angiography to be performed to assess the vessel and assess the bleeding point.

- The rate of death, re-operation, or re-intervention after 48 h post Melody implantation has been quoted as 4.2% per person per year [6, 16]. The US Melody Valve Investigational Device Exemption Trial quoted a 5-year freedom from intervention of 76.4%, and explantation 92%. Further long-term evaluations are ongoing.

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43 SAPIEN XT Valve Implantation in the Pulmonary Position

Noa Holoshitz and Ziyad M. Hijazi

43.1 Introduction

The SAPIEN XT valve is made up of three bovine pericardial leaflets, hand sewn into a cobalt–chromium, balloon expandable stent (Fig. 43.1). A fabric skirt covers the lower end of the stent to achieve a seal with the calcified conduit and prevent paravalvular leak. The valve has been designed to reduce leaflet stress and maximize coaptation. The pericardial tissue is processed with ThermaFix anti-calcification treatment, which is the same treatment used in the surgical valve, the Carpentier-Edwards PERIMOUNT Magna valve. The SAPIEN XT valve is currently available in 23, 26, and 29 mm diameter with heights of 14.3, 17.2, and 19.1 mm, respectively. It can therefore be used in conduits measuring up to 29 mm at the time of transcatheter valve replacement.

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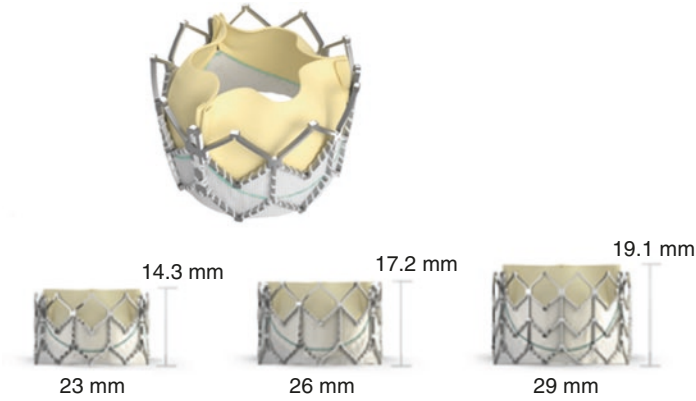

Fig. 43.1 The Edwards SAPIEN XT transcatheter heart valve. Top, the valve shown enface and bottom, from the side is available in 23, 26, or 29 mm diameter. It consists of three bovine pericardial cusps mounted into a cobalt-chromium balloon expandable stent

The delivery system used with the SAPIEN XT valve is the Novaflex+. It has a tapered nose cone-shaped balloon catheter with a deflectable tip (Fig. 43.2) to facilitate delivery to the RVOT. It requires either a 14 Fr expandable sheath for the 20, 23, and 26 mm valve or 16 Fr expandable sheath for the 29 mm valve. With this delivery system, the valve is crimped proximally to the balloon tip, which allows for a smaller delivery profile. Once the delivery system is past the sheath, in a straight part of the vena cava, the balloon is pulled back onto the crimped valve. The delivery system includes a flex wheel for articulation of the flex catheter and a flex indicator on the handle. A valve alignment wheel allows for fine adjustment of the valve between the valve alignment markers after it is retracted onto the balloon.

Currently, the next generation of the SAPIEN valves, the S_3, is being used routinely in the aortic position in the USA. This valve was designed to reduce post-procedural paravalvular leak by the addition of an outer skirt at the distal part of the prosthesis. In addition, the S3 valve is also available in a 20 mm size and the profile of the stent was reduced slightly. While the S3 valve is not currently approved in the US for use in the pulmonic position, it

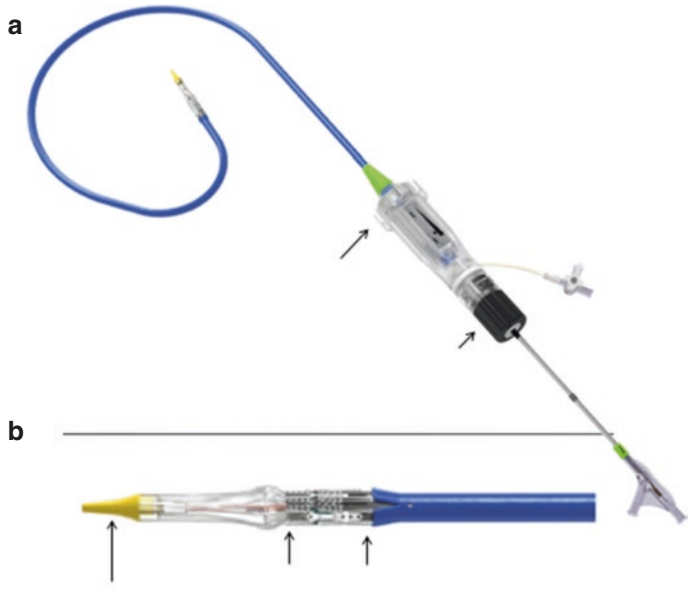

Fig. 43.2 (**a**) The Novaflex plus delivery system. Long black arrow shows the rotating wheel used to flex the catheter and may facilitate passage through tortuous tract. Short black arrow denotes the wheel used to align the valve across the balloon. (**b**) close-up view of the nose cone (long black arrow) and where the valve is crimped on the shaft of the catheter (two short black arrows) proximal to the balloon to decrease the profile of the delivery system

is being used "off label" in certain patients who are thought to benefit and it is widely used in Europe and Canada. Short-term (two-year) data on 82 patients from Europe and Canada showed good procedural hemodynamic outcomes and low complication rate (only 2 out of 82 patients) [1]. The COMPASSION S3 trial is currently recruiting patients in the US to evaluate the efficacy and safety of S3 use in the pulmonic position.

43.2 Anatomic Description and Pathophysiology

Congenital heart disease affects up to 1 in every 100 live births in the USA, and the tPVR procedure has provided a less invasive option to many of these patients instead of an additional cardiac

surgery. It is performed primarily in patients with an RV to PA conduit and/or a bioprosthetic pulmonic valve.

In patients with a congenital RVOT obstruction, surgical implantation of an RV to PA conduit has allowed for the treatment and palliation of complex congenital heart disease that was previously untreatable. It has thereby contributed to the current survival rate of over 85% of congenital heart disease patients into adulthood. Patients with cardiac anomalies afflicting the RVOT including pulmonary atresia with ventricular septal defect, tetralogy of Fallot, and truncus arteriosus need surgical correction with a conduit in the early neonatal period to improve blood flow to the lungs. Conduits are also used in patients with congenital aortic valve abnormalities, when undergoing the Ross procedure (autotransplantation of the native pulmonic valve in the aortic position and placement of a conduit between the right ventricle and pulmonary artery instead of the pulmonic valve used for the aortic position).

tPVR may also be performed in patients whose native RVOT was repaired surgically with a patch, such as patients with less severe forms of tetralogy of Fallot. The native RVOT can be stented, thereby creating a "conduit" between the RV and PA prior to valve implantation. It is this group of patients we believe that constitutes the largest population who may benefit from such technology.

43.3 Clinical Scenarios

Conduit degeneration and prosthetic valve dysfunction are indolent disease processes. Symptoms of RVOT obstruction may develop slowly over the course of several years, if at all, before intervention is indicated. Symptoms typically include shortness of breath, fatigue, and symptoms of heart failure. Patients may also present with dizziness, syncope, or even sudden cardiac death if arrhythmias are present. However, many patients who are followed routinely by a cardiologist may develop conduit or prosthetic valve dysfunction and remain asymptomatic for many

years. It is then up to the treating cardiologist to determine the optimal time for tPVR or surgery, based on the indications outlined in the next section.

43.4 Indications and Patient Selection

The 2010 American Heart Association statement on the Indications for Cardiac Catheterization and Intervention in Pediatric Cardiac Disease was expanded to include a class IIa indication for tPVR [2]. It recommends that "It is reasonable to consider percutaneous pulmonary valve replacement in a patient with an RV-to-PA conduit with associated moderate to severe pulmonary regurgitation or stenosis provided the patient meets inclusion/exclusion criteria for the available valve. (Level of Evidence: B)."

The inclusion and exclusion criteria for the SAPIEN valve trial are summarized in Table 43.1. These criteria were based on surgical

Table 43.1 Inclusion and exclusion criteria for the Edwards SAPIEN valve trial

Inclusion criteria
Weight >35 kg
In situ conduit >16 and <24 mm
Dysfunctional RVOT conduit:
>3+ PR by transthoracic echocardiogram
Pulmonary regurgitant fraction >40%
With or without pulmonic stenosis
Exclusion criteria[a]
Active infection requiring antibiotics
History of or active endocarditis
Intravenous drug abuse
Preexisting prosthetic heart valve in any position
Pregnancy
Severe chest wall deformity
Echocardiographic evidence of intracardiac mass, thrombus, or vegetation
Known intolerance to aspirin or heparin

[a]Multiple exclusion criteria, please see http://clinicaltrials.gov for full list

indications for RVOT revision. However, it is important to note that there is some controversy regarding the optimal timing of surgery to prevent irreversible RV damage. The typical criteria that we use for asymptomatic patients include a pulmonary regurgitant fraction of >40%, RV ejection fraction <40%, and an indexed RV end-diastolic volume >150 ml as determined by cardiac MRI. However, if the patient is symptomatic due to severe pulmonary regurgitation or stenosis, then such criteria is not strictly enforced. Furthermore, the QRS duration in patients with severe pulmonary regurgitation should be taken into account. A QRS duration >180 ms is associated with ventricular arrhythmias, and sudden death, and is considered an indication for intervention.

43.5 Treatment Options

In adults, conduit replacement becomes necessary on average 10–15 years postsurgical implantation, but in children this time interval may be considerably shorter. Therefore, patients who had their first conduit placed during infancy may require four or more operations over their life span. Given the significant morbidity and mortality involved in redo operations in the setting of RV failure, a less invasive alternative is desirable.

tPVR is therefore a good option for patients requiring pulmonic valve intervention who meet the criteria listed above. It is important to remember, however, that even when patients meet criteria for tPVR and are seeking out a less invasive alternative to surgery, if their anatomy is not suitable (i.e., conduit or RVOT is too large or too small for available valves or not a long enough landing zone between the MPA and branch PAs), surgical conduit or valve replacement is still the gold standard.

There are currently two approved available valve systems for tPVR in the USA. Other than the SAPIEN XT valve, which is the focus of this chapter, the Melody valve is widely used in the USA for tPVR. The Melody valve (previously described in this book) is made of a bovine internal jugular vein and valve, sewn inside a platinum-iridium stent.

Both available valve systems have their unique benefits and drawbacks as summarized in Table 43.2. The SAPIEN XT valve is available in larger sizes than the current Melody system and therefore may be appropriate for placement in larger conduits, which may be found in older patients. It is also important to remember that it is not the original conduit size, but the degree of narrowing which determines the final size of the valve implanted. The SAPIEN XT valve has a shorter height than the Melody valve, which may be beneficial in certain anatomies; however, pre-stenting is necessary in order to give an adequate landing zone. The Melody delivery system, however, is less bulky, and the retractable sheath protects the valve until it is deployed in the desired location. The bulkier delivery system of the SAPIEN XT valve makes it potentially more difficult to implant, especially in patients with a tortuous RVOT. Careful consideration must be given to the likelihood of procedural success before attempting valve implantation because the SAPIEN XT system does not use a covering sheath; therefore, once it exits its short delivery sheath (35 cm) positioned in the inferior vena cava, it may be difficult to retract inside the sheath. Some operators have been using a 22–24 Fr, 65 cm DrySeal sheath (WL Gore) for valve delivery. This ensures that the valve is housed inside the sheath until the target zone is reached.

Table 43.2 Comparison of Melody and the SAPIEN valves

Characteristic	Melody valve	SAPIEN XT valve
Stent material	Iridium 10%, platinum 90%	Cobalt-chromium
Valve material	Bovine jugular vein	Bovine pericardium treated with ThermaFix
Available size (diameter)	18–22 mm	23, 26, 29 mm (SAPIEN 3 available in 20 mm)
Stent height	34 mm	14.3, 17.2, 19.1 mm
Delivery sheath size	22 French	14 French (23 and 26 mm), 16 French (29 mm)

43.6 Pre-procedural Imaging

Echocardiography (echo) is usually the first imaging test performed in patients who may be candidates for tPVR. From the initial echo, the patients' right and left ventricular function can be evaluated as well as the amount of pulmonic insufficiency using color and continuous wave Doppler. If there is concern that PV intervention may be indicated, cardiac magnetic resonance imaging (MRI) is the next imaging test which should be ordered. Cardiac MRI has become a vital component of patient selection for tPVR. It is important that centers performing these procedures have access to a team of physicians and radiology technicians who have been trained at performing and interpreting congenital cardiac MRI. The MRI can help evaluate the degree of pulmonic valve dysfunction by calculating the regurgitant fraction, RV ejection fraction, and end-diastolic dimensions. Moreover, valuable information about the patient's anatomy can be obtained by MRI such as native RVOT dimensions, degree of conduit stenosis, and distance of the coronary arteries from the outflow tract or conduit, which is a critical step in the evaluation.

43.7 Technique (Step by Step)

In the USA, tPVR is typically performed under general endotracheal anesthesia. However, the procedure has been performed under conscious sedation in Europe with good results. The femoral vein is the preferred route of delivery, but it is also possible to deliver the valve through the internal jugular vein. We typically start off with a 7 French venous sheath, which is later upsized to the larger delivery sheath, based on the size of the valve chosen. Arterial access is also obtained (5 or 6 French) for aortic root or selective coronary angiography. Once access has been established, intravenous heparin is administered for a goal activated clotting time of >200 s. The research protocols also include starting the patients on 81 mg of aspirin (for adult patients) the night

prior to the procedure; however, this is not something that we routinely do in our practice. All patients should be given antibiotic prophylaxis per protocol.

1. Standard right heart catheterization is carried out to evaluate the baseline hemodynamics and the pressure gradient across the dysfunctional conduit.
2. Angiographic evaluation of the RV–PA conduit is performed through a side hole catheter with biplane fluoroscopy to assess the degree of pulmonary regurgitation, the shape of the conduit, and presence of calcifications (Fig. 43.3a).
3. The minimum diameter of the conduit is measured by inflating a sizing balloon across the pulmonic valve.
4. Aortic root angiography or selective coronary angiography is carried out with simultaneous balloon inflation in the RVOT to evaluate for coronary artery compression (Fig. 43.3b). This step is crucial given the higher prevalence of coronary artery origin anomalies in patients with congenital heart disease. It is important to assure that the final conduit diameter will not impinge on the coronary blood flow. For this reason, certain operators may suggest inflating a stiff balloon of the same diameter that will be used for the final stent implantation, to insure safe distance from the conduit to the origin of the coronary arteries. Others are satisfied with inflation of compliant sizing balloons and assuring presence of at least 10 mm from the margin of the inflated balloon to the origin of the coronary arteries. The drawback of inflating a high pressure in the conduit to assess distance to coronary arteries is the small possibility of causing conduit dissection and rupture, especially if the lab is not equipped with covered stents to bail the situation out. Therefore, our approach has been to use the compliant balloon and see how far the distance is, and then deploy a bare metal stent as a landing zone [3]. Further, it is crucial to look at the coronary arteries with balloon inflation in at least two orthogonal views. From our experience, one view can show normal origin and no evidence of compression, yet the other view shows compression.

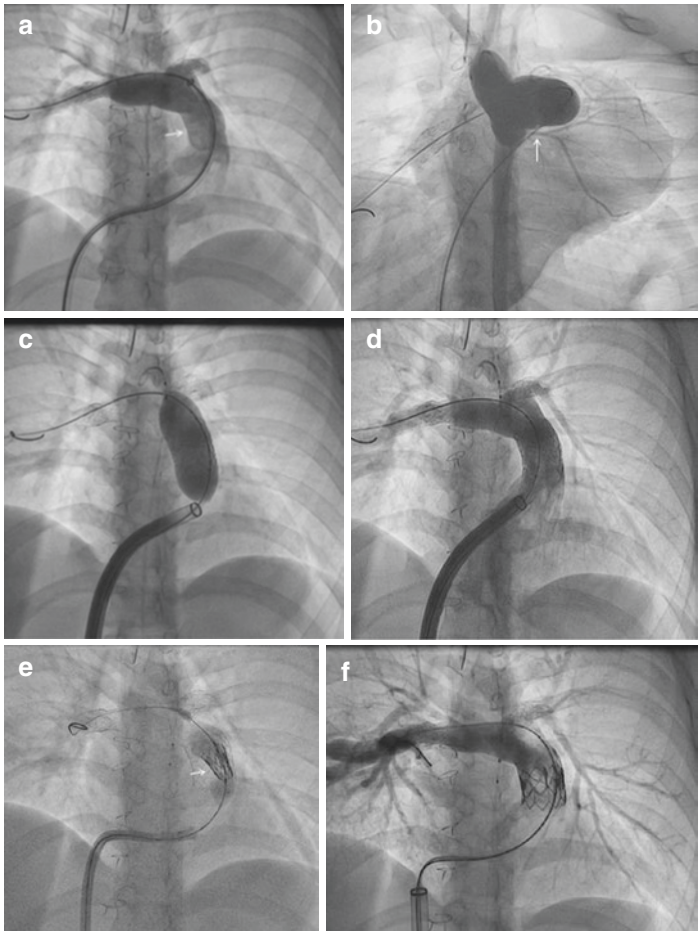

Fig. 43.3 Angiographic stepwise approach to tPVR. (**a**) Angiography of a dysfunctional conduit showing pulmonic regurgitation (*arrow*). (**b**) Simultaneous balloon inflation and aortic root injection in steep caudal angulation to assess distance from the conduit to the coronary arteries (*arrows*). (**c**) Positioning of the 35 mm bare-metal AndraStent mounted on a 24 mm BiB catheter. (**d**) Repeat angiogram after stent deployment showing stent in good position and no complications. (**e**) Deployment of a 26 mm Sapien XT valve inside the conduit, (arrow). (**f**) Angiography in MPA above the new valve showing good valve position and trivial regurgitation

5. Given the short height of the Edwards SAPIEN XT valve, bare-metal stent implantation (pre-stenting) as a landing zone is performed routinely. The stent is deployed on a BiB (balloon-in-balloon) catheter (NuMED Inc., Hopkinton, NY, USA) over a stiff guidewire placed in one of the pulmonary arteries, preferably in the left pulmonary artery, but the right may on occasion be the better branch (Fig. 43.3c–d). Generally, it is recommended to inflate the balloon to a diameter up to 2 mm less than the original conduit size in stenotic conduits or slightly larger in regurgitant conduits with no stenosis. In the case of significant recoil of the stent after balloon deflation, post dilation with a high-pressure balloon may be required, or in certain cases multiple stents may be implanted to create a suitable landing zone for the valve. In heavily calcified conduits, which are at a higher risk for rupture, a covered stent may be used in place of a bare-metal stent.
6. The final valve size is determined by the size of the stent used for pre-stenting. It is important to measure the fully expanded stent diameter in two dimensions (utilizing biplane fluoroscopy) to ensure uniform stent expansion.
7. The valve stent is crimped symmetrically using a specialized crimping tool proximally to the balloon.
8. Once the delivery system is past the sheath, in a straight part of the vena cava, the balloon is pulled back onto the crimped valve.
9. The valve is then delivered across the pre-stented RVOT over a stiff guidewire (Meier wire or Lunderquist). If the area is not pre-stented, angiograms are performed prior to balloon inflation to assess for proper positioning of the valve. However, if there is a stent, it is easy to position the valve in the middle of the stent. Inflation of the valve is slow (Fig. 43.3e).
10. Valve performance is evaluated either angiographically (Fig. 43.3f) or by intracardiac echocardiography. Continuous wave Doppler and color Doppler are used to evaluate the gradient and assess for any regurgitation, either valvular or paravalvular (Fig. 43.4a–f).

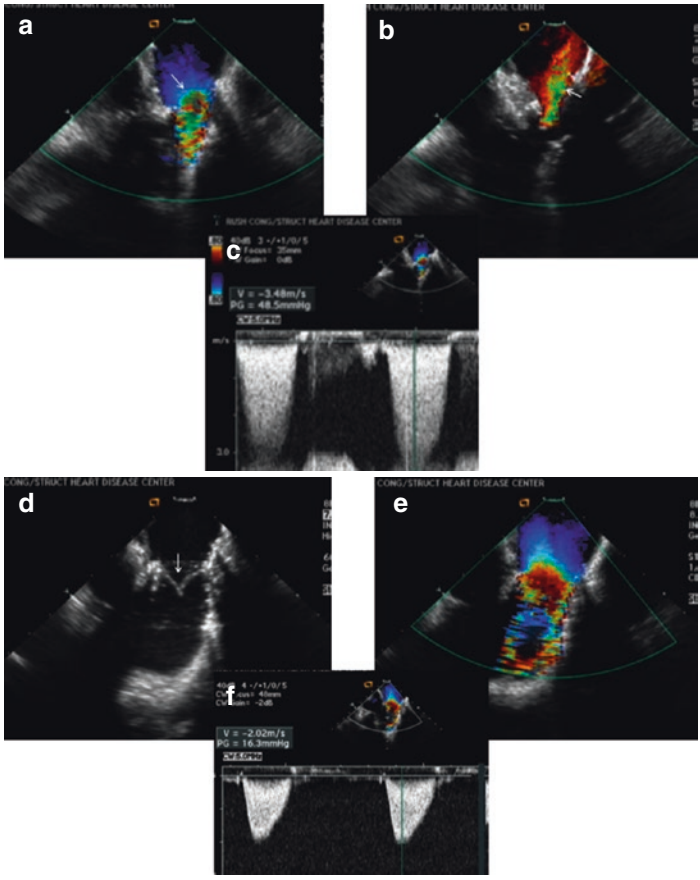

Fig. 43.4 Intracardiac echocardiography. At baseline showing a degenerated conduit with Color Doppler revealing turbulence across conduit (arrow) (**a**) and regurgitation (arrow) (**b**) and CW Doppler showing obstruction (**c**). After placement of a 23 mm Sapien valve showing valve leaflet (arrow) (**d**), no regurgitation (**e**), and CW Doppler no obstruction (**f**)

Given the large size of the sheath, it is recommended that venous hemostasis be achieved by utilization of a vascular closure device such as two Perclose sutures (Abbott Vascular, Abbott Park, IL, USA) placed at the beginning of the procedure. However,

we frequently utilize the "figure of 8" suture effectively. The Vicryl suture approximates the soft tissue around the access site to form a "pressure dressing" over the puncture site. The stitch is removed the next morning.

43.8 Expected Results

The Congenital Multicenter Trial of Pulmonic Valve Regurgitation Studying the SAPIEN Interventional Transcatheter Heart Valve (COMPASSION) trial published in 2011 was the first multicenter trial looking at the use of the SAPIEN valve in the pulmonic position. It showed a significant reduction in RVOT gradient with reduction in clinical symptoms and maintenance of pulmonary valve competence at 6-month follow-up [4]. The study included 36 patients from three US centers and one European center. Valve implantation was attempted in 34 of the patients (the two cases in which implantation was not attempted were because of unfavorable anatomy in one and stent embolization in another) and was successful in 33 (97.1%). Migration of the SAPIEN valve occurred in three patients. In two of those cases, surgical retrieval was necessary, but in the third, the valve was successfully deployed in the inferior vena cava. Other complications included pulmonary hemorrhage ($n = 2$), ventricular fibrillation, and stent migration. There was a significant reduction in RV/aortic pressure ratio from 0.6 ± 0.2 to 0.4 ± 0.1 ($p < 0.001$). At 6-month follow-up, there were no deaths, and in 97% of the patients, pulmonary regurgitation was <2+. One patient required elective placement of a second valve due to conduit-induced distortion of the initial implant. In the 3-year follow-up data of the COMPASSION trial, freedom from all-cause mortality was 98.4%. Freedom from reintervention was 93.7% and from endocarditis was 97.1%. There were no observed stent fractures [5].

More recent European case series with the Edwards XT [6] as well as the use of S3 in the pulmonic position [7] have continued to show excellent success rate and very low rate of complications. In the French registry [8], early major complications occurred in four patients (5.6%). Complications included 1 death, 1 coronary

compression, and 2 pulmonary valve embolizations. Three of the four major complications occurred in the first 15 operated patients. Hemodynamics were excellent with no significant regurgitation after the procedure. Transpulmonary gradient was significantly reduced from 34.5 to 10.5 mm Hg ($p < 0.0001$). No patient died during a 1-month follow-up period. At 1-year follow-up, the death rate was 2.9%, and three patients had undergone surgical reintervention (4.4%).

The most recent data on tPVR with the SAPIEN valve is from a European series of 22 patients [9]. The group reported a 95.5% procedural success rate (21 of 22 patients). There were three procedural complications including one stent embolization, inability to pass the valve past the inferior vena cava due to severe occlusion, and one plexus injury. Hemodynamic results were favorable with RV systolic pressure decreasing from 61.2 mmHg ± 23.1 to 41.2 mmHg ± 8.6. There was a substantial reduction in the degree of pulmonary regurgitation, with only one patient having mild regurgitation following valve implantation.

43.9 Tips and Tricks

The RVOT can be a tricky area to navigate. The Novaflex + delivery system does have a lower profile and more flexion compared to the prior delivery model which has helped in valve delivery. Experience is one of the most important determinants of success for this procedure. There are however a few tips that should be kept in mind. Wire positioning is critically important to ensure the valve will cross into the right position. Using a stiff wire (such as a Meier wire or Lunderquist) is preferred, and it is important that the wire tip is positioned as distally as possible. We have found that positioning the wire in the distal left lower lobe gives us the best rail for stent and valve delivery. Since these wires are so stiff, the operator must be meticulous with constant visualization of the tip of the wire to prevent perforations. We also recommend that all wire exchanges or removals be done through an end-hole catheter as to not damage the vasculature or the valve once it has been positioned. Availability of the DrySeal sheath (22 Fr and 24 Fr)

[WL Gore] has been very helpful and as long as this sheath is positioned distal to the landing zone, delivery of the valve assembly has been easy.

We believe that availability of covered stents is extremely important in the event a dissection or frank perforation occurs. In-house availability of self-expanding covered stents (Gore, Cook, and Medtronic) is important. Finally, in-house availability of a congenital cardiac surgeon who is familiar with the anatomy and is willing to operate in case of complications is also important. Last, availability of coronary guiding catheters and wires and even intravascular ultrasound (IVUS) is very desirable in questionable cases, and collaboration with adult cardiologist for assessment of coronary flow/distance during balloon inflation in the RVOT is a must.

43.10 Complications

Serious complications associated with tPVR are very rare but are devastating when they happen. In the US multicenter SAPIEN study (COMPASSION), the rate of serious complications was as high as 19.4% in the initial 36 procedures attempted [4]. The European multicenter registry reported a major complication rate of 13.6% in the first 22 procedures performed [6]. As we have seen in more recent case series [9], complication rates are now reported to be as low as 2.4% and will continue to decrease as operators become more experienced with the use of the SAPIEN XT valve. Expected complications can be broken down into several categories which are discussed below.

43.10.1 Vascular Complications

Given the large caliber of the delivery sheath of the SAPIEN XT valve, there is potential for serious vascular complications including femoral vein thrombosis, perforation, or hematoma. Using a vascular closure device such as the Perclose device (preclosure with two sutures placed at the beginning of the case) has been

advocated as a way to reduce these complications and may even be used in children. With the Sepien XT valve, the smaller sheath size reduces that risk significantly. There have been no reports of vascular complications in the COMPASSION study or more recent case series [9].

43.10.2 Coronary Artery Compression

The potential for coronary compression is not an uncommon occurrence; approximately 4% of the US Melody valve cohort had unsuitable anatomy and therefore did not undergo valve implantation [2]. This catastrophic complication can be avoided by thorough evaluation of the coronary anatomy prior to the procedure by noninvasive imaging such as CT or MRI. Furthermore, nonselective aortic root angiography or selective coronary angiography with a simultaneous balloon inflated in the RVOT in multiple views has become standard of care prior to valve implantation. When available, three-dimensional rotational angiography may complement traditional angiography to further assess the distance from the RVOT to the coronary ostia.

43.10.3 Conduit Rupture

This is a life-threatening complication, which may require conversion to an open surgery. However in the right patient, utilization of a covered stent as a bailout may be an effective way to avoid surgery in this situation. We believe that laboratories performing tPVR should have the appropriate-sized covered stents available for use in case of conduit rupture.

43.10.4 Valve Embolization

Because of the shorter height of the SAPIEN valve, embolization is a real possibility. Valve migration and embolization can be successfully treated with percutaneous device retrieval and redeployment of the valve in one of the great vessels. Alternatively, surgical

valve retrieval is an option if the valve cannot be moved to a safe location percutaneously, such as when it is caught in the subvalvular apparatus or Eustachian valve. The rate of valve migration in the COMPASSION trial was 8.8% and 4.5% in the European cohort [4–6]. In the recent French registry, the rate of valve embolization was as low as 2.9% [8].

43.10.5 Pulmonary Artery Obstruction

It is possible to obstruct the branch PAs either during pre-stenting or during implantation of the valve. A thorough angiographic evaluation of the landing zone prior to stent placement and careful assessment of valve position prior to deployment is crucial. Deploying the stent or valve across either branch PA may lead to decreased flow and difficulty in accessing the branch PA if future interventions are required.

43.10.6 Pulmonary Artery Hemorrhage

Perforation of the PA with either a stiff guidewire or a hydrophilic wire can easily occur. This is especially true for arteries, which have been subjected to high pressure and have become friable over time. For this reason, very careful attention must be paid to the tip of the guidewire during the procedure. Luckily, most bleeding is self-limited and manifests as a small amount of blood in the endotracheal tube. Nevertheless, major pulmonary artery bleeding can lead to hemodynamic compromise, which may require an open thoracotomy and is associated with a high mortality rate. A cardiothoracic surgeon with experience in congenital heart disease should always be available on-site in case of such complications.

43.10.7 Stent Fracture

Stent fracture has been a significant limitation of the Melody valve, with rates of stent fracture initially shown to be between 12% and 28% [2]. Pre-stenting of the RVOT with a bare-metal

stent is thought to reduce the rate of stent fracture. There have not been any stent fractures reported with the Edwards SAPIEN valve or the SAPIEN XT, most likely because of the shorter stent height, high radial strength, and routine use of pre-stenting.

43.10.8 Endocarditis

In Melody follow-up studies, five patients (3.2%) were diagnosed with endocarditis over a mean follow-up of 5 months [2]. Slightly lower rate has been seen with the Sapien valve following implantation in the pulmonic position. The updated AHA guidelines recommend continuing lifelong endocarditis prophylaxis for patients who have a conduit.

43.11 How to Manage Complications

The most important part of managing complications is to think ahead and have a plan in the situation that a complication arises. It is crucial that operators who perform tPVR procedures either have access to the right equipment in their lab or equipment is readily accessible, so that life-threatening complications may be treated in a timely fashion. Specifically, we stock our catheterization laboratory with a small supply of Gore Excluders (W.L. Gore, Newark, DE), which are typically used to treat abdominal aortic aneurysms endovascularly. We, however, have used them as a covered stent during the occurrence of a conduit rupture. We also stock our laboratory with the necessary coronary guides, wires, and stents, which could be used in the setting of coronary occlusion. It is also important to use all available resources in the hospital including vascular surgeons, coronary interventionalists, and interventional radiologists who may have more experience using this equipment and are an invaluable asset at a time of need.

43.12 Post-procedural Care

Patients are usually kept for observation overnight and discharged home the following day on 81 mg aspirin for 1 year. Prior to discharge, a chest radiograph and an echocardiogram are performed for a baseline assessment. Careful attention should be paid to the access site prior to discharge to make sure there is no hematoma or excessive tenderness.

43.13 Follow-Up

Follow-up examination and echocardiography are performed at one, 6, and 12 months and yearly thereafter. Chest radiograph is obtained before discharge and at 6 months to look for valve/stent position and any potential stent fracture. It is important that patients and their families understand that routine follow-up is important and that dental prophylaxis should be continued lifelong for patients with conduits. For patients with native RVOT, this can be reduced to 6 months.

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44 Percutaneous Tricuspid Valve Implantation (PTVI)

Andreas Eicken and Peter Ewert

44.1 Introduction

Severe primary congenital tricuspid valve dysfunction is rare. Secondary tricuspid valve dysfunction may occur after various surgical or catheter interventional procedures. If tricuspid valve surgery is indicated, usually a surgical valve plasty is performed and only if this fails tricuspid valve replacement is performed. In patients with congenital heart disease, a biological valve prosthesis is often chosen in tricuspid position, which has a limited durability. Once repeated tricuspid valve replacement is necessary, percutaneous tricuspid valve implantation (PTVI) is a safe alternative to surgical redo. The previously implanted bioprothesis is a perfect landing zone for a percutaneous valve (valve-in-valve implantation). The largest experience with PTVI exists with two commercially available percutaneous valves, the Melody valve (Medtronic Inc., Minneapolis, MN) and the Edwards SAPIEN

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XT/Sapien three valves (Edwards Lifesciences LLC, Irvine, CA) [1]. Both valves have been described in detail previously and received FDA approval for implantation in pulmonic position (Melody valve and Sapien XT valve). Since until today there is no approval for percutaneous tricuspid valve implantation, this intervention is performed as an off-label procedure, which needs to be discussed with the patients or their guardians.

44.2 Patient Selection and Pathophysiology

A bioprosthesis in tricuspid position may develop progressive stenosis, regurgitation, or a combination of both. Echocardiography allows grading of valve regurgitation and assessment of the mean Doppler diastolic inflow gradient through the tricuspid valve. In tricuspid valve dysfunction, right atrial and right ventricular enlargement may develop. If left untreated, right heart failure (atrial flutter/fibrillation, liver congestion, effusions) with reduced exercise capacity and reduced NYHA functional class may result. At cardiac catheterization a diastolic inflow gradient >5 mmHg (atrial a-wave to right ventricular end diastolic pressure) may be present indicating valve stenosis, or a prominent right atrial v-wave is the hallmark of severe tricuspid valve regurgitation. Serial examinations (clinical, echocardiography, cardiac MRI with indexed right atrial and right ventricular volumes and right ventricular ejection fraction, exercise test with VO_2 max) help to determine the optimal time for PTVI. The AHA/ACC guideline for the indication for tricuspid valve replacement was updated recently [2], and the ESC guidelines are currently rewritten.

44.3 Procedure

Femoral venous access is the preferred route for percutaneous tricuspid valve implantation. However, the jugular veins (preferably the right jugular vein) may also be used. During continuous sedation, the femoral vein and artery are cannulated. Intubation and controlled ventilation are not necessary. Heparin (in adults

5000 units, in children 100 units/kg up to a maximum of 5000 units) is given to keep the activated clotting time above 200 s. A dose of a first- or second-generation cephalosporine (for example, cefuroxime) is given followed by two additional doses 8 and 16 h later after successful PTVI. A complete hemodynamic assessment with pressure registration in the right atrium, right ventricle, aorta ascendens, and the left ventricle is performed. If no shunt is present, oximetry may not be necessary. Angiographic depiction of the right ventricle in two planes is performed (Fig. 44.1a, b). Then, a Super Stiff guidewire (i.e., Meier wire Boston scientific Corp, Quincy MA, USA; Amplatz Ultra-Stiff 0.035′ Boston scientific Corp; Lunderquist 0.035′ Cook medical, Bloomington IN, USA) is positioned distally into the left or the right pulmonary artery. If the wire tip is located within the right ventricle, a safe and controlled tricuspid valve delivery is harder to achieve, since the wire position is not as stable as in a pulmonic position.

Then, although a valve-in-valve delivery can be performed without a "balloon-test" if the bioprosthesis is well known, we tend to perform a balloon test for the final decision which percutaneous valve is best suited. If the tricuspid valve prosthesis is distensible (homograft), a selective injection into the right

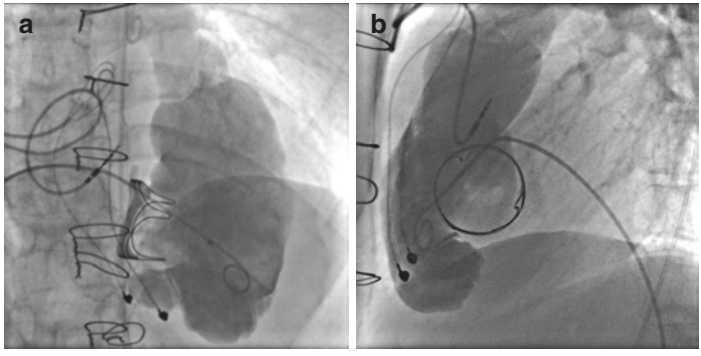

Fig. 44.1 (**a**, **b**) Angiography into the right ventricle with a 6F pigtail catheter in PA and lateral position. In the PA-plane (**a**), the system is rotated strictly perpendicular to the plane of the biological tricuspid valve (in this case, an Edwards Perimount prosthesis 33; Edwards Lifesciences LLC, Irvine, CA)

coronary artery should be performed during the balloon test. The balloon should be at least 1–2 mm larger than the suspected internal diameter of the bioprosthesis. The balloon is advanced over the wire into the tricuspid valve and inflated until a waist is visible (Fig. 44.2).

In general, if the bioprosthesis is well known and a Sapien valve is chosen, pre-stenting is not necessary. The selected percutaneous valve should also be at least 1–2 mm larger than the known internal diameter of the "landing zone" bioprosthesis. Then a valve is selected. The venous sheath is upsized (14 Fr for Sapien 23 and 26 mm valves and 16 Fr for the Sapien 29 mm valve). Care needs to be taken for correct orientation of the Sapien valve in tricuspid position, since in PTVI the valve needs to be positioned 180° opposite to the regular orientation in a TAVI aortic procedure. The venous sheath is positioned into the inferior vena cava. Then the NovaFlex+ delivery system is inserted through the sheath via the loader until the valve exits the sheath. The balloon is then pulled back until the valve is positioned

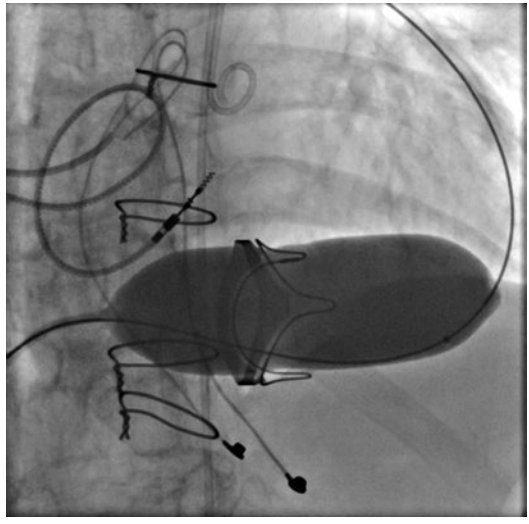

Fig. 44.2 Balloon test with a VACS balloon 30 × 60 mm (Osypka AG, Rheinfelden, Germany). An indentation is seen at 27 mm diameter

properly on the balloon between the two markers and the system is fixed. The valve is then advanced into the bioprosthesis. Once the Sapien valve is located across the bioprothesis, slow balloon inflation is done by the second operator to ensure optimal positioning of the valve across the bioprosthesis. The first operator can manipulate the valve position during slow inflation by pushing or pulling on the guidewire and by keeping the valve in a horizontal orientation. This enables optimal positioning. At final valve delivery, the proximal Sapien stent struts should peak into the right atrium, and most of the Sapien valve should point towards the right ventricle (Fig. 44.3).

A final angiogram is done to document the result (Fig. 44.4).

Finally, right atrial, right ventricular, and aortic pressures are assessed. A transesophageal echocardiogram may be done to depict valve function and additionally to assess the diastolic Doppler inflow gradient through the newly implanted valve (Fig. 44.5). However, the valve can be assessed by TTE as well.

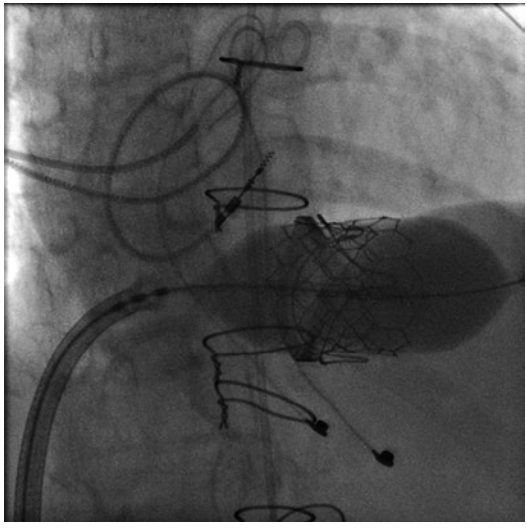

Fig. 44.3 A Sapien S3 valve 29 mm is inflated within the Perimount 33 prosthesis. The RA struts are just peaking into the right atrium, and the valve is securely fixed in the bioprosthetic valve

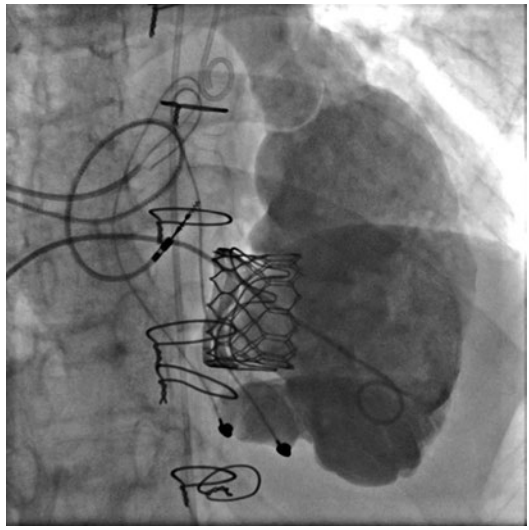

Fig. 44.4 Final angiogram into the right ventricle showing excellent position and function of the Sapien S3 valve

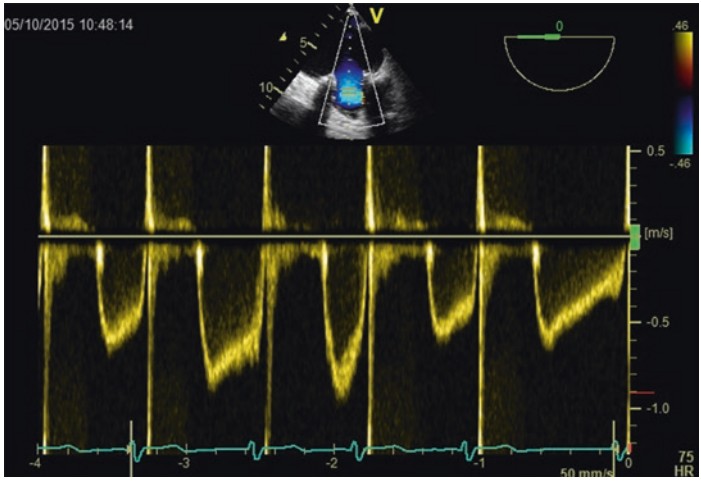

Fig. 44.5 (Patient 1): Transesophageal echocardiography directly after PTVI with a Sapien S3 29 mm into a Carpentier-Edwards Perimount 33 mm valve, showing unrestricted diastolic TrV inflow and depicting the absence of any tricuspid regurgitation

If a Melody valve is used, the vein needs to be dilated with a 22 Fr dilator to accommodate the 22 Fr delivery system. If the valve diameter is not known, a balloon test together with selective RCA depiction is indicated preceding PTVI (Figs. 44.6 and 44.7). For the Melody valve, pre-stenting is usually done to create a safe landing zone for the valve (Figs. 44.8 and 44.9). If the landing zone is short, the Melody valve may be folded and thus shortened to prevent coronary arterial compression (Figs. 44.10, 44.11, 44.12, 44.13, and 44.14).

The implantation of multiple covered CP stents (Numed, Hopkinton NY, USA) may help to downsize the internal tricuspid valve diameter of a large bioprosthesis, since the outer diameter of a Melody valve is only 24 mm on a 22 mm BiB delivery system.

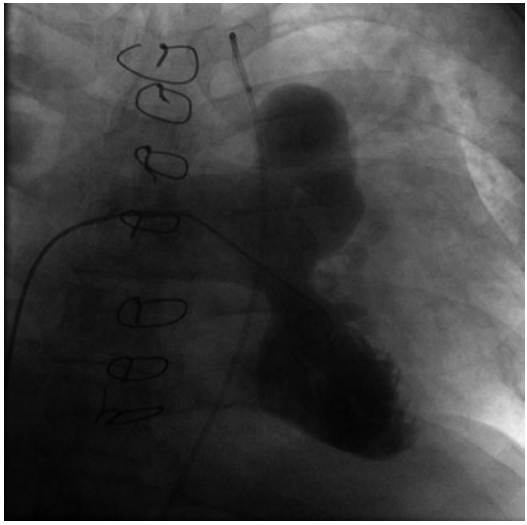

Fig. 44.6 (Patient 2): 38-year-old patient with tricuspid atresia after a Fontan RA–RV anastomosis (Björk modification) with a homograft done in 1994. In 2007, the stenotic and calcified homograft was exchanged for a 25 mm homograft. Now the patient presents in NYHA III with atrial fibrillation, maximal "tricuspid" regurgitation with severe right atrial enlargement. RAP is $v = 21$, $m = 18$ mmHg. RVP is 27/5/11, hence severe stenosis and regurgitation of the homograft is assessed. Repeated surgery was classified to be at an increased risk

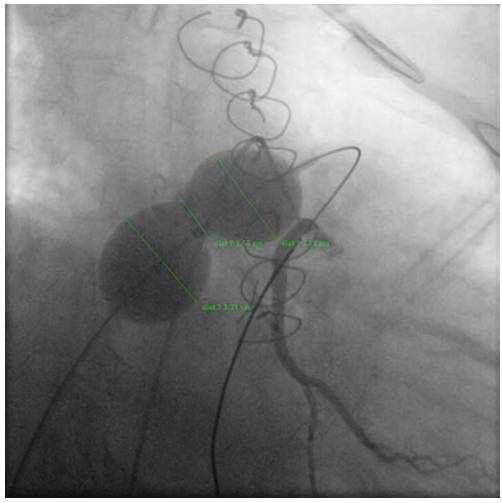

Fig. 44.7 (Patient 2): A balloon test was done with a 34 mm sizing balloon (St. Jude Medical St Paul, MN, USA) on a 0.035 Super Stiff guidewire. It shows a waist at 16 mm together with a rather close neighborhood of the right coronary artery

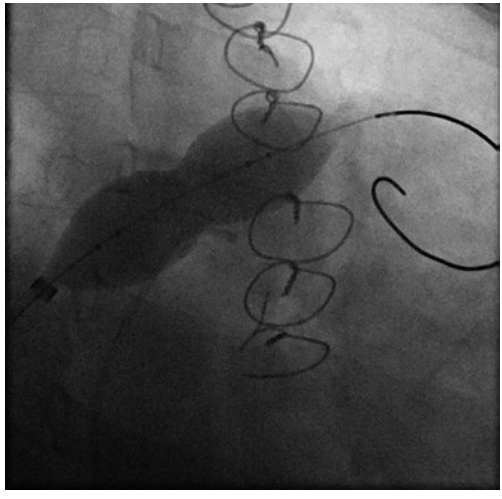

Fig. 44.8 (Patient 2): An EV3 Max LD 26 mm (Medtronic Inc., Minneapolis, MN) stent was mounted on a 22 mm balloon-in-balloon (Numed, Hopkinton NY, USA) and implanted into the stenotic homograft

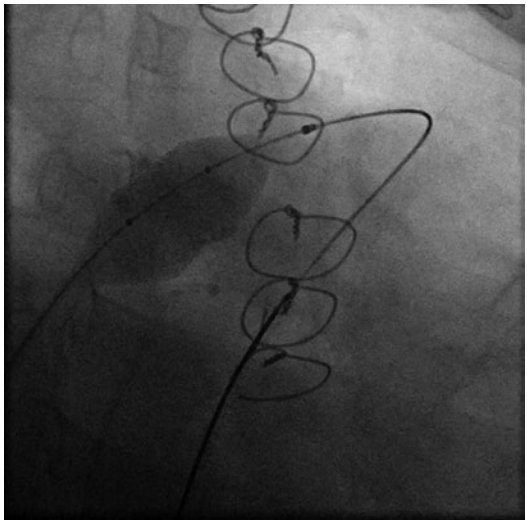

Fig. 44.9 (Patient 2): The stent was dilated with a 22 × 20 mm Atlas high-pressure balloon (Bard Tempe, AZ, USA)

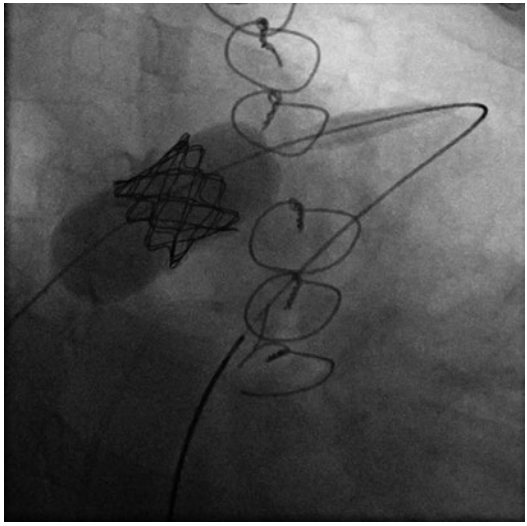

Fig. 44.10 (Patient 2): A doubly folded Melody valve was crimpled on a 22 mm delivery system and implanted into the Max LD stent

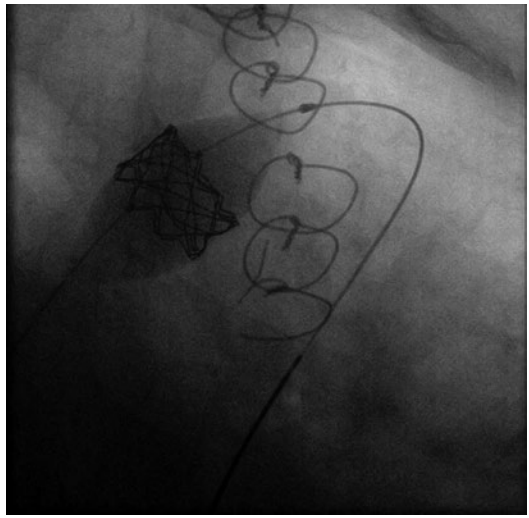

Fig. 44.11 (Patient 2): The Melody valve was dilated with a 24 × 20 mm Atlas balloon

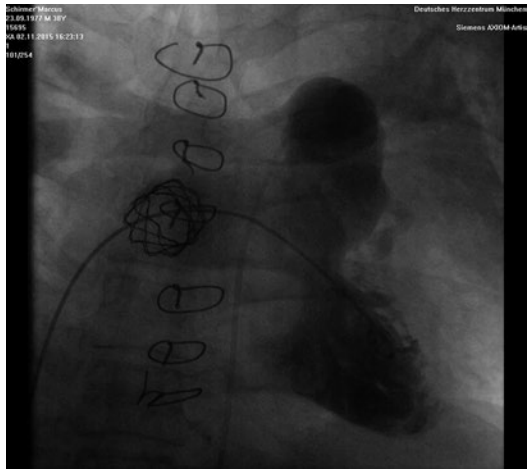

Fig. 44.12 (Patient 2): After the intervention, no residual tricuspid regurgitation is present

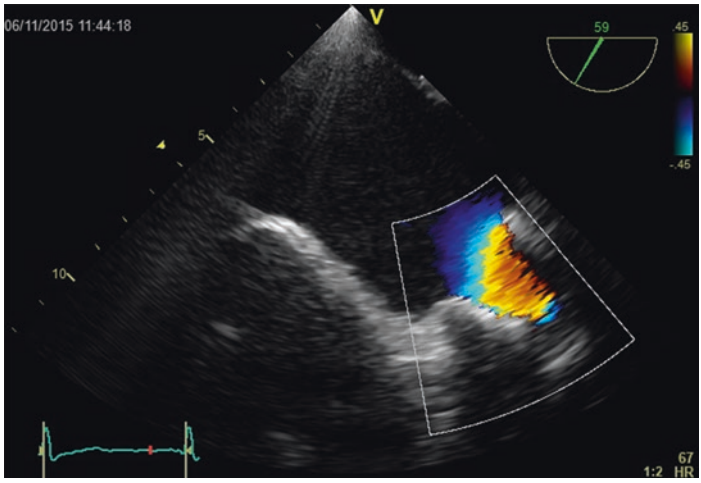

Fig. 44.13 (Patient 2): Transesophageal echocardiography shows normal diastolic TrV inflow and absent tricuspid regurgitation

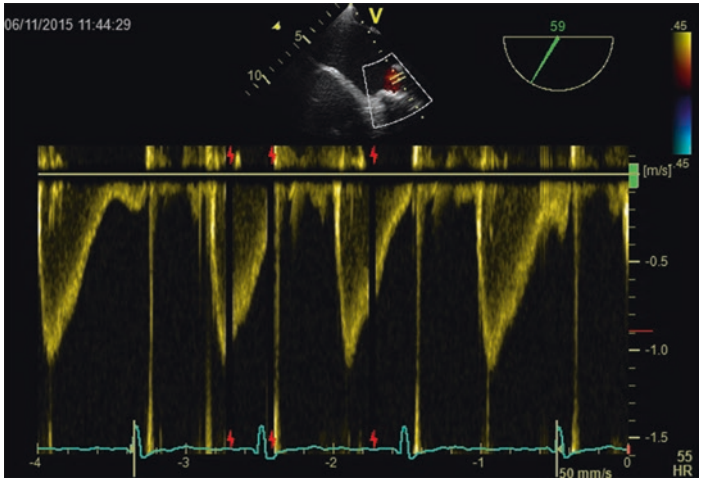

Fig. 44.14 The Doppler inflow signal has normalized after Melody valve implantation into the stenotic homograft. A repeated TEE 14 months later shows good Melody valve function

In most of our patients, the Melody valve was delivered with a 22 mm delivery system into the tricuspid valve bioprosthesis. Folding of the distal and proximal Melody stent struts shortens the valve and adds another 1–2 mm to the valve profile. It may be cumbersome to load the Melody valve into the delivery system after folding. Dilatation of the ensemble helps to solve this issue. Delivery of the Melody valve on a 24 mm balloon-in-balloon results in an external diameter of 26 mm, but is not generally recommended because valve regurgitation may result at this valve diameter. However, good valve function is documented in patients in whom a Melody valve was dilated to 24 mm internal valve diameter, as in our patient example.

44.4 Post- procedural Care

After PTVI, we keep our patients on Heparin (10,000 units/m^2, aPTT 40–60) until the next day. Then aspirin (100 mg/d) is initiated for 6 months and two doses of a cephalosporine are given. On the next day echocardiography is performed. After 6 months, an exercise test with assessment of VO_2 max is scheduled together with a cardiac MRI examination. There is some discussion whether patients after PTVI should be kept on warfarin or NOACS to prevent valve dysfunction due to thrombosis.

44.5 Outcome

So far only few patients are treated in every center. Our experience comprises 25 patients with PTVI so far, and the initial results were published in the year 2015 [3] and a review was written in 2018 [4]. 24/25 patients are alive and reinterventions (valve-in-valve procedure) were necessary only in 2/24 (8%).

The largest multicenter series was published by Doff McElhinney in 2019 on 306 patients after PTVI, with 64% of patients being alive and without reinterventions at 3 years. This series included our patients.

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45 Novel Self-Expanding Pulmonary Valves and Devices

Tomohito Kogure and Shakeel A. Qureshi

45.1 Introduction

Various congenital heart defects involving the right ventricular outflow tract (RVOT) abnormalities, such as tetralogy of Fallot (ToF), often have residual pulmonary regurgitation (PR) after the initial surgical repair. This may progress over time, requiring additional interventions to establish competent pulmonary valve, necessary for improved long-term quality of life. Historically, most patients, in whom there was felt to be an indication to abolish pulmonary regurgitation, were referred for surgery, which often involved prosthetic valve implantation at the time of surgery. However, prosthetic valves need repeated operations because of inevitable degeneration of the artificial tissue and the

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development of clinically significant pulmonary stenosis and pulmonary regurgitation. Reoperation may be associated with significant morbidity and mortality [1]. Percutaneous pulmonary valve implantation (PPVI) using balloon-expandable valves, such as Melody valve (Medtronic, Minneapolis, MN, USA) and Edwards SAPIEN series of valves (Edwards Lifesciences, Irvine, CA, USA), have been replacing surgery. However, balloon-expandable PPVI was limited to patients, who had right ventricle to pulmonary artery conduits or bioprosthetic valves [2–4]. These commercially available valves are usually unsuitable for non-conduit, native RVOTs in which a transannular patch has been used. This results in variable geometry and enlarged and expansile RVOT, and therefore there is a need for PPVI options. Novel designs using self-expanding technology have been developed for larger sizes of valves [5]. This review summarises the new self-expanding pulmonary valves that have been used clinically or are undergoing evaluation.

45.2 Venus *P*-Valve

45.2.1 Device and Delivery Systems

The Venus P-valve (Venus MedTech, Shanghai, China) is a self-expanding percutaneous valve, comprising a nitinol stent and a tri-leaflet porcine pericardial tissue valve, hand-sewn inside the nitinol frame and designed to be implanted into the RVOTs after previous transannular patch repair (or other similar reconstruction). The valve stent is flared in its distal and proximal parts. The flared distal portion of the stent allows anchoring at the pulmonary artery bifurcation, whilst the uncovered struts at the distal flare allow unobstructed branch pulmonary artery flow (Fig. 45.1). The proximal flare is covered and positioned in the RVOT. Currently, the Venus P-valve is available in diameters from 16 to 36 mm with 2 mm increments, with each diameter available in 20, 25, 30 and 35 mm straight section lengths. The valve assembly is delivered through a long 19 to 24 Fr delivery sheath (Fig. 45.2). The valve can be implanted in the RVOTs,

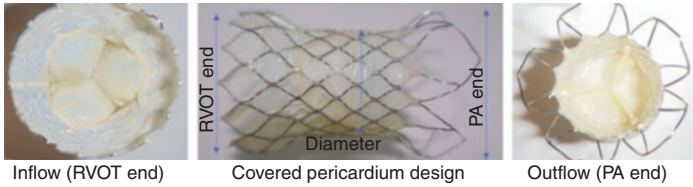

Fig. 45.1 Venus *P*-valve. The Venus *P*-valve (Venus MedTech, Shanghai, China) has proximal and distal flares that anchor the valve in the right ventricular outflow tract. The diameters of the proximal (RVOT end) and the distal (PA end) flares are 10 mm larger than middle segment. The PA end is not covered, permitting unobstructed flow into the branch pulmonary arteries

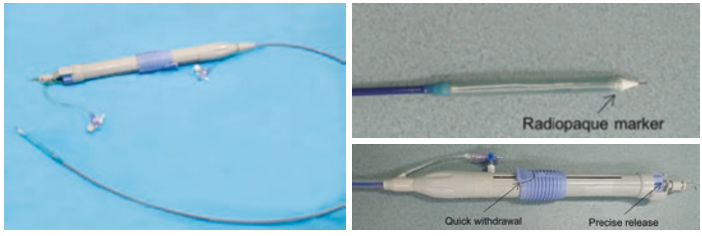

Fig. 45.2 The Venus *P*-valve delivery system**.** Note the handle with a knob for slow and controlled release of the valve and the distal capsule with a crimped and loaded valve inside

whose narrowest diameter is less than 34 mm. The 34 mm narrowest diameter may be suitable for the 36 mm valve. These valves have been implanted with acceptable safety and maintained valve function (Fig. 45.3).

45.2.2 Efficacy Outcomes

The first implantation of a Venus *P*-valve was reported by the Chinese group in 2014 [6]. Promphan et al. [7] reported early clinical experience outside China in six patients with severe pulmonary regurgitation following surgical repair of ToF with a transannular patch. The Venus P-valve was successfully implanted in all the patients with implanted valve diameters ranging from 24

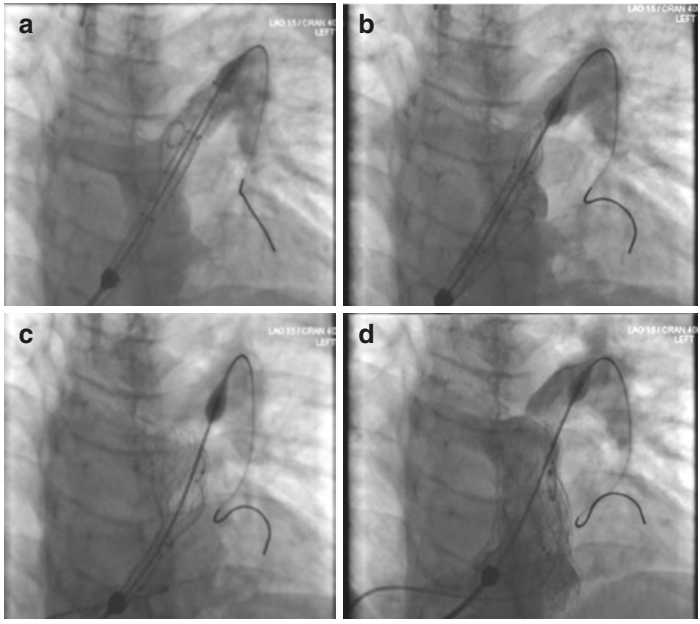

Fig. 45.3 Deployment of the flared Venus P-valve. (**a**) The deployment starting position in the proximal LPA. After exposing the distal flare, the system is pulled free from the LPA origin (**b**, **c**) before gradually deploying the rest of the valve frame (**d**), using intermittent low-volume check angiography from a pigtail catheter placed in the RV for fine adjustments

to 32 mm. None of the patients had significant outflow tract gradient or pulmonary regurgitation immediately after valve implantation and 6 months follow-up. Recently published data from Morgan et al. [8] and Zhou et al. [9] reported procedural success rates of 94.7% and 98.1%, respectively. Freedom from reintervention after PPVI has been reported above 95% during follow-up of 12 months with less than mild pulmonary stenosis and pulmonary regurgitation in all patients. The cohort of patients demonstrated a statistically significant reduction in pulmonary regurgitation fraction (PRF) and indexed right ventricular end-diastolic volumes (RVEDVi) at 6 and 12 months. At the latest MRI follow-up, the overall mean PRF had reduced from 48% before to 4% after the

valve implantation. The RVEDVi decreased from a mean of 151 ml/m^2 before to 112 ml/m^2 after valve implantation [8].

45.2.3 Complications and Mortality

The above-mentioned studies reported valve migration in two cases and one case, respectively, and one case in each study required urgent surgical procedure. Morgan et al. reported eight out of 30 (26.6%) of the valve frames had wire fractures during follow-up fluoroscopic evaluation. All the stent fractures were in the region of the proximal flare, which was in the contractile part of the RVOT. The fractures have not affected the function of the valve or frame integrity or stability and there has been no need for further reintervention related to stent fracture. The Chinese study reported infective endocarditis in four patients (7.2%) during follow-up of 12 months and one patient required surgical pulmonary valve replacement and one died 3 months after the PPVI. The study of Morgan et al. demonstrated no mortality during average follow-up of 25 months.

45.2.4 Summary

Clinical experience shows satisfactory procedural as well as short-term and mid-term outcomes for the Venus P-valve. Venus P-valve is undergoing further evaluation in a CE (Conformité Européene) multicentre study of 81 cases.

45.3 Pulsta Valve

45.3.1 Device and Delivery System

The Pulsta valve (TaeWoong Medical Co., Ltd., Gimpo-si, Gyeonggi-do, South Korea) is a self-expanding transcatheter valve, which consists of a nitinol stent covered with porcine pericardium. The valve is made of treated tri-leaflet α-Gal–free

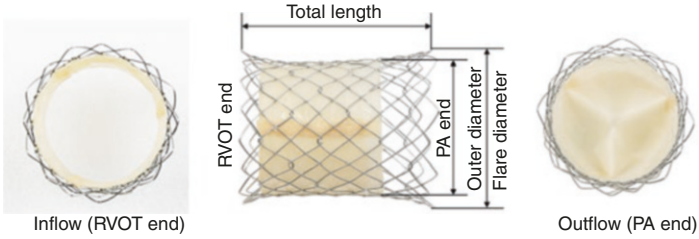

Fig. 45.4 Pulsta valve. The Pulsta valve (TaeWoong Medical Co. Ltd). The outer diameter of valve ranges from 18 to 32 mm with 2 mm increase with knitted nitinol wire backbone. Total length is 28 to 38 mm. Both ends of the valve stent are flared to 4 mm wider than the valve diameter itself

pericardium and hand-sewn into the stent (Fig. 45.4). The valve diameter ranges from 18 to 32 mm with 2 mm increments. Both ends of the valve are flared 4 mm wider than the central portion of the stent for stable valve adaptation to various RVOT geometries. The total length of the valve is 28 to 38 mm depending on the diameter of the valve. Transcatheter delivery system is 18 Fr for up to 28 mm valves and 20 Fr for 30, 32 mm valves. By hooking the proximal end of the nitinol wires at the hook block for attachment, controlled deployment and subsequent good positioning of the valve at the target area are possible (Fig. 45.5).

45.3.2 Efficacy Outcomes

The first-in-human experience of a Pulsta valve was reported by Kim et al. in 2017 [10]. They conducted feasibility trial on ten patients for native right ventricular outflow tract lesions [11]. According to the study, all ten patients had successful implantation of a Pulsta valve without any periprocedural complications. At the six-month follow-up, RVEDVi was dramatically decreased from 176.7 ± 14.3 mL/m^2 before to 126.3 ± 20.3 mL/m^2, and the mean value of peak instantaneous pressure gradient between the right ventricle and the pulmonary artery decreased from 6.8 ± 3.5 mmHg before PPVI to 5.7 ± 6.7 mmHg without significant pulmonary regurgitation.

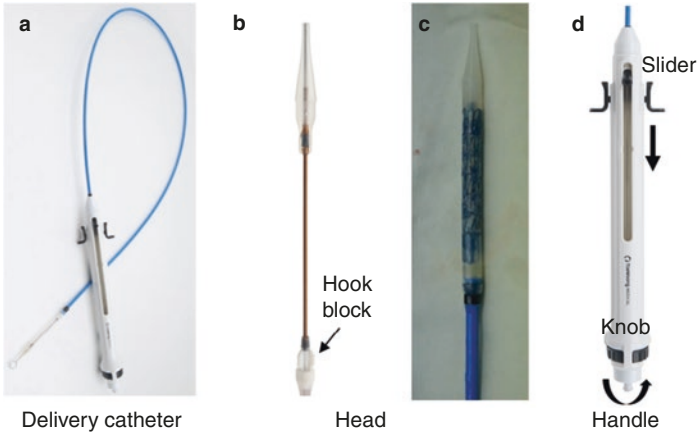

Fig. 45.5 Delivery catheter profile. The total usable length of delivery cable is 110 cm (**a**). Head portion of valve loading has a 17 mm conical tapered tip and hook block to perform controlled deployment at the target area (**b**). The outer profile of valve loading area is 18 Fr for 28 mm valve, 20 Fr for 32 mm and the shaft of delivery cable is 12 Fr (**c**). Valve is partly exposed by turning the knob clockwise (curved arrow) and deployed fully by pulling the slider (straight arrows; **d**)

45.3.3 Complications and Mortality

There were no adverse events associated with the valve. There was no stent fracture on fluoroscopy examination at 6-month follow-up.

45.3.4 Summary

A feasibility study of the Pulsta valve for native right ventricular outflow tract lesions demonstrated good short-term results without serious adverse events. CE certification approval study has currently started in six countries in Europe.

45.4 Harmony Transcatheter Pulmonary Valve (hTPV)

The harmony transcatheter pulmonary valve (Medtronic, Minneapolis, Minnesota, US) is a porcine pericardial tissue valve mounted on a self-expanding nitinol frame. The device has an outer diameter of 23.5 mm at the valved section and is approximately 55 mm in length. The TPV is treated with an alpha-amino oleic acid antimineralisation process to mitigate leaflet calcification, and a 0.2% glutaraldehyde sterilant (Fig. 45.6). The delivery system is a 25 Fr coil-loading catheter with an integrated sheath. The loading funnel collapses the valve to facilitate mounting on the delivery system, and the retractable sheath helps to control self-expansion of the TPV during deployment (Fig. 45.7).

45.4.1 Efficacy Outcomes

Animal studies using the hTPV in an ovine model of pulmonary regurgitation were firstly reported in 2016 [12]. The first Food and Drug Administration–approved early feasibility study has been reported by Bergersen et al. [13]. According to the study, the procedure was successful in all 19 cases, including one case, in whom

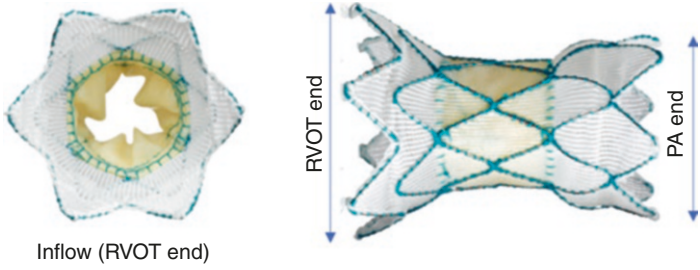

Fig. 45.6 Harmony TPV. This porcine pericardial tissue valve is mounted on a self-expanding nitinol frame. It has an outer diameter of 23.5 mm at the valved section and is approximately 55 mm long. The outflow diameter is 34 mm and the inflow diameter is 42 mm

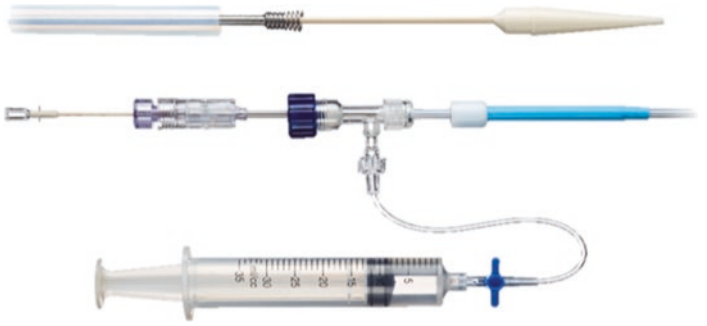

Fig. 45.7 The delivery system. 25 Fr coil-loading catheter with an integrated sheath. The loading funnel collapses the valve to facilitate mounting on the delivery system, and the retractable sheath helps to control self-expansion of the valve during deployment

the valve migrated proximally toward the RV after release, which was left in place and functioned adequately. Freedom from unplanned intervention was 90% during the follow-up of 6 months. At 6 months, mean RVOT gradient was 15 ± 6 mmHg. There is no more than mild PR including two patients with mild paravalvular leak at the 6-month visit.

45.4.2 Complications and Mortality

In one patient, the device migrated distally within 24 h. In this patient, the TPV was surgically explanted within 48 h of catheter procedure. They reported three cases of type I stent fracture (stent fracture without loss of stent integrity) in 6 months. One patient complained of persistent fatigue and exercise intolerance at 1-month follow-up. Transthoracic echocardiography indicated an increased RVOT gradient, and fluoroscopy demonstrated a type II stent fracture with associated partial frame collapse. These findings were confirmed at cardiac catheterisation then the device was surgically explanted during the same visit. There was no evidence of infective endocarditis and mortality during six-month follow-up.

45.4.3 Summary

Early publications and short-term outcomes in the Harmony Feasibility study have been encouraging with good procedural success rates and favourable valve function at early follow-up. An international, multicentre, prospective human feasibility study is ongoing [14].

45.5 Summary of Novel Self-Expanding Pulmonary Valves

Table 45.1 shows device specification. Table 45.2 summarises clinical studies of the commercially approved valves and the new self-expanding valves reported in the literature. The novel valves demonstrated promising early and mid-term results.

45.6 Others

45.6.1 Med-Zenith PT-Valve

The Med-Zenith PT-Valve (Med Zenith, Beijing, China) is a self-expanding transcatheter valve (Fig. 45.8) which consists of a nitinol stent fully covered with porcine pericardium. The first human study demonstrated that transcatheter implantation of this new self-expanding pulmonary valve in a patient with non-conduit pulmonary regurgitation was feasible with good immediate results [15].

45.6.2 Alterra Adaptive Prestent

The Alterra Adaptive Prestent (Edwards Lifesciences, Irvine, CA, USA) is a new concept of a self-expanding, partially covered stent (Fig. 45.9), which was designed to internally reconfigure the

Table 45.1 Device specification

Device	Company	Country	Tissue	Available diameters	Available lengths	Delivery sheath
Venus P-valve	Venus MedTech	China	Porcine pericardium	16–36 mm	20, 25, 30, 35 mm	19–24 Fr
Pulsta valve	TaeWoong Medical	South Korea	Porcine pericardium	18 to 32 mm	28, 31, 33, 38 mm	18–20 Fr
Harmony TPV	Medtronic	United States	Porcine pericardium	23.5 mm	55 mm	25 Fr

Table 45.2 Summary of clinical studies in percutaneous pulmonary valve implantation

First author, year (Ref. #)	Country	Time period	Valve	*N*	Age (years)	Weight (kg)	Follow-up (months)	Valve size (mean)	Procedure success[a] (%)
McElhinney et al., 2010 [17]	US	January 2007 to August 2009	Melody	30	19	61	12	NA	29 (96.6%)
Kenny et al., 2011 [18]	US, UK	April 2008 to May 2010	SAPIEN	36	30	73	6	NA	35 (97.2%)
Morgan et al., 2019 [7]	UK, others	October 2013 to April 2017	Venus P	38	24	59	25	NA	36 (94.7%)
Zhou et al., 2019 [8]	China	NA	Venus P	55	28	53	12	29.7 mm	54 (98.1%)
Kim et al., 2018 [10]	South Korea	NA	Pulsta	10	21	59	6	27.0 mm	10 (100%)
Bergersen et al., 2017 [12]	US, Canada	May 2013 to May 2015	Harmony	20	28	72	6	23.5 mm	20 (100%)

Complications							
Valve migration	RVOT rupture	Stent fracture (SF-r[b])	Infective endocarditis	Moderate- PS or PR	Unpland intervention	Mortality (%)	Reason of death
0	1	8 (3)	0	5	Surgical 1Transcatheter 4	0	
3	0	0	0	1	Surgical 4Transcatheter 1	0	
2	0	8 (0)	0	0	Surgical 1	0	
1	0	0	4	0	Surgical 2	1 (1.7%)	Infective endcarditis
0	0	0	0	0	0	0	
2	0	4 (1)	0	1	Surgical 2	0	

[a]Defined as the percentage of subjects with a transcatheter pulmonary valve placed with no more than mild PR, an RV-PA peak-to-peak gradient <35 mm Hg by angiography

[b]SF-r: stent fractures requiring reintervention

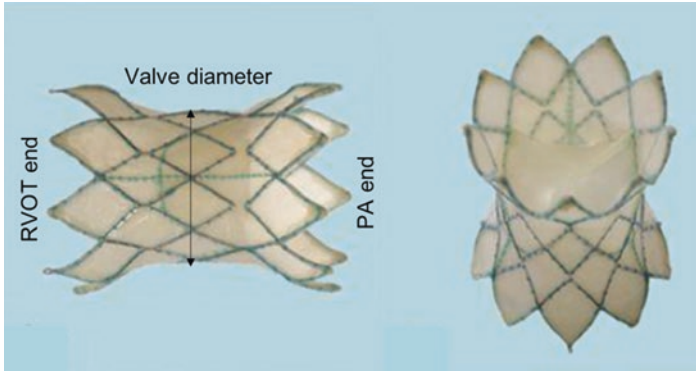

Fig. 45.8 Med-Zenith PT-Valve. The Med-Zenith PT-Valve (Beijing Med-Zenith, Beijing, China) is a porcine pericardial tissue valve mounted on a symmetric, self-expanding nitinol frame fully covered by porcine pericardium. The outflow and inflow part of the frame has five sizes (range 28 to 44 mm), and the overall length varies from 38 to 54 mm. The valve diameter in the middle portion is (20, 23, or 26 mm)

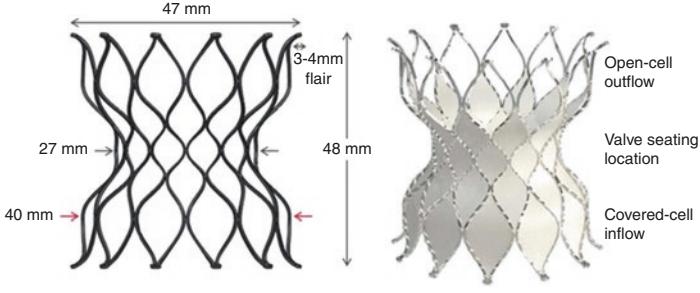

Fig. 45.9 Alterra Adaptive Prestent. It is comprised of a self-expanding nitinol frame assembly and polyethylene terephthalate (PET) fabric covering. The device has a symmetrical frame design with the inflow and outflow diameters equal to 40 mm and the central section 27 mm which aid in device stability. The uncovered distal row of cells allows for placement of the device into the orifice of the branch pulmonary arteries without causing obstruction to flow

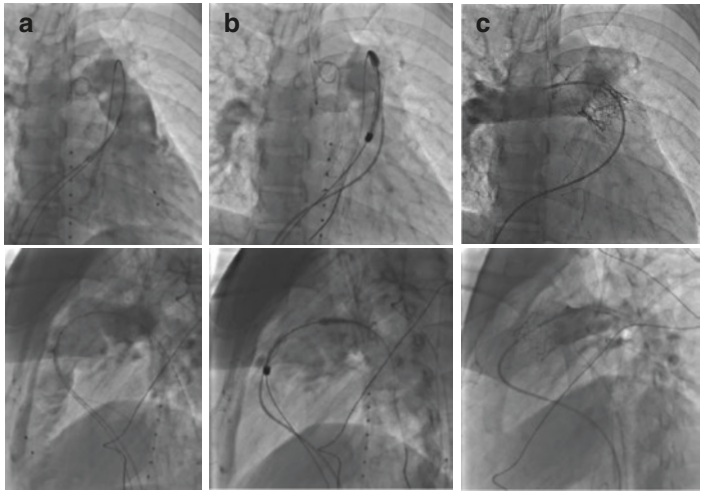

Fig. 45.10 Implantation of the Alterra Adaptive Prestent and SAPIEN 3 valve. (**a**) Right ventricular angiogram prior to the Alterra deployment. (**b**) Partial deployment of the Alterra. (**c**) Pulmonary angiogram following delivery of the SAPIEN 3. (Fig. 45.10 with courtesy and kind permission of Dr. Evan Zahn)

native dilated RVOTs, such as to make them suitable for implantation of a commercially available balloon-expandable heart valve, the SAPIEN S3 transcatheter heart valve (Edwards Lifesciences, Irvine, CA, USA). The device has a symmetrical frame design to provide a rigid "landing zone" for a SAPIEN S3 (29 mm). Zahn et al. reported the first human implantation which has shown encouraging result [16] (Fig. 45.10).

45.7 Conclusions

The management of pulmonary regurgitation after surgical repair of ToF has evolved over the last 20 years. Until recently, only about 25% of patients were suitable for transcatheter valves. With

further developments in the technology, a majority of patients may be potentially treatable by transcatheter valve technology within this decade. The number of patients in whom transcatheter valve therapy may be an option has expanded with the advent of novel valves designed specifically for use in the larger, non-conduit, outflow tracts. However, each of these valves is currently at varying stages of clinical trials, and it remains to be seen whether the promising early and mid-term results translate into long-term outcomes comparable to those of the commercially available, smaller diameter valves or surgical valve implantation or both.

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46 Approaches to Large or Complex Right Ventricular Outflow Tract

Younes Boudjemline

46.1 Introduction

Percutaneous pulmonary valve implantation (PPVI) has been widely accepted as a suitable alternative to surgery in selected patients. It is now considered the treatment of choice for patients with hemodynamically dysfunctional right ventricular outflow tract (RVOT) [1, 2]. The number of patients undergoing PPVI has been increasing over the years. Initially limited to patients with stenotic conduits, indications have extended to a large number of patients with dysfunctional RVOT including those with complex anatomy or pure pulmonary regurgitation. Indications, patient selection, and standard approach have been discussed in previous chapters for the two commercially available transcatheter valves. In this review, we will discuss approaches developed over the years to insert a valve in patients with large or complex right ventricular outflow tract.

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46.2 Patient Selection

The most important besides imaging is clinical history. The information about the type and size of the valve if present in all operative notes should be extracted and analyzed in regard to mode of degeneration. In obstructive lesion with or without PR, the question is not to know if a transcatheter could be implanted but if the RVOT can be opened enough to reduce the systolic right ventricular pressure. Therefore, initial diameter and most importantly the nature (i.e., elasticity) of the valve implanted surgically are very important to know and understand. In the other hand, in purely regurgitant lesion, the main question is to know if a transcatheter can be anchored safely in the RVOT. Here again, the nature of the RVOT is important to consider. If a surgical valve is present, instruction for use of the manufacturer will provide inner diameter. Otherwise, the compliance of the RVOT should be tested as part of the assessment prior to valve implantation. It is also important to analyze both pulmonary arteries as they can be used to better anchor a valved stent.

46.3 Technical Modifications for Challenging RVOT

A variety of technical modifications have been described to cover all clinical situations. Those techniques were essentially developed to extend the indications of Melody valves (Medtronic Inc., Minneapolis, MN, USA). The techniques will be reviewed in details providing indications and a full description.

46.3.1 Folded Technique (Fig. 46.1) [3, 4]

The Melody® valve is made of an original bovine jugular vein containing a valve. The valve is sutured to the frame of a balloon expandable stent (CP 8Z34, Numed Inc., Hopkinton, NY, USA). When expanded to 22 mm, the total length of the valved stent is 24.6 mm. In comparison, Edwards valve (Sapien XT©, Edwards

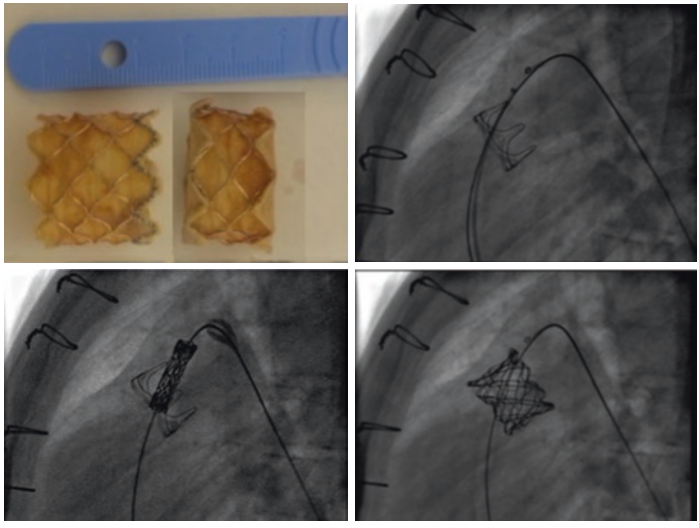

Fig. 46.1 Folded technique. In vitro imaging showing Melody before and after folding the two extremities. Example in a patient with a bioprothesis. Look at the aspect of the two extremities on fluoroscopy. Radiopacity of the extremities is increased compared to the middle part. After deployment, the Melody is seating just inside the bioprosthesis

Lifesciences, Irvine, CA, USA) is a manufactured valve made of pericardium mounted in a cobalt-chromium stent shorter than the Melody (14.3–19.1 mm). While the length of the Melody is appropriate for most of the cases, having a shorter valve might be useful in certain conditions specially when the landing zone is too short, or in short, complex RVOTs with early bifurcation or when there is a potential of coronary artery obstruction by the stent edge. While Sapien valve might be the valve of choice in this condition nowadays, it was not available worldwide for long time and interventionists had to find a way to deal with those patients. The Melody can also be modified and used in such clinical conditions. The modification is performed as followed: the terminal stent struts of the Melody is hand-folded over a syringe before crimping from inside out at one or both ends. Folding reduces the number of rows of the stent which drops from six to five if single extremity is folded or to

four if both extremities are folded. This manipulation reduces the length of the Melody from 24.6 mm to 16.7 mm when expanded on a 22 mm balloon. The folded valve is then loaded, crimped on standard Ensemble delivery system, and delivered using conventional technique. Because the thickness of the valve may vary, it can be difficult to load in the delivery system. In that case, one or two stent struts can be unfolded back to their original condition. After doing that, loading is easier. In our experience, around 10% of the valves need partial unfolding. This folding technique can be also used for surgical mitral valve placement of Melody valve.

46.3.2 One-Step Procedure (Fig. 46.2) [5]

A stepwise approach is needed during any PPVI, which includes balloon interrogation followed by insertion of a BMS to create a safe landing zone for the valve to be implanted. Pre-stenting is

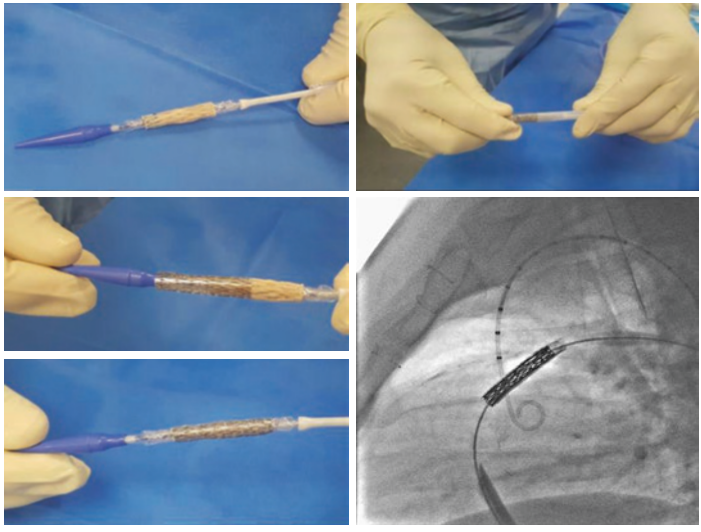

Fig. 46.2 One-step technique. Ex vivo images showing steps of preparation of one-step assembly. First Melody is crimped on an Ensemble delivery system, then EV3 LD max stent positioned and crimped on the Melody. Aspect in fluoroscopy of the one-step assembly

mandatory in most patients to reinforce the RVOT and significantly reduce the risk of valved stent fractures. However, this approach tends to lengthen the procedure time, increase radiation exposure, and expose patients to various complications, such as stent embolization and crushing while advancing the delivery system. In patients with purely regurgitant RVOTs, most interventionists would perform the pre-stenting in a different setting postponing valve implantation about 3 months later. Recrossing the bare-metal stent might be challenging especially if hanging in the RVOT. The experience with the folded Melody technique described earlier showed that a thickened valve can be loaded in the same delivery system. Some interventionists have been using this feature to deliver bare-metal stents and Melody at the same time. In practice, Melody is crimped on an Ensemble delivery system using the usual fashion. When done, one to three EV3 LDmax stents are prepared as described below. A 10- or 12-mm vascular balloon (any brand) is used to expend the EV3 stents on the shelf. The expanded EV3 is then manually crimped on the Melody valve. Of note, crimping of the Melody is more efficient as the EV3 prevents the recoil of the Melody. Up to three stents can be crimped over the Melody valve and delivered within the 22-mm Ensemble catheter without any problem. Crimping, sheathing, unsheathing, advancement, and delivery are performed using the standard fashion. This technique reduced the procedural and fluoroscopic times, and radiation exposure, compared to the standard twosteps technique. It eliminated the risk of BMS embolization and crushing while advancing the delivery system. Valvulation can be done during the same setting. It is of note that we have been using the same technique to deliver multiple BMS in one time in the RVOT prior to PPVI for highly mobile RVOT (or during Russian Dolls technique, see below) thus reducing recoil, and limiting the risk of stent fractures. This technique can also be used with Sapien valves but recent literature seems to demonstrate that presenting is not required for this particular valve to limit the risk of stent fracture.

46.3.3 Extending the Indications up to 26 Mm

Melody valve is intended to be positioned in stenotic RVOT associated or not with pulmonary insufficiency. However, it has been used in native or patched RVOT to treat pure pulmonary regurgitation. When dealing with patched RVOT, diameter and compliance of the landing zone is very important and should be tested as a prerequire before proceeding with valvulation. Imaging performed prior to the procedure (CT or MRI) might be useful but tend to underestimate the real diameter of the RVOT and do not appreciate the compliance of the vessel. Recent introduction of 4D-MRI might be a game changer providing better understanding of the target zone but its use remains confidential and most of interventionists will still rely on balloon calibration to decide on feasibility and to select the type of valves and its diameter. Similar to surgical valves, the manufacturers do not talk the same language. Medtronic talks about inner diameter while Edwards refers to outer diameter. A 22-mm Melody valve has an outer diameter of 24 mm and can theoretically be implanted in RVOT up to 24 mm. However, it has been recognized for long time now that the bovine jugular vein valve can be over-dilated up to 24 mm (inner diameter) keeping an excellent function. This is only possible because the height of coaptation is high compared to other valves. As a result, a Melody can be inserted in RVOT with a diameter up to 26 mm. However, delivery systems are only available up to 22 mm. Some interventionists have been using conventional Mullins sheaths (18 or 20 Fr) or DrySeal (Gore) to deliver a Melody crimped on a 24-mm BIB catheter [6]. Because those sheaths are quite stiff, a very stiff wire should be used to be able to advance this sheath (either Lunderquist or Back-up Meier wire). An alternative option is to modify the Ensemble catheter by exchanging the balloon provided by either a BIB or a Nucleus balloon or any balloon that would fit a 14 Fr sheath (size of the proximal part of the Ensemble catheter). Because in order to remove the balloon from the Ensemble, cutting the blue nose is required, and the introduction at the skin level would be possible (sparing the nose is difficult, and regluing ex vivo is not efficient)

only through a short sheath (DrySeal 22 Fr or 24 Fr). If the RVOT exceeds 25–26 mm, use of Melody is still possible but requires dedicated techniques (see below).

46.3.4 Russian Doll Technique and/or Branch Pulmonary Artery Jailing for Anchor in Large Patched RVOTs (Fig. 46.3) [7, 8]

If the RVOT measured between 25 and 30 mm, multiple stents with decremental diameters may be deployed with full overlapping in the RVOT region to reduce its size and allow proper seating of the Melody valve. The stent of choice for this method (Russian doll technique) is usually a high-profile stent (CP10Z or 8Z45 covered stent, Numed Inc).

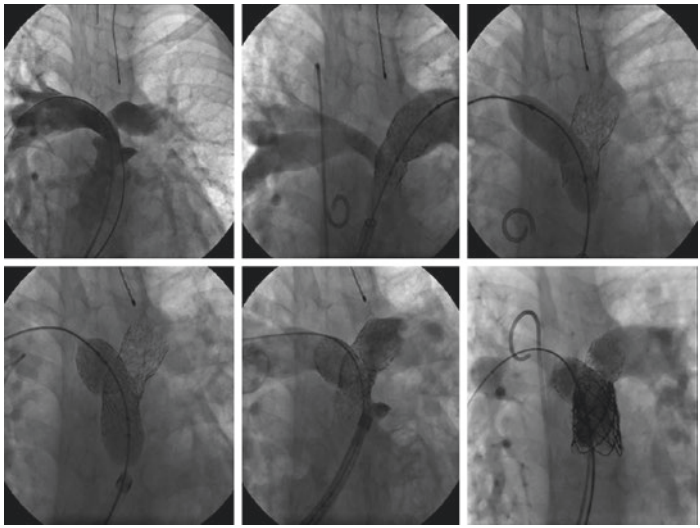

Fig. 46.3 Jailing technique. Patient with stenosis of the origin of both PAs. Creation of a Y stent. First stent is placed in the left pulmonary artery. Mesh of the stent is opened after placing a wire in the right PA. Another stent is positioned in the origin of the right PA. A Melody is then placed in the RVOT

46.4 Step by Step. What Technique to Use in Patients?

In practice, right and left heart catheterizations are performed the usual way. Angiograms are performed in multiple projections (four chambers, lateral and RAO views). It is important to measure both RVOT and PA branches in each projection to define the proper landing zone for the valve. The RVOT and the PAs are then sized using either sizing balloons or low-pressure balloons such as Tyshak II (Numed) to access their compliance. Balloon diameter should be selected 3–5 mm larger than the baseline diameter. To make sure to capture the right measurement, two tricks can be used. First, the balloons are inserted inside long sheath allowing dye injection at the time of balloon inflation. No or minimal flow across the RVOT (or PA) should be seen. If it is not the case, balloon should inflate more or a larger one should be used. Second, arterial line is connected to pressure recording. During balloon inflation in the RVOT, the aortic pressure should go down as a result of reduced pulmonary blood flow. If it is not happening, then it is a sign of inappropriate balloon inflation. The selected balloon should inflate more or a larger one should be used. The balloon testing allows to select the ideal landing zone, decide on the technique to use, and make sure that the coronary arteries will not be compromised. Below various case scenario will be discussed. Decision-making depends on valve availability in your country and hospital.

1. MPA diameter within the range of valve size availability. No special technique is required. One should think about one-step procedure to reduce procedural time and radiation exposure. In short RVOTs or if coronary closed to the landing zone or in order to stay far from the sternum, think about using a short valve or folded Melody.
2. MPA diameter below the range of valve size availability. No special technique is required. It is important to make sure that the RVOT (conduits, bioprosthesis, or any substrates) can be opened with balloon interrogation (use high-pressure balloons) before proceeding with valve implantation. Placing a valve in

none expandable RVOT will end-up leaving significant gradient putting the patient at risk for early reintervention, endocarditis, and impairment of valved stent.

3. MPA diameter exceed the range of valve size availability. Patient can be referred to surgeon for surgical replacement or special technique can be used.
   (a) Both pulmonary artery diameters are within the range of valve size availability.

   The origin of both PAs can be used as a landing zone. Valved stents are implanted using conventional technique starting with the most stenosed PA. Position of the stent is crucial. Stent placed too low in the RVOT will make placement of the second-valved stent more complex with risk of crushing or compressing the previous stent. If it occurs, a second vein should be punctured and balloons should be inflated simultaneously in both PAs to avoid compressing the stents. In case of doubt, leave a wire across the first valve stent with a deflated balloon across; expand the second valve stent in the other PA; if a compression occurs, simply inflate the balloon to reopen the previous stent while maintaining the balloon inflated in the other stent; if no compression occurs, simply remove the wire and balloon.

   Alternatively, a Y stent or a trouser-like stent can be created. First, start by placing a long stent in the more stenosed PA. The stent should extend from the selected PA to the RVOT jailing the opposite PA. Ideally, an open cell design bare-metal stent (EV3 LD max 36 mm) or hybrid such as AndraStent XXL should be used. It is important to note that the diameter of the balloon used to open the stent should stay in the range of valve availability (usually a maximum of 22 mm). After placing the first stent, the mesh of the stent is crossed with a 0.035 wire. If not possible, coronary wire can be used as well. A pre-dilatation of the strut is usually needed to facilitate the advancement of a larger balloon. If large wire has been used, vascular balloons such as Mustang balloons can open the strut. If coronary wire was only able to cross, then use of coronary

balloon (5 or 6 mm by 20, Maverick XL for example) is interesting. Ideally those balloons should be used with a guiding catheter and a frontloading technique can be used to exchange the wire with an appropriate 0.035 wire. In practice, the balloon is inflated in the strut at first, it is then deflated and reinflated again, and the guiding catheter is advanced and pushed with the balloon as an assembly inside the PA. If not working, the balloon can be slowly deflated while advancing the guiding catheter. But doing that the balloon will be recaptured inside the guiding and the guiding will be advanced within the PA. A last option is to use an over-the-wire low-profile balloon that takes a 0.035 wire. The balloon can be advanced deeply within the PA; balloon is then inflated; 0.014 wire is then removed from the lumen of the balloon and exchanged with an 0.035 wire; balloon is then deflated and pulled-back and reinflated to open the strut of the stent. While in good position with the 0.035, the strut of the stent can be opened with a larger balloon using, for example, Atlas Gold 16 to 20 mm. This is followed by the implantation of an additional stent is the contralateral PA. Here again, the stent will extend from the PA to the RVOT creating the Y shape. If needed, the mesh of the stent covering the first PA can be opened using the same technique. The base of the Y will be the landing zone for the valve stent to be implanted. If the base of the Y is not applied to the RVOT wall, Russian dolls can be used to limit the risk of paraprosthetic leak. As many as needed, stents are positioned in the base of the Y until the stents are applied to the wall of the RVOT.

(b) One pulmonary artery diameter is within the range of valve size availability. A jailing technique can be used. The first steps are very similar to the one described with the creation of a Y stent but this time only one PA is used. A long stent is positioned from the PA to the RVOT. Here again the choice of the balloon to expand the stent is very important. One should remember that the stent will be used to secure the valved stent (landing zone). Balloon diameter should not exceed 22 mm initially. If Sapien

valve is used, the stent laying in the RVOT can be expanded to a larger diameter. If bigger balloons are used, extending the length of the landing zone by placing an additional stent might be needed to compensate stent foreshortening. Here again, if the stent is hanging in the RVOT not applied to the wall, Russian dolls technique can be used. As many as needed, stents are positioned within RVOT stent until the stents are applied to the wall of the RVOT. The balloon used to implant the first stent (for example, BIB 22 mm, Numed Inc) is used to implant the subsequent stent. This assures that the stent will be stable and will not embolize. Because the same balloon is used, the inner diameter of the stent assembly remains the same but the outer diameter increases after each stent. Use of covered stents (CCP, Numed, Inc) is very useful as it will limit the risk of paraprosthetic leak and increase outside diameter faster as they have increased thickness compared to bare-metal stents.

(c) The diameter of both pulmonary arteries exceeds the range of valve size availability.

This is of course the most complication complex scenario. However, solutions exist.

Diabolo shape stent (Fig. 46.4) [9]

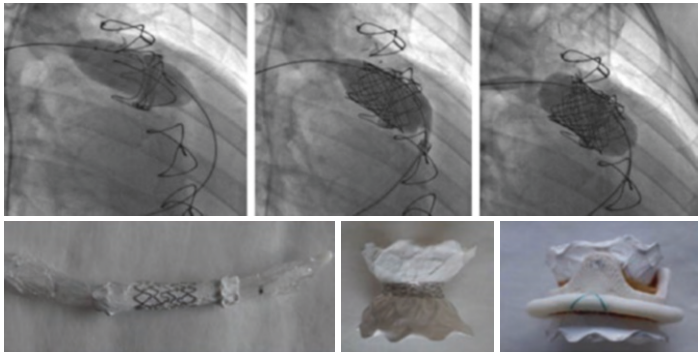

Fig. 46.4 Diabolo procedure. Placement of a Melody in a 27 mm bioprosthesis. Ex vivo images showing preparation of diabolo stent and ex vivo aspect after inflation of stent assembly

A diabolo-shaped stent can be created. Two techniques can be used. First a 20-mm snare can be positioned and trapped between a stent and a balloon. The expansion of the middle part of the stent will as a result be limited to the size of the snare. The second option is to use the property of two different stents used simultaneously with one fitted on the second one. A bare-metal stent with limited expansion (EV3 LD mega, S17-26, EV3, Covidien) allows a restrictive region in the middle part in stent assembly during inflation. This stent is crimped on the covered stent with more expansion capabilities allowing anchoring to the wall. In large RVOTs, long covered stents (10Z, Numed Inc. or Bentley Begraft aortic stent) are used. Stent assemblies are crimped over large balloons (BIB or Nucleus balloons, Numed Inc., Canada; Balt). Noted that Bentley stent being premounted on a balloon of maximum 24 mm, the stent should be re-crimped ex vivo on a larger balloon. A short EV3 LD Mega is then crimped over and on the middle part of the covered stent. According to manufacturer charts, the LD Mega cannot expand over 22 mm in diameter and foreshortening is maximal at this diameter. The balloon is slowly inflated and subsequently deflated. The stent assembly as the shape of a diabolo. The middle "restricted" area will shelter the valve to be implanted while the extremities will hold the device in position. This technique is particularly useful in large tubular RVOT and can be used with Melody as well as Sapien valves. Of course, the choice of balloons and stents will depend on the availability of transcatheter valves.

Side-by-side technique (Fig. 46.5) [10]

The technique was described for cases where no other techniques can be used because RVOT is very large. RVOTs up to 34 mm in diameter have been valved using a 22 mm Melody valve. The combination of the 29 mm Sapien valve and a side stent having a nominal diameter of 22 mm would allow to implant valves in patients with RVOT larger than 36 mm. This technique is only transcatheter. No surgical access is required. The first steps of the procedure are similar to those described

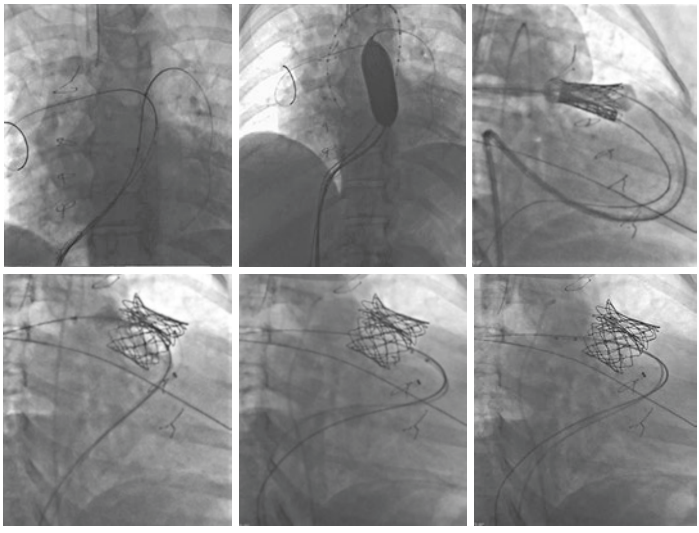

Fig. 46.5 Side-by-side technique. Example of a side-by-side technique in a patient with 34 mm RVOT. Pictures showing balloon sizing, simultaneous placement of Melody, and covered stents, followed by crushing of the non-valved stent and closure with a vascular plug

previously. Two femoral or jugular veins are needed. Balloon interrogation is performed to measure the diameter of the RVOT and to assess the position of the coronaries. When a Melody valve is used, two Ensemble 22 mm delivery catheters are prepared. In the first one, a Melody is firstly crimped on the outer balloon followed by crimping of an EV3 LD max stent 36/12 using the same technique described for the "one-step technique." In a second Ensemble delivery catheter, a CP8Z39 covered stent (Numed Inc) and an EV3 stent over it are crimped on the outer balloon. Lunderquist or Back-up Meyer wires are positioned distally in two different PAs. The Ensemble catheter is advanced in the RVOT. The covered stent (non-valved stent) is firstly deployed. The Melody is then inflated maintaining the outer balloon of the covered stent inflated until full apposition of the Melody to the wall.

While the Melody is applied to the wall, the balloons of the covered stent are deflated slowly and delivery system is removed. The balloon of the Melody is fully inflated crushing the covered stent. The delivery system of the Melody is left in place until the end of the procedure. A guiding catheter or a long sheath is placed through the covered stent. The lumen of the covered stent is then filled with a vascular plug II (St Jude Medical, USA) of appropriate size. Guiding catheter and delivery system are finally removed. This technique can be used as a bailout technique if a valved stent embolized using conventional technique. In this case, a wire is positioned in the contralateral PA using a different venous access. Over this wire, an Ensemble delivery catheter containing an EV3 mounted on a covered CP is advanced in the RVOT. A wire is then placed across the embolized valved stent. A balloon is inflated in the valve and pulled back in the RVOT. The covered stent is then inflated in the RVOT to block the instable valve in good position. After the valve is fixed to the wall, the balloon is deflated. The lumen of the covered stent is then filled using a vascular plug.

46.5 Conclusion

Multiple techniques are available to enlarge the indications of transcatheter pulmonary valve. Choosing between the various techniques depends on the anatomy of the RVOT. Newer self-expandable valves (Venous P valve, MedTech, China; Harmony, Medtronic, USA) will not fully cover the full RVOT anatomy we are dealing with [11, 12]. There are some spaces for research and development of devices to cover all RVOT anatomies.

Disclosure Statement The author has no conflict of interest to disclose.

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Part VIII

Step-By-Step Procedures: Principles of Hybrid Approach

47 Hybrid Approach in Hypoplastic Left Heart Syndrome (HLHS)

Dietmar Schranz and Hakan Akintuerk

47.1 Anatomic Description and Pathophysiology

Hypoplastic left heart syndrome is a rare congenital heart defect in which the left heart structures are underdeveloped; the right or subpulmonary ventricle has to support the systemic and pulmonary circulation. HLHS is accounting for 2–3% of all congenital heart defects [1]. The etiology of hypoplastic left heart (HLH) structures range between a flow-related "borderline" development of left ventricle (LV) [2] and a multigenetic and genetically heterogenous background of an HLH syndrome [3]. The morphology of HLHS is subdivided in mitral and/or aortic atresia or stenosis, respectively [4]. Specificities of hypoplastic left heart structures and its consecutive hemodynamics dictate the postnatal therapy. Untreated HLHS is a fatal congenital heart defect. Neonates with HLHS, but even HLHC, remain stable as long as

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a parallel circulation is balanced or still in part assured. Risk factors influence procedure-related mortality. Risk analysis includes age, prematurity, birth weight, weight at surgery, prenatal diagnosis, lowest preoperative pH, organ dysfunction, complexity of additional abnormal cardiac morphologies, the exact cardiac diagnosis of HLHS or variants, and the presence of genetic or chromosomal abnormalities. The exact cardiac diagnosis of HLHC has to be determinate in assessment of an early corrective surgery or Norwood palliation, but also before hybrid procedure. Independent of the made treatment decision, time and mode of the follow-up corrective or palliative surgeries are hypothesized not at least for fair parental counseling. Current operative mortality for the staged procedure also for the hybrid strategy has to be included. The hybrid approach consists of placing a bilateral pulmonary banding (bPAB), duct stenting, and manipulation of the atrial septum, if necessary [5]. Provided the parents written consent, newborns with hypoplastic left heart syndrome (HLHS) follow the classical three-step algorithm with final creation of a Fontan circulation [5, 6]. Meanwhile, the hybrid procedure has moved from a rescue approach to an alternative modality of a Norwood palliation [7–10]. The hybrid approach expands also the options for neonates with "borderline" left heart structures, summarized as an HLH-(C)-complex [11–14]. In this context, hybrid procedure facilitates dealing with ccTGA with a small systemic right ventricle (RV), DORV and unbalanced AVSD with small LV, tricuspid atresia combined with d-TGA, long segment subaortic stenosis, arch hypoplasia, or some variants of IAA (interrupted aortic arch). The goal of hybrid approach is a complete or partial provided systemic blood flow (SBF) via a right-to-left shunting Ductus arteriosus (DA) combined with an unrestrictive or slightly restrictive left-to-right shunt at the atrial level both (interatrial septum, IAS), essential for postnatal clinical stability and for survival. Therefore, postnatal condition of neonates with HLHS or HLHC depends on the morphology-related pathophysiology, the quality of pulmonary vein drainage, the atrioventricular valve competence, as well as the quality of blood flow guaranteeing cerebral and coronary perfusion. The

goal of the postnatal hybrid procedure is to assure or reestablish, but balancing the complex interaction of the pulmonary (Qp) and systemic circulation (Qs). A normal patient's breath rate is the leading symptom of a balanced lung and body perfusion; tachypnoea is the earliest clinical symptom of a circulatory imbalance. Components of the hybrid approach are also the most effective measures to treat an unstable newborn with HLHS or HLHC. The hybrid procedure is suitable for almost all cardiac lesions with DA-dependent SBF, regardless of age (premature, infancy), weight, or hemodynamic stability, in particular treating cardiogenic shock.

47.2 Clinical Scenarios

Prenatal, fetal echocardiography improves the postnatal outcome of newborns with HLHS. Prenatal counseling of the parents avoids postnatal heart failure or even cardiogenic shock. If a restrictive atrial communication excluded, a prophylactic prostaglandin infusion in a low dosage of about 5 [10] ng/kg × min can be safely applicated by a continuous infusion; by this drug regime, PGE1 treatment has almost no side effects (Chap. 25). Considering the written consent of the parents, a stable neonate can be prepared for a surgical procedure, based on three-stage strategy, independent if a Norwood or hybrid approach is chosen. In term of the patient's condition, each component of the hybrid approach—bilateral pulmonary banding, duct stenting, atrial septum manipulation—might also be high effectively used as a high-urgency procedure; even for bridging to heart transplantation.

47.3 Indications and Patient Selection

Independent of the dramatic improvements of surgical and intensive care for newborns with HLHS, lastly the parents have to decide freely among three options that include a surgical, surgical-interventional treatment, or compassionate therapy.

This therapeutic choice should be done after an intense communication and discussion. Sensitive counseling after prenatal diagnosis might have the best chance for the parents to opt the best for their child. In any case, the heart defect by itself and its current and long-term consequences have to be explained in detail. Following consent of the parents, hybrid approach is currently performed as the procedure of our first choice in all newborns with HLHS and in particular for treating high-urgency situations of newborns with HLHS. In case of prostaglandin refractory duct obstruction (metabolic acidosis), duct stenting is the treatment of choice; in case of a systemic low cardiac output due to pulmonary runoff, immediately performed surgical pulmonary branch banding is the best therapeutic option. Transcatheter creation of a sufficient atrial septal communication is performed with various techniques in any compromised heart–lung interaction caused by a missing or severe obstructive interatrial communication.

47.4 Pre-procedural Imaging

Echocardiography (ECHO) is the imaging of choice to detect and to subclassify HLHS in a suspected newborn. ECHO examination needs to collect all morphologic and functional data. In HLHS, atrioventricular, ventriculoarterial arrangement is mostly normal, but ccTGA with hypoplastic systemic right ventricle has postnatally the same pathophysiological and therapeutic consequences. Imaging of the interatrial communication, pulmonary venous flow characteristics, the atrioventricular valve (tricuspid valve), systemic ventricular function, the duct morphology in relationship to the aortic arch, and the assumed coronary and cerebral needs to be performed in clinical context; in particular, in patients with an atretic aortic valve (Chap. 25). In complex anatomy, cardiac MRI gives additional insights of the cardiovascular anatomy and function; like catheterization, cMRI is routinely performed without anesthesia only in a light sedation (Fig. 47.1).

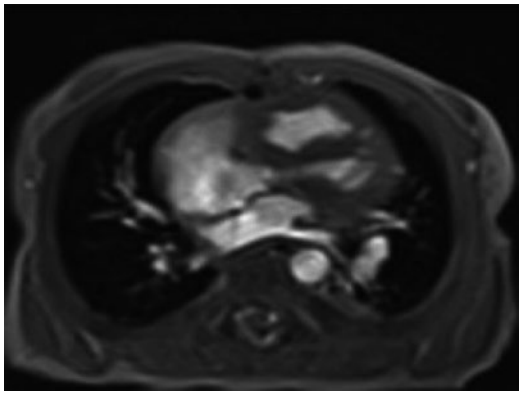

Fig. 47.1 depicted is four-chamber view performed by a 3 Tesla MRI machine; left ventricular cavum is almost missing, the atrial septum restrictive, dominant is the anterior positioned enlarged, functionally single-right ventricle

47.5 Giessen Hybrid Features

The "Giessen hybrid (GH)" stage I approach is a sequential procedure, which differs from other hybrid strategies (i.e., Columbus, [9]), in which duct stenting is performed by a transpulmonary access, immediately after bilateral pulmonary artery banding. Considering the GH, bPAB is performed as an exclusive open-chest, "off-pump" surgery; duct stenting and atrial septum manipulation are performed as an elective percutaneous transcatheter approach later on [10, 12]. The GH is meanwhile used for almost all types of HLHS and variants as well as selected newborns with HLHC; the principle of the GH is also translated to older patients, even adults suffering on a similar pathophysiology, both, as a definitive treatment or bridging to transplant [15, 16].

HLHS stage I, including the interstage to comprehensive stage II, can be offered with a mortality of less than 10%. Prerequisites of a successful interstage are a) closed controls by experienced physicians and b) medication for improved interstage aiming a

sufficient body weight gain during the interstage by balancing Qp/Qs and optimizing the DO2 to VO2 ratio. Therefore, we avoid diuretics and fluid restriction, instead we applicate a β1-selective blocker (B, bisoprolol) in almost all patients, and a tissue ACE-inhibitor (L, lisinopril) if the coronary perfusion pressure is not jeopardized; so, nearly 90% receive a β1-blocker and 70% of the newborns after hybrid approach receive a tissue ACE inhibitor; the mineralocorticoid blocker (S, spironolactone) is used under the assumption to influence right ventricular fibrosis [17]. We change our B-L-S concept only in case of persistent atrial tachycardia, to an unselective β-blocker (propranolol), which was necessary in less of 10% of our cohort.

The best interstage monitoring is performed by enlightened parents monitoring the ventilation rate during sleep of their babies. Heart rate at rest of less than 120 or even 110/min is usually related to a well-balanced circulation in a young infant after stage I. The long-acting β1-specific bisoprolol and tissue ACE inhibitor lisinopril can mostly be applicated in a similar dosage of 0.1–0.2 mg/kg once per day. While, spironolactone is applicated in a less-diuretic dosage of 1–2 mg/kg also only once per day; respecting dosages and applications once per day, the recommended medical combination is very attractive for the parents achieving a high compliance. In case of judgment between the triple drug therapy, the β1-receptor blocker seems to be most beneficial one in term of cardiac economizing, heart rate is reduced, and diastolic ventricular filling time prolonged [17].

Comprehensive stage II is performed at an age of 4–6 months currently with a surgical mortality of about 5% [10, 18, 19]. Comprehensive stage II combines removal of bilateral PAB and DA stent, reconstruction of a Norwood-like aortic arch, and bidirectional Glenn shunt (BCPC). TCPC with a Fontan circulation is the final step, usually performed in an age of 2–4 years; an elective fenestration was in the past performed in almost all patients despite utilizing a preferred extra-cardiac conduit; currently in about 50–60% of the patients [20]. Since June 1998, almost 260 patients received a stage I procedure as an initial approach treating newborns with HLHS and HLHC. Following initial hybrid

approach; the 20 years survival probability for HLHS and variants (n = 169) represents 76%; Fontan completion, performed meanwhile in almost 100 patients, can be offered with a surgical mortality of less than 2%. Following an initial hybrid approach, biventricular repair for HLHC patients is performed after the neonatal period, mostly between 3 and 9 months of age; an age, in which surgery can be performed with the lowest mortality for complex congenital heart defects [12–14]. HLHC patients (n = 94), staged by initial hybrid approach, have an acturial survival of almost 94% in a follow-up of almost 17 years. Several centers utilize a neonatal hybrid approach [21, 22], also for bridging to a Norwood surgery beyond the neonatal period [23]; a reverse BT-shunt was also introduced as a hybrid variant in case of a significant aortic arch obstruction [24].

47.5.1 Bilateral Pulmonary Banding

Elective hybrid stage I usually starts with bPAB as an open-chest approach in general anesthesia via a median sternotomy [7, 9]. Following points have to be pointed out: (A) before bPAB is performed, a wide-open duct needs to be imaged; in case of an HLHS, a relevant aortic coarctation and a significant obstruction across the IAS have to be excluded. The anesthesiologist has to avoid any overtreatment by ventilation, oxygen supplementation, or too high dosages of anesthetic drugs as a single dosage. Prostaglandin E1 infusion is preferentially used in a low dosage of 5–10 ng/kg/min. Milrinone is continued in in a dosage of 0.5–1 μg/kg/min, if already applicated prior to surgical bPAB. (B) before the pericardium is opened, volume depletion needs to be excluded. Left PA-branch isolation is slightly hindered and PAB placement associated with a greater risk for distortion. Right branch band is positioned between superior caval vein and ascending aorta, left branch banding fixed close to the pulmonary trunk, both sided PAB fixed at the adventitia avoiding band migration. Usually, the bands are fixed by utilizing a 6/0 Prolene® suture (Fig. 47.2a–c). An already stented duct is considered as

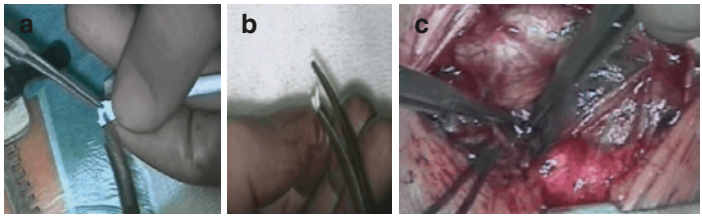

Fig. 47.2 (**a**–**c**) shows preparation of right pulmonary branch banding by utilizing a 3.5 mm PTFA E tube; 2a shows the cut of the tube to a small strip, (**b**) the cut of the 1–2 mm strip for placing around the pulmonary branch (**c**) the strip is placed around the PA-branch and sutured

an additional risk factor and should be avoided, if possible. Therefore, duct stenting prior to bPAB should only be performed as a high-urgency procedure in patients with duct obstruction despite PGE-1 treatment. There are multiple advantages of a surgical bPAB performed during only PGE-1 infusion. The surgical approach is easy, safe, short, and effective; any stenting-related hemodynamic instabilities can be avoided; mortality can be avoided. Based on Galantowicz technique, pulmonary branch banding was facilitated by utilizing a 3.5 mm or 3 mm PTFE tube graft (3 mm PTFE tube for neonates with a body weight <2.5–3 kg) cut to a 1–2 mm wide strip [9]. Small bands with a width of 1–2 mm have the advantage avoiding pulmonary branch hypoplasia, but instead forcing post-stenotic dilatation. Considering an unobstructed DA, the efficacy of bPAB is immediately observed by an increase of systemic blood pressure and ideally by a slightly decrease of the arterial oxygen saturation. There is no need to determinate the pressure gradient across the placed branch bands. Echocardiography-based Doppler-related measurements are sufficient to analyze the systolic and diastolic Doppler flow pattern; therefore, it is recommended to measure simultaneously the systemic blood pressure. Presupposed, there is no pressure gradient across the arterial duct, the systolic Doppler pressure gradient let assume the postcapillary pressure level, related to the left atrial or pulmonary vein (confluence) pressures. The diastolic Doppler component reflects the quality

of PAB. An end-diastolic flow pattern above 50–75% of the systolic Doppler flow curve can be associated with a too narrow PAB. The surgical procedure focused on bPAB does not consume more than 20 min; the time in the operation theater takes almost 2–3 h.

47.5.2 Atrial Septum Manipulation

Newborns with HLHS have in almost 6% a complete intact atrial septum and up to 22% a severely restrictive atrial septum [25]. Both anatomic entities are associated with an increased mortality rate and a need for an immediate postnatal atrial septum manipulation. The adequacy of the atrial septal communication is determined on echocardiographic and by invasive hemodynamic data. The strategy differs between a neonate born with a HLHS and HLHC. In HLHS patients, an almost nonrestrictive atrial communication is necessary; in HLHC, oftentimes, a sufficient preload of the subaortic ventricle is important, guaranteeing growth of left heart structures. In case of a too restrictive or even absent atrial communication, a balloon atrial septostomy (Rashkind procedure) or deployment of an atrial septal stent is performed. The decision for one of both measures needs to be made first, because stenting of the IAS even after an insufficient Rashkind maneuver has a higher risk in terms of stent embolization, despite that self-expandable stents with an open cell design are used; for IAS stenting, we use preferentially a sinus SuperFlex DS (15/18 × 8/9 mm). Some, very diseased newborns need an IAS manipulation by Brockenbrough or alternatively high-frequency technique, followed by ballooning, but usually stenting of the IAS. Considering a high-urgency measure, total anomalous venous return (TAPVR) with a severe obstruction can also be treated by transcatheter techniques; both by stenting an obstructed vertical vein or by perforating the wall between pulmonary vein confluence and left atrium followed by stent placement [26].

47.5.3 Elective Percutaneous Duct Stenting (See Chap. 25)

Following bPAB, DA stenting is performed under stabilized hemodynamics as an elective, precise, and even low-risk percutaneous transcatheter approach. BPAB without previous DA stenting, but continuous PGE1 infusion stabilize also a slightly obstructed aortic isthmus or avoid a significant aortic coarctation (CoA). Considering an unobstructed wide-open arterial duct, it should be mentioned that a stented duct does not necessarily improve hemodynamics or add patient's stability; duct stenting is performed because of out-clinic bridging to comprehensive stage II, and avoiding side effects of PGE1 even by mid-term application.

Stenting of the ductus arteriosus carried out by a percutaneous transcatheter approach was facilitated by novel designed self-expandable sinus SuperFlex DS stents with the CE mark for duct stenting in newborns (OptiMed, Karlsruhe, Germany). Access by 4 Fr sheath allows duct stenting via femoral vein as well as artery. The arterial access made duct stenting to an easy approach [10, 12, 16]. Stent size and positioning within the duct is based on a right lateral oblique 30° and 90° lateral angiogram. Choice of stents is largely influenced by the ductal anatomy and the morphology of the duct–aortic junction. Balloon-expandable stents are currently used only in case of an obstructed arterial duct despite prostaglandin treatment. One main reason, why we prefer percutaneous duct stenting is based on the variability of the duct-descending aortic junction. An additional narrowed aortic isthmus or aortic coarctation can be treated, if necessary, before or after duct stenting. Some patients receive only a "prophylactic" balloon dilatation of the slightly obstructed aortic isthmus before stent placement within the duct, other patients need an additional stenting of an aortic coarctation after duct stenting. Therefore, a narrowed descending aortic arch is not per se considered as a contraindication for hybrid approach among patients with aortic atresia. As described in Chap. 25, prostaglandin is not stopped before duct stenting, rather continued for 24–48, together with heparin infusion.

Taking into account the institutional experience, technical and material details, percutaneous duct stenting can be performed with almost no mortality [10].

47.6 Follow-Up Care

The hybrid stage I is not finished, when bPAB and IAS manipulations are performed and the arterial duct successfully stented. Out-clinic follow-up prior to stage II is routinely performed at 1–2-week intervals or earlier depending on the clinical condition during interstage I. History, clinical examination (respiratory, heart rate) including systolic and diastolic (never mean!) blood pressure measurements at the right arm and of the leg, which is not used by catheterization before as well as pulse oximetry at right arm and one leg (HLHC!) are routinely performed. Following these mandatory information, echocardiographic examination has to rule out any potentials for immediate hemodynamic imbalance. Therefore, pulmonary vein flow, atrial communication, atrio-valve competence, systolic–diastolic single or bi-(inter) ventricular function, pulmonary valve competence, pulmonary flow pattern within the trunk, assessment of the bPAB, and stented duct flow as well as the retrograde (antegrade or concurrent) aortic flow quality need to be analyzed, including the flow quality within the coeliac trunk as well as (right) anterior cerebral artery (Fig. 47.3).

Routine in-hospital interstage monitoring is not necessary and should be avoided or minimized in interest also of the psychosocial development of the affected baby. The interstage mortality can extremely be reduced by very well-educated parents and a strict surveillance program, but a sudden death can never be fully excluded. By this information, the parents are discharged home and well educated to observe their baby's breathing pattern and rate during sleep or at rest in addition to the baby's feed and thrive behavioral and in particular weight gain development. Considering the persistence of a parallel circulation after hybrid stage I, the patients are also carefully instructed on chronic treatment with 1 × 0.1–0.2 mg/kg bisoprolol and lisinopril and

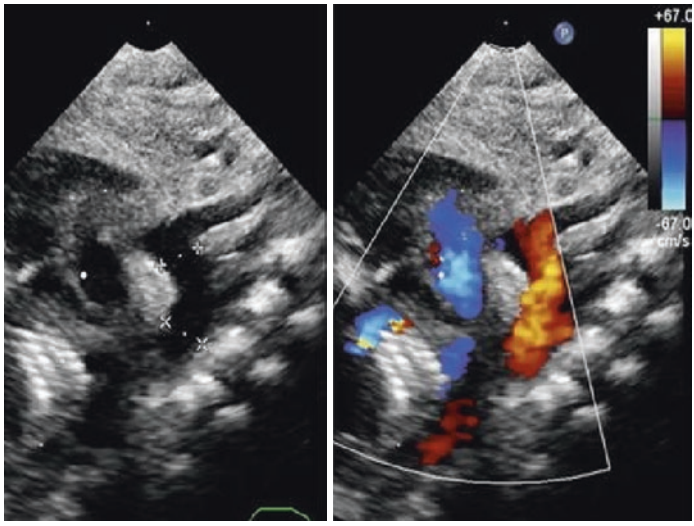

Fig. 47.3 Echocardiographic suprasternal short axis shows an unobstructed aortic arch in 2D and by color Doppler in a newborn with HLHS (MA, AA) after hybrid stage I consisting of surgical pulmonary artery banding, percutaneous duct, and atrial septum stenting, respectively

spironolactone; digoxin is sometimes additionally applicated in case of a still too high heart rate despite β1-receptor blocker; diuretics, as furosemide is usually avoided. The two main indications utilizing this drug regime are aimed to reduce oxygen consumption by avoiding unnecessary high heart rate, and consecutively breath rate, the systemic vascular resistance without endanger coronary and cerebral perfusion pressures and to increase the diastolic filling time. Additionally, blocked neurohumoral activation reduces diastolic left-right shunt across the stented duct (Fig. 47.4). Effectiveness of this drug regime is best monitored by the body weight gain before stage II.

Prior to comprehensive stage II, patients with a hybrid stage I approach are not referred for an elective invasive hemodynamic and angiographic evaluation, however only if an issue is suspected. CMRI is additionally used for echocardiography assessment, if any unanswered diagnostic question remains open prior

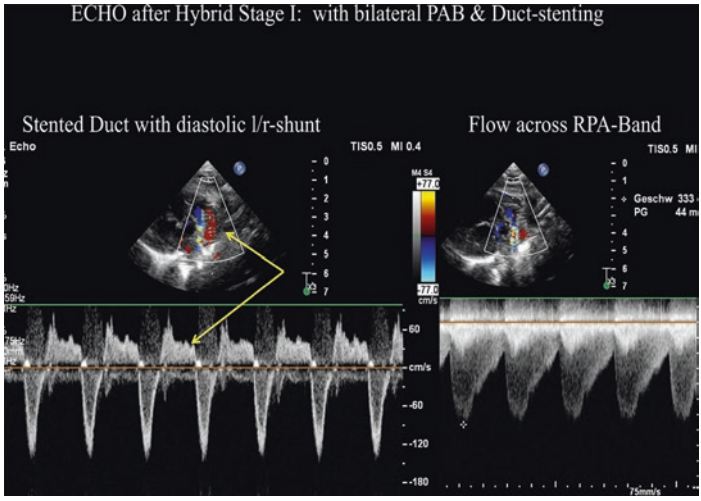

Fig. 47.4 depicts the echocardiographic of a stented duct (left side) and the CW Doppler across a right branch pulmonary artery band (right side); the red color of the 2D, color-Doppler corresponds with the diastolic backwards flow (PW-Doppler) through the stented duct. On the right side of the Fig. 47.1, an optimal Doppler-flow pattern is shown obtained by continuous wave Doppler across the right pulmonary branch band. The systolic gradient was measured about 44 mmHg, the diastolic pattern demonstrates an effective, but not a too tight PAB

to provide stage II; it has to be noticed that cMRI, like an elective heart catheterization, is performed only in sedated and spontaneous breathing patients; general anesthesia is routinely avoided because it is considered dangerous for a routine procedure, which is true for all patients with duct-dependent systemic blood flow (Fig. 47.5).

47.7 Tips and Tricks

Tip 1: *hybrid mentality* between pediatric cardiac surgeon, anesthesiologist, and pediatric cardiologists is mandatory for successful hybrid approach. A non-experienced institution should usually

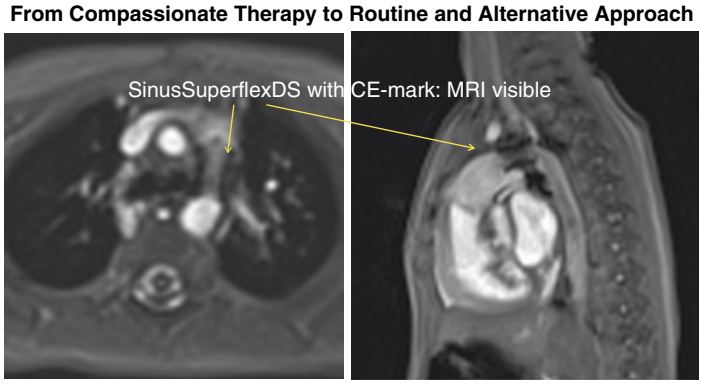

Fig. 47.5 cardiac MRI shows an arterial duct with a well-visualized nitinol sinus SuperFlex duct stent. The stent is exact positioned, the descending aortic arch free of a stent

not start with HA in complex or high-risk HLHS patients; in interest of the patients, it seems to be beneficial starting the hybrid program with lower risk patients or neonates with HLHC. It has to considered that the hybrid approach has its own learning curve with an immediate impact on the patient's outcome. Therefore, institutionalizing a hybrid program, in particular replacing the Norwood-procedure, needs a careful institutional preparation.

Tip 2: *safety* of patients with duct-dependent systemic blood flow depends on wide-open duct, high pulmonary vascular resistance despite an unrestrictive interatrial communication. Considering postnatal fall of Rp, usually bilateral PAB is indicated within the first 2–5 days. Before bPAB is not sufficiently performed, anesthesia, intubation, and controlled ventilation remain critical. High pulmonary vascular resistance as well as bilateral PAB defend against hemodynamic instability caused by pulmonary run-off. Therefore, when IAS manipulation becomes necessary as the first and urgent part of the hybrid procedure, surgical bPAB should additionally be prepared to perform any time after successful atrial septostomy.

Tip 3: *duct stenting* is an elective approach in the majority of neonates with HLHS and HLHC. There is no need for duct

stenting as long as the arterial duct remains wide open with low dosages of prostaglandin ("Japanese hybrid strategy" without duct stenting). Based on the institutional decision-making, duct stenting can be performed as a true hybrid approach by transpulmonary access or as an elective percutaneous transcatheter approach independent of the surgical bPAB. However, it has to be mentioned that the "Columbus" hybrid [8] combining bPAB with transpulmonary duct stenting does also have its own, but different learning curve. Weighing benefits and pitfalls of both the "Columbus" and "Giessen" hybrid, we, the authors, opted for performing bPAB as a sole surgical approach followed by duct stenting and IAS manipulation as a separate percutaneous transcatheter procedure. However, our recommendation is also related to local availability of materials in particular stents. An established institutional experience of percutaneous duct stenting treating duct-dependent systemic blood flow has additional advantages: it can be performed as a high-urgency approach in case of a prostaglandin refractory duct obstruction; it gives the chance for precise stent placement and analyzing the aortic isthmus before and after duct stenting and further; IAS manipulation can additionally performed, if necessary.

Tip 3: *the use of a certificated self-expandable stent systems (sinus SuperFlex DS, sinus-Repo-DS)* improves and facilitates percutaneous duct and IAS stenting as well as stent placement within a retrograde aortic obstruction, if necessary. Therefore, "Giessen hybrid" expanded options for newborns with HLHS and HLHC. Further, utilizing self-expandable nitinol stents reduce the risk of an artificial duct obstruction during a follow-up bPAB surgery.

Tip 4: *postsurgical hybrid stage I* treatment needs also to follow a well-prepared protocol; early extubation, nonventilated, spontaneous breathing neonate with focus on a balanced Qp/Qs and VO2/DO2 should be the preferential aim; therefore, following surgical bPAB, the patient needs to be carefully monitored and early extubated without the risk for reintubation; oxygen demand and consumption are best guided by monitoring the heart rate and systolic diastolic blood pressure measurements at the right upper and left lower limb. Alpha-2 receptor agonist

clonidine (dexmethomidine) is preferentially used because of its reducing effects on heart rate and systemic vascular resistance as well as sedative properties. Inotropic treatment is usually not necessary, if indicated the inodilator milrinone is drug of choice. Duct stenting and/or IAS manipulation can be performed immediately or as an elective approach after extubation and stabilized conditions.

47.8 Pitfalls (See also Chap. 25)

Pitfalls and complications of duct stenting are described in Chap. 25.

Anesthesiologic pitfalls are based on neglecting the very sensitive parallel circulations. Induction of the anesthesia, technical problems during intubation, hyperventilation, and application of too much oxygen are dangerous because systemic low-cardiac output can easily be induced, in particular, if the bPAB is still not performed.

Surgical-dependent pitfalls are cardiac ischemia or artificially induced arrhythmias by too much manipulation in particular in context of a very tiny ascending aorta. Pulmonary branch banding needs to be carefully performed in close communication to the anesthesiologist. Bleeding, caused by a left atrial appendage injury, is one surgical complication by placing the band around the left pulmonary artery; it needs to be avoided, as any other risk for bleeding (transpulmonary duct stenting) or reasons to imbalance the fragile univentricular circulation.

Intensive care pitfalls are also related to neglecting the sensitive parallel circulation as well as the immense pressure and volume work-loaded right-single ventricle.

47.9 How to Manage Complications

Dealing with complications of duct stenting is described in Chap. 25.

Anesthesiologic-, surgical-, and intensive care-related complications are rare in experienced centers and in particular if surgery is concentrated on left and right branch PAB.

Artificial aortic coarctation following stent placement within the duct should immediately be treated by placing a coronary 4–5 mm or sinus-Repo-DS™ stent. Continuing prostaglandin infusion during the first 24–48 h after duct and aortic isthmus stenting is hypothesized as an additional factor for reducing the incidence of acute but may be also follow-up complications. The duct tissue is hyperreactive to any foreign body, but remains prostaglandin-sensitive; therefore, we applicate PGE1 in a dosage of 5 ng/kg/min together with a heparin infusion in a dosage of 300 E/kg/24 h. Then, heparin is replaced by oral clopidogrel (0.2 mg/kg, once per day) if two or more stents were placed within the duct or an aortic isthmus stent became additionally necessary; oral application of cyclooxygenase inhibition is avoided as an anti-aggregative drug, minimizing the risk for additional duct constriction.

Stenting of the interatrial septum has a high incidence of embolization; utilizing self-expandable sinus SuperFlex DS reduced the embolization rate dramatically; however, if a stent embolized, the nitinol designed stent can easily be snared both for retrieving and repositioning.

47.10 Postprocedural Care

Following successful hybrid stage I, the patient needs a close immediate and mid-term surveillance until the comprehensive stage II is dated. Usually, the clinical condition after HA allows to care the baby by their parents at home. Ideally, respiratory rate should be less than 40 (60)/min, and the hemodynamic data have to be stable: systolic blood pressure at the right arm above 65 mmHg, pressure difference to the not catheterized leg not more than 15 mmHg, and oxygen saturation less than 90% and above 75%; echocardiography should show an unrestricted or only slightly restricted interatrial communication and a stented duct flow of less than 2.5 m/s; the Doppler flows across the bilateral PAB should show a systolic–diastolic pattern; retrograde flow in the aortic arch, truncus coeliacus flow, and flow in the arteria cerebri anterior need to be monitored by color and PW Doppler.

HLHS patient after hybrid stage I should have an acceptable right ventricular function, almost competent tricuspid, as well as pulmonary valve function. The numbers mentioned should not be understood dogmatically, but serve as a reference point.

47.11 Follow-Up

During the interstage, an experienced pediatric cardiologist should be responsible for the patient. Together with the pediatric cardiologist at home, the patient needs to be closely monitored until comprehensive stage II is successfully performed usually in an age of 4–5 months. Considering, that the hybrid stage I is a palliative approach, patients with fully duct-dependent systemic blood flow have a high mortality risk independent of the cause of duct obstruction. The same is true for any significant obstruction within aortic arch or atrial communication. The medical art consists of assuming any complication and not waiting for a complication. Therefore, close follow-up control is mandatory in any patient until the next therapeutic step is performed.

Since 1998, when hybrid stage II was first time successfully performed by Akintuerk et al. [7], several surgical and catheter-based modifications were conducted. The stage II surgical reconstruction consisted of amalgamation of the proximal ascending aorta with the main pulmonary artery, removal or resection of the ductus/stent complex, aortic arch reconstruction, atrial septectomy, removal of the branch pulmonary artery bands with routine angioplasty or left pulmonary artery patching or prophylactic stenting in some, and superior cavo-pulmonary connection. Hypothermic cardiovascular arrest is avoided; some patients underwent a comprehensive stage II even without cardiac arrest during permanent beating heart [10]. An ideal intensive course treating a patient after comprehensive stage II was recently described in detail [27]. Completion of Fontan circulation is routinely performed in an age of 2–4 years. The total cavo-pulmonary connection is also performed without circulatory or cardiac arrest by utilizing an extra-cardiac conduit in most, but not all with surgical fenestration. Transcatheter fenestration is performed during

an acute or late follow-up, if necessary [28]. The variants of biventricular repair with the special respect of the diversity morphologies as well as the surgical technique of heart transplantation were previously described [8, 12, 29].

47.12 Summary

Prenatal diagnosis of prenatally HLHS and HLHC improves the postnatal management. Neonates are not further admitted in cardiogenic shock because of severe obstruction of the arterial duct, pulmonary run-off in unrestricted interatrial communication or by severe atrial septum restriction. Surgical options are based on a three-staged procedure, delayed biventricular repair or heart transplantation (HTX). Independent of the improvements of surgical, interventional, and intensive care for newborns with HLHS, the parents have to decide for classical Norwood stage I, surgical-interventional treatment (hybrid-stage I), and HTX or compassionate therapy after an intense repetitive communication. Hybrid stage I or the components of the approach might be life-saving; presupposed, the pediatric heart team is familiar with a hybrid strategy and with any surgical and interventional step of the approach. In this term, the hybrid approach gives the chance to avoid neonatal high-risk operations utilizing cardiopulmonary bypass with or without cardiac arrest. Hybrid approach per se has not to be associated with mortality. From the current available techniques and materials at least in Europe, there is almost no reason for death from the procedural point of view, as it was in the past, when the hybrid procedure started [5, 30]. We emphasize that the outcome of newborns with HLHS is strictly dependent on straightforward decision-making and based on the goal to offer an effective, but "gentile medicine" in term of minimal invasiveness, which completely differes from the situation at the beginning [30].

Perspective: already today, hybrid stage I can be performed in a spontaneous breathing, well sleeping newborn only based on percutaneous transcatheter techniques. The surgery can exclusively focus on the comprehensive stage II; followed by Fontan completion, again as a minimal invasive transcatheter approach.

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48 Hybrid Approach: Defect Closure

Gareth Morgan and Eric Rosenthal

48.1 Anatomical Considerations

The hybrid approach to ventricular septal defect (VSD) closure is applicable in a wide range of muscular VSDs. While hybrid closure is possible in most muscular VSDs, the anatomical position will influence the approach to closure and may limit the ability to appropriately position a closure device [1–3].

48.2 Indications and Patient Selection

Whenever a hybrid approach to VSD closure is considered, it should always prompt careful review of all the approaches to VSD closure including transcatheter, hybrid and traditional surgical closure.

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48.3 Indications

1. *The patient is too small to consider transcatheter closure of the septal defect.* Transcatheter VSD closure is usually reserved for patients greater that 10 kg. Most children with haemodynamically significant VSDs (causing heart failure or a risk of pulmonary vascular disease) require closure within the first 6 months of life and are too small to allow predictable morbidity-free success with a transcatheter approach.
2. *There is a relative contraindication to cardiopulmonary bypass.* This may be due to an ongoing neurological concern or a thrombotic or thrombophilia tendency. In the vast majority of cases, hybrid VSD closure is performed without cardiopulmonary bypass.
3. *The anatomical location of the defect is such that a surgical or transcatheter approach may be difficult.* Defects whose RV exit points are placed in the more extreme regions of the ventricular septum, such as apical and anterior mid-muscular and those closely associated with the moderator band, may be more amenable to a hybrid approach.

48.4 Typical Clinical Scenario

A 4.8 kg infant following surgical repair of a perimembranous VSD is unable to progress from ITU respiratory support despite maximal anti-failure treatment. The chest radiograph is consistent with a large left-to-right shunt. The echocardiogram shows a dilated left side of the heart, a surgically repaired perimembranous VSD and a significant left-to-right flow across the muscular ventricular septum. A haemodynamically significant additional muscular VSD is found apical to the moderator band and measures 5 mm. The tricuspid valve regurgitant velocity suggests an RV pressure at least 75 % of systemic pressure.

48.5 Treatment Options

This patient should be considered for muscular VSD closure when appropriate aggressive medical management has failed.

Option 1: surgical device closure. The patient is within 2 weeks of their initial cardiopulmonary bypass run. The position of the defect in such a small infant is likely to provide a major challenge to the surgeon.

Option 2: percutaneous transcatheter device closure. Although theoretically feasible, the practicalities at this weight in this clinical setting are unfavourable. Even if the defect can be crossed from the left side with a wire and catheter, manipulating a delivery sheath through the right side of the heart without major haemodynamic instability and accurate device delivery is likely to be impossible.

Option 3: hybrid periventricular VSD device closure. Given the patient's weight, position of the defect and the clinical condition, this is an attractive option.

48.6 Pre-procedural Imaging

Adequate imaging is usually possible with high-quality transthoracic echocardiography alone at this age. Transoesophageal echocardiography (TOE) and 3D echo imaging can add useful information in delineating the shape of the defect and allowing a better understanding of its orientation on the septal surfaces. Angiographic delineation of the ventricular septum may be particularly useful in larger patients with complex multiple defects, but is unlikely to add much at this age. The key features which need to be recognised and discussed are:

1. Size and position of the target lesion.
2. Relationship to structures such as the moderator band (the defect may straddle the moderator band), the tricuspid valve and its septal attachments and the mitral valve apparatus.

3. Proximity to the apex and the cavity size on either side of the defect (i.e. how much space is available to deploy the left and right discs).
4. The presence and significance of any additional defects – do these also require closure? If not, then they need to be recognised to ensure that the correct defect will be crossed.

The imaging data is carefully scrutinised by the interventionist, the surgeon and the echocardiographer to plan the procedure and the equipment inventory.

48.7 Technique (Step-by-Step)

1. The ideal place is in a fully specified hybrid operating facility. A full description of this can be found elsewhere; however, in brief biplane angiographic imaging equipment should be available in case angiography becomes necessary during the case. The room should have full cardiopulmonary bypass and deep hypothermic circulatory arrest capabilities. TOE imaging is the key imaging modality in these cases and angiography is rarely necessary. Epicardial echocardiography can provide additional useful guidance (Figs. 48.1 and 48.2).
2. When the cardiac position and connections are normal, a sternotomy will usually be the correct approach; however, a thoracotomy or subxiphoid approach may be used in cases where the anatomical orientation is favourable. Exposure of the right ventricular surface is usually adequate, allowing a "limited" sternotomy to be used. Cardiopulmonary bypass should not be necessary in uncomplicated cases.
3. After locating and delineating the defect on TOE, the correct position to puncture the right ventricle is identified. A combination of angle towards the septum, cavity space for device deployment, proximity to the moderator band and the space constraints for the operators to manipulate the catheters and sheaths needs to be considered. Practically, this is done by indenting different parts of the RV free wall with a finger while observing the TOE image.
4. Prior to puncturing the RV, the occlusion device is selected, prepared and loaded, ready for insertion into the sheath. The

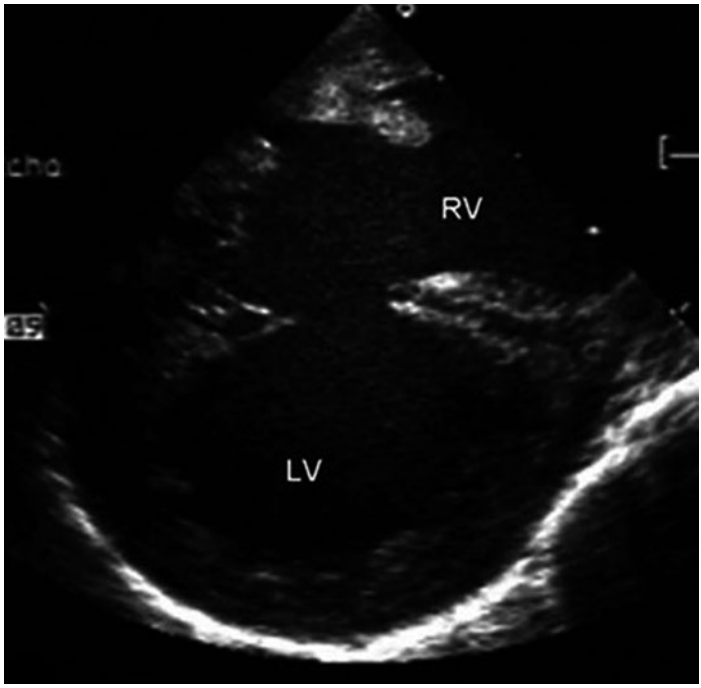

Fig. 48.1 A large mid-muscular VSD delineated with epicardial echocardiography in a 4.8 kg patient. Epicardial echo can be useful as the probe can be used to mimic the desired angle and direction for the wire and sheath passage. *RV* right ventricle, *LV* left ventricle

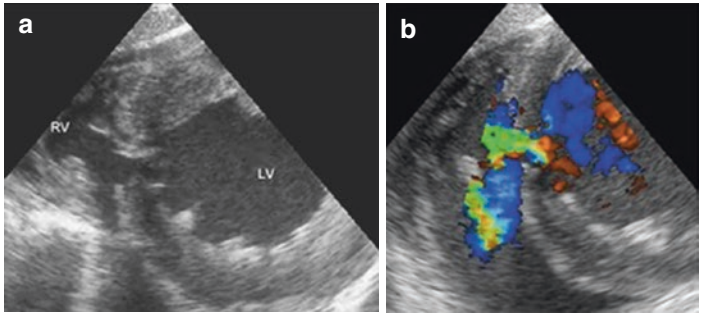

Fig. 48.2 TOE view of a moderate mid-muscular VSD with 2D imaging (**a**) and colour flow Doppler (**b**). Note that the TOE view gives a less 'surgical orientation' of the defect and requires more spatial awareness from the operators compared with the epicardial scan. *RV* right ventricle, *LV* left ventricle

correct device size usually has a waist diameter of 2 mm larger than the maximum measured diameter of the defect. The most frequently used device is the St. Jude AMPLATZER Muscular VSD Occluder; however, VSD occluders by other manufacturers are available. In certain anatomical variants, other device designs such as that used for patent ductus occlusion may be more appropriate although this would be "off-label" use.

5. A purse string is placed on the RV free wall and heparin 100 units/kg is administered. Under TOE guidance the RV is punctured with an 18 g needle and a 0.035″ Terumo J-Tip hydrophilic guidewire guided across the defect into the LV cavity. The guidewire is ideally directed out the left ventricular outflow tract to avoid interference with the mitral papillary muscles and away from the posterior wall of the LV. A short (7.5–15 cm) sheath, large enough (usually 6–10 F) to accommodate the chosen device, is advanced over the wire and across the VSD to the LV cavity (Fig. 48.3). Depending on the anatomy, the VSD may be difficult to cross with the puncture needle and wire. Although attempting to direct the wire with a catheter and wire combination, it is likely that the RV free wall

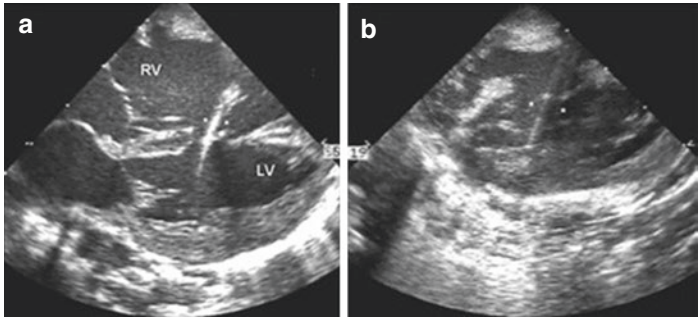

Fig. 48.3 With epicardial imaging the defect shown in Fig. 48.1 has been crossed with the sheath and wire and this has been followed with the dilator and sheath, delineated by the *asterisks* (**a**). After the dilator and wire have been removed, the 'train-track' appearance of the empty sheath is seen (**b**). *RV* right ventricle, *LV* left ventricle

puncture point is suboptimal and needs to be redone. A perpendicular approach from the free wall to the ventricular septum is required so as not to distort the anatomy and enable successful deployment.

6. Using TOE guidance, the LV disc is deployed in the mid-cavity and withdrawn to oppose the disc onto the septum (Fig. 48.4). The waist of the device and subsequently the RV disc are uncovered by withdrawal of the sheath. Several attempts may be needed to conform the RV disc correctly; it is therefore important not to pull the sheath out of the RV during the initial deployment. Indeed the RV disc may not completely conform on the RV septal aspect due to trabeculations, moderator band and limited chamber size near the apex. The operators must then decide whether the RV disc has formed adequately to allow defect occlusion and device stability even if it looks constrained (Fig. 48.5).
7. When the device is in the appropriate position on TOE and not interfering with cardiac function, the device is detached from the delivery cable.
8. The sheath is then withdrawn and the purse string tightened.

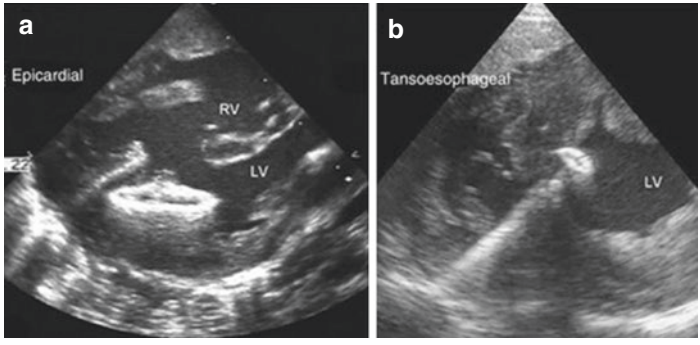

Fig. 48.4 The difference in orientation between epicardial (**a**) and TOE (**b**) guidance is demonstrated here, with deployment of the left ventricular disc. Note that careful planning and good imaging have allowed the disc to be opened free in the LV cavity in both cases. *RV* right ventricle, *LV* left ventricle

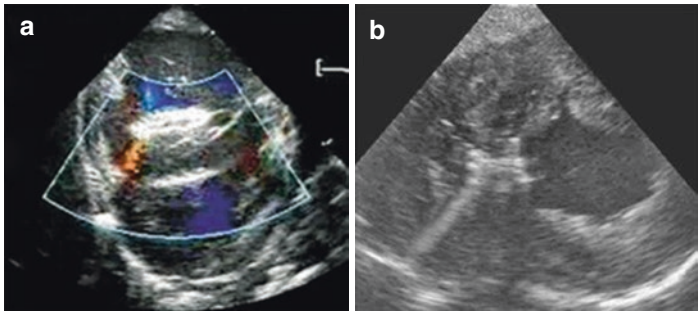

Fig. 48.5 Assessment of the conformation of the waist and RV disc is one of the most important steps. Time should be taken to assess the device and possibly multiple modalities including fluoroscopy can be used. Here we see satisfactory conformation of both discs on the epicardial echo (**a**) and TOE (**b**) – the waist of the device can also be seen to have conformed well (**b**)

48.8 Tips and Tricks

1. Perforation of the posterior wall of the LV with the sheath and dilator is a recognised complication. There are two practical ways of decreasing this risk. Firstly, placing the wire in the aorta should deflect the sheath away from the posterior structures during advancement. Secondly, the dilator should be withdrawn from the sheath until just before the transitional "shoulder" to minimise the length of dilator that needs to be advanced into the LV (Fig. 48.6).
2. Some operators have advocated soaking the device in the patient's blood prior to insertion to decrease the risk of postoperative haemolysis.
3. On occasions where the stability of the RV disc is uncertain, we have sutured the RV disc to the RV trabeculations with a brief period on cardiopulmonary bypass and a limited ventriculotomy.
4. Echocardiography may be supplemented by angiography at any stage although this is rarely necessary with high-quality TOE imaging.

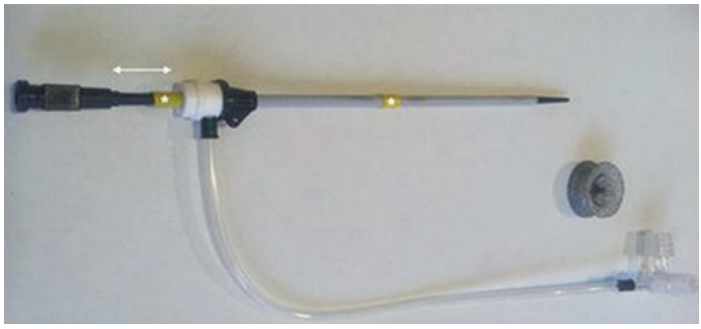

Fig. 48.6 Modification of the sheath to decrease excursion of the stiff dilator towards the LV posterior wall and managing carefully the depth of the sheath into the ventricle can be done by placing two rubber shods onto the ensemble. The first goes onto the dilator and limits its protrusion from the soft sheath to just at the shouldered transition point (*arrow*). The second goes at the point which describes the maximum insertion depth into the heart and can be used as a marker for purse-string placement, avoiding crushing or kinking the sheath at the ventricular puncture site. An AMPLATZER Muscular VSD Occluder is seen on the right of the image

48.9 Expected Results

With careful patient selection, complete occlusion of the muscular VSD or occlusion with only a minimal residual shunt should be achieved. Although no minimum weight has been defined, the size limitation is usually related to the RV and LV cavity being large enough to accommodate the conformed device discs.

48.10 Pitfalls

1. Accepting a suboptimal angle from the RV free wall may result in failure. As mentioned, major difficulty in crossing the VSD should be addressed by relocating the RV access site.
2. Perforation of the LA posterior wall. This has been addressed above and depends on scrupulous communication between the TOE operator and the interventionist.

3. Poor RV disc conformation. This may be unavoidable as described. Usually occlusion is not dependent on apposition of the RV disc; but device stability may be affected and it is important to conform the RV disc with the least tension and distortion possible.
4. Failure to identify additional defects which may be haemodynamically significant may render a difficult and expensive procedure fruitless, and a pragmatic approach must be taken if multiple VSDs, which cannot all be closed, are found at any stage in the assessment or during the procedure.

48.11 Complications

Device embolism, heart block, LV wall rupture, tricuspid valve or mitral valve support apparatus damage, air embolism, thromboembolic stroke and haemolysis are all possible. These complications can be minimised by taking into account the steps and tips above.

48.12 How to Manage Complications

The keys to managing the significant complications are preparation for conversion to cardiopulmonary bypass and the availability of angiographic fluoroscopic imaging. Complications occurring while performing these types of procedures without the necessary personnel and infrastructure back-up may lead to avoidable morbidity and mortality.

48.13 Post-procedural Care and Follow-Up

This should involve an appropriate period of recovery in a cardiac ICU. Careful confirmatory imaging of the implanted device and assessment of any residual shunt should be made over the first 24–48 h. Aspirin at a dose of 3–5 mg/kg should be administered

for 6 months after the procedure to aid non-thrombotic endothelialisation of the device. Intermittent assessment of heart rhythm and RV function should be continued along with monitoring of any concomitant cardiac defects.

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49 Hybrid Approach: Stent Implantation

Ralf J. Holzer and Alejandro Torres

49.1 Introduction

Hybrid therapies that involve the cooperation between cardiothoracic surgeon and interventional cardiologist have increased significantly over the last 25 years [1]. This applies even more so intraoperative stent placement, which has seen a vast expansion in its utilization since the late 80s and early 90s [2, 3]. Stents (as well as transcatheter valves) are now implanted using a hybrid approach in many different locations, including pulmonary arteries, aorta, RVOT on neonatal Tetralogy, atrial septum, pulmonary veins, venous baffles, and many more [4–11]. A CCISC survey conducted

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by Dan Gruenstein in 2012 documented that almost 75% of centers were placing intraoperative stents using a hybrid approach. Most intraoperative stents are placed in the pulmonary arteries, and a variety of articles have reported on institutional experiences of intraoperative stent therapy [12, 13]. While surgical patch angioplasty has often been considered the "gold standard," results have frequently been disappointing, resulting in the need to evaluate other treatment modalities, such as hybrid therapy.

However, the engagement in intraoperative stent therapy should not be regarded as a replacement of traditional transcatheter stent therapy, nor does it replace surgical patch angioplasty. Each of the three approaches to vascular rehabilitation—surgical, transcatheter, and hybrid—has its own indications, advantages, and disadvantages. As such, hybrid therapy complements rather than competes with the more traditional surgical and transcatheter therapies.

Stent therapy, whether hybrid or percutaneous, can avoid the need for extensive surgical dissection and also shortens procedure and CPB times. In addition, the results of stent therapy in the presence of a vascular kink or external compression are superior to surgical therapy. The same "radial force" characteristics that can provide advantages over surgical patch augmentation in the presence of a vascular kink can also potentially create harmful compressions of adjacent airways and other vascular structures [14].

Potential advantages of hybrid stent delivery as opposed to percutaneous transcatheter approaches include the avoidance of long sheaths and stiff wires, reduced hemodynamic instability, a fairly "straightforward" technique with shorter procedure and fluoroscopy times, better control of potential vascular complications, the ability for stent modifications such as "shortening" a stent as well as the ability to place adult-sized stents irrespective of patient size. Furthermore, hybrid stent delivery makes it easier to deal with some technical shortcomings, such as, for example, the protrusion of stent meshwork into the main pulmonary artery, which can easily be molded surgically, thereby facilitating any future percutaneous transcatheter therapy. In addition, delaying stent placement until the time of open-heart surgery allows the surgeon to inspect the lesion and decide upon the most desirable therapy that addresses the vascular lesion as well as the additional

surgical therapy that needs to be performed (such as conduit replacement). This approach maintains all therapeutic options and avoids a stent placed preoperatively in the catheterization laboratory potentially being an obstacle at the time of surgical therapy.

There are however also disadvantages of a hybrid approach as opposed to standard percutaneous stent therapy. Hybrid techniques that do not include the use of angiography make it more difficult to be certain about the distal wire and stent position. Furthermore, if a freshly dissected vessel is expanded, it can be much more fragile and vascular complications such as vascular tears are more common in a thin-walled dissected vessel, especially in smaller patients, than would be the case in a closed chest with a percutaneous approach, where the vessel is protected by surrounding (scar) tissue. Finally, it is important to choose the therapeutic modality in the overall context of a specific patient. Clearly, taking a patient to the operating room to perform a median sternotomy and solely stent a left pulmonary artery stenosis using an "open" approach with CPB would be inappropriate, unless additional surgical therapies such as pulmonary valve replacement are needed.

Hybrid stent delivery can be performed using direct visualization (with or without) endoscopy, fluoroscopy with angiography, or a combination of both approaches. Occasionally, modified intraoperative stent delivery with guidance via transesophageal echocardiography can be utilized for intracardiac locations, such as stenting of the intra-atrial septum. What technique is used depends on specific case scenarios, but also the institutional setup, such as the availability of a hybrid OR, hybrid catheterization laboratory, c-arm, and intraoperative endoscopy. This section will provide examples and technical descriptions for each of the more common approaches.

49.2 Hybrid Stent Delivery Using Direct Visualization

Hybrid stent therapy using direct visualization is probably the most common form of hybrid stent therapy. Holzer and colleagues reported its use in about 75% of cases in which hybrid pulmonary

artery stent therapy was performed [12]. The most common scenario is a patient that requires pulmonary valve or conduit replacement, and who has a concomitant kink or stenosis of proximal pulmonary artery, more common on the left than the right (Fig. 49.1). For direct visualization to work, it is important to review recent imaging data such as CT, MRI, as well as cardiac catheterization. This data provides appropriate vascular measurements, which allow choosing the correct size of the stent and balloon. Available imaging should be reviewed to identify the dimension of the stenotic segment and the adjacent normal vessel, length of the lesion as well as distance to vascular side branches. If a patient is taken to the cardiac catheterization laboratory prior

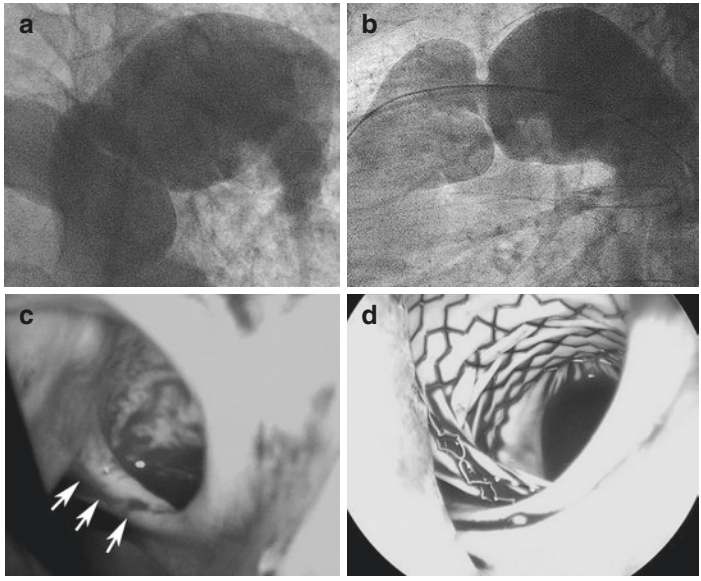

Fig. 49.1 Adult patient with a proximal LPA stenosis (**a**, **b**) undergoing hybrid stent therapy using an endoscopic approach with direct visualization, as well as surgical pulmonary valve replacement. (**c**) Documents the ridge/kink at the proximal LPA while (**d**) documents the same lesion after placement of an intraoperative stent

to any scheduled elective surgery (such as valve replacement), and a vascular stenosis is identified during the procedure, this should be discussed "there and then" with the cardiothoracic surgeons to make a decision whether a percutaneous stent delivery is to be performed or whether an intraoperative hybrid therapy is the preferred therapeutic option.

Using previous imaging data, stent and balloon choices should be made in advance to intraoperative stent placement, and the chosen diameter is additionally evaluated intraoperatively using variety of Hagar dilators. Endoscopic guidance is extremely helpful in aiding stent delivery (Fig. 49.2). Once the vessel is exposed, the endoscope is advanced until the first side branch is visualized, thereby obtaining a good estimate of the maximum length to avoid jailing of those side branches. In addition, the endoscope can facilitate advancing a wire into the correct vessel, rather than mistakenly entering a smaller side branch distally. This endoscopic

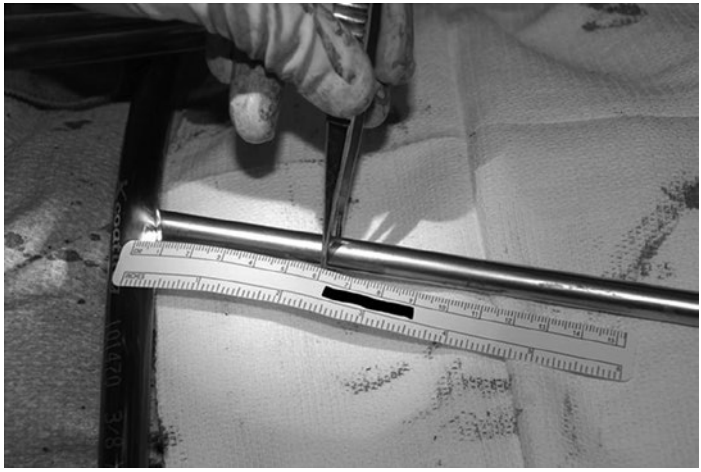

Fig. 49.2 Endoscope used during intraoperative stent placement via direct visualization. The endoscope facilitates imaging of the side branches distal to the stenotic lesion and allows estimate the distance to those branches

approach is even more important when X-ray guidance is not being utilized to visualize stent expansion. Inadvertently tearing a distal vessel due to inappropriate wire/balloon positioning is a major concern and could significantly prolong the length of the surgical procedure. In fact, a small tear may not even be immediately apparent until the vessel is closed and filled with blood.

Soft wires are preferable and stiff wires as well as long sheaths are usually not required. Prepping the stent on the balloon should be performed in standard technique, even though there is usually no need to add contrast to the balloon unless expansion is performed under fluoroscopic guidance. Furthermore, de-airing the balloon vigorously is of course less crucial when a stent is being deployed in an open vessel.

In small patients, if a stent that can be expanded to adult size is too long, it can be shortened during the procedure using standard sterilized wire cutters or strong scissors [15]. Importantly though, this should only be performed for closed-cell design stents, as cutting or shortening open cell design stents can be associated with a loss of stent integrity and radial strength. Once a wire has been placed in appropriate position, the balloon/stent is advanced over the wire and positioned by visualizing the proximal end of the stenotic lesion. If there are any concerns about wire or balloon positioning, it can be helpful to use a c-arm during stent expansion as this will show if a stent expands unequally or if the balloon/stent may be trapped in a smaller distal vessel. However, if there is sufficient distance of the stenosis to any side branch from available preoperative imaging, and/or the wire position has been adequately confirmed using intraoperative endoscopy, then there is no need for intraoperative fluoroscopy. This is in particular important for centers that do not have a dedicated hybrid OR, where the usage of an intraoperative c-arm may add considerable time to the intraoperative stent placement. Once the stent is fully expanded, the endoscope can be utilized to evaluate the entire stent position and stent lumen (Fig. 49.1). Any struts that expand beyond the proximal end of the vascular lesion can be folded-over by the surgeon using some stronger pick-ups, thereby creating a smooth adherence of the struts to the vessel

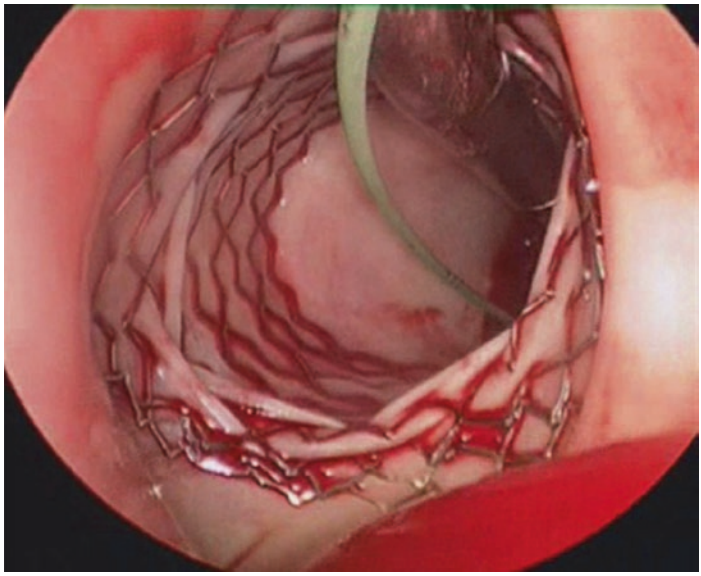

Fig. 49.3 Stent positioned in proximal LPA using direct visualization with endoscopy. Note the folded meshwork of the stent that is bent over the proximal LPA ridge to create a smooth entry site into the LPA

wall (Fig. 49.3). In addition to visual and endoscopic inspection, in selected cases it may be beneficial to perform an exit angiography at the end of the procedure to evaluate the results of stent placement angiographically.

49.3 Hybrid Stent Delivery Using Angiographic Guidance

Hybrid stent therapy using angiographic guidance is less commonly performed than direct visualization with endoscopic guidance. This technique is usually reserved for patients where a percutaneous transcatheter approach has not been successful in treating a specific vascular lesion, or patients where a standard

vascular access route is either not available, or where its usage may potentially lead to hemodynamic instability, or in patients/scenarios where there is very little opportunity for direct visualization, such as residual arch obstructions identified during exit angiography after a comprehensive arch repair (Fig. 49.4), or a stenosis of the branch pulmonary arteries in the presence of a large pseudoaneurysm (Fig. 49.5). In addition, angiographic guidance is advantageous in patients where there is very little preexisting imaging data, or in smaller vascular structures where the

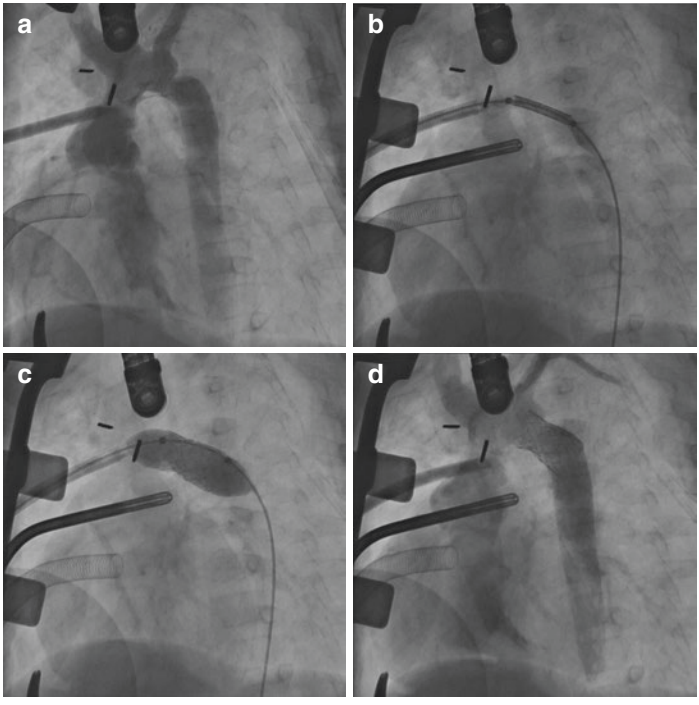

Fig. 49.4 Six-month-old infant with hypoplastic left heart syndrome undergoing a bidirectional Glenn procedure. There was a preexisting concern about an arch obstruction. Intraoperative angiography (**a**) documented a narrowing distal to the left subclavian artery. An intraoperative stent was placed through a sheath that was advanced over a wire through a purse-string in the ascending aorta (**b**: stent positioning, image **c**: stent expansion, image **d**: final angiogram)

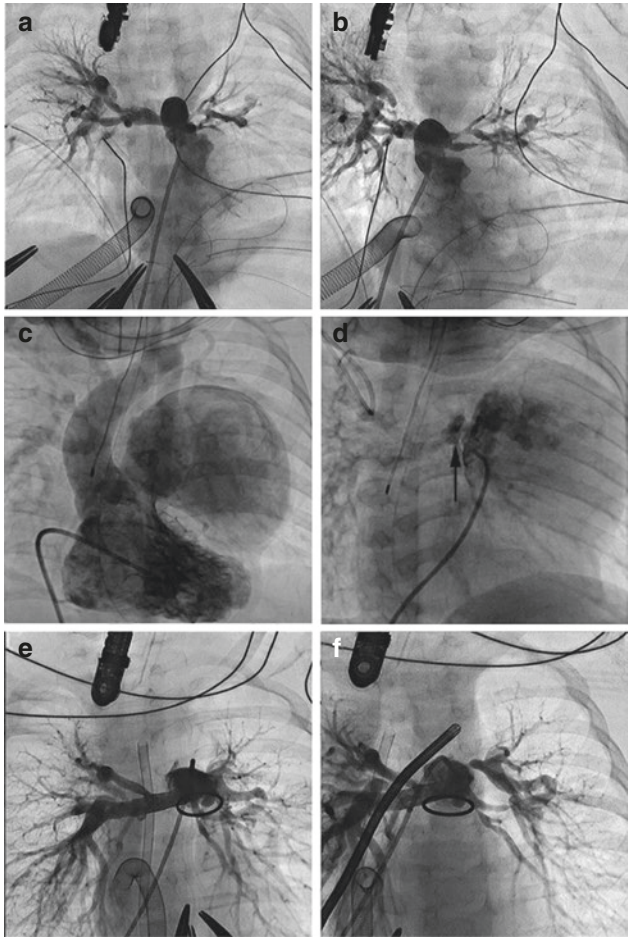

Fig. 49.5 Six-month-old with PAVSD and multifocal pulmonary blood supply with a history of unifocalization and placement of an RVPA conduit. A completion angiogram demonstrated multiple stenoses of both right and left pulmonary arteries (**a**, **b**). The patient underwent cardiac catheterization 3 months postoperative, which documented a large RVOT aneurysm (**c**) with a stenosis at the distal end of the conduit and multiple stenoses of both branch pulmonary arteries (**d**, arrow). LPA branches were extremely hypoplastic and compressed by the aneurysm. He therefore underwent surgical resection of the aneurysm and conduit replacement, combined with intraoperative "hybrid" delivery of a stent to the RPA and balloon angioplasty of two LPA branches (**e**, **f**)

use of endoscopy would not be feasible in evaluating the distal vessel. Furthermore, this approach is beneficial in critically ill postoperative patients where a longer percutaneous procedure may not be well tolerated, especially if stents are desired that can be expanded to adult size, which would require larger delivery sheaths and stiffer wires. A direct approach in these patients avoids the use of stiff wires and long sheaths and is hemodynamically often a lot better tolerated. In addition, if there are concerns of creating vascular injury in a freshly dissected vessel in a postoperative patient, performing this procedure with an open chest and cardiopulmonary bypass on standby provides an additional safety net. It is important though to emphasize that stenting even of freshly operated vascular lesions can be safely performed in the majority of patients [16].

Technically the procedure is fairly straightforward and requires the use of a portable c-arm, a hybrid operating room, or hybrid catheterization laboratory. The vascular access point is chosen ideally to provide a "straight shot" and approach towards the lesion that requires treatment (Fig. 49.5). It is important though to choose the entry site with some distance to the lesion, as to allow expansion of the balloon and its shoulders. Once an entry point has been identified, a purse string is placed and an adequately sized short sheath inserted just 2 mm into the vessel (the tip of the sheath can be marked with a silk suture). As this technique is often utilized in smaller patient, most angiographic acquisitions are obtained as small hand injections, and a power injector will rarely be necessary. The angiographies should provide all the measurements necessary to choose the correct balloon and stent size. Once the vascular lesion has been visualized, an appropriate wire is utilized to cross the lesion and positioned in the more distal vasculature. If necessary, a catheter can be advanced over the wire and the wire exchanged as needed. Stiff wires though are usually not required if an appropriate entry site has been chosen. An exception are larger patients that undergo hybrid pulmonary valve implantation, in whom a stiffer wire still offers considerable benefits. Given that the distance between the sheath and the lesion is not too long, it is usually feasible to advance the stent and balloon directly over the

wire without necessarily having the sheath positioned far distally. An exception are fresh suture lines where it is important not to create any injury by advancing the uncovered stent/balloon combination with its somewhat sharper edges across, and therefore ideally the sheath should be advanced sufficiently to avoid this problem. On occasions, partially inflating the balloon without expanding the stent allows creating a more smooth distal end of the assembly, which may allow advancing the stent without needing to position a longer sheath across the lesion. In most circumstances though, the stent/balloon will still be positioned partly within the short sheath when crossing the stenotic lesion, allowing some degree of pushability without worrying of the stent migrating off the balloon. Once the stent is expanded under fluoroscopic guidance, an angiography is performed through the short sheath to evaluate the result of stent placement.

49.4 Hybrid Stent Delivery Using Combined Approaches

Combined approaches generally have very similar indications as hybrid therapy using solely angiographic guidance. They are usually required when there is a need for angiographic guidance to accurately place the stent, specifically in small patients with small vessel diameters, especially when preexisting imaging data is somewhat limited. When additional cardiac lesions require surgical correction, then stents can be placed initially using angiographic guidance, and then further adjusted once the vessel is opened surgically with the patient on cardiopulmonary bypass.

A very good example would be a patient who had undergone surgical correction of pulmonary atresia with VSD but who developed a large false aneurysm as well as branch pulmonary artery stenosis (Fig. 49.6). Especially if hemodynamically compromised, the false aneurysm may make transcatheter stent therapy very difficult and often poorly tolerated. In that scenario, a direct per-MPA/conduit approach can be used to treat the branch pulmonary artery stenosis using a hybrid approach with angiographic

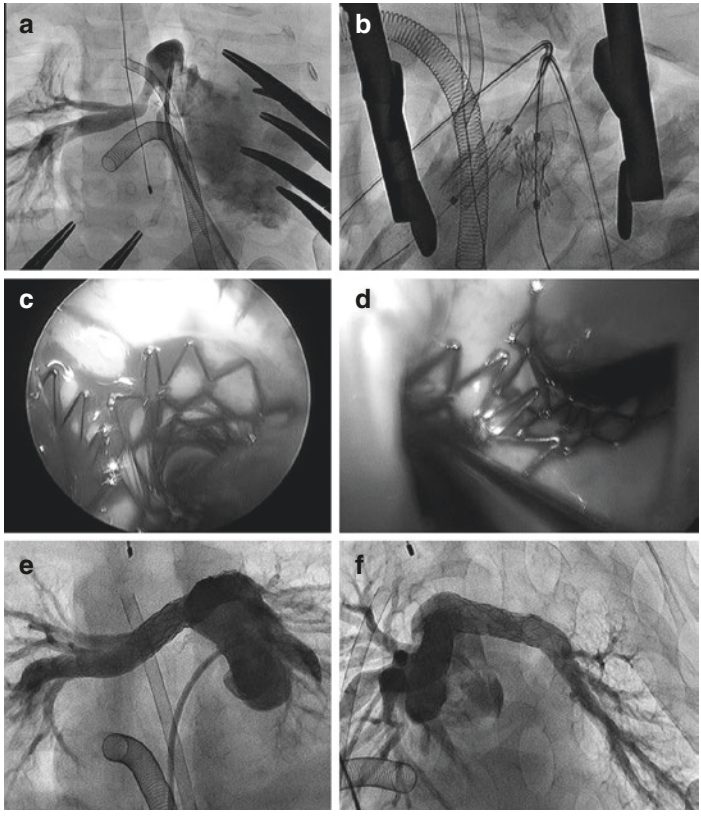

Fig. 49.6 Patient with pulmonary atresia and VSD after surgical correction who developed a large false aneurysm as well as bilateral branch pulmonary artery stenosis. (**a**) Bilateral branch PA stenosis delineated with intraoperative angiography. (**b**) Intraoperative stent expansion of RPA and LPA stent simultaneously. (**c**, **d**): Stent meshwork visualized after resection of the false aneurysm before (**c**) and after (**d**) manual folding of the mesh. (**e**, **f**) Exit angiography documenting excellent relief of the RPA and LPA stenosis

guidance in the operating room, before going on cardiopulmonary bypass and replacing the conduit as well as folding any protruding stent material to facilitate subsequent intervention. As with all hybrid techniques, the approach will need to be modified for each individual patient.

49.5 Other Hybrid Stent Deliveries

In general, intraoperative stents can be placed virtually anywhere within the heart or vascular structures, but intracardiac stent placement can be a little more difficult to visualize compared to vascular stent placement. Very rarely will this technique be necessary though. Figure 49.7 provides an example of a late diagnosed six-month-old infant with DORV, hypoplastic left ventricle, and a

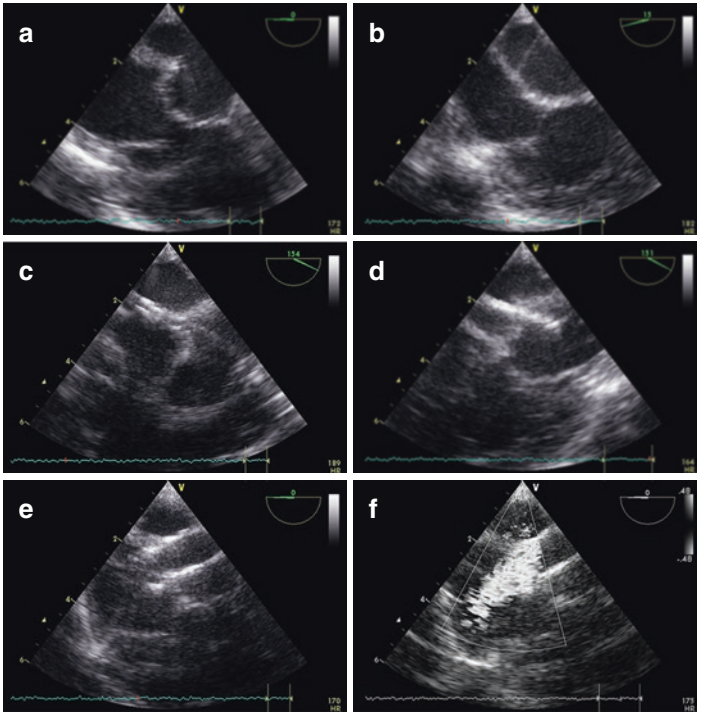

Fig. 49.7 Six-month-old infant with DORV, hypoplastic LV, mitral stenosis, and an intact atrial septum. Intraoperative stent placement across the atrial septum is performed at the time of pulmonary artery banding using guidance via transesophageal echocardiography. (**a**) Needle tenting the atrial septum. (**b**) Wire crossing the atrial septum. (**c**) Sheath across the atrial septum. (**d**) Stent positioning across the septum. (**e**) Stent fully expanded across the septum. (**f**) Laminar color-flow across the atrial septal stent

stenotic mitral valve as well as an intact atrial septum. This patient required surgical banding of the main pulmonary artery as well as creation of an ASD to reduce the left atrial hypertension. Under TEE guidance, an atrial puncture entry site was determined directly perpendicular to the atrial septum, and once a purse string had been placed, a needle was advanced through the purse string directly across the atrial septum. This allowed advancing a wire into the left atrium and then followed by advancing a short sheath into the LA. Transesophageal echocardiography is usually able to visualize the wire and sheath and once the stent is advanced it becomes visible within the sheath as a structure with increased echo brightness. It is important to position the stent as central as possible. Once the stent has been expanded balloon and wire will need to be removed carefully to avoid dislocating the atrial septal stent in the process.

With the limited (approved) availability of transcatheter pulmonary valves for the dilated native right ventricular outflow tract, a variety of hybrid approaches have been explored that facilitate this technique using presently approved devices. What these approaches have in common is either reducing the size of the MPA in which the valve will be implanted or creating a specific landing zone that allows the valve to be securely anchored inside a larger space [7–11]. On occasions, hybrid pulmonary valve implantation may be required after a failed percutaneous approach (Fig. 49.8).

49.6 Exit Angiography

Any time an intraoperative vascular stent has been placed, there is a benefit to consider evaluating the result using an exit angiography at the end of the procedure. This is in particular important, when there is question about the intraoperative result that may impact further management. Exit angiography is not only able to better delineate the vascular structures, but it also has an important yield of identifying vascular pathology that would have otherwise been left undetected using standard diagnostic approaches such as

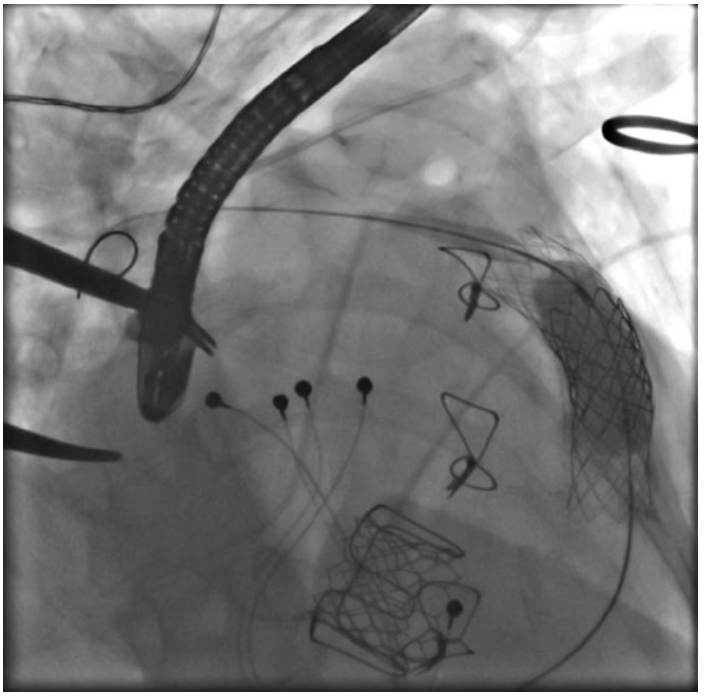

Fig. 49.8 18-year-old female with PAVSD who underwent transcatheter pre-stenting of an RVPA conduit as well as transcatheter tricuspid valve-in-valve implantation of a Sapien S3 valve. Transcatheter pulmonary valve implantation was unsuccessful as the angle of the tricuspid valve and the pre-stented conduit did not allow advancing the valve into appropriate position. The valve was then implanted 4 weeks later using a minimal subxiphoid incision and a periventricular approach

transesophageal echocardiography. Holzer and colleagues published a single-institutional experience using exit angiography and were able to show that exit angiography was able to identify lesions that required either surgical revision or hybrid therapy in 10% of cases [17] (Fig. 49.9). Exit angiography is fairly straightforward and can be performed just with a C-arm, an angiographic catheter, and a power injector.

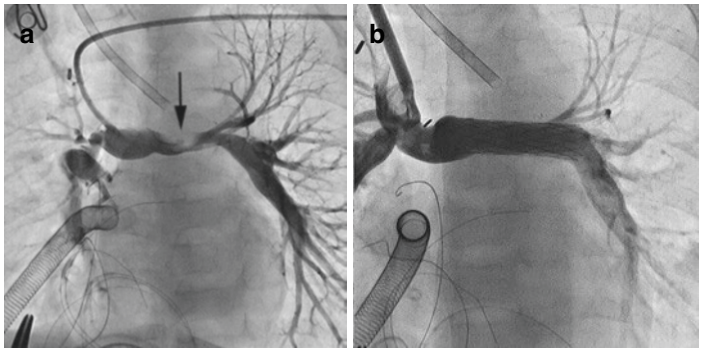

Fig. 49.9 Exit angiography after comprehensive stage II palliation in a 5-month-old infant with hypoplastic left heart syndrome documenting a stenosis of the left pulmonary artery. (**a**) on the left, arrow points to the LPA fold. (**b**) on the right depicts an angiography after intraoperative placement of a stent to the left pulmonary artery

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Part IX

Step-By-Step Procedures: Miscellanea

Retrieval Techniques

50

Rui Anjos, Inês C. Mendes,
and Duarte Martins

50.1 Introduction and Clinical Scenarios

Removal of foreign bodies from vessels or cardiac structures has become more frequent over the last decades, as the result of widespread use of indwelling catheters, leads, guidewires, and devices.

Foreign bodies requiring intravascular or intracardiac retrieval are usually the consequence of iatrogenic events. Frequently, a lost device is an immediate complication of an interventional procedure, diagnosed and retrieved during the same procedure, but in a significant number of cases, lost objects are an incidental finding on imaging studies. In fact, most patients are asymptomatic [1, 2]. Successful endovascular retrieval has been achieved in over 90% of cases in the literature [1, 2]. A small number of patients will require a combined open and endovascular approach. Unsuccessful retrievals requiring surgery are more frequent with

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large devices, usually implantable valves, atrial septal or patent arterial duct devices and in many of these cases a sternotomy, frequently with cardiopulmonary bypass, will be required.

50.2 Embolized Materials Requiring Retrieval

The most frequently embolized material requiring retrieval in clinical practice is by far a fragment of catheter [1], usually as the result of a fractured central venous catheter, frequently a long-term indwelling catheter. Fracture and embolization of central venous catheters, inserted in large veins or peripherally, occur frequently at the time of catheter removal and can be recognized immediately, or long after the event, as an incidental finding. Many other embolized devices and materials have been reported in the literature, including therapeutic devices (coils, atrial septal defect or patent arterial duct occluders, stents, valves, and venous filters), guidewires, and pacing wires.

In most cases, embolization occurs in the systemic venous system, as in the case of fractured long-term venous catheters. Embolized, fractured, or mispositioned devices and balloons generally migrate into the systemic veins, right heart, or pulmonary arteries. The systemic arteries are involved less frequently, when malpositioned devices migrate into the left heart and arteries, as with stents or implantable valves. This is also the case when coils or devices used to embolize anomalous arteries migrate into other arteries or when stents fall off balloons or are undersized for the target artery, migrating distally.

50.3 Indications and Patient Selection: Should It Be Retrieved?

As a rule, embolized catheters and devices should be retrieved. Earlier studies reported a high incidence of complications and even mortality after foreign body intravascular or intracardiac embolization, probably reflecting the fact that in early publications, diagnosis and retrieval were made predominantly in symp-

tomatic patients [1]. There are reports of severe complications occurring with embolized fragments of catheters, devices, stents, pacing wires, and venous filters. These include arrhythmias, cardiac and vessel perforation, myocardial infarction, vascular occlusion with ischemia or congestion and secondary infection, and sepsis. Even so, leaving certain embolized foreign bodies in place may be acceptable in selected patients. This is the case with very small fragments positioned in difficult access locations, stents or valves left in vessels with low hemodynamic consequences or in patients with a very low life expectancy.

50.4 X-Ray Equipment

Removal of a foreign body in the cardiac catheterization laboratory requires adequate equipment. The use of biplane X-ray equipment is very useful to locate a foreign body in a three-dimensional spatial orientation and facilitate precise and purposeful catheter maneuvering to and around it. Nevertheless, foreign body retrieval can be achieved with single-plane imaging, bearing in mind that using a single plane may result in catheter movement that appears to be directed to the foreign body in one plane when actually it is moving away from the target.

50.5 Planning the Procedure

Careful planning of the procedure is of paramount importance. Adequate planning is one of the most essential parts of the retrieval technique. It is important to know the exact position and dimensions of the embolized catheter or device so that an appropriate selection of retrieval sheaths and devices is made right from the beginning of the procedure. The major initial decisions concern the vascular access, guiding catheter or sheath (size, length, type, and eventual modifications), and selection of the retrieval device.

A good rule for selection of the percutaneous access is to use the largest vessel available, even when this is not the original

access vessel for the interventional procedure. Usually, the best venous approach is via the femoral vein, but in some cases, the jugular or subclavian veins are better alternatives, depending on the location of the foreign body. If arterial access is required, the femoral artery is usually the best approach, but alternative routes, especially in the patient with congenital heart disease, should be considered (such as a venous approach and access to the descending aorta via a patent arterial duct in case of a coil embolized into the aortic bifurcation). In some cases, the contralateral femoral vessel is the best approach for patients with a device embolized and impacted into an iliac artery or vein.

A second venous or arterial access point can be useful in some occasions, allowing for repeat small volume contrast injections through a catheter positioned near the target vessel, directing the manipulation of the retrieval system [3]. Alternatively, contrast injections can be performed through the side arm of a sheath positioned in a proximal artery. A second access sheath can also be valuable for advancing a catheter to hold and stabilize a device, facilitating snaring.

Most embolized or lost catheters and devices should be retrieved into a long sheath placed as close to the device as possible and then safely removed, minimizing the risk of injury to nearby structures, such as veins, cardiac valves, or atrial chambers. Recovering the device into a long sheath also minimizes the risk of losing it during extraction. Catheters and other foreign bodies, particularly larger and bulkier devices, should not be captured and pulled across cardiac valves or small vessels. All sheaths used for retrieval procedures should have distal radiopaque bands to identify the precise location of the tip [3].

One of the most important decisions is selection of the length and size of the sheath, which should usually be at least two French sizes larger than the original delivery sheath or the embolized catheter. Very frequently, a lost device is captured but not retrieved because the sheath is not large enough to accommodate the device. Soft sheaths should be avoided, as the pressure applied when pulling out a foreign body is frequently higher than the resistance of the sheath to compression, causing a longitudinal or concertina deformation. This leads to loss of torque control and maneuver-

ability on the sheath. The standard long sheaths (e.g., from Cook or Cordis) are usually adequate for most retrieval procedures. For the recovery of large devices, when the use of a significant force is anticipated, a good option is to use stiffer sheaths, like the Arrow Flex® (Arrow) or Flexor® (Cook), which have good resistance to compression, but maintain adequate flexibility. For very large devices and cardiac leads dedicated sheaths can be used as the inner (12 Fr) and outer (16 Fr) Femoral Introducer Sheath Set® (Cook). Large sheaths should be maneuvered very carefully, especially after the introducer has been removed, as they have a high potential for damaging vascular and cardiac structures.

When the embolized catheter or device is not very large (e.g., a fragment of catheter), a guiding catheter with an internal diameter ranging from 0.058″ (5 Fr) to 0.090″ (8 Fr) can be used as an alternative to a long sheath. It is introduced through a short sheath, easily handled, and available in a wide selection of curves (straight, right and left coronary, 3D, etc). A guiding catheter provides enough support for removal of most coils, guidewires, and small catheter fragments.

50.6 Available Retrieving Systems and Devices

50.6.1 Snares

Snares are the most frequently used devices for retrieval procedures, employed in over 80–90% of the cases [1–3]. Therefore, catheterization laboratories dealing with device implantation, therapeutic embolization, and retrieval procedures should have available a selection of retrieval snares.

50.6.1.1 Single-Loop Snares

Homemade snares are rarely used nowadays but can be useful when standard snares are not available. They can be made very easily inserting a 0.014″ coronary or a 0.018–0.025″ teflonated exchange length guidewire into a large inner diameter catheter (5 Fr guiding catheter or 6 Fr multipurpose). The guidewire exits from the distal tip and is reinserted until it exits again from the

proximal end of the catheter. The loop formed at the distal end of the catheter can be angulated. Another alternative is to use a balloon catheter with a cutoff tip, attaching one of the extremities of a guidewire to the balloon lumen and inserting the other extremity through the catheter lumen until it exits the proximal end of the catheter. The snare diameter can be adjusted by advancing or pulling the guidewire.

Commercially available snares are offered in several diameters and support the traction force required in most extractions. They have good torque control and are flexible, with a kink-resistant loop with an excellent X-ray visibility. They are therefore better options than homemade snares. The snare is usually supplied with a catheter or coaxial sheath system with a radiopaque band at the tip and a torquing mechanism that facilitates rotation of the device and tightening of the loop after capture.

The simplest single-loop snare is the straight Curry Intravascular Snare® (Cook). Most of the available simple-loop snares are angulated, including the Angled Wire Loop Retriever® (Cook), Lassos® (Osypka), Sympro Elite® (Teleflex), Argon Medical Single-Loop® snare, and Bard Snare Retrieval Kit® (BD Interventional). The Amplatz GooseNeck® (Medtronic) with loop diameters ranging from 5 to 35 mm, inserted through 4–6 Fr catheters and the ONE Snare® (Merit Medical), with similar diameters and catheters, have a small fold incorporated in the loop, theoretically providing a better grasping of the foreign body. The Andrasnare® (Andramed) has a pre-angled tip and a shapeable introducer. Microsnares are small single-loop snares, available from Medtronic, PFM, Merit Medical and Andramed, with 2–7 mm loop diameters, inserted through 2–3 Fr catheters, intended for very small vessels, usually in the neuroradiology setting.

The Expro Elite Snare® (Teleflex) is a snare with a smooth helical loop ranging from 5 to 35 mm diameter, with the potential advantage of a smaller distal diameter with a longer reach than right-angle loops. It has a 0.035″ profile, allowing insertion through a conventional diagnostic catheter, thus eliminating the need for exchanges when recovering very small devices as

microcoils. The Sympro Elite is a similar snare with a 90° loop, and 5–35 mm diameters. The Micro Elite Snare® (Teleflex) is a smaller version of this catheter with a 0.014″ profile and 2–7 mm diameters.

Some snares are designed for a specific type of procedure as the Gunther Filter Retrieval Set® (Cook), for IVC filter retrieval.

50.6.1.2 Multiple-Loop Snares

Multiple-loop snares have several loops opening simultaneously. The two-loop Multi-Snare® (PFM) has a main loop and an additional lateral loop, forming an orthogonal dual-plane system, with variable loop sizes. Snare diameters range from 5 to 40 mm, introduced through 4–6 Fr catheters. The Multi-Snare Micro Sets®, also from PFM, with 2–6 mm diameters, are inserted through 3 Fr catheters.

Other multiple-loop snares have a slightly more complex configuration, with 3–4 loops which come off simultaneously from the catheter as petals. They are designed with interlaced loops to increase the probability of capture and manipulation of foreign objects, covering a higher vessel area. These devices are particularly useful for retrieval of inferior vena cava filters, occlusion and embolization devices, but are also a good option for other foreign bodies. They have a good resistance to kinking and to the pressure required to manipulate and retrieve the larger and bulkier devices.

Three-looped snares include the EN Snare® (Merit Medical), the Vascular Snare® (Argon Medical), and the Atrieve Vascular Snare® (Argon). They are available in 2–8 mm diameter (mini version) requiring a 3.2 Fr catheter, or 6–45 mm diameter (standard version), introduced through 6–7 Fr catheters.

Four-loop snares include the Indy OTW Vascular Retriever® (Cook), with overlapping loops that open at right angles to the catheter. They have a 40 mm diameter and requires an 8 Fr sheath. The CloverSnare four-Loop Vascular Retriever® (Cook) is introduced through a 6 Fr catheter, advanced through an inner (8 Fr) and outer (10 Fr) coaxial introducer sheaths, all locking together in order to provide a good transition and resistance.

50.6.1.3 Snares for Retrieval of Closed-End Catheters and Wires

A special type of snare was designed to retrieve catheters or guidewires without a free end. The Needle's Eye Snare® (Cook) was specifically built for pacing wire retrieval, with an excellent resistance to traction. It has an open-curved snare (13 or 20 mm diameter) that is positioned around the foreign body and a second smaller looped wire that closes the circuit when it is advanced on the opposite side of the catheter or wire to be retrieved. It is delivered by a flexible 12 Fr sheath inserted coaxially within a larger 16 Fr outer sheath, the Femoral Introducer Sheath Set® (Cook).

The Loopmaster-Sochman Snare® (Andramed) has a 25 mm open-loop curved snare that is positioned around the catheter or wire to be retrieved, the loop being closed by advancing a straight guidewire. Its smaller profile, with an 8 Fr introducer sheath, compares favorably with the previous device particularly for pediatric patients.

50.7 Graspers and Forceps

The Vascular Retrieval Forceps® (Cook), used especially for intravascular retrieval of coils but also for catheters, guidewires, or other foreign objects, requires a 4–5 Fr sheath or a 5–6 Fr guiding catheter. The Alligator Retrieval Device® (Medtronic) is designed for coil retrieval from small vessels, typically from the cerebral circulation and requires a 3 Fr microcatheter. Catcher® (OSYPKA) is a forceps catheter expandable up to 12 mm in width. Although these graspers are appealing, in reality, their use is generally limited to small vessels and their preferential use is for coil or catheter retrieval. Biopsy forceps are used to grasp foreign bodies, particularly coils and catheters, but there is some potential for vascular or cardiac injury. They are also used to stabilize a free-floating device in order to facilitate capture by another device. Note that the potential for capturing or grasping a device with a biopsy forceps is low when using the regular oval, spoon, or cup jaws, with regular edges, and much more effective with alligator or teeth jaws.

50.8 Baskets

Helical baskets are the only devices capable of engaging and retrieving a spherical or ovoid object (like a bullet) and can be very useful for extraction of plugs, PDA occlusion devices, catheters, and coils. The Dotter Intravascular Retrieval® (Cook) is a four-wire helical-loop basket, with 7 cm length and 3 cm diameter. The catheter shaft is 8 Fr, requiring a sheath, the size of which depends on the size of the device to retrieve. The Andra basket® (Andramed) has a lower profile and is available in 25–30 mm diameters, mounted on 5–7 Fr catheters.

The basket is placed along or advanced distally to the foreign body. When the retrieving system is rotated or removed, the foreign body is drawn into the basket and then pulled into the sheath. The major disadvantage of the basket systems is that they are bulky and rigid, with potential injury to vascular or cardiac structures.

50.9 Step-by-Step Approach

50.9.1 Retrieval of a Fragment of Catheter (or a Coil) Embolized

A catheter fragment embolized into the systemic veins, the right heart, or the pulmonary arteries is the most frequent situation requiring foreign body retrieval (Fig. 50.1).

The approach should follow these steps [1, 3, 4]:

- Obtain a thorough history and ascertain the type of catheter (length, diameter, structure) and likely time following the embolization. Determine the exact position of the catheter by echocardiography and biplane chest X-ray. If inconclusive, consider other imaging techniques.
- Select the retrieval device. The best choice for a catheter fragment is usually a single- or double-loop snare. In most cases, a 10–25 mm loop diameter snare is selected, depending on the

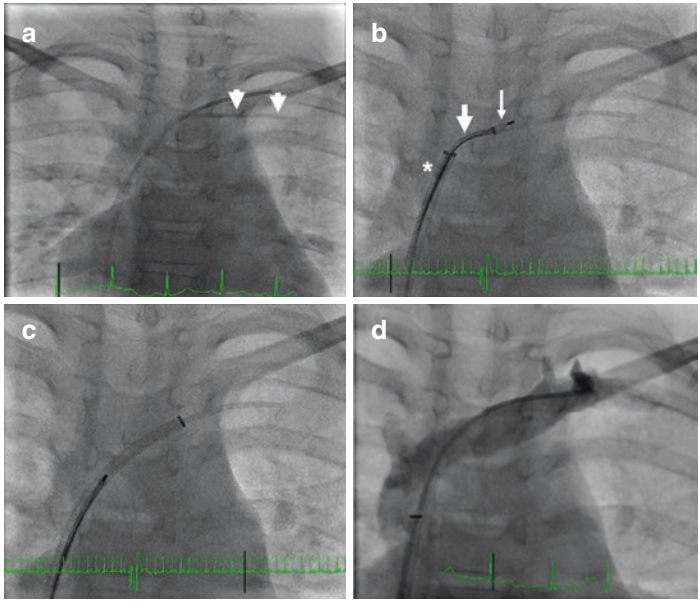

Fig. 50.1 (**a**) A fragment of a long-term indwelling catheter is identified in the innominate vein. (**b**) A snare was used to capture the proximal part of the catheter (small arrow) supported by the snare catheter (large arrow). A large sheath (*) provides additional support, allowing for full recovery of the device into the sheath prior to extraction (**c**), avoiding inadvertent embolization to other location or damage to the heart or vessels. A final angiography documents integrity of the vein (**d**)

size of the vessel (a larger snare is more appropriate for a large vessel). Other initial options for retrieving a catheter fragment include a multiple-loop snare or a basket.

- Select the guiding catheter or sheath to use. For flexible indwelling catheter fragments up to 5 Fr, a 6 or 7 Fr guiding catheter will accommodate the fragment; for 7 Fr fragments, select a 9 Fr guiding catheter or an 8–9 Fr valved sheath with a tip marker, long enough to reach the catheter fragment.
- Obtain informed consent.
- Access the femoral vein percutaneously with a short introducer. If you are using a guiding catheter, choose an introducer

with the same size. If you are planning to use a sheath, use a 6 Fr short introducer, which will be replaced later by a long sheath. Obtain invasive arterial pressure monitoring whenever you predict the procedure is long, complex, or with potential for vascular or cardiac injury.

- Heparinize (100 IU/kg iv).
- Advance a diagnostic catheter to the desired location under fluoroscopic control. Obtain biplane angiography of the vessel or chamber into which the fragment is lodged. In some cases, angiography may not be required.
- Advance an exchange wire beyond the fragment position and exchange the diagnostic catheter for the selected guiding catheter (or sheath).
- Attach a hemostasis valve with a sideport (Tuohy-Borst or similar) to the guiding catheter, purge and flush the system.
- Pull the snare loop inside the snare catheter and introduce it through the hemostatic valve.
- Advance the snare catheter until it exits the guiding catheter or sheath and position it proximal to the indwelling catheter.
- Then advance the snare until the loop is around the proximal end of the foreign body. The snare loop can be rotated by turning the torquing device which is firmly attached to the central core.
- By advancing the snare catheter, the loop is closed, grasping the foreign body.
- Tension between the central core and the snare catheter must be maintained, by advancing and tightening the torque device close to the snare catheter.
- To retrieve the indwelling catheter, maintain tension and pull the central core and snare catheter while advancing gently the guiding catheter or sheath.
- After retrieval, hospital standard of care should be followed for removing the sheath and providing hemostasis to prevent bleeding at vascular access site.

Tips and Tricks: It is always preferable to withdraw the captured catheter or coil into a guiding catheter or sheath, immediately after device capture, avoiding trauma to the heart or vessels.

50.9.2 Retrieval of an Embolized Occluder Device

Follow the general steps indicated in the previous section.

Tips and tricks and particular details for occluder devices [3, 4]:

- Determine the exact position of the embolized device by fluoroscopy and echocardiography.
- For large devices, a good option is to use a *multiple coaxial system*, formed by a snare, a guiding catheter, and a large valved sheath (Fig. 50.2). The large sheath is positioned near the embolized device. A guiding catheter with a curve best suited to the position of the device is then advanced through the sheath. Maneuver the guiding catheter so that it faces the device hub or attachment pin. The snare is advanced through the guiding catheter. With the multiple coaxial system, the

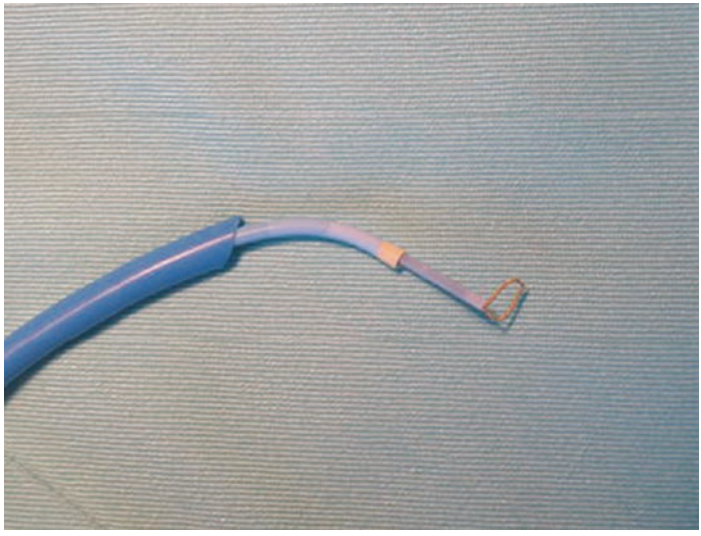

Fig. 50.2 A multiple coaxial system, formed by a snare, a guiding catheter, and a large valved sheath. The maneuverability and orientation of a guiding catheter to direct the snare are coupled with the resistance and dimension of the sheath. A coaxial system provides extra support to retrieve large devices

maneuverability and orientation of a guiding catheter to direct the snare to an optimal position are associated with the support of a large sheath.

- The most commonly selected retrieval device for an occluder is a snare catheter. Multiple-loop snares are excellent options, increasing the probability of capture (Fig. 50.3). The target is the device hub or attachment pin. Capturing the central part (stent) of an atrial occluder device with a snare will make it impossible to retrieve it into a sheath.

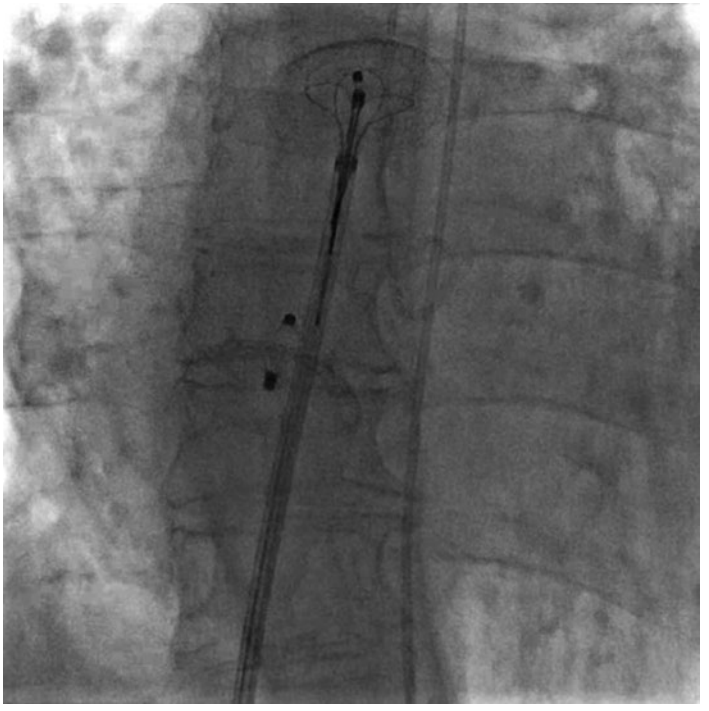

Fig. 50.3 A multiple-loop snare retrieving a large Amplatzer device embolized into the left atrium. The multiple loops increase the probability of capturing and when used on a frontal approach, as shown in this figure, has a self-centering mechanism

- The sheath selected for retrieval of an occluder device should be at least 2 Fr larger than the size of the original delivery sheath. When retrieving large atrial septal defect occluders, it can be useful to bevel the tip of the sheath at 30–40°, before introducing and advancing the system. The beveling technique is applicable to non-armored sheaths such as the Cook Mullins Check-Flo. The main purpose of beveling is to provide a wider profile of the sheath to retrieve the snared hub of a large device. The best alignment for pulling the snared device into the beveled sheath is obtained by rotating the sheath or the snared device, so that the free end of the snared hub is aligned with the most proximal part of the beveled tip (Fig. 50.4). A beveled tip is sharp and so can potentially cause damage to the vascular and cardiac structures, therefore care must be taken when advancing the sheath without the protection of a dilator or a coaxial catheter.

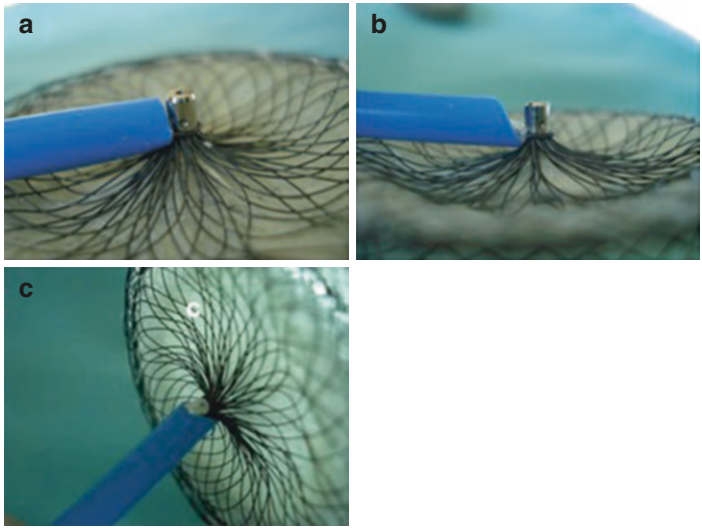

Fig. 50.4 Capture of the hub of a large Amplatzer device. Retrieval into the sheath is not possible as the hub is long and perpendicular to the sheath (**a**). Beveling the tip of the sheath (**b**) allows for hub retrieval into the sheath (**c**), as this increases the sheath extremity profile

- In some cases, it is useful to position a super stiff guidewire adjacent or beyond the embolized device, stabilizing the sheath during capture and reducing the risk of trauma. The guidewire should be removed after capture, before retrieval of the device into the sheath.
- Maintain tension and pull the snare and guiding catheter while maintaining the position or advancing gently the sheath. Retrieve the entire device into the sheath before removing it from its original position.
- In case of difficulty snaring the device hub, a technique using a coronary guidewire advanced through the outer border of the device and snaring of the distal tip of the guidewire has been described [5]. The device is captured into a large sheath, pulling it through the external rim of one disk, requiring complete deformation of that disk. This technique is probably reserved to small devices, as capture of a large deformed device may be difficult to achieve even with large sheaths.

50.9.3 Retrieval of an Embolized Stent

Tips and tricks and particular details for stents [3]:

- If a stent embolizes (due to an undersized stent, dislodged stent from the balloon, or a large rupture of the balloon), the most important rule is to maintain the guidewire position through the stent in a very distal and secured position.
- If the stent is displaced and cannot be repositioned with the balloon in the target area, expand it and fix it in a more distal location in the vessel.
- Withdrawal of a stent over a balloon out of the pulmonary artery through the right ventricular outflow tract is not safe and should never be considered when the struts are proud from the balloon.
- If a stent is partially expanded and positioned over a balloon, it can be recompressed over the balloon with a snare. This is particularly useful for stents in venous position. The snare can be

positioned around the external part of the wire and balloon catheter and advanced along it, through the delivery sheath until it reaches the stent/balloon assembly. It is then tightened slightly around the balloon/stent, loosened, and moved a number of millimeters up the stent. The tightening is repeated along the entire length of the stent. If it is possible to compress the stent to its original diameter, it can be withdrawn into the sheath and removed. If not, the balloon/stent assembly can be pulled to a peripheral location, and if it is not possible to recover it through the puncture site, it can be extracted through a small cut down.

- The snare can also be inserted through a very large sheath via a separate access sheath, capturing the tip of the wire and then maneuvered until it reaches the balloon/stent assembly. The compressed stent can be recovered via this alternative access route, by pulling the snare and pushing the balloon catheter into the sheath.

50.9.4 Retrieval of a Fractured Balloon

Tips and tricks and particular details for a circumferential balloon fracture:

- A circumferentially ruptured balloon may be difficult to handle as the distal part of the balloon will open as an inverted umbrella, preventing extraction at the insertion. This will be even more difficult if the shaft of the catheter breaks.
- Always maintain guidewire position in the balloon.
- Remove the proximal portion of the balloon and catheter.
- Insert a multipurpose catheter over the guidewire and advance it until it reaches the distal part of the balloon.
- Insert an introducer larger than required for the balloon in the contralateral femoral or the jugular vein.
- Through this access, introduce a snare catheter and capture the tip of the guidewire.

- Retrieve the guidewire and the distal part of the balloon by pulling the snared guidewire while pushing the multipurpose catheter and distal end of the balloon. As the pointed distal end of the balloon will be directed to the new introducer, it will be easily withdrawn via this approach.

50.10 Complications of Retrieval

The most important complications related to retrieval techniques are tears, perforation, or injury to vascular or cardiac structures. Other complications include arrhythmias, device embolization to another location, device entrapment, and thromboembolic complications, such as stroke, ischemia, pulmonary embolism, and myocardial infarction.

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Pericardiocentesis

51

Maarten Witsenburg

51.1 Introduction

The pericardial space normally contains several ml of serous fluid. Due to diseases, external or iatrogenic trauma, the fluid volume may increase, either acutely or chronically. The increase of volume and intrapericardial pressure may compress cardiac chambers and restrict filling, which may lead to a decrease in cardiac output and cardiac tamponade. Rapid accumulation of pericardial fluid may produce tamponade at much smaller volumes than when an accumulation occurs over a longer period of time.

51.2 Diagnosis of Cardiac Tamponade

Pericardial effusion may present as an incidental finding on routine echocardiography or may be suspected because of a large heart contour on X-ray or a low-voltage ECG. The other extreme of the spectrum is the patient with acute low output due to cardiac

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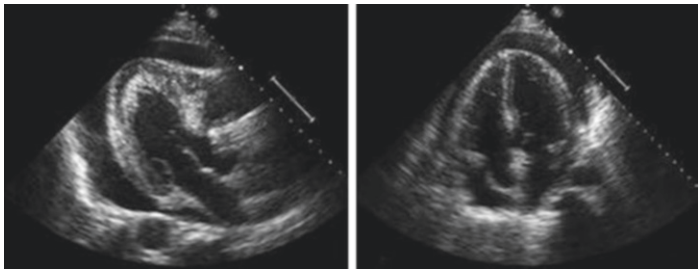

Fig. 51.1 Echocardiographic four-chamber and long-axis view showing moderate pericardial effusion

tamponade where pericardial drainage is potentially lifesaving procedure. Clinical signs include low blood pressure, tachycardia and muffled heart sounds. Echocardiography will reveal pericardial effusion (Fig. 51.1). Right atrial collapse in late diastole, increased tricuspid E-wave velocity during inspiration and decreased mitral E-wave velocity confirm the diagnosis of tamponade.

The urgency for drainage depends on the clinical picture, echo findings and patient history.

51.3 Indication for Pericardiocentesis

These are the indications for pericardiocentesis:

- Cardiac tamponade
- Impending cardiac tamponade
- Recurrent or persistent pericardial effusion
- Relief of symptoms due to pericardial effusion
- Need for diagnostic culture or fluid analysis

51.4 Complications

Pericardicentesis should be performed by an experienced operator. Echo guidance is essential and has proven to reduce the complication risk. Potential complications include visceral perforation, pneumothorax, haemothorax, coronary artery laceration and

cardiac perforation (the inferior vena cava, right atrium, right or even left ventricle). Arrhythmias may occur, as well as transient hypotension and low cardiac output.

51.5 Contraindications for Pericardiocentesis

There is no absolute contraindication for pericardiocentesis in acute cardiac tamponade, but a variety of conditions may increase the risk of the procedure. Do realize that surgical drainage may be a superior alternative in some instances. Special caution should be taken in case of a traumatic bleed, bleeding diatheses and suspected purulent effusion. A small or posteriorly located effusion will be difficult to reach, and in the presence of multiple septa, a simple puncture is likely to fail.

If in adults tamponade or haemopericardium is associated with aortic dissection, emergency surgery is the only reliable approach.

51.6 Preparation

In a non-urgent procedure, the patient and/or parents should be informed about the procedure and possible complications and give consent.

Depending on local practice, the pericardiocentesis is performed either in the ICU or in the cath lab, with echo standby. Patient's ECG, heart rate, blood pressure and oxygen saturation are monitored continuously. The echo machine should be running and pericardiocentesis package prepared.

In children, general anaesthesia by a dedicated anaesthesiologist is helpful, as long as one realizes that induced changes in body position as well as vascular resistance may compromise the haemodynamic condition. Close collaboration between anaesthesiologist and cardiologist is essential, and the puncture should be performed directly after induction of anaesthesia.

Echocardiography is used for the confirmation of the appropriate puncture site for pericardial drainage and helps to assess at what depth the effusion is to be expected. Subsequently, it shows the position of the drain and relief of fluid volume.

51.7 Access and Drainage

A pericardial puncture set is prepared (Fig. 51.2).

Positioning the patient in a 30° head up angle may help pooling the effusion at the inferior site of the heart. With the help of echocardiography, the location of the effusion is reconfirmed and the appropriate puncture site is marked. The patient is draped and cleansed with an aseptic solution. The skin and subcutaneous tissue are infiltrated with a local anaesthetic. The needle (appropriately long for patient size) is slowly advanced through the skin at an angle of 15–30° pointing at the left shoulder (Fig. 51.3).

Mild negative pressure with a 5–10 ml Luer-Lok syringe is applied. The patient monitor is continuously checked for arrhythmias. Passing the parietal pericardium into the pericardial space may be felt as a pop, and then it should be possible to gently

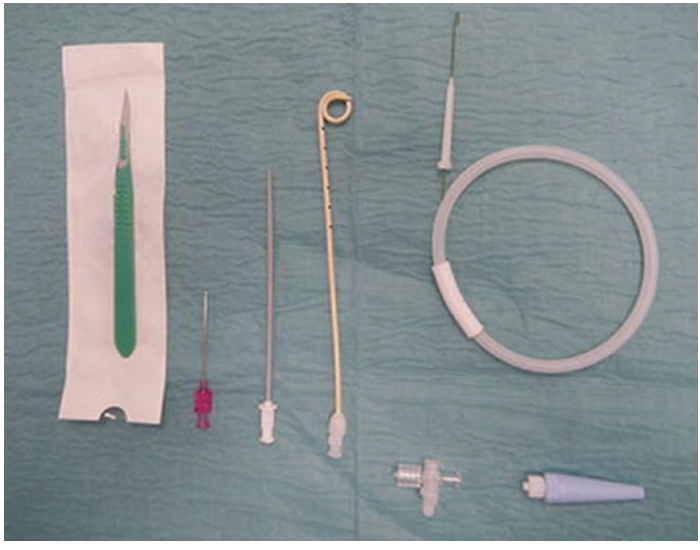

Fig. 51.2 Paediatric pericardial drainage set with scalpel (William Cook Europe, Bjaeverskov, Denmark)

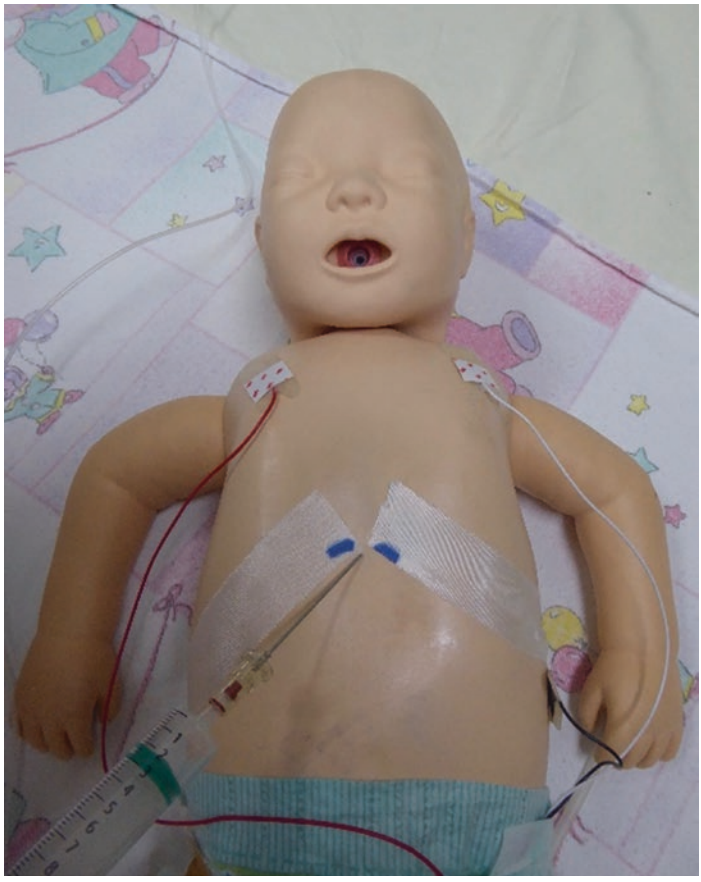

Fig. 51.3 Subcutaneous approach for pericardial drainage, with needle directed at the left shoulder at a 15–30° angle

aspirate fluid. When necessary, the access can be echocardiographically confirmed by injecting some agitated saline.

Alternatively, but nowadays less frequent used techniques include ECG monitoring from aspiration needle, pressure monitoring from aspiration needle, contrast injection and/or observation of wire curve once introduced during fluoroscopy.

Depending on the urgency and indication for pericardial drainage, some more fluid is aspirated and a J-wire is inserted. The wire advance should not be forced against resistance. The entry site is dilated with a 6–8 F dilator, and a (pigtail) catheter with multiple side holes is advanced for continuing drainage. Fluid is collected for laboratory analysis and culture. A three-way stopcock is connected with a 20–50 ml syringe and collection bag. The drain can be sutured if continued drainage is expected. In case of haemorrhagic fluid aspiration, a rapid comparison of the fluid and whole blood haematocrit may confirm the proper drainage site.

51.8 Monitoring After Drainage

Following the pericardiocentesis, vital signs of the patient are closely monitored. An X-ray will confirm drain position and rule out pneumothorax. Drain volume is noted. Echocardiography should be repeated before drain removal and in case of suspicion of inappropriate fluid drainage.

Depending on the cause of the effusion, anti-inflammatory agents and antibiotics may be started. The management of chronic pericardial effusion is beyond the scope of this chapter.

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Endomyocardial Biopsies

52

Anders Christensen, Davide Marini, and Audrey Marshall

52.1 Indications

Right ventricular (RV) endomyocardial biopsy (EMB) remains the gold standard for in vivo diagnosis of rejection in cardiac allograft patients [1]. It is performed as routine surveillance, in the setting of reduction of immunosuppression, and also in cases of suspected acute rejection. Beyond 2 years post heart transplantation, asymptomatic acute cellular rejection is extremely rare, thus long-term

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routine surveillance EMB might be avoided in these patients, especially in those transplanted as neonates or toddlers [2].

EMB may also have a supplemental role in the diagnosis of myocarditis, cardiac tumours, or infiltrative myocardial diseases (e.g. amyloidosis, sarcoidosis, Fabry disease and arrhythmogenic right ventricular dysplasia) [3]. Consensus opinion as to appropriate indications for EMB in evaluation of these other diagnoses is lacking, particularly in children. Relevant literature typically cites EMB in the discussion context of a specific cardiac disease, while the decision to pursue EMB is generally made on the basis of clinical presentation [4]. When performed for diagnosis of acute myocarditis, EMB may aid in identifying causative viral agents and identifying potential outbreaks. The use of EMB as a component of evaluation for infiltrative disease is uncommon, given the low incidence of disease in children and use of alternative diagnostic tests.

52.2 Clinical Scenarios

In 2007, the American Heart Association (AHA), the American College of Cardiology (ACC) and the European Society of Cardiology (ESC) published guidelines regarding the role of EMB in the management of cardiovascular diseases in adults, providing a list of clinical scenarios where EMB might be considered [4].

In scenarios 1 and 2 (sc. 1, 2), the aim of the EMB is to detect more aggressive forms of myocarditis, such as giant cell myocarditis and necrotizing eosinophilic myocarditis, in which immunosuppressive treatment may improve outcomes. In sc. 3, EMB aims to reveal sarcoidosis or idiopathic granulomatosis myocarditis and in sc. 4 hypersensitivity myocarditis; all of which may respond to treatment with corticosteroids. EMB is reasonable in cases of heart failure associated with suspected anthracycline cardiomyopathy (sc. 5) or restrictive cardiomyopathy (sc. 6), in which case it may reveal either a specific infiltrative disorder, for example amyloidosis or haemochromatosis, or myocardial fibrosis and myocyte hypertrophy consistent with idiopathic restrictive cardiomyopathy. In cases of suspected cardiac tumour (sc. 7),

with the exception of typical myxomas, EMB may be considered if the diagnosis cannot be established by non-invasive techniques, or the tissue diagnosis will influence the course of therapy. EMB is reasonable in the setting of unexplained cardiomyopathy in children (sc. 8). Rarely, drug-refractory arrhythmias may be the first manifestation of an underlying myocarditis (sc. 13) [5].

52.3 Preparation of the Exam and Vascular Access

In preparation for the procedure, laboratory exams are reviewed to ensure normal platelet counts and coagulation parameters. Females of reproductive age should be given a urine pregnancy test. Baseline conduction should be documented by EKG. Previous catheterization reports can provide valuable information about the number of former biopsies, vascular access and anaesthesia issues, and prior complications.

Preprocedural echocardiography can alert the operator to myocardial dysfunction, as well as documenting its severity and regional distribution. The echocardiogram will document baseline tricuspid regurgitation and will also identify and localize any existing pericardial effusion. It may be possible to increase the diagnostic sensitivity of EMB by targeting the biopsy to areas in which MRI has detected inflammatory disease or cellular necrosis [6].

EMB can often be performed using local anaesthetic at the access site, along with necessary levels of anxiolytic or sedation. The most common access sites are the internal jugular and femoral veins. The route of access impacts the anaesthetic management, as many patients (particularly infants and young toddlers) tolerate neck access poorly. These patients may require general anaesthesia, with an airway via laryngeal mask or endotracheal intubation. Femoral access (used most commonly in infants or toddlers) can be less stimulating and uncomfortable. Standard monitoring includes cardiac rhythm, non-invasive blood pressure and pulse oximetry.

Either venous access site suffices for RV septal sampling (the most common site). Optimally, samples are obtained from a

variety of sites on the septal surface, while avoiding the septal leaflet of tricuspid valve, as well as very apical positions. Very few centres perform left ventricular (LV) EMB in children, for rare indications [6]. When performed, the femoral artery is usually chosen for retrograde access to the LV, although a transseptal route could also be used.

EMB is routinely done under fluoroscopic guidance. Single plane or biplane may be used depending on operator preference and available facilities. Use of a minimal frame rate (e.g. <7.5 frames per second) reduces radiation exposure by >50% without sacrificing necessary visualization or safety in heart transplanted children [7].

52.4 Materials

The bioptomes commonly employed are single-use, with sharpened stainless-steel cusps designed to pinch rather than to cut the myocardial muscle (Fig. 52.1) (Table 52.1). Bioptomes are manufactured in two general forms: [1] with both a preshaped and a stiff distal end and [2] with an unshaped distal end and a flexible shaft. In general, preshaped bioptomes are used for the jugular

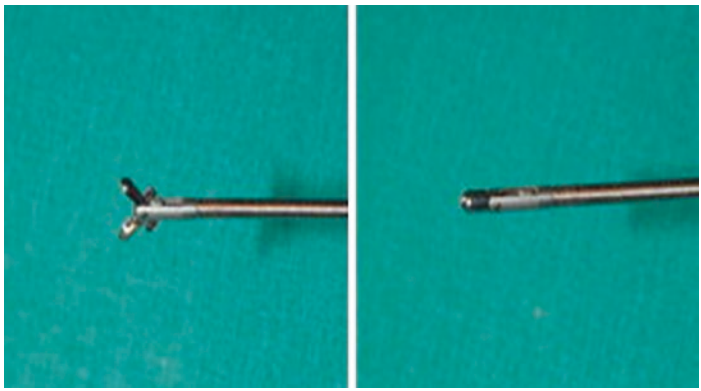

Fig. 52.1 The profile of the biopsy forceps (Cordis®) when the jaws are open (*left panel*) and closed (*right panel*). Note that the bioptome has sharpened cusps designed to pinch rather than to cut the myocardial muscle

Table 52.1 Common bioptomes of different size and shaft length

Model	Size (fr)	Shaft length (cm)
Cook®	3.0, 5.2	0 and 120
Cordis®	5.5, 7.0	50 and 104
Sparrow Hawk®	5.0, 6.0, 7.0	50 and 105
Novatome™	6.0, 7.0, 8.0, 9.0	50 and 100
Argon®	5.0, 5.5, 6.0, 7.0,7.5	50 and 105

approach in order to avoid a very posterior or apical sample site. Flexible bioptomes requiring the use of a long sheath or a guiding catheter are used for both arterial and venous EMB. The long sheath directs the bioptome clear of the tricuspid valve and expedites reintroduction to the RV.

We normally perform EMB of the right ventricle with a bioptome through a long sheath which has been positioned in the RV. Summarizing, the following materials are required:

- A short introducer
- A balloon end-hole catheter
- An exchange-length standard guidewire
- A bioptome — 5 or 6 Fr — is usually sufficient to obtain adequate samples
- A long sheath (shorter than the selected bioptome)
- Containers for the specimens

52.5 Technique

Before using a flexible bioptome, either from the internal jugular vein or the femoral vein, a long sheath with angulated tip is advanced to the pulmonary artery. This can be accomplished by floating a balloon end-hole catheter to the PA and then performing an over-the-wire exchange for the long sheath with its dilator. Removal of the wire and dilator, and slight withdrawal of the sheath then leaves the sheath tip in the midcavity of the RV. This technique has the advantage of a single passage through the orifice of the tricuspid valve. Some operators prefer to use the preshaped and stiff bioptome from the internal jugular vein. In this case, the

bioptome is manoeuvred independently and advanced into the RV without the protection of a long sheath. Thus, it recrosses the tricuspid valve each time a specimen is obtained, possibly increasing the risk of tricuspid valve damage. For EMBs from the left ventricle, a curled-up guidewire is positioned in the left ventricle and the long sheath used usually has a straight tip.

The standard fluoroscopic views are 0° anteroposterior and 90° lateral for the right ventricle, and 30° right anterior oblique and 60° lateral anterior oblique for the left ventricle. Additionally, transthoracic or transesophageal echocardiography may be used if specific myocardial lesions are targeted (i.e. cardiac tumours).

Approximately 5 or 6 samples, each of 1–2 mm in size, should be obtained from different regions of the interventricular septum, according to guidance from the requesting service and pathology lab. This may reduce the risk of myocardial perforation as well as increase the chance of obtaining a diagnosis in disease processes that do not affect the myocardium uniformly. If the sheath falls back to the right atrium, it should be repositioned by reintroducing the balloon catheter. Sample sites can be changed by advancing or retracting, or torqueing the sheath slightly. Sites can also be varied by modifying the curve on the bioptome. Between acquisitions, clearing and flushing of the sheath will minimize air embolization or thrombus accumulation.

Postprocedural ventricular angiograms to exclude extravasation of contrast are infrequently useful in the absence of clinical suspicion. Postprocedural echocardiograms are not routinely performed unless indicated for other reasons [8].

52.6 Tips and Tricks

Obtaining biopsy specimens from the RV or LV free wall risks perforation and cardiac tamponade; hence, the samples should be taken from the interventricular septum. This is mandatory in patients with dilated cardiomyopathy, in which the RV free wall is thin and friable. Before the insertion of the flexible bioptome, it is useful to shape the distal end of the bioptome to guide it posteriorly (to the septum) and to the midcavity of the RV. Experimentation

with a variety of curves and combinations allows for targeted sampling in challenging cases. Repeated sampling of the same site can reduce test sensitivity, increase the risk of complications and decrease yield (due to scarring) on future biopsies.

We use a straight long sheath (Flexor® Check-Flo® Introducer® Cook®) that is heated up with warm air and shaped so the distal end is pointing posterior towards the interventricular septum (Fig. 52.2).

It is recommended to open the jaws of the bioptome within the ventricular cavity before engaging the muscular septum (Fig. 52.3). If the jaws do not fully open, bioptome and long sheath should be slowly pulled back, since they might already be "buried" in muscle. Then, the bioptome must be advanced slowly until feeling mild resistance, or "buckling" of the shaft. When the bioptome engages the septum, it usually triggers some ectopy. It is important to keep gentle forward pressure on the bioptome while closing the jaws. With the jaws firmly closed, the bioptome may be retrieved, while leaving the long sheath tip in the ventricu-

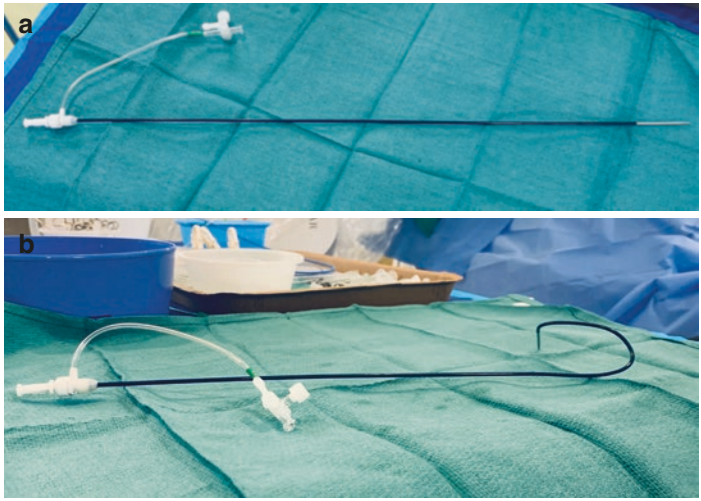

Fig. 52.2 Flexor® Check-Flo® Introducer® (Cook®) straight long sheath before (**a**) and after (**b**) shaping under hot air. The distal portion is curved towards the tricuspid valve and then posterior to reach the interventricular septum

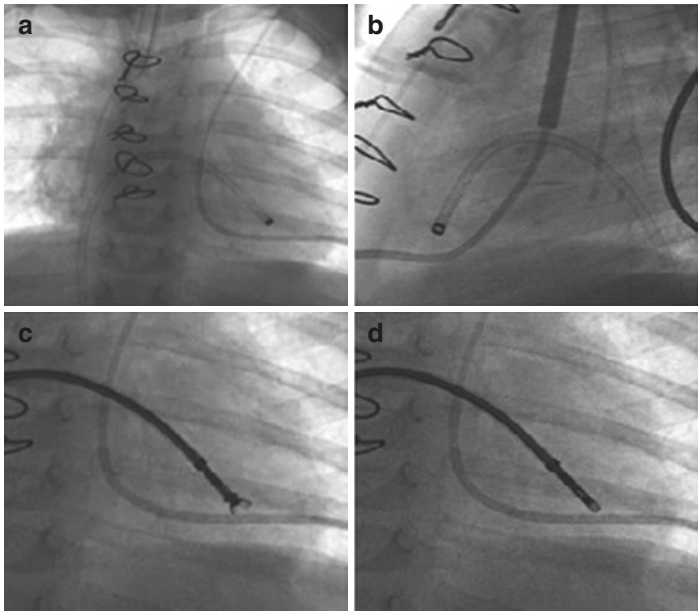

Fig. 52.3 Anteroposterior view (**a**) and lateral view (**b**) showing the position of the long sheath from the femoral venous approach. The jaws of the bioptome should be fully open before reaching the muscular wall (**c**). The jaws are closed to pinch off a sample from the interventricular septum (**d**)

lar cavity. Mild resistance is normally perceived when the bioptome pulls away from the septum. Pushing slightly on the long sheath can ensure it stays within the ventricular cavity.

52.7 Limits and Potential Complications

It is worth highlighting that, since EMB consists of taking only a few random samples from the myocardial muscle of the septum, its diagnostic sensitivity may be limited [9]. Inflammation of the myocardium may have a patchy pattern or involve segments inaccessible to the bioptome. Thus, when EMB is done to prove myocarditis or to detect post-transplant rejection, only positive findings are considered diagnostic [9].

A well-recognized drawback of EMB is the risk of procedural complications and long-term sequelae. Complications may be related to sheath insertion, to the biopsy itself or to the clinical status of the patients. Major complications include myocardial perforation with pericardial tamponade and need for pericardiocentesis, haemopericardium, permanent AV block and severe valvular damage [6, 10]. Other complications of potentially major long-term consequence are tricuspid regurgitation and creation of coronary artery fistulae. Minor complications are considered pericardial effusion not requiring drainage, transitory arrhythmias, conduction anomalies (most often a right bundle branch pattern) and vascular site complications.

Myocardial perforation with pericardial tamponade and need for pericardiocentesis occurs in <1% of the patients. Altogether, minor complications occur in <10% of the patients [6, 11]. Major complications are more common in children with cardiomyopathy than in children undergoing post-transplant biopsy [12]. Infants constitute a distinctly higher risk cohort, with complication rates two or three times higher than those of older children and adolescents [6, 10]. Typically, the risk of EBM is highest in young patients with poor ventricular function, a dilated thin-walled ventricle, high right ventricle pressure and requiring inotropes [6, 10, 13]. Complications following LV EBM may be three times more common than after RV EMB [6].

Procedural mortality is exceedingly rare and in most cases is a result of perforation and tamponade in young patients with cardiogenic shock or unstable ventricular arrhythmias.

52.8 How to Manage Complications

In cases of pericardial tamponade, immediate pericardiocentesis should be performed, with concomitant volume resuscitation. Consideration should be given to prompt return of the aspirated blood through the vascular introducer, if donor products are unavailable. The standard manoeuvres of resuscitation, surgical repair of the lesion and implantation of a ventricular assist device may be required.

52.9 Postprocedural Care

The samples can be removed from the bioptome using a sterile needle and should be placed on sterile gauze as appropriate for transport to the laboratory. The type of storage depends on the clinical question [4]. Some standard fixatives and techniques of analysis are listed in Table 52.2.

Table 52.2 Some standard fixatives and techniques of analysis for common clinical questions

Fixative	Technique	Clinical question
10% neutral-buffered formalin at room temperature	Light microscopy	Transplant rejection
		Myocarditis
		Infiltrative/ unexplained cardiomyopathy
		Tumours
	Immunohistochemistry	Transplant rejection
		Tumours
	Polymerase chain reaction	Myocarditis
4% glutaraldehyde at room temperature	Transmission electron microscopy	Unexplained cardiomyopathy (amyloidosis, glycogen storage diseases, lysosomal storage diseases or mitochondrial disease)
		Anthracycline-induced cardiotoxicity
Flash-frozen tissue transported on ice or fixative like RNA later (Ambion, Austin, TX) or snap frozen in OCT-embedding medium or liquid nitrogen	Culture, polymerase chain reaction (PCR), reverse transcriptase PCR (rtPCR)	Myocarditis
	Immunofluorescence	Storage diseases
	Immunohistochemistry	Tumour typing
	Molecular studies	Amyloid classification
		Muscular dystrophies

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53 Evaluations Before Partial and Total Cavopulmonary Connections

Gabriella Agnoletti
and Giuseppe Antonio Mazza

53.1 Anatomic Description and Physiopathology

Patients with single ventricle physiology have a variety of complex heart diseases that are not suitable to biventricular repair. Fontan circulation can be obtained by performing an atriopulmonary connection or a total cavopulmonary connection (TCPC). Both allow the passive flow of the systemic venous blood into the lungs. In the completed Fontan state, the pressures in the caval and pulmonary circulations must be high enough to ensure flow through the lungs and adequate preload of the systemic ventricle while avoiding high-pressure venous congestion. This goal is achieved when pulmonary arterial pressure (PAP) is between 10 and 14 mmHg. Partial cavopulmonary connection (PCPC), with or without additional pulmonary blood flow (PBF), usually

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precedes conversion to TCPC. PCPC forces 50% of cardiac output to bypass the heart and directly enter the lungs, increasing oxygen saturation.

53.2 Clinical Scenarios

Patients with single ventricle physiology can have duct-dependent pulmonary or systemic circulation, and a protected or non-protected pulmonary vascular bed.

(a) Patients with duct-dependent pulmonary circulation or insufficient PBF need either a BT shunt or a ductal stenting.
(b) Patients with duct-dependent systemic circulation need a Norwood operation or a hybrid treatment (Chap. 39).
(c) When obstruction to the systemic outflow exists, a Damus–Kaye anastomosis bypasses the subaortic obstruction, by creating an aortopulmonary anastomosis.
(d) Patients with excessive PBF require pulmonary artery banding.

During follow-up there can be different scenarios:

1. Neonates with adequate PBF can reach the age of 6 months without a neonatal palliation and receive a PCPC as a first intervention.
2. Neonates with pulmonary banding or BT shunt-dependent pulmonary circulation display progressive cyanosis and generally need PCPC at the age of 4–6 months.

   An additional source of PBF such as BT shunt or native antegrade flow can be occluded or left in place at time of PCPC, in order to prepare the child for a TCPC at an older age.
3. TCPC is generally performed after the age of 3 years, depending on the degree of systemic desaturation. At the time of TCPC, any additional source of PBF is removed either surgically or in the cardiac catheterization laboratory.

53.3 Indications and Patient Selection

PCPC and TCPC are often preceded by cardiac catheterization aimed at measuring PAP, assessing pulmonary artery size, and treating possible associated anomalies.

Catheterization is performed in either all or selected patients, according to the team policy and depending on the availability of high-quality noninvasive imaging.

Patients in whom catheterization is commonly performed prior to PCPC include:

- Patients with hypoplastic left heart syndrome (having received either Norwood I or hybrid treatment).
- Patients in whom the anatomy of pulmonary arteries needs to be clarified or in whom PAP might be high.

Prior to TCPC a cardiac catheterization should be considered in:

- Patients with hypoplastic left heart syndrome having had Norwood II (surgical or hybrid treatment).
- Patients with additional PBF.
- Patients in whom the anatomy of pulmonary artery needs to be clarified or in whom PAP might be high.

Commonly associated anomalies that can be treated percutaneously are aortic recoarctation, aortopulmonary collaterals, restrictive foramen ovale, and stenosis of pulmonary arteries or of the superior vena cava.

53.4 Imaging

At present, the need for a routine pre-Glenn and pre-Fontan cardiac catheterization is questioned, and there is a mounting interest to replace it with noninvasive methods. In addition to possible acute adverse events of catheterization, a growing awareness of

hazardous long-term effect of ionizing radiation has spread. In fact, the risk of radiation-related cancer in patients exposed to radiation during childhood seems to be dose-related, and more prominent in children irradiated early in life. Furthermore, the risk for solid tumors persists throughout life.

Therefore, in patients with functional single ventricle before PCPC operation, several clinical trials compared cardiac magnetic resonance (CMR) versus cardiac catheterization, demonstrating similar short- and long-term post-Glenn outcomes.

Even before TCPC operation an algorithm based on clinical, hematological, and echocardiography findings proved to be useful in selecting patients not needing catheterization, with a high negative predictive value, further increased by CMR [1].

Noninvasive imaging can provide definitive information on pulmonary artery anatomy (Fig. 53.1).

It can offer information on intracardiac and extracardiac structures, display unsuspected anomalies and offer accurate data on ventricular function.

Radiologic imaging, in order to be valuable, has to be of high quality.

Operators should be skilled at obtaining and interpreting the images of the hearts with congenital anomalies.

However, despite CMR can provide valuable anatomical information, it does not offer hemodynamic data, that could be essential to predict mortality and negative long-term outcomes in selected patients. In addition, prospective studies comparing anatomical data derived by catheterization versus CMR are deficient, and the cost effectiveness yet needs to be investigated [1].

53.5 Pre-PCPC Catheterization

Vascular access, technique, and materials can vary in accordance with the anatomy.

The aim of the exam is to measure PAP, assess the anatomy of pulmonary arteries and rule out or treat associated anomalies.

Intravenous heparin at a dose of 50–100 UI/kg should be administered.

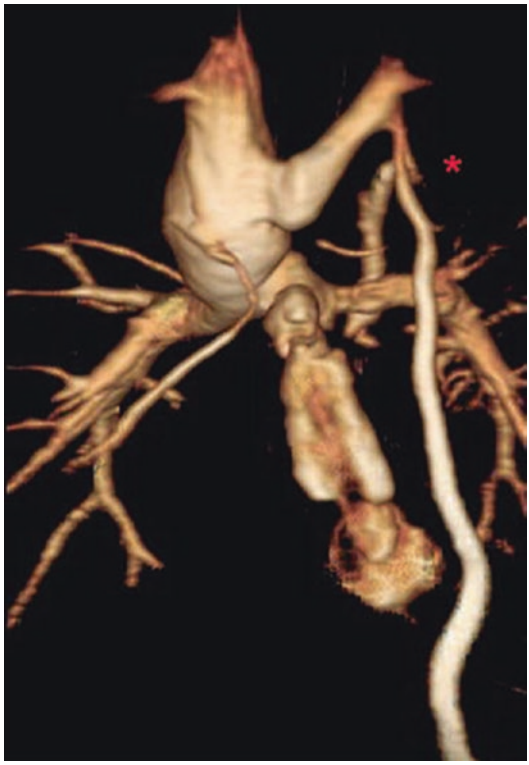

Fig. 53.1 3D MRI in AP view shows a dilated superior vena cava, the presence of antegrade flow and a venovenous collateral

Arterial access is generally needed to visualize pulmonary arteries in patients with pulmonary atresia and a BT shunt.

Angiography rules out the presence of aortic coarctation and aortopulmonary collaterals.

If the shunt cannot be entered, PAP can be measured via pulmonary veins [2]. We penetrate a pulmonary vein until the catheter is wedged and the shape of the pressure curve changes and a transpulmonary gradient appears.

The same approach is used in patients with inadequate antegrade PBF and a BT shunt.

In our view, it is easier and safer to measure PAP via pulmonary veins rather than via a small BT shunt or a small native pulmonary outflow.

Patients with hypoplastic left heart syndrome are complex patients who need a complete and precise evaluation.

In subjects after a Norwood I operation, venous and arterial access should be obtained.

In some cases, it is possible to obtain a complete evaluation via venous access alone. The catheter enters the right atrium, and then the left atrial pressure and pulmonary vein wedge pressure are obtained. Then ventricular pressure is measured, the catheter enters the neoaorta and is guided into the descending aorta. If difficulty is encountered when trying to enter the neoaorta or the child shows instability, this approach should be abandoned, and the arterial approach should be adopted.

An angiography should be performed into the BT shunt to visualize pulmonary arteries. It is often difficult to enter the brachiocephalic artery using venous access. Arterial access should be used in those cases. Possible coexisting problems are recoarctation and aortopulmonary collaterals. Both conditions can be treated, if necessary.

Patients with hypoplastic left heart syndrome having received a hybrid treatment are potentially fragile and can display several associated anomalies such as restrictive atrial septal defect, proximal or distal displacement of the ductal stent, stent thrombosis, and preductal or postductal coarctation. The treatment of these lesions, including stenting of the atrial septum and of the native aortic arch, will be discussed in the dedicated chapter.

In these patients, the neoaorta provides pulmonary, systemic and coronary circulations. High-volume angiography (2–3 ml/kg) and appropriate views are necessary to obtain an adequate visualization of the banded pulmonary arteries, the stented arterial duct, and the native aortic arch.

We rarely manage to enter a banded pulmonary artery and generally prefer to measure pulmonary pressure via pulmonary vein wedge pressure [2]. If PAP is high, we need to know if this is due to excessive flow, restrictive atrial septal defect, obstructed

pulmonary venous return, recoarctation, or ventricular dysfunction. The pressure measurement should be repeated once the associated lesions are treated.

53.6 Pre-TCPC Catheterization

Catheterization is performed, according to the presence of forward flow to the lungs, via the femoral vein or the internal jugular vein [3].

Arterial catheterization can be needed to rule out or treat aortic recoarctation and aortopulmonary collaterals.

If a pulmonary banding or a BT shunt is left in place, the pulmonary artery can be entered via femoral venous access or femoral arterial access, respectively.

However, it is generally easier and rapid to reach pulmonary arteries via the internal jugular vein. PAP and either wedge or left atrial pressure are measured.

Pulmonary angiography is performed in a four-chamber view.

If some washout is found, aortopulmonary collaterals should be suspected and aortography should be accomplished. Aortopulmonary collaterals in single ventricle can be found in more than 80% of cases, and their impact on clinical outcomes is not yet fully understood. Despite some authors suggested that they lead to significant energy loss, increased single ventricle end-diastolic pressure, worsening single ventricle function, prolonged pleural effusion and hospitalization duration in the immediate period after Fontan completion, other ones reported no significant adverse effects on the immediate post-operative outcomes and suggested that they can rather have a positive effect in maintaining an adequate preload of the systemic ventricle. Thus, significant practice variability continues to be present, and no current consensus exists regarding the indications of their occlusion. However, during catheterization it must be considered that the presence of significant aortopulmonary collaterals makes the accurate evaluation of pulmonary vascular resistance difficult [1].

If a rapid opacification of pulmonary veins is noticed, pulmonary fistulae should be ruled out. The presence of microfistulae can be confirmed by injecting microbubbles obtained by rapidly mixing 80% blood with 20% air in both distal pulmonary arteries. Simultaneously transthoracic or transesophageal echocardiography is performed. If pulmonary fistulae are present, generally massive opacification of the left atrium is seen.

Post-PCPC patients can have right and left superior vena cava and as such a bilateral bidirectional PCPC may have been performed.

In these patients, the relative size of the superior vena cava should be established. Competitive flow can sometimes prompt the thrombosis of the smaller vena cava that can, if necessary, be reopened using balloon angioplasty and stenting.

Disconnection of pulmonary arteries can also be observed in patients with additional competitive flow (Fig. 53.2).

Any stenosis in the PCPC system must be treated, and collateral vessels connected with the inferior vena cava may need occlusion if they are large before measuring a reliable pressure.

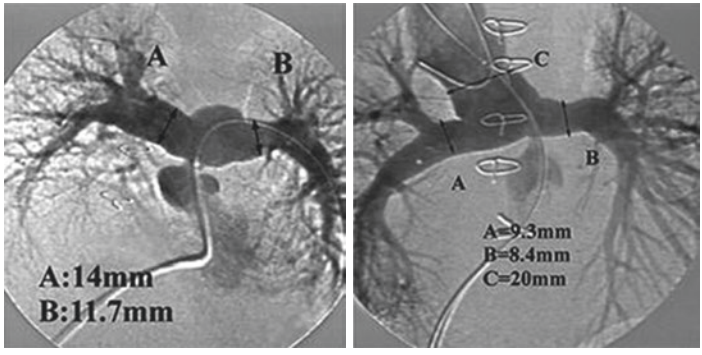

Fig. 53.2 Measurement of pulmonary artery (double-headed arrows) size in two different patients before (*left*) and after (*right*) PCPC. In both cases, the catheter enters the pulmonary artery in an antegrade way. In the patient with PCPC, a multi-track catheter (notice the guidewire parallel to the catheter) attains the superior vena cava

When stenosis of the superior vena cava involves the origin of one or both pulmonary arteries, the lesion can be treated percutaneously, using open-cell stents, but can also be treated surgically, at the time of TCPC.

Before TCPC, however, the measurement of pulmonary pressure has to be reliable; therefore, confounding factors that either lower pulmonary pressure (presence of venovenous collaterals and pulmonary fistulae) or increase pulmonary pressure (obstruction to pulmonary venous return, restrictive atrial septal defect, ventricular dysfunction, aortic coarctation, additional flow to the lungs, stenosis/hypoplasia of pulmonary arteries) should be looked for and, if possible, treated.

Depending on the quality of the available noninvasive imaging, the right and left pulmonary artery diameters can be measured immediately before their first branches and used to calculate cross-sectional areas (Fig. 53.2).

The most useful index to measure pulmonary artery size is the Nakata index.

Accepted values for patients scheduled for TCPC are >200 mm^2/m^2 [4].

53.7 Materials

Diagnostic catheterization in pre-PCPC and pre-TCPC patients is performed using standard catheters and guidewires, in accordance with the experience of the operators. Open-tip catheters can be more easily manipulated when the anatomy is unusual. However, flow-directed catheters can provide safe manipulation and offer high-quality imaging.

If interventional catheterization is necessary, various materials are needed.

To occlude the venovenous collaterals, pulmonary fistulae, and aortopulmonary collaterals, we use coils, particles, plugs, or various devices, according to their anatomy and size.

To treat a restrictive atrial septal defect or an intact atrial septum, radiofrequency, balloons, cutting balloons, blade, and stents are required.

To treat aortic coarctation and stenosis of pulmonary arteries, we use balloons, cutting balloons, and stents.

53.8 Expected Results

The ideal pre-PCPC patient has low PAP and normal pulmonary artery size.

He has non-obstructed pulmonary venous return, non-restrictive atrial septal defect, normal ventricular function, and non-obstructed ventricular outflow, and does not have aortopulmonary collaterals.

The ideal pre-TCPC patient has mean PAP <14 mmHg, normal pulmonary artery size, normal ventricular function, and competent atrioventricular valve(s).

He has no venovenous collaterals or pulmonary fistulae, non-obstructed pulmonary venous return, non-restrictive atrial septal defect, and non-obstructed ventricular outflow, and does not have aortopulmonary collaterals.

Real patients are often very different from ideal patients. Some degree of ventricular dysfunction, incompetence of atrioventricular valve(s), and small aortopulmonary collaterals can be tolerated. TCPC can be performed also in patients with occluded inferior vena cava. Stenoses in the PCPC anastomosis must however always be treated, either in the catheter laboratory or at the time of surgery.

53.9 Tips and Tricks

Do not forget that PAP can be measured via pulmonary veins. Always obtain pressure measurement before performing a pulmonary angiography to avoid increasing PAP.

Be aware that PAP changes in accordance with aortic pressure.

The simultaneous measurement of pulmonary vein wedge pressure and end-diastolic ventricular pressure can rule out stenosis of pulmonary veins.

Simultaneous angiography in disconnected pulmonary arteries allows the measurement of the distance between disconnected segments (Fig. 53.3).

Patients with a single ventricle who had a prolonged stay in intensive care unit can lose femoral venous access.

In patients with antegrade pulmonary flow, an arterial retrograde cardiac catheterization can be performed from the venous approach.

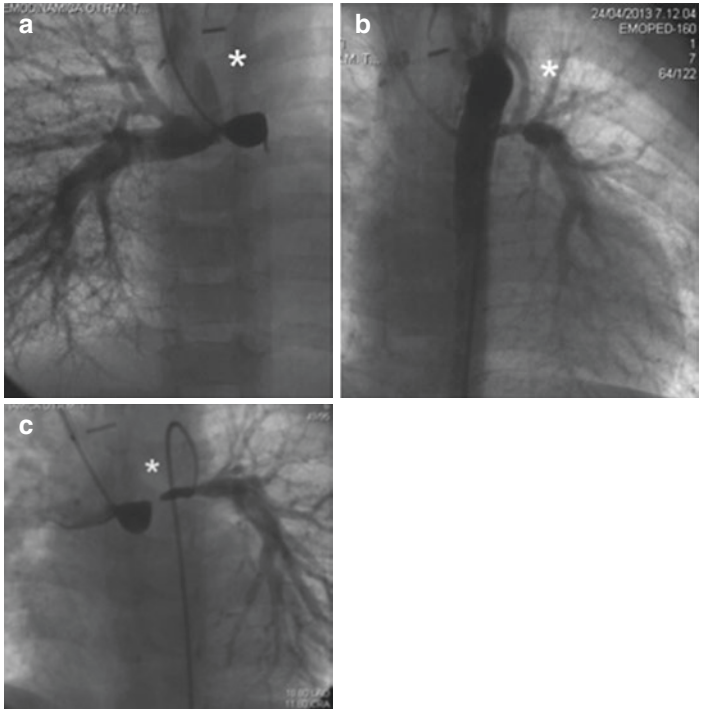

Fig. 53.3 Disconnected pulmonary arteries in a patient with competitive flow. The right pulmonary artery is fed via the superior vena cava (**a***) and the left pulmonary artery via a BT shunt (**b***). Simultaneous injection allows to appreciate the distance between the pulmonary arteries (**c***)

In patients in whom a persistent left superior vena cava is suspected, a hand injection in a vein of the left arm will easily demonstrate this condition.

To obtain a reliable measure of PAP, we can perform a balloon test occlusion of any additional source of PBF. We have to be aware that we need two vascular accesses or two catheters to measure pressure during test occlusion, unless we use Berman and reversed Berman catheters. However, these catheters have a small balloon and rarely provide a stable occlusion.

53.10 Pitfalls

PAP can be unusually low even when pulmonary arteries are small. In this case, always look for anomalies able to lower PAP such as pulmonary fistulae and venovenous collaterals.

53.11 Complications

Patients in whom a diagnostic catheterization is performed have the general risks of any cardiac catheterization.

Jugular catheterization is burdened by the risk of arterial puncture and bleeding.

Rarely, transient atrioventricular block occurs when the catheter is manipulated from the ventricle into the aorta. When this occurs, the catheter should be removed from the vein and an arterial approach used. Ventricular pacing is rarely necessary.

Patients in whom an interventional catheterization is performed are subject to the risks of the respective interventions. There are generic rules and specific rules to manage complications. Generic rules are be quiet, be logical and avoid useless maneuvers. Follow the sequence A-B-C-D (A, airway; B, breathing; C, circulation; D, drugs). Specific rules are read in the chapters referring to the respective interventions.

53.12 Post-procedural Care and Follow-Up

Patients having had a diagnostic catheterization should undergo standard follow-up. In anti-aggregated patients having had a jugular catheterization, particular attention should be paid to the risk of bleeding. After interventional catheterization, post-procedural care varies in accordance with the intervention performed.

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54 Hemodynamics in Pericardial and Myocardial Diseases

Maria Giulia Gagliardi, Mario Panebianco, Roberto Formigari, and Giacomo Pongiglione

54.1 Pericardial Disease

54.1.1 Anatomy and Function

The term *pericardium* ("around the heart") outlines a complex structure composed of:

1. Visceral layer
2. Parietal layer
3. Pericardial fluid

The normal total volume of pericardial fluid surrounding the heart is around 30 ml.

The function of the pericardium is to:

1. Protect the heart within the chest
2. Preserve myocyte function under stress and to limit acute distention of the heart chambers

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3. Distribute hydrostatic forces over the heart
4. Exclude extracardiac and/or intrathoracic disease from extension into the heart [1]

54.1.2 Physiology and Physiopathology

The inflammation of pericardium ("pericarditis") from any cause can be followed by three hemodynamic complications:

1. A pericardial effusion under pressure, resulting in cardiac tamponade
2. Progressive pericardial fibrosis and thickening, causing constrictive physiology
3. A combination of both [1]

The normal *intrapericardial pressure* (IP) is slightly negative (−3 mmHg) and it is affected by:

1. Pleural pressure
2. Intrapericardial volume
3. Intracavity pressure [1]

The IP is an important determinant of the transmural filling pressure of cardiac chamber:

$$\textit{Transmural filling pressure} = \text{intracavitary pressure}\left(\text{EDP}\right) - \text{IP}.$$

The normal value is >5 mmHg. As IP elevates above zero, the recorded intracavity pressure becomes less representative of the cavity's transmural filling pressure.

For example, LVEDP = 10 mmHg and IP = 8 mmHg, the left ventricular transmural pressure will be just 2 mmHg; this will start to affect the right atrial filling. When the IP exceeds the intracavity pressure, the chamber wall compresses or collapses in mid-diastole.

Ventricular interdependence imparted by the pericardium: The consequence of limitation of cardiac filling by the pericardium is that overfilling of one ventricle will reduce filling of the other ventricle because the parietal pericardial layer limits overall cardiac volume. This phenomenon is referred to as *ventricular interdependence* [1] and can be seen in constrictive pericarditis, pericardial tamponade, effusion-constrictive pericarditis, right ventricular infarction, and tumor encasement of the heart.

Under normal conditions, with inspiration, negative intrathoracic pressures cause increased systemic venous return to the right heart, but there is also even a larger increase in the capacity of the pulmonary vascular bed. This leads to a reduction in left-sided output, despite the increase in systemic venous return to the right heart (Fig. 54.1).

In cardiac tamponade, the physiological behavior is emphasized: right ventricular filling is maintained at the expense of restricted left ventricular filling. In inspiration, the increased volume of blood accommodated by the pulmonary vascular bed, coupled with reduced left ventricular filling, results in a greater reduction in systemic output [1].

In constriction, there is discordance of LV and RV pressure changes in inspiration: the LV pressure falls, and the RV pressure rises.

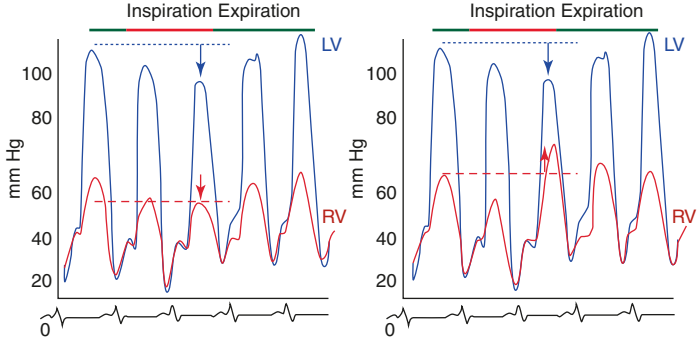

Fig. 54.1 Left tracing, normal; right tracing typical of constriction. Normally there is a concordant fall in LV and RV pressures in inspiration

54.2 Clinical Scenarios

54.2.1 Acute Pericarditis and Pericardial Effusion

There is a wide spectrum of causes of pericarditis associated with pericardial effusion or constrictive disease: infections (e.g., viral, bacterial, protozoal), immune-inflammatory disorders (systemic lupus erythematosus, rheumatoid arthritis, drugs), postradiation therapy, and neoplasia (primary and secondary) [1].

Clinical presentation is strongly influenced by the acute setting of the disease.

Physical examination: Regarding pericarditis, a pericardial friction rub is a typical finding, best heard when the patient is bent forward and accentuated at the end of expiration.

In tamponade, the combination of the classic findings known as Beck's triad (hypotension, jugular venous distention, and muffled heart sounds) occurs in only 10–40% of patients. Tachycardia, tachypnea, and hepatomegaly are common. Pulsus paradoxus (explained below) is relatively nonspecific and insensitive.

Pulsus paradoxus (or Kussmaul's pulse): It is defined as a fall in arterial systolic pressure of >10 mmHg during normal inspiration.

Oximetry tracing often reveals the finding in the absence of direct arterial pressure line.

Diastolic pressure is not supposed to fall, thus reducing the difference between systolic and diastolic pressure with a consequent weakening of the pulse.

Beware that Kussmaul's pulse is not specific for pericardial disease. It can occur in any condition with exaggerated inspiratory effort (pulmonary disease, pulmonary embolism, pleural effusion, congestive heart disease) [1]. It may also be obscured if the following conditions are also present: aortic insufficiency, atrial septal defect, and mechanical ventilation with positive end-expiratory pressure (PEEP). These are all cases that normalize the degree of left ventricular filling by favoring left ventricle volume load [2].

In the presence of tamponade, the right atrial and right ventricular pressure tracings reveal a blunted or an absent y descent; hence there is no dip-and-plateau waveform ("square root sign") and no Kussmaul's sign.[1]

Jugular venous pressure: The *x* wave in the venous pressure trace is produced by atrial relaxation but predominantly by the systolic descent of the atrioventricular plane from ventricular contraction.

In cardiac tamponade, the *x* is steepened. The *y* is the result of early ventricular filling and lowering of the atrial venous pressure. It is prominent in conditions such as constriction and is attenuated in tamponade. In case of effusion–constriction, the *x* and *y* waves are usually similar producing an M or W pattern in right atrial tracing (Fig. 54.2). Also, this pattern is not specific for constrictive pericarditis but is also seen in heart failure, from restrictive cardiomyopathy and right ventricular infarction [2]. In constrictive disease, because of chronic elevation of right atrial pressure, hepatic congestion and dysfunction with ascites and peripheral edema are frequently encountered.

ECG findings: Typically, the electrocardiographic changes evolve through four stages characterized by diffuse ST-segment elevation and PR depression (seen in >80% of patients), to normalization of the ST and PR segments, to widening of the T-wave. In cases where there is a moderate to severe pericardial effusion, low voltages may be seen and electrical alternans, a cyclic variation in QRS amplitude, when there is excessive motion of the heart within the fluid-filled pericardial space (swinging heart) [3].

Imaging: Echocardiography is the main diagnostic tool used for the diagnosis and characterization of the pericardial effusion, but it adds little where there is constrictive disease.

MRI is an optimal tool to study the morphologic characteristics of the pericardium and also allows the identification and characterization of other pathology [1].

[1] *Kussmaul's sign:* It is a paradoxical rise in jugular venous pressure on inspiration and it is usually indicative of limited right ventricular filling.

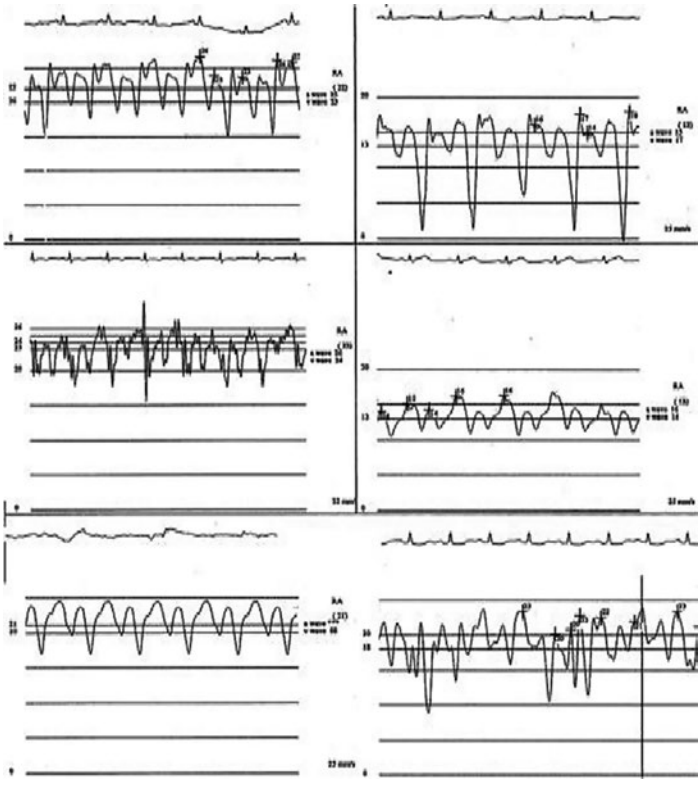

Fig. 54.2 Right atrial pressure in proven constriction cases. The mean pressures are all elevated. Although the *x* and *y* descent are all exaggerated, it can be seen how variable the depth of the *x* and *y* descent may be. In most cases, the magnitude of the *x* and *y* descents are similar, producing an M or W pattern. In the right upper tracing, the y descents are greatly dominant because of unusually vigorous recoil of the constrictive pericardium. In the right lower image, the inspiratory increase in *x* and *y* descents is depicted over two respiratory cycles. In later inspiration, the mean pressure rises as the right-sided heart compliance is exceeded by increased venous return—Kussmaul's sign. It is better appreciated by a mean pressure tracing

54.2.2 Constrictive Disease

Constrictive physiology may develop after pericarditis. It is found in approximately 0.2% of cases after open heart surgery, presenting a mean of 2 years postoperatively, and it is notable for occurring with underlying abnormal hearts (due to residual valve disease and/or infarction). Radiation therapy-induced constriction almost always displays concurrent fibrotic restrictive cardiomyopathy and surgical *pericardiectomy* is far less effective in respect to other causes of constriction. In constrictive disease, the pericardium is usually thickened, sometimes calcified, but it can be apparently normal.

54.2.2.1 Clinical Scenarios

From the clinical point of view, symptoms are often vague and their onset is insidious; they include malaise, fatigue, and decreased exercise tolerance. Classical signs of right heart failure are typical (peripheral edema, nausea, abdominal discomfort, ascites).

Physical examination: Jugular venous distention is frequent and Kussmaul's sign can be present even though it is sensitive but nonspecific for constriction. Auscultation reveals muffled heart sounds and occasionally a characteristic pericardial knock (60–200 ms after the second heart sound), caused by sudden termination of ventricular inflow by the encasing pericardium.

ECG findings: The ECG does not show specific findings, but low voltage may be seen.

Echocardiographic findings: Inflow Doppler analysis usually demonstrates mitral E wave reduction during inspiration due to inability of the left ventricle to generate a proper diastolic pressure because of the thick pericardium. The tissue Doppler findings will usually be normal excluding a myocardial muscle disease.

Cardiac catheterization: Ideally in constrictive pericarditis, a catheter study should be performed using mild sedation in order to minimize interference with respiratory physiology.

Arterial and venous accesses and two pressure transducers are needed to simultaneously record right and left pressures. Required measurements include:

1. Right atrial pressure
2. RV pressure
3. Pulmonary pressure
4. Wedge pressure

All these measurements should be recorded using an end-hole catheter (e.g., Swan–Ganz).

Then a pigtail catheter or an end-hole catheter should be positioned in the left ventricle.

First, observe and record simultaneous wedge pressure and LVEDP across the respiratory phases. A normal gradient between these two pressures is considered to be <5 mmHg.

The right-sided catheter should then be positioned in the RV and recorded simultaneously with the left, with overlap of the two pressure lines in order to highlight the ventricular interdependence (reduction of the LV systolic pressure and increase of the RV systolic pressure during inspiration). An irregular heart rhythm may obscure this finding [2].

In summary, typical findings for constrictive pericarditis are:

1. Elevated and equilibrated left and right ventricle end-diastolic pressures.
2. RV diastolic pressure (RVDP)>1/3 RV systolic pressure (RVSP).
3. Ventricular interdependence: *Dissociation of RVSP from LVSP.*
   On the first–second cardiac cycle during inspiration, there is a rise in RVSP and a fall in LVSP.
4. Inspiratory increase in PW pressure: LVDP gradient (>5 mmHg) [3].
5. The ventricular pressure tracings show an early diastolic dip in pressure followed by a plateau phase due to rapid early diastolic filling and subsequent restriction in filling (*square root sign or dip-and-plateau sign*).
6. Right atrial waveforms show a preserved *x* and prominent *y* descent, often with equal *a* and *v* waves (M or W sign).

Differentiating constrictive pericarditis and *restrictive cardiomyopathy* is important because treatment is radically different. Patient history is often useful but sometimes their presentation and course overlap in many aspects [2–4].

Restrictive cardiomyopathy can be caused by a number of diseases and normally is associated with systemic diseases. Often, the cause is unknown. The rigidity of the heart walls may be caused by fibrosis, the replacement of muscle cells with tough, fibrous tissue. Examples are amyloidosis, hemochromatosis, and sarcoidosis.

In respect to constrictive pericarditis, in hearts with a restrictive physiology the impact of the phasic respiratory cycle is limited, with a much lower degree of ventricular interdependence. This may be unveiled by the blunting of the "x" descent in the right atrial pressure curve and a less pronounced inspiratory fall of left ventricular systolic pressure [5, 6].

Computerized tomography is a useful tool for detection of abnormal pericardial thickness, calcifications, and assessment of volume and function of the cardiac chambers.

More recently, MRI has emerged as a powerful diagnostic tool, providing insights of the myocardial structure, and allowing a precise picture of infiltrative cardiomyopathies resulting in a restrictive physiology [1, 4, 5].

In Table 54.1, there are echocardiographic and hemodynamic data helping differentiate between constrictive pericarditis and restrictive cardiomyopathy.

54.3 Myocardial Disease

Cardiomyopathies can be classified into five groups: dilated cardiomyopathy (DCM), hypertrophic cardiomyopathy (HCM), restrictive cardiomyopathy (RCM), arrhythmogenic right ventricular cardiomyopathy (ARVC), and unclassified cardiomyopathies.

Myocarditis is an inflammation of the myocardial tissue and may need a catheterization for endomyocardial biopsy (see Chap. 44).

Table 54.1 Clues to differentiate constrictive pericarditis from restrictive cardiomyopathy

Restrictive cardiomyopathy		Constrictive pericarditis
	ECHO	
LVH, RVH	Ventricle morphology	Normal ventricles
Typical, biatrial	Atrial enlargement	Possible LA enlargement
Normal systolic	Ventricle function	Inspiratory septal bounce
Low tissue velocity	Tissue Doppler	Normal velocity (TDI mitral lateral wall >8 cm/s)
Normal	Doppler LV inflow velocity	Fall by 25% with inspiration
	CATH	
Absent	Paradoxical pulse	Present (1/3 of the cases)
LV at least 3–5 mmHg>RV	RVEDP and LVEDP	Equal
Common	LVEDP >25 mmHg	Rare
Variable	Square root sign	Present
Common	PA systolic pressure >60 mmHg	Absent
Normal	Inspiratory variation	Exaggerated

LVH left ventricle hypertrophy, *RVH* right ventricle hypertrophy, *TDI* tissue Doppler imaging, *LVEDP* left ventricle end-diastolic pressure, *RVEDP* right ventricle end-diastolic pressure, *LA* left atrial, *LV* left ventricular

Thanks to the improvements in noninvasive diagnostic techniques (i.e., MRI, CT), cardiac catheterization is usually only required to:

1. Exclude secondary etiology (i.e., ischemic disease in DCM, possible viral etiology, ALCAPA)
2. Pre-transplant assessment

In this section, we will describe the *DCM and HCM hemodynamic findings*. RCM has been discussed above and compared with constrictive disease.

54.3.1 Pre-catheterization Study

- History (family history, other comorbidities)
- Symptoms onset (in utero, after respiratory/gastrointestinal infections, after chemotherapy treatment, other)
- NYHA functional class
- Clinical examination focused on heart failure (HF) signs
- Medical therapy
- ECG (check for sinus rhythm, atrial enlargement signs, AV conduction, QRS morphology, and repolarization anomalies)
- Check laboratory analysis (positive C-reactive protein, high white blood count) and abnormalities of hemoglobin, electrolytes, and blood glucose levels

54.3.2 Catheterization Laboratory

For accurate assessment of pressures and resistances, the catheter study should ideally be performed using local anesthesia or sedation.

Vascular access: Femoral vein (or jugular if an endomyocardial biopsy is also to be performed) and femoral artery.

Materials:

- Introducer size appropriate to patient weight:
- Babies (weight between 2 and 15 kg): femoral artery (3/4 Fr); femoral vein (5 Fr)
- Children (weight between 15 and 40 kg): femoral artery (4 Fr); femoral vein (6 Fr)
- Children and adolescents (weight >40 kg): femoral artery (5 Fr); femoral vein (6 Fr)

Catheters: Balloon-tipped wedge catheter (Swan–Ganz), pigtail, Judkins right and left. Guidewires according to catheter size: a standard guidewire, Nitinol hydrophilic guidewire, and thermodilution (*e.g.,* Vigilance®) gas analysis machines to calculate cardiac output.

Two pressure transducers in order to measure left- and right-sided pressures simultaneously.

Procedure: Right- and left-heart catheterization are performed. Ensure *zeroing is accurate*; otherwise all the pressure parameters will be unreliable.

Focus on mean pulmonary pressure, wedge capillary pressure, and *LV end-diastolic pressure* (LVEDP) simultaneously evaluated with wedge pressure.

Pull back with an end-hole catheter from the LV to descending aorta to exclude aortic valve stenosis and/or coarctation if MRI or CT not available.

Routine measurement of oxygen saturation in blood sample taken from superior vena cava and pulmonary artery to detect unsuspected shunts [2].

54.3.3 Specific Hemodynamic Findings in DCM

Left and right ventricular filling: In symptomatic patients, they are usually elevated. However, it is possible that ventricular filling pressure is normal at rest especially in those patients who have been treated intensively with diuretics. A stress test (supine bicycle or chronotropic drug/volume challenge) may outline a rapid increase in atrial pressure [2].

Cardiac output (CO): It is generally depressed but might be normal or slightly abnormal in milder cases.

Beware that in the pediatric population, the normal range of CO may differ consistently.

Therefore, it is preferred to use the cardiac index (CI = CO/BSA) as a functional parameter. Normal CI ranges are:

- In neonates = 4–5 l/min/m^2.
- In children = 3–4.5 l/min/m^2.
- In adolescents and young adults = 2.5–4 l/min/m^2 [3].

Left ventricular pressure waveform: The rate of both rise and fall of left ventricular pressures are slow. The ventricular pressure

wave has a *triangular appearance.* This deformity accounts for the brief duration of systolic ejection and this is due to a reduction of left ventricular isovolumic pressure (d*F*/d*t*) [2].

Elevation of early diastolic (protodiastolic) pressure: In a normal heart, early diastolic (protodiastolic) pressure in the ventricles is = 0 mmHg. In extreme situations (e.g., hypovolemic or adrenergic states), protodiastolic pressures may become negative (*diastolic suction*). Especially in a severely depressed heart, early diastole which reflects heart relaxation is slow and incomplete so that the early diastolic pressure is always above zero [2].

Pulmonary vascular resistance calculation: Augmented mean pulmonary pressure is a common finding such as increased pulmonary vascular resistance (defined as TPG/cardiac output that exceeds 2.5–3 UW m^2). Pulmonary hypertension in DCM (PH) is usually "post-capillary," characterized by an elevated PCWP (>15 mmHg). Initially, in PH associated with left-sided heart failure, the transpulmonary gradient is normal, though over time it increases (>10 mmHg). The hemodynamic progression of PH is typically characterized by a progressive rise in transpulmonary gradient and PVR over time. A vasodilatation test is normally recommended when the transpulmonary gradient exceeds 12 mmHg. However, caution is advised in patients with very high left atrial pressure [2] (for vasoreactive tests, see Chap. 9).

54.3.4 Specific Hemodynamic Findings in HCM

In pediatric populations, HCM can be present in a wide variety of cardiac diseases [3]. In familial cases, it is usually transmitted as an autosomal dominant trait. It can affect both ventricles but more often affects only the LV. From the functional point of view, there are obstructive (HOCM) or nonobstructive forms. Therefore, it is possible to assess an *intraventricular gradient* (usually midventricular or at the left ventricle outflow tract level).

Left ventricular end-diastolic pressures: The left ventricular end-diastolic pressure can be in the normal range but is usually high due to a reduction in left ventricular distensibility (stiffness of

the thick wall, decreased rate, and extent of myocardial relaxation) [2]. This finding is particularly obvious if there is mitral regurgitation (systolic anterior motion of the posterior mitral leaflet).

Cardiac output: In general, it is normal or increased.

Intraventricular pressure gradient: Most patients with HCM do not have a systolic pressure gradient at rest. This finding is highlighted by exercise and Valsalva maneuvers. The presence at rest of a systolic gradient is typical of asymmetric hypertrophic cardiomyopathy (hypertrophy of the interventricular septum at the outflow level).

The *Brockenbrough–Braunwald sign*: It is a typical spike and dome configuration in the arterial pressure waveform following an extrasystolic beat. The decrease in pulse pressure after a premature ventricular contraction is due to reduced stroke volume caused by increased dynamic obstruction, which is due, in turn, to a post-extrasystolic potentiation beat. The presence of this sign in HOCM is usually indicative of worsening of obstruction [2].

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55 Imaging and Treating Coronary Arteries in Children (2019 Revised)

Teiji Akagi

55.1 Normal Coronary Artery Anatomy

The right and left coronary arteries originate from the right and left sinuses of Valsalva of the aortic root, respectively. The posterior sinus rarely gives rise to a coronary artery and is referred to as the "noncoronary sinus." The locations of the sinuses are anatomic misnomers: The right sinus is actually anterior in location and the left sinus is posterior. The myocardial distribution of the coronary arteries is somewhat variable, but the right coronary artery (RCA) almost always supplies the right ventricle (RV), and the left coronary artery (LCA) supplies the anterior portion of the ventricular septum and anterior wall of the left ventricle (LV).

The RCA arises from the right coronary sinus somewhat inferior to the origin of the LCA. After its origin from the aorta, the RCA passes to the right of and posterior to the pulmonary artery and then emerges from under the right atrial appendage to travel in the anterior (right) atrioventricular (AV) groove. In about half of the cases, the conus branch is the first branch of the RCA. In the

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other half, the conus branch has an origin that is separate from the aorta. The conus branch always courses anteriorly to supply the pulmonary outflow tract. Occasionally, the conus branch can be a branch of the LCA, have a common origin with the RCA, or have dual or multiple branches.

In 55% of cases, the sinoatrial nodal artery is the next branch of the RCA, arising within a few millimeters of the RCA origin. In the remaining 45% of cases, the sinoatrial nodal artery arises from the proximal left circumflex (LCx) artery. In either case, the sinoatrial nodal artery always courses towards the superior vena cava inflow near the cephalad aspect of the interatrial septum. As the RCA travels within the anterior AV groove, it courses downward towards the posterior (inferior) interventricular septum. As it does this, the RCA gives off branches that supply the RV myocardium; these branches are called "RV marginals" or "acute marginals." They supply the RV anterior wall. After it gives off the RV marginals, the RCA continues around the perimeter of the right heart in the anterior AV groove and courses towards the diaphragmatic aspect of the heart.

The LCA normally emerges from the left coronary sinus as the left main coronary artery. The left main coronary artery is short, passes to the left of and posterior to the pulmonary trunk, and bifurcates into the left anterior descending (LAD) and LCx arteries. Occasionally, the left main coronary artery trifurcates into the LAD artery, the LCx artery, and the ramus intermedius artery.

The LAD artery runs in the anterior interventricular sulcus along the ventricular septum. Commonly, the LAD artery may be embedded within the anterior myocardium forming an overlying myocardial bridge. Myocardial bridging is seen more often on CT than described in the coronary angiography literature. Most myocardial bridges are asymptomatic, although rarely myocardial bridging can be associated with ischemia. The LAD artery has branches called "septal perforators" that supply the anterior ventricular septum. It also has diagonal arteries that course over and supply the anterior wall of the LV. The diagonals and septal perforators are numbered sequentially from proximal to distal.

The LCx artery runs in the posterior AV groove analogous to the course of the RCA on the opposite side. The major branches

of the LCx artery consist of obtuse marginals. Obtuse marginals branches supply the lateral wall of the LV. They are numbered sequentially from proximal to distal.

55.2 Angiographic Projections in Normal Coronary Artery

During coronary angiography, the heart is viewed in a variety of projections, each of which is a two-dimensional representation of a three-dimensional structure. A given coronary artery thus appears to "rotate" and change its position relative to other structures. This change, together with the fact that the heart continues to beat during the contrast injection, makes identification of the coronary difficult, at first. Moreover, the contraction of the left ventricle occurs apex to base with rotation of the lateral wall. One must therefore acquire the skill of reconstituting the three-dimensional anatomy of the coronary vessels from a series of different, two-dimensional views. Conventionally, the orientation of the X-ray tube with respect to the patient is described using two angles, each of which may be positive or negative. The first angle refers to "rotation." It describes the position of the image intensifier around the longitudinal axis of the patient. Zero degrees is vertically above the patient, positive angles are towards the patient's left and negative angles are towards the patient's right. The second angle refers to "angulation." It describes the position of the image intensifier in the short axis of the patient. Zero degrees is directly above the patient's head, positive angles are towards the patient's head, and negative angles are towards the patient's legs.

3D coronary artery imaging using the latest computed tomography (CT) and magnetic resonance imaging has excellent spatial resolution and is widely used to diagnose coronary artery anatomy instead of conventional coronary angiography [1, 2]. However, high heart rate and respiratory chest motion in pediatric population may influence on decreasing imaging quality for coronary imaging. Appropriate sedation and oral beta blocker administration support for improvement of imaging quality.

55.3 Kawasaki Disease

55.3.1 Introduction

Kawasaki disease is an acute, self-limited vasculitis of unknown etiology that occurs predominantly in infants and young children. This vasculitis frequently affects on small- to mid-size arteries especially coronary arteries. In Japan, nationwide surveys have been conducted every 2 years since 1970 and >200,000 patients have been registered. Although no nationwide outbreak has been observed since the outbreak in 1986, the incidence rate has gradually increased over the past 20 years. The disease has been reported in all over the world. Currently, the incidence of coronary artery abnormality is about 3–5% even in the appropriate high dose intravenous immunoglobulin treatment.

The most striking feature of coronary artery abnormality in Kawasaki disease is the change of size or shape of aneurysm. About 50% of coronary aneurysms regress within 2 years. On the other hand, coronary artery stenosis occurred in 4% of all patients, or in 20% with coronary aneurysms in the follow-up period. In general, no coronary artery stenosis has developed in patients with regressed aneurysms. Thrombotic occlusion may be seen relatively soon after onset in medium-sized or larger aneurysms. Occlusion is seen at arteriography in 16% of cases of coronary artery injury during follow-up, and in 78% of these cases occlusion is confirmed at arteriography less than 2 years after onset. While sudden death may be one outcome, asymptomatic occlusion accounts for about 2/3 of cases of occlusion seen at follow-up coronary arteriography. In the majority of cases, improvement of myocardial ischemic findings is seen as a consequence of post-occlusion recanalization and the development of collateral circulation routes.

However, patients whose original aneurysmal size is larger than 4 mm, revealed thickened intima and media. Pathological or

pharmacological studies on vascular function suggested that coronary artery lesion may be long-term coronary risk factors, even in the regressed aneurysms, and other conditions [3].

55.3.2 Procedure

During past decades, the clinical experience of catheter interventional treatment in Kawasaki disease has been gradually increasing. These are including balloon angioplasty, stent implantation, rotational ablation, and transluminal coronary revascularization. However, the experiences in Kawasaki disease are still limited compared to coronary intervention in adults, which provided satisfactory therapeutic results. The coronary artery stenosis in Kawasaki disease commonly involves severe calcification, in contrast with adult coronary artery lesions, which consist primarily of atherosclerosis. Therefore, the indication of catheter intervention for adult patients cannot be directly employed in Kawasaki disease patients, mostly in pediatric population.

It is clear that coronary artery bypass surgery is the fundamental therapeutic option for ischemic heart disease after Kawasaki disease. However, even in internal thoracic artery bypass surgery, the long-term coronary graft patency rate is not perfect, especially when the patient is younger than 12 years old at the time of surgery (Fig. 55.1).

Balloon angioplasty is effective in many situations, particularly in patients without severe calcification or in patients with a relatively short interval (within 6 years) between the onset of the disease and the intervention. Based on recent improvements in balloon catheters, this procedure may be used even in small children.

Therefore, balloon angioplasty may become a first-line procedure in younger children with significant coronary artery stenosis.

Stent implantation is preferable, because it may prevent new aneurysm formation and restenosis. If patients have severe calcified coronary stenosis, percutaneous coronary rotational ablation

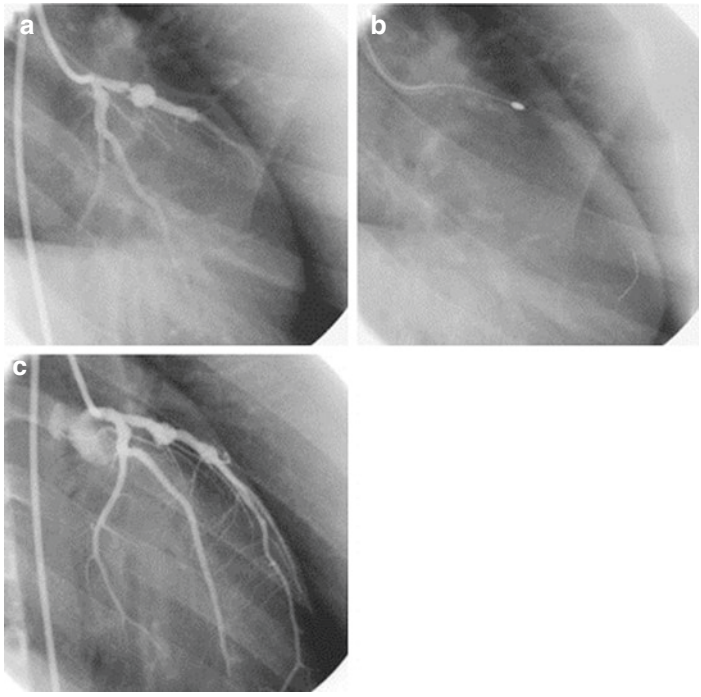

Fig. 55.1 Coronary rotational ablation in Kawasaki disease. (**a**) before the intervention, (**b**) rotational ablation, (**c**) post-rotational ablation. Coronary stenosis is completely resolved

(PTCRA) may be the only effective treatment. Excellent acute results for PTCRA were observed in previous studies. Although use of this procedure is still limited, PTCRA may be the most appropriate catheter intervention for Kawasaki disease. The advantage of PTCRA is the high success rate, even in patients with calcified coronary artery stenosis. The limitation of this procedure is the need for larger arterial access for the metal burr (Fig. 55.2).

For this reason, this procedure can only be performed in older patients. Anti-coagulation and anti-platelet medication should be continued for their life long. Intravascular ultrasound imaging

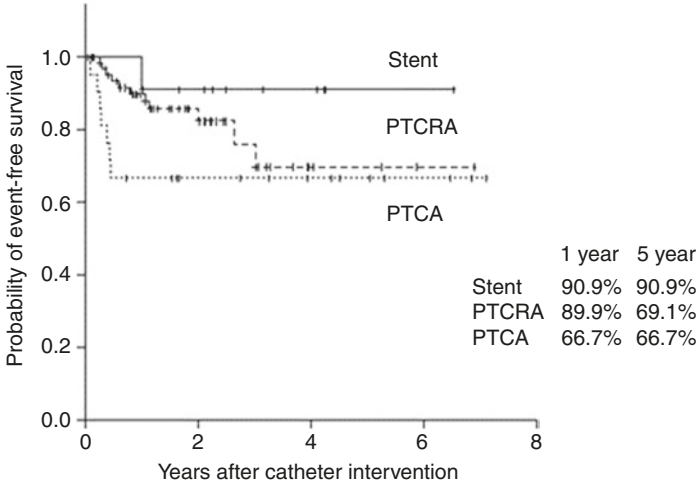

Fig. 55.2 Long-term outcome after catheter intervention in Kawasaki disease (PTCA: percutaneous coronary angioplasty, PTCRA: percutaneous coronary rotational ablation)

provides valuable information for the selection of the appropriate interventional procedure and early detection of vascular complications.

55.3.3 Other Special Conditions

- Tetralogy of Fallot

  The most frequent abnormality seen in coronary artery branching in tetralogy of Fallot is the presence of a coronary artery crossing the right ventricular outflow tract. This can be a left anterior descending coronary artery from the right coronary artery with an anterior course [4].
- Post-arterial switch evaluation

  First of all it is mandatory to know the spectrum of coronary artery variations seen in d-TGA (Fig. 55.3) and the surgical report.

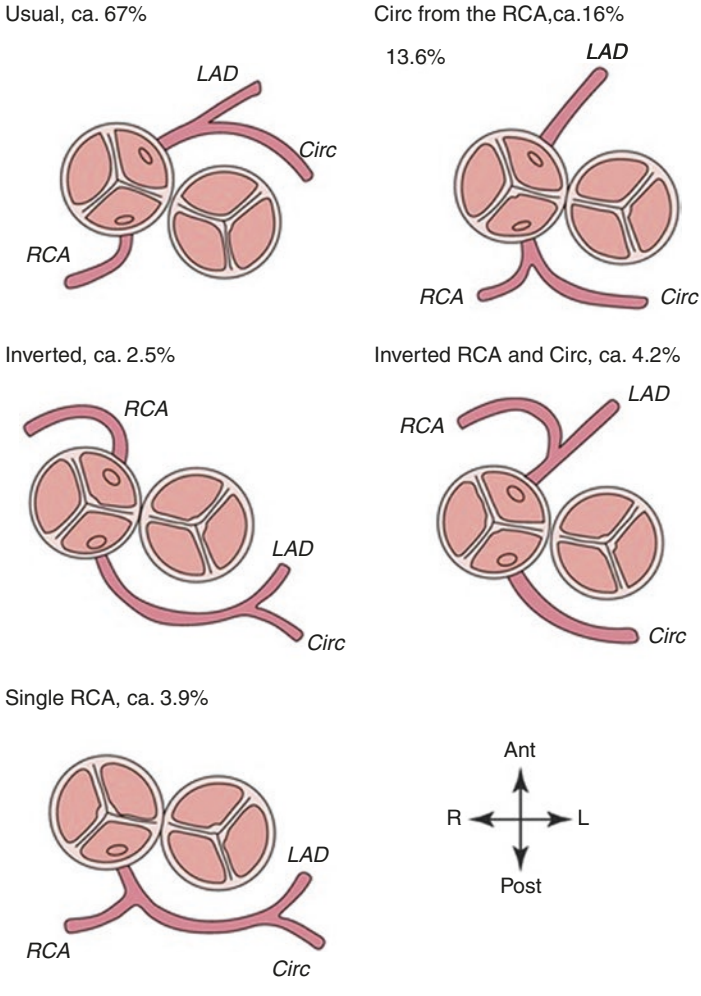

Fig. 55.3 Coronary artery anatomy in d-TGA. (From Wernovsky and Sanders [2])

Secondly usually the coronary arteries are re-implanted anteriorly on the wall of the neo-aorta. Therefore, they are better looked after in the lateral view by using a right judkins or amplatzer coronary artery catheter.

It is important to show a clear reflow of contrast towards the aorta in order to clearly delineate the surgically created ostium.

- Pulmonary atresia and intact ventricular septum

  See Chap. 19.

- ALCAPA (Anomalous connection of the left coronary artery from the pulmonary artery)

  Selective angiography of the normally connected right coronary artery usually shows a dilated right artery while the left is vascularized retrogradely. Finally a flow of contrast in the pulmonary artery can be clearly seen.

  Usually, the connection to the pulmonary artery is made at the part facing the PA sinus, but it may also be made at the level of the main pulmonary artery or one of the two proximal branches.

- Coronary artery disease post-transplantation

  It consists in a concentrical myointimal proliferation involving the entire vessel.

- Possible complications

  Air embolism

  Usually this is a self-limiting problem. However, if hemodynamic abnormalities of arrhythmias occur they have to be treated aggressively. The patient should be sedated, receive drugs for pain relief and receive oxygen 100%.

  Coronary artery spasm

  It can occur because of a catheter tip advanced too far into the vessel. If this occurs, the catheter should be retrieved. Usually it resolves quickly. If it does not occur, infusion of nitroglycerin has to be given.

  Coronary artery dissection

  It is caused by the forced injection of contrast media into the coronary arteries, especially into the smaller branches.

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56 Transcatheter Treatments in PA-VSD-MAPCAs

Diego Porras

56.1 Introduction

Angiography and transcatheter interventions play an integral role in the management of tetralogy of Fallot with pulmonary atresia and major aorto-pulmonary collaterals (TOF/PA/MAPCAs). This role starts with a complete diagnostic investigation that delineates the anatomy of each source of pulmonary blood flow, as well as the vascular health of each segment of lung. Transcatheter interventions are also used to eliminate unnecessary or potentially harmful sources of pulmonary blood flow, to maximize the health of the distal vasculature, and to optimize the anatomy after surgical repair has been completed. In addition, the majority of these patients will have a right ventricle-to-pulmonary artery conduit as part of their surgical repair, and this will require transcatheter interventions to treat conduit dysfunction in most patients. In this chapter, we will focus on the role of cardiac catheterization in the

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diagnostic evaluation of TOF/PA/MAPCAs and the role of transcatheter interventions as part of an integrated strategy to manage this complicated disease.

56.2 The Role of Cardiac Catheterization and Angiography in the Diagnostic Evaluation of TOF/PA/MAPCAs

The main goal of the management of a patient with TOF/PA/MAPCAs is to establish cardiopulmonary physiology that is as close to normal as possible. This, in general terms, is defined as:

1. Two-ventricular circulation
2. Normal systemic oxygen saturations
3. No residual intracardiac shunts
4. Antegrade flow to all lung segments
5. Normal or near-normal pulmonary artery and right ventricular pressures

Therefore, it is of paramount importance to incorporate as much pulmonary vascular surface area to the repair as possible and to do this in a timely fashion in order to avoid the development of vascular disease [1]. In order to appropriately plan the best-suited surgical approach for a particular patient, the goals of the initial diagnostic evaluation are to:

1. *Characterize the sources of pulmonary blood flow*: By definition, patients with TOF/PA/MAPCAs have several sources of pulmonary blood flow. In general, major aorto-pulmonary collaterals (MAPCAs) are enlarged bronchial arteries that preserve primitive connections to the native pulmonary arteries (PAs) [2]. It is important to delineate the anatomy of each of these sources of pulmonary blood flow, including their anatomic relationship to other structures in the chest, in an effort to provide intraoperative guidance. It is especially useful for the surgeons if the anatomy of each source of pulmonary blood

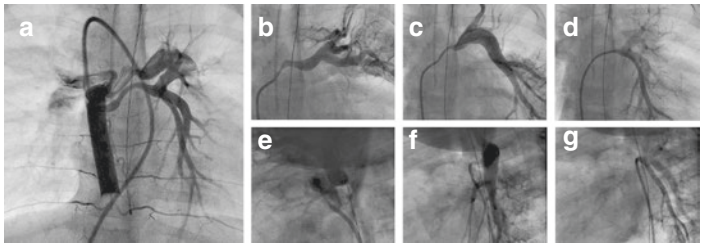
flow can be delineated in relation to other anatomic structures that are easily identifiable in the operating room, like the pulmonary veins, the carina, the main stem bronchi, etc. Therefore, the description of an APC should include:

(a) Origin: from what vessel does it originate (descending thoracic aorta, subclavian artery, internal mammary artery, thyrocervical trunk) and from what part of that vessel (anterior/posterior, left/right, proximal/distal). In the case of the aorta, it is important to specify if it originates from the underside of the distal aortic arch, in which case the vessel may be a patent ductus arteriosus (PDA) instead of a MAPCA and its patency may respond to intravenous infusion of prostaglandin E. It is important to remember that there may be bilateral PDAs and that at least one PDA can originate from the innominate artery. When it is unclear if the vessel being characterized is a PDA or not, but its origin is anatomically consistent with a PDA, it can be referred to as a "*duct-like collateral.*"

Since the majority of true APCs are enlarged bronchial arteries, their origins and trajectories follow certain predictable patterns. The majority arise from the descending thoracic aorta, near the carina and the main stem bronchus ipsilateral to the arch sidedness. However, they can also originate from the innominate artery, the subclavian arteries and their branches, and even from the abdominal aorta and its branches [2].

(b) Level of origin: If it originates from the descending thoracic aorta, at what level does it originate and close to what structures (Fig. 56.1). Describing the level of thoracic vertebra at which the APC arises is of limited utility, since this is not a landmark that can be identified in the operating room.

(c) Trajectory (Fig. 56.1): This includes the detailed description of the course of each collateral, especially in relation to other anatomic structures.



(d) Branching: Many APCs give off several branches (Fig. 56.3f). It is important to describe this in detail, so that the surgeon can be aware of where these branches originate and whether each one needs to be unifocalized separately.
(e) Segments supplied by each APC, as detailed below and in Figs. 56.1, 56.2, 56.3, 56.4, 56.5, and 56.6.

2. *Establish the presence or absence of central mediastinal pulmonary arteries* (Figs. 56.2, 56.3, and 56.4): Central mediastinal PAs, even when diminutive, have enormous growth potential once exposed to higher pressure and flow. Therefore, whenever possible, it is important to incorporate them into the repair as early as possible. In many cases, the central mediastinal PAs may not be readily identifiable, and certain techniques are required to establish their presence. These include selective injections into APCs (Figs. 56.1, 56.2, and 56.3) and pulmonary venous wedge injections (Figs. 56.4 and 56.6). It is also important to delineate the anatomy of the central mediastinal PAs if they are present, including their size, presence of discrete stenoses and/or atretic segments (continuity vs. lack of continuity) (Figs. 56.4 and 56.6).
3. *Characterize how each segment of each lung is supplied and which segments have single vs. dual blood supply* (Fig. 56.5): Each segment of each lung should be accounted for. Some segments may not have direct flow at the time of the catheterization and will therefore not fill from APC injections or aortograms because the APCs supplying them have become

Fig. 56.1 Defining major aorto-pulmonary collaterals. (**a**) Balloon occlusion aortogram: a Berman catheter is advanced from the femoral vein, through the RA, RV into the aorta and around the aortic arch into the descending thoracic aorta, positioned behind the cardiac silhouette. The balloon, which is distal to the angiographic holes in the catheter, is inflated to occlude the distal thoracic aorta and a power injection of approximately 1 cc/kg is given at a rate of 1 cc/kg/s. The resulting angiogram gives adequate opacification of the collaterals proximal to the balloon, and avoids run-off of contrast to undesired parts of the aorta. In this example, three different major collaterals that supply the left lung are seen arising from the thoracic aorta, below the level of the carina. There are also two collaterals going towards the right lung. (**b–g**) Detailed delineation of the vessels supplying the left lung using selective injections into each collateral: in order to delineate the anatomy of each collateral, including which segments of the left lung each supplies and whether these segments have dual supply, selective injections in straight frontal (**b**, **d**, **f**) and lateral (**c**, **e**, **g**) projections are necessary. (**b** and **c**) show that the collateral that arises from the anterior aspect of the thoracic aorta, below the carina, travels in front of the left main stem bronchus, then over it. The relationship of this vessel to the left upper pulmonary vein can also be seen in this angiogram, since the vein is lightly opacified in the levophase of the injection. The relationship of each collateral to these structures (carina, main stem bronchus, pulmonary veins), which are easily identifiable for the surgeon, are important in the description of the anatomy of these collaterals. The anatomy of this particular collateral suggests that this collateral supplies the native proximal LPA (it travels from anterior to posterior and right to left as it goes over the left main stem bronchus). However, it only supplies the anterior segment of the left upper lobe and the superior lingula. (**d** and **e**) show a collateral that arises at the same level of the descending thoracic aorta, but from the lateral aspect. It travels under the left main stem bronchus and connects to what appears to be part of the native left lower lobe pulmonary, supplying some of the basal segments of the left lower lobe (lateral, medial, and anterior basal left lower lobe segments). It also gives a branch proximally, that supplies the apical left upper lobe and one that supplies the inferior lingula. (**f** and **g**) show injection in a collateral that arises lower in the thoracic aorta, from its posterior and leftward aspect and supplies the posterior and medial basal left lower lobe segmental branches, as well as the superior segment of the left lower lobe. With this kind of detailed analysis, we can account for the supply to each segment of the left lung and we can also say that all of these three collaterals are the single source of pulmonary blood flow for their corresponding segments. Therefore, unifocalization of all of them is paramount in preserving the maximum amount of lung vasculature possible at the time of repair. A similar delineation is carried out for the collaterals supplying the right lung (not shown)

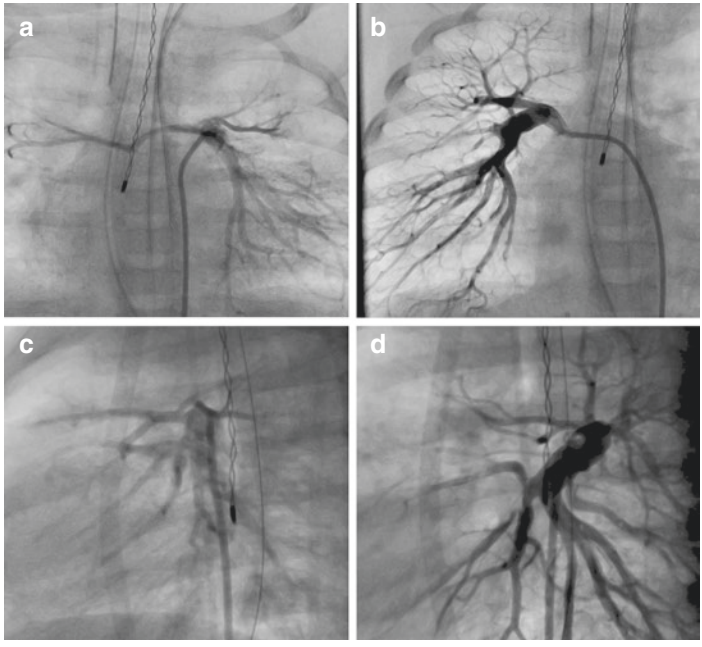


Fig. 56.2 Determination of the presence of native mediastinal pulmonary arteries. (**a** and **b**) show the frontal and lateral projections of a selective injection into a collateral that supplies the entire left lung vasculature. The native mediastinal LPA is seen filling retrograde, then connecting to the MPA and RPA. These mediastinal branch pulmonary arteries are in continuity and are quite small, measuring approximately 1.5 mm in diameter. It is also evident in A that the mediastinal RPA only supplies a few segments of the right lung, at least under the present conditions. Further exploration is necessary to determine the sources of pulmonary blood flow to the rest of the right lung. (**c** and **d**) show a selective injection into a collateral that arises from the rightward aspect of the descending thoracic aorta, under the carina. This collateral supplies flow to the majority of the right lung vasculature. Therefore, in this case, this vessel must be unifocalized to the native branch pulmonary arteries in order to preserve flow to the majority of the right lung

atretic. Pulmonary venous wedge injections are useful in identifying these segments, by forcefully filling them with contrast in retrograde fashion (Fig. 56.6). Once identified, their anatomy, especially in relation to other pulmonary arteries and

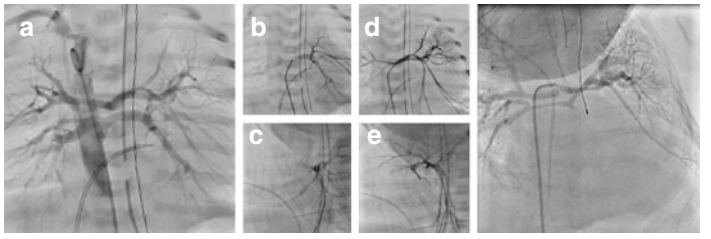


Fig. 56.3 How do you define native mediastinal branch pulmonary arteries? (**a**) AP projection of a descending thoracic aortogram showing at least three vessels that could potentially be the native mediastinal branch pulmonary arteries. (**b** and **c**) show selective injection in the lowest collateral seen in (**a**). The lateral projection (**c**) shows that the vessel that goes leftward stays posterior and inferior to the left main stem bronchus. This vessel supplies the superior, lateral, and medial segments of the left lower lobe, but is not connected to the central mediastinal pulmonary arteries. (**d** and **e**) show injection into the collateral that arises from the anterior aspect of the descending thoracic aorta, immediately below the carina. This vessel provides flow to the rest of the left lower lobe, the lingula, and the entire left upper lobe. On the lateral it can be seen that it also connects to what may be a mediastinal LPA, because it is seen traveling anteriorly over the left main stem bronchus and going across the midline to the mediastinal RPA. This is confirmed in F, with cranial angulation. This angulation makes it clear that this collateral supplies the mediastinal LPA which is in continuity with the MPA and the mediastinal RPA. The retrograde filling of the LPA to the MPA and then towards the RPA forms the classic "seagull" appearance in the cranial angulation, which is characteristic of mediastinal pulmonary arteries that are in continuity. In this particular case, the same collateral bifurcates early into a branch that goes rightward and is the sole supply to the majority of the right upper lobe, without connecting directly with the native RPA

APCs should be clearly delineated since they will need to be found and unifocalized in future surgeries. Once each segment has been identified, it is important to determine how each segment is supplied:

(a) *Single supply*: only supplied by a single APC and does not connect in any way to the rest of the native pulmonary arterial tree. Once the APC is interrupted, no other blood flow will go to this segment unless it is surgically unifocalized.

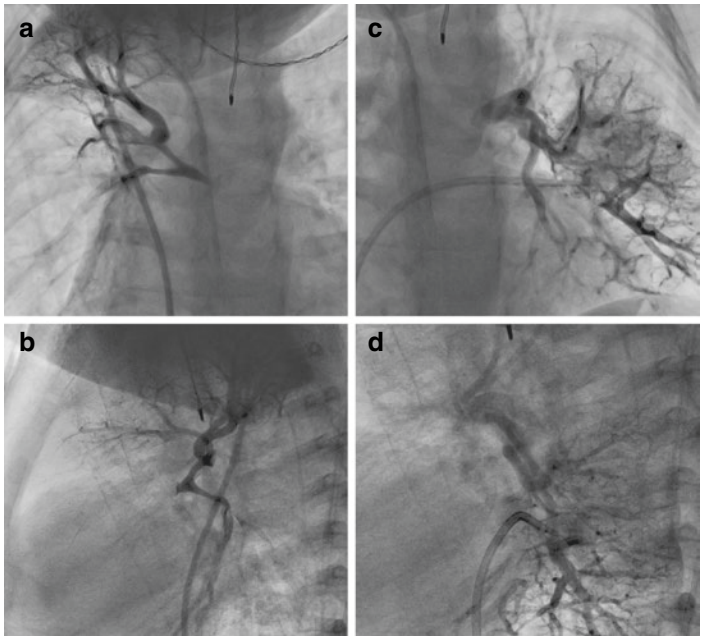


Fig. 56.4 Present, but discontinuous native mediastinal pulmonary arteries. (**a** and **b**) show the frontal and lateral projections of a pulmonary venous wedge injection in the right upper lobe. The right upper pulmonary artery is seen filling and connecting to the native mediastinal RPA which connects to the rest of the right pulmonary artery branches and also fills the proximal native mediastinal RPA to a diminutive MPA and a miniscule LPA that ends blindly almost immediately. When a similar injection is carried out in the left lower lobe (shown in **c** and **d**), the native LPA is seen filling retrograde, traversing over the left main stem bronchus and then ending blindly. This patient therefore has mediastinal pulmonary arteries, but they are not in continuity because the native mediastinal LPA is interrupted between its proximal portion, just after the take-off from the MPA, and the hilum of the left lung

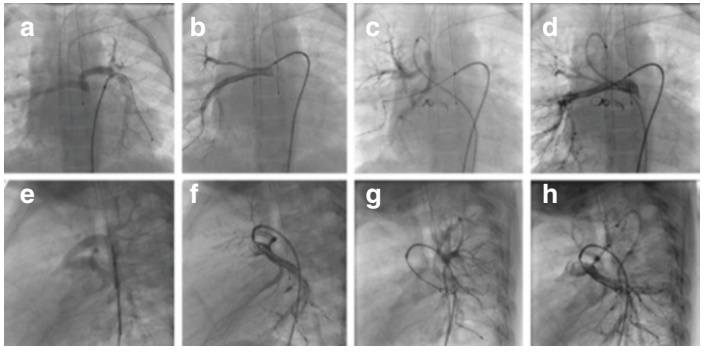


Fig. 56.5 Delineation of dual vs. single supply. Top panels (**a–d**) show frontal views of selective angiograms. Bottom panels (**e–h**) show the corresponding lateral view for each injection. (**a**, **e**) Collateral from the leftward aspect of the descending thoracic aorta connects to the native left lower lobe pulmonary artery, which fills the mediastinal pulmonary arteries in retrograde fashion. On the lateral projection (**e**), it is clear that the vessels that cross the midline are the mediastinal PAs (LPA, MPA, RPA) because they travel anteriorly and form the classic seagull appearance. (**b**, **f**) The long sheath has been advanced to the mediastinal RPA and injection shows that this vessel supplies the anterior and apical segments of the right upper lobe, the medial segment of the right middle lobe, and the medial and anterior segments of the right lower lobe. It may also supply the lateral and posterior segments of the right lower lobe. However, these segments appear to "washout" with non-contrasted blood, suggesting that they have dual supply from another collateral. (**c**, **g**) Collateral from the right subclavian artery supplies the posterior right upper lobe and gives another branch that supplies the superior segment of the right lower lobe, as well as the posterior and the lateral segments of the right lower lobe. (**d**, **h**) Angiogram in native RPA while the collateral from the right subclavian artery is balloon occluded confirms that the lateral and posterior segments of the right lower lobe are supplied both by the native mediastinal RPA and by the collateral from the subclavian artery, since now these segments opacify fully without washout (compare to **b**, **f**). This is important for surgical planning, because it means that the branch of the collateral from the subclavian that supplies the posterior right upper lobe must be unifocalized to the native RPA because it is its only direct supply. However, the branch that supplies the lateral and posterior segments of the right lower lobe can be ligated

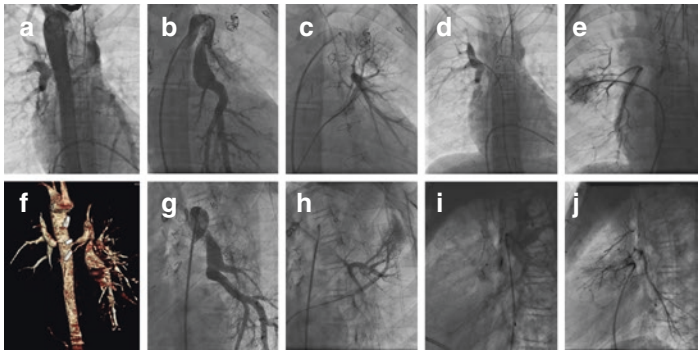


Fig. 56.6 Pulmonary venous wedge injections to delineate the anatomy of vessels that don't have antegrade flow. (**a**) Anteroposterior projection of aortogram in a patient with TOF/PA/MAPCAs, showing two large APCs from the descending thoracic aorta, one to each lung. F shows the same information from a three-dimensional reconstruction from a CTA. In both images there appear to be entire segments of both lungs that are not perfused. (**b**, **g**) Injection into the collateral to the left lung shows that it provides flow to medial and posterior segments of the left lower lobe, but no flow to the upper lobe or any anterior segments of the left lung. (**c**, **h**) AP and lateral projections of a pulmonary venous wedge injection performed in a branch of the left upper pulmonary vein. Contrast is seen filling the superior segment of the left lower lobe in retrograde fashion. It also fills the lingular branches, all of which are missing segments in the injection of the collateral to the left lung (**b**, **g**). (**d**, **i**) AP and lateral projections of selective injection into collateral to the right lung showing that this collateral supplies apical and posterior right upper lobe, as well as the superior segment of the right lower lobe. (**e**, **j**) AP and lateral projections of pulmonary venous wedge injection into a branch of the right upper pulmonary vein, filling the right intermediate pulmonary artery, which is connected to part of the right lower and middle lobes as well as anterior right upper lobe. Putting all of this information together, it is evident that the vessels that are not receiving antegrade flow will have to be unifocalized to the vessels fed by the large collaterals in order to have sufficient pulmonary vasculature to be able to complete a full repair in the future

(b) *Dual supply*: These are segments that are supplied by the central mediastinal PAs, which at the time of the catheterization are supplied by other collaterals. In addition, these segments are supplied by one or more separate collaterals, meaning that once the collaterals feeding the

central mediastinal PAs are interrupted and the central mediastinal PAs are given a new source of blood flow (RV-PA conduit or shunt), these segments will continue to receive flow from other APCs. This flow will, therefore, be redundant and competitive, which can lead to hypertension and lack of growth of the proximal vessel. Therefore, sources of dual supply should be eliminated before the surgery.

(c) *Potential dual supply*: These are segments that are supplied by native pulmonary arteries, which are in turn supplied by a MAPCA. This means that once the central mediastinal PAs are given a new source of pulmonary blood flow (shunt or RV-PA conduit) the MAPCA will provide dual supply, which can be detrimental. These APCs cannot be interrupted before surgery, because at that time they are the only source of pulmonary blood flow to those segments. Therefore, they should be ligated during surgery or, if surgical ligation is not possible because of the anatomic location of the collateral, then interrupted using transcatheter techniques shortly after surgery.

4. *Delineate the anatomy of each lobar, segmental, and subsegmental pulmonary artery branch, including size of the vessels in the lung parenchyma and the presence or absence of discrete stenoses in the peripheral branches*: Some of these vessels can be highly abnormal and can have discrete stenoses that will need to be addressed surgically or using transcatheter techniques in the future. Knowing this in advance will help guide decisions related to the type of surgical approach taken, as well as decisions regarding closure of the ventricular septal defect.
5. *Characterize the health of the vascular bed for each segment of lung*: Because each segment may be supplied by different vessels under different conditions, the health of each segment can be highly variable. Ideally, the health of each segment would be determined by understanding the resistance within each segment. Practically, this is not possible, because in order to know the resistance, one would have to know not only the

pressure drop across the segment (which can often can be determined), but also the exact amount of flow going through the segment (which is impossible to determine using currently available techniques). Therefore, this is usually a very subjective assessment and it is based on the angiographic appearance of the vascular bed, the subjective assessment of flow through that particular vascular bed, and the pressure in that segmental artery. Some of the angiographic findings consistent with pulmonary hypertension and vascular disease include [3]:

(a) Paucity of distal vessels: described as a tree in winter, as opposed as a tree in summer which would be the appearance of a healthy pulmonary vascular bed.
(b) Tortuosity of the branches.
(c) Pulsatility of distal branches.
(d) Rapid tapering of distal branches: Healthy vascular beds are characterized by smooth, subtle tapering of the vessel size as it goes from proximal to distal. Diseased pulmonary vascular beds are characterized by the proximal vessels being somewhat dilated and then rapidly becoming narrow (rapid taper).
(e) Decreased intensity of background capillary haze.
(f) High mean pressures in the distal vasculature: The pressure depends on the amount of flow going into that particular vascular bed and the resistance through the vascular bed. Ideally, segments should have mean pressures under 20 mmHg. Segments that have mean pressures above 30 mmHg have a high likelihood of having rapidly progressive vascular disease. However, it is important to keep in mind that this is entirely dependent on the amount of flow. For example, a segment with a mean pressure of 25 mmHg may be quite healthy if it is receiving a very large amount of flow and it is still able to accommodate it without significant hypertension. On the other hand, a very diseased segment that has the same pressure but is receiving a negligible amount of flow would be unsalvageable.

56.3 Special Angiographic Techniques Used to Delineate Anatomy in TOF/PA/MAPCAs

1. *Balloon occlusion aortogram* [4] (Fig. 56.1): A Berman angiographic catheter or similar is advanced from the femoral vein, through the RA, RV into the aorta and around the aortic arch into the descending thoracic aorta, positioned behind the cardiac silhouette. The balloon, which is distal to the angiographic holes in the catheter, is inflated to occlude the distal thoracic aorta and a power injection of approximately 1 cc/kg is given at a rate of 1 cc/kg/s. The resulting angiogram gives adequate opacification of the collaterals proximal to the balloon, and avoids run-off of contrast to undesired parts of the aorta. If the balloon of the Berman catheter is not large enough to occlude the aorta and prevent run-off of contrast, a dual-catheter balloon occlusion aortography technique may be used [5]. In this technique, a soft balloon of equal size to the aorta may be inflated from the arterial access while the Berman catheter or a pigtail catheter is used to inject contrast proximal to the inflated balloon. If using this approach, it is important not to overinflate the balloon and to choose a balloon that is soft and will not damage the aortic wall. It may also be necessary to simultaneously perform rapid pacing of the ventricle, in order to stabilize the balloon, instead of using overinflation of the balloon as the stabilization technique.
2. *Selective injections into each independent collateral and/or its branches*: In order to delineate the anatomy of each collateral, including which segments of the left lung each supplies and whether these segments have dual supply, selective injections (Fig. 56.1) in straight frontal (b, d, f) and lateral (c, e, g) projections are necessary. These injections can generally be performed by "hand-injection." However, if the collateral is large, power injections may be necessary in order to provide the necessary anatomic detail. If performing power injections in these vessels, it is important to have a guide-wire through the angiographic catheter of sheath to ensure the tip of the catheter or

sheath is not in contact with the vessel wall, which may result in a sub-intimal injection and dissection of the vessel. In some cases, it may be necessary to balloon occlude one collateral while injection is performed in another collateral to prove that there is dual supply to one or more segments/lobes of the lung of interest (Fig. 56.5).

3. *Selective pulmonary venous wedge injection* [6]: This technique consists of advancing a wedge catheter into the distal pulmonary vein branches in retrograde fashion. Once the catheter is wedged, an injection of approximately 3 cc of contrast layered with approximately 7 cc of heparinized saline solution behind it is injected. This forces the contrast retrograde up the pulmonary veins, to the capillaries, arterioles and finally to the pulmonary artery (Fig. 56.4). This is a useful technique to visualize the native mediastinal pulmonary arteries if they are not seen when collaterals are injected. It is also useful to find the native lobar pulmonary arteries of lobes that have no flow at the time of catheterization (Fig. 56.6), usually because the APC supplying it has become atretic over time. It is important to avoid this technique in lobes that are supplied by collaterals at a high pressure or with high flow, since trying to overcome this pressure/flow may result in rupture of the small vessels (capillaries/arterioles/venules) and may cause pulmonary hemorrhage. Therefore, collateral injections should be tried first to delineate the anatomy and the central pulmonary arteries. If this is insufficient, then pulmonary venous wedge injections may be used, choosing a lobe that was at particularly low pressure or flow if possible.

It should also be noted that most of the components of the preoperative diagnostic evaluation detailed above can be characterized in great detail using non-invasive axial imaging. In fact, in recent years, CTA has become widely available and used as the initial preoperative diagnostic modality in these patients, sometimes followed by cardiac catheterization to refine and confirm the diagnosis, anatomy, and physiology. CTA does provide

excellent anatomic data and can be used as the only preoperative imaging modality in a subset of patients. Some criteria to select patients that can forego preoperative catheterization include [1]:

1. CTA clearly demonstrates all MAPCAs provide dual supply, as defined above.
2. Normal PA arborization.
3. No lung segments receive sole blood supply from a MAPCA that is not otherwise connected to the rest of the pulmonary circulation.
4. Physiologic data is considered unnecessary: In young patients in whom the CTA clearly shows stenotic MAPCAs and pulmonary arteries that appear to be at low pressure based on appearance and clinical presentation (oxygen saturations below 90%, no signs or symptoms of heart failure and/or pulmonary edema), it may be determined that obtaining the physiologic information provided by the catheterization is unnecessary.

Similar level of anatomic definition, in terms of extravascular structures, can be obtained in the catheterization laboratory using three-dimensional rotational angiography (3DRA) (see Fig. 56.7). This modality provides excellent definition of the anatomy and allows for creation of 3D models similar to those created with standard CTA. The disadvantage is that it is invasive and that it requires more radiation than a regular CTA. However, in patients undergoing cardiac catheterization for preoperative planning and interventions, it may help reduce the total amount of contrast and radiation, and provides excellent anatomic definition. Because the patient is already in the catheterization laboratory, it also allows for further interrogation of areas of the anatomy that may be unclear, in addition to the hemodynamic and interventional advantages described in this chapter.

A reasonable strategy would be to obtain a cardiac CTA on all TOF/PA/MAPCAs as an initial study for preoperative planning, and then use this information to decide whether a cardiac catheterization is necessary based on the criteria described above. This

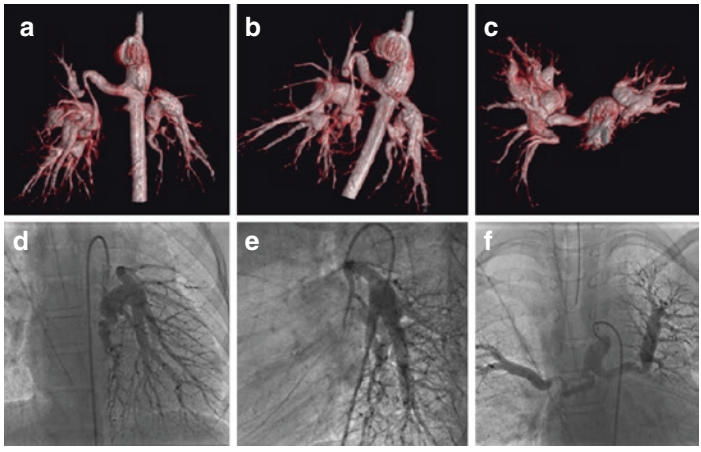


Fig. 56.7 3D CT Rotational angiography to delineate the anatomy of major aorto-pulmonary collaterals. (**a–c**) 3D reconstruction obtained from CT rotational angiography in the catheterization laboratory. This kind of image can be obtained by injecting half-diluted contrast through a pigtail catheter while performing rapid pacing of the ventricle. The rapid pacing decreases the cardiac output during the injection, so that the contrast opacifies the area of interest, but is not "lost" to other parts of the circulation while the image is being obtained. The image is obtained by a rapid >180° rotation of the C-arm around the area of interest. The data from this injection and angiography is processed immediately by specialized software to produce a CT angiography and automatic 3D reconstruction of the area of interest. This 3D reconstruction can then be adjusted to produce images like the ones seen on (**a–c**). This model can be manipulated to see different angles and understand the anatomy of the vessels in three dimensions. In this patient, there are two large APCs with severe stenoses along their length. The APC to the right lung gives off two branches. The first branch is highly stenotic and supplies the posterior right upper lobe. The second branch continues caudally and posteriorly and supplies a dilated right lower lobe branch, which connects to the rest of the right lower lobe branches. It also connects, in retrograde fashion, to the right intermediate PA which gives off right middle lobe branches anteriorly (**c**), then a right upper lobe branch that supplies the apical and anterior segments of the RUL, and finally has a blind end anteriorly at the hilum. The CT images can also be used to improve understanding of the relationship of each vessel to other structures like the airway, pulmonary veins, etc. For example, in this case, the native RPA which fills retrograde from the right lower lobe, could be seen ending blindly immediately in front of the right mainstem bronchus, at the level of the right upper lobe bronchus take-off (not shown). Evaluating the

(continued)

anatomy of the APC to the left lung on the 3D model, it is noted that the APC arises at the same level as the larger APC to the right lung, but straight anteriorly and is markedly stenotic along its length. In similar fashion to the APC to the right lung, it connects to a dilated branch to the left lower lobe, which in turn connects to the rest of the left lower lobe and, in retrograde fashion, fills the native left lower lobe pulmonary artery, ending blindly as it turns over the left mainstem bronchus, at the hilum. It is important to keep in mind that only the vessels that receive contrast will be seen in the 3D model. In this case, it would be easy to miss the supply to the lingula or the left upper lobe on the 3D model, for example. In (**a–c**), no vessels are seen supplying any of the lingula or the left upper lobe. (**d**, **e**) Follow-up selective injections of the collateral to the left lung seen in (**a–c**) show that, if contrast is forcefully injected, the lingula is also supplied by this collateral. (**f**) The supply to the left upper lobe was found by injecting a separate collateral that arose from the underside of the arch (and therefore was not filled by the descending thoracic aorta injection). This is important, because this collateral is the single supply to the left upper lobe and, therefore, will need to be found and unifocalized to the native LPA during surgery. The collateral that arises from the underside of the arch (**f**) also bifurcates and gives a branch to part of the right upper lobe that was also supplied by the larger APC to the right lung. Because this collateral represents dual supply, it may be ligated during surgery and does not need to be unifocalized. This case illustrates the importance of accounting for flow to each and every segment of both lungs. It is important to note that some segments may not be receiving flow under normal circumstances, but these vessels can still be found with techniques like selective injections, like in this case, or other techniques illustrated in prior images like pulmonary venous wedge injections and balloon occlusion injections

would allow for excellent definition of the anatomy with less radiation and less contrast exposure, while avoiding an invasive procedure. Importantly, avoiding instrumentation of the femoral arteries during the neonatal period may reduce the risk of loss of patency of these vessels, in patients in whom multiple catheterizations may be needed throughout a lifetime. Of course, all of this needs to be weighed against the inherent risk related to incomplete diagnostic information. Therefore, the threshold to perform a cardiac catheterization to confirm, or further delineate the anatomy and physiology, should be low.

56.4 Delineation of Coronary Anatomy

The coronary anatomy in patients with TOF/PA/MAPCAs can be variable [7, 8], and certain anatomical variants can affect the way the surgery is performed and/or can place the coronaries at risk during surgery or future transcatheter interventions. Therefore, it is important to characterize the coronary anatomy in detail during the preoperative evaluation. While this can be performed with echocardiography and/or CTA in the majority of patients, some may require aortic root angiography and/or selective coronary angiography to confirm the anatomy or give further details required to plan surgery. Also, since the majority of these patients will have an RV-PA conduit as part of their full repair, it is important to avoid placing the RV-PA conduit near the coronary arteries, when possible. Therefore, a detailed understanding of the coronary arteries is essential in the overall management of these patients.

56.5 The Role of Transcatheter Interventions in the Management of Patients with TOF/PA/MAPCAs

Once the anatomy and physiology have been fully delineated and understood, transcatheter interventions can play an important ancillary role in the management of these patients, before and after surgery. Some of the interventions that are often performed in this patient population include:

1. *Balloon and stent angioplasty of MAPCAs* (Fig. 56.8): It is important to remember that APCs are primitive sources of pulmonary blood flow that are programed to go away. The risk of losing blood flow to a segment supplied solely by a highly stenotic APC is high. Therefore, if these are identified, and surgical incorporation of the vessel is not feasible in a timely manner, consideration should be given to balloon and/or stent angioplasty of these vessels in order to preserve flow to these segments, and ideally, to augment flow and promote growth of the distal vasculature to increase the chances of surgical

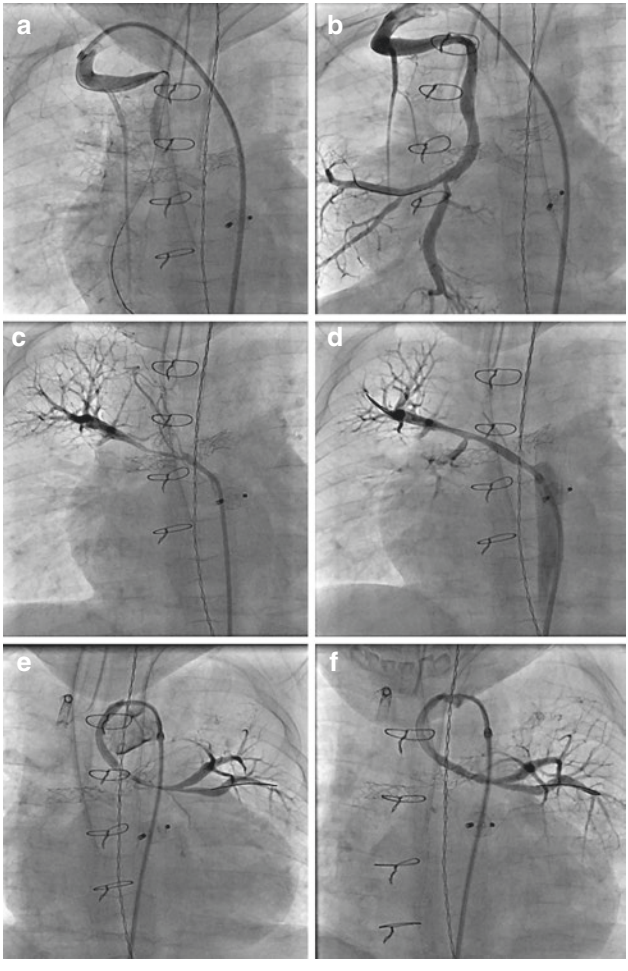


Fig. 56.8 Stenting of stenotic aorto-pulmonary collaterals. In some patients with multiple, small or obstructed aorto-pulmonary collaterals, it may be necessary to stent the APCs in order to maintain flow to the segments supplied by them and get some growth of the distal vasculature before they can be successfully unifocalized at a later surgery. In this patient, three APCs were the sole supply to segments in both lungs. (**a**, **c**, and **e**) show each collateral before stenting, and (**b**, **d**, **f**) show the corresponding collateral after stenting

success once the segments are brought in to the pulmonary circulation. These vessels are highly abnormal and can be resistant to high pressure balloons. Therefore, cutting balloons are often required to achieve any kind of meaningful result [9]. Once the vessel has undergone successful angioplasty, stenting can ensure a longer lasting result. It is important to avoid primary stenting of these vessels, since they have highly resistant stenoses, and it is unlikely for the stent to open the stenosis sufficiently without the prior use of a cutting balloon. Finally, it is important to perform the angioplasty in a very gradual manner, to ensure that the distal pressure in the vessel does not rise to unsafe levels that can lead to acute pulmonary hemorrhage (reperfusion edema) and/or pulmonary hypertension in that segment.

It is also important to keep in mind that approximately 2/3 of the patients with TOF/PA/MAPCAs have retroesophageal MAPCAs [10]. Approximately half of these vessels course through some muscular fibers of the esophagus (intraesophageal MAPCAs) and can even be submucosal. Retroesophageal MAPCAs arise from the side of the aorta that faces the esophagus (as opposed to other types, which arise from the anterior aspect of the thoracic aorta) and go to the lung that is contralateral to the side of the aortic arch. While other types of MAPCAs often have stenosis at or near their origin, the majority of retroesophageal MAPCAs have mid-segment stenosis as they course behind or through the esophagus. It is important to keep this in mind when performing interventions on retroesophageal MAPCAs because a tear of the MAPCA could result in massive and possibly catastrophic bleeding into the esophagus/hematemesis. When interventions on these vessels are carried out, it would be wise to consider avoiding instrumentation of the esophagus (e.g., transesophageal echocardiography, endoscopy, feeding tube placement) in the postoperative period.

2. *Balloon and stent angioplasty of pulmonary artery branches* (Fig. 56.9): This includes unifocalized APCs, as well as reconstructed or native branch PAs, lobar, segmental and subsegmental branches of the pulmonary arteries. These

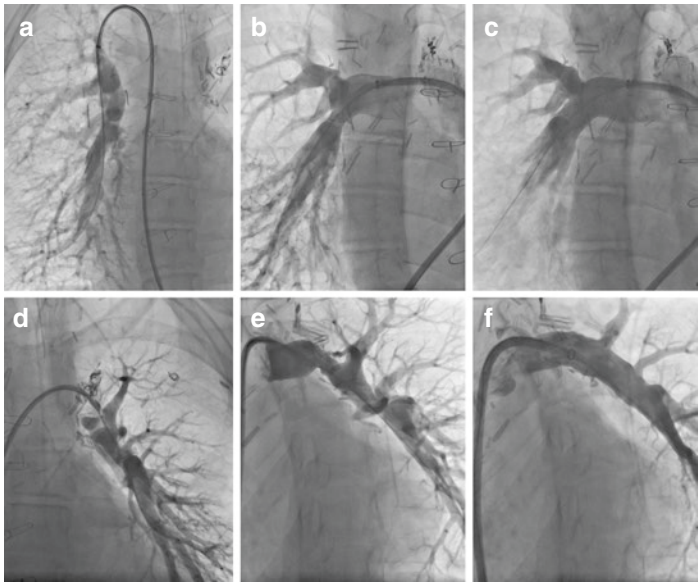


Fig. 56.9 Balloon and stent angioplasty as a part of the management of patients with repaired TOF/PA/MAPCAs. This patient is the same patient shown in Fig. 56.6 at different stages of the repair. (**a**, **d**) AP projections in the shunted, unifocalized pulmonary arteries to the right (**a**) and left (**d**) lungs. (**b**, **e**) After another surgery consisting of unifocalization of these arteries to an RV-PA conduit with closure of the VSD and takedown of the shunts, angiograms into the RPA (**b**) and LPA (**e**) show good distal vasculature and some areas of discrete stenosis, mainly of the proximal left pulmonary artery, the left lower pulmonary artery, the RULPA, and the right intermediate PA. (**c**, **f**) After balloon and stent angioplasty, the vessels have a much better caliber and no discrete stenoses

interventions are reviewed elsewhere, and some examples are given in the figures in this chapter.

3. *Establishment of antegrade flow across a functionally or anatomically atretic pulmonary outflow* (Fig. 56.10): In some cases, there may be plate-like atresia of the pulmonary valve or functional atresia of the valve (see Fig. 56.10), which prevents opening of the valve because of the combination of sub-PS and pressurized MPA due to APC flow. In these cases, it is possible

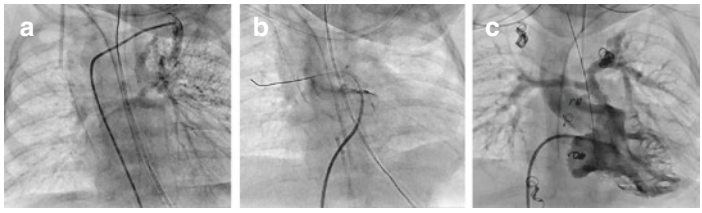


Fig. 56.10 Opening and stenting of a functionally atretic right ventricular outflow tract followed by coiling of APCs allows for transcatheter establishment of antegrade pulmonary blood flow, with the goal of establishing pulmonary artery growth in preparation for future, single-stage full surgical repair. (**a**) AP projection of selective angiogram into an APC that supplies the left pulmonary artery branches and the central mediastinal pulmonary arteries in retrograde fashion. (**b**) Even though there was no visible antegrade flow across the pulmonary valve, probing the outflow tract resulted in a wire crossing the valve and going into the RPA. (**c**) The valve and infundibulum were balloon dilated and stented and the APCs were coil occluded. Follow-up angiogram in the RV shows good antegrade flow across the stented pulmonary outflow with continuous mediastinal PAs of reasonable size supplying all segments of both lungs. Over time, the expectation would be that the pulmonary arteries will grow sufficiently to allow a single-stage full repair of tetralogy of Fallot

to establish antegrade flow across the pulmonary outflow by balloon and stent angioplasty, either using percutaneous or hybrid transcatheter techniques [11]. Once this antegrade flow is established, the segments that have dual blood supply from APCs may be occluded to promote antegrade flow through the native pulmonary arteries. Over time, this will promote growth and may facilitate a full, single-stage surgical repair.

4. *Coil or device embolization of APCs* (Figs. 56.11 and 56.12): As described above, APCs that provide dual supply should be occluded to prevent competitive flow, which may result in pulmonary hypertension in the affected segments, as well as underdevelopment of the native pulmonary arteries supplying those segments.
5. *Device closure of atrial or ventricular septal patch fenestrations, and/or residual defects*: These techniques are also reviewed elsewhere and can be an important part of the management of these patients.

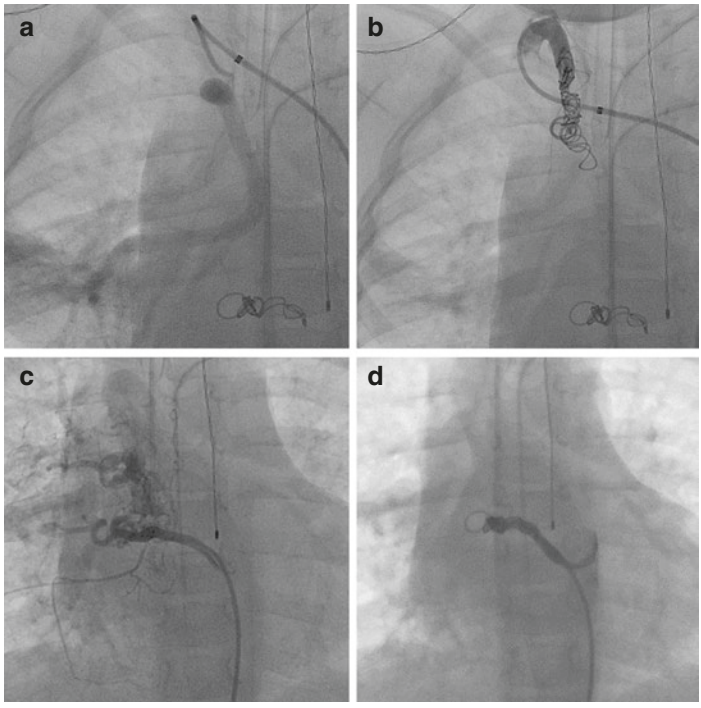


Fig. 56.11 Coiling of collaterals that supply vasculature that is at high pressure and will not be unifocalized. (**a**) Selective injection into collateral from the right thyrocervical trunk to the right middle lobe. The pressure in this lobe was significantly elevated and the vasculature appeared very hypertensive and damaged. Therefore, the vessel was coil occluded (**b**) to avoid unnecessary inefficient pulmonary blood flow that provides no physiologic benefit to the patient and may be at risk for causing hemoptysis in the future. (**c** and **d**) Small APC providing single supply to small portions of the right lung. The vasculature is quite abnormal and the collateral is too remote and small to be unifocalized, so it was coil embolized (**d**)

6. *Interventions to treat RV-PA conduit dysfunction*: including balloon and stent angioplasty of conduits, as well as transcatheter pulmonary valve replacement; also reviewed in other chapters and will not be addressed here.

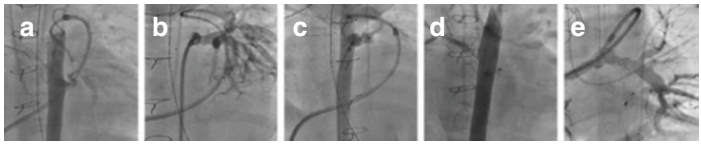


Fig. 56.12 Occlusion of aorto-pulmonary collateral providing dual supply to the left lung. (**a**) AP view of an aortogram showing a collateral that connects to the previously unifocalized left lower lobe pulmonary artery. There is a catheter in the left lower lobe PA and a wire goes from this catheter, through the collateral into the descending thoracic aorta, proving that this collateral provides flow to the same vessels supplied antegrade by the LLPA, which is now connected to an RV-PA conduit. (**b**) The wire in the LLPA catheter has now been moved to a lower lobe branch and there is a wire through a long sheath in the descending thoracic aorta, through the collateral into a LLPA branch. Contrast is injected into the collateral which fills the entire left lower lobe and part of the proximal LPA in retrograde fashion. (**c**) A vascular plug is deployed in the collateral (still attached to the delivery cable) and an angiogram confirms adequate position. (**d**) After the device is deployed, repeat angiogram shows complete occlusion of the APC. (**e**) Angiogram after stenting the proximal LPA shows good antegrade flow into the left lower lobe, without any competitive flow ("washout") from the occluded collateral shown in (**a–d**)

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57 Stenting of the Right Ventricular Outflow Tract as Initial Palliation for Fallot-Type Lesions

Oliver Stumper, Daniel Quandt, and Gemma Penford

57.1 Introduction

The initial management of severely cyanosed patients with tetralogy of Fallot-type lesions remains challenging. True neonatal repair of these lesions remains the exception [1–3]. The creation of a Blalock-Taussig (BT) shunt is well established but continues to have a high early and late complication rate and mortality [4]. Earlier attempts at transcatheter interventions (either balloon pulmonary valvuloplasty or stenting the right ventricular outflow tract) were rather high risk or yielded unpredictable results [5–8]. It is only recently that several groups have revisited stenting of the

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right ventricular outflow tract [9–12] in the initial palliation of symptomatic patients with Fallot-type lesions.

At Birmingham Children's Hospital we have performed close to 100 of such procedures until the end of 2015, and this report summarizes our experience focusing on technical tips and advice.

57.2 Patient Selection and Imaging

Tetralogy of Fallot is amongst the most common cyanotic congenital cardiac lesion. Some children may progress to acquired pulmonary atresia within a few weeks after birth. Others remain stable until weighing more than 5 kg in body weight when one-stage complete repair should be the preferred treatment option. Yet, associated cardiac lesions such as a complete AVSD, hypoplastic pulmonary arteries, double outlet right ventricle arrangement, or abnormal systemic or pulmonary venous return or the co-existence of associated syndromes or co-morbidities may make initial palliation preferable, rather than an attempt at early complete repair.

The diagnosis of Fallot is universally made on cardiac ultrasound. CT angio may be useful in patients with suspected coronary artery anomalies or collaterals. If the patient requires palliation prior to complete surgical repair, cardiac ultrasound is very accurate in defining the relative contribution of subvalvar, valvar, and supravalvar obstruction and assessment of the branch pulmonary arteries. The most reliable views to assess these are subcostal images of the RVOT and parasternal short-axis views. Diagnostic cardiac catheterization in the initial diagnosis of Fallot-type lesions is nowadays rare—its mayor impact being the delineation of the peripheral pulmonary arteries, the exclusion of associated collateral arteries, and anomalous coronary distribution.

57.3 Catheter Stenting of the RV Outflow Tract

57.3.1 Patient Selection

Indications for consideration of RVOT stenting in symptomatic Fallot-type lesions are all cases in whom one-stage complete

repair is considered high risk or where there are significant associated cardiac lesions or morbidities which make delay of bypass surgery preferable.

57.3.2 Procedural Preparation

Cases should be discussed in a multidisciplinary team meeting. Detailed informed consent has to be obtained. All patients should be cross-matched for blood products, and there has to be thorough briefing of the whole team prior to sending for the patient. Emergency drugs are prepared, and the cardiac surgical and intensive care team should be made aware about the timing of the procedure.

57.3.3 Cardiac Catheterization

The standard vascular approach should be from the right femoral vein. This largely facilitates entry into the right ventricular outflow tract using either right Judkins or Cobra preshaped catheters. The right internal jugular venous approach may be beneficial in neonates weighing less than 2 kg due to vessel size (Fig. 57.1). Ideally the patient should be draped to allow for peri-procedure ultrasound scanning from subcostal and parasternal projections. A sterile ultrasound probe should be available for the operators to use.

Baseline angiograms are performed in 30 RAO + 20 cranial and lateral projections.

57.3.4 Measurements

Ultrasound measurements of the RVOT are generally the most reliable. Angiography is always likely to underestimate required stent length due to foreshortening. Echo measurements should be made to cover the entire length of the RVOT and should ideally be

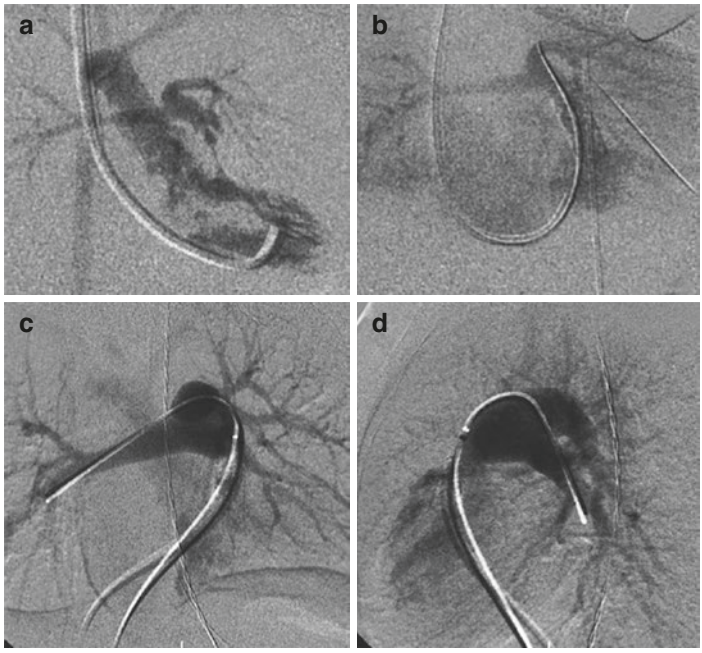

Fig. 57.1 RVOT stenting in a 1.7 kg premature (28 weeks +5) baby at 3 weeks of age. Digital subtraction angiographic stills. (**a**) From a right internal jugular venous approach, a diagnostic angiogram is performed in 30 RAO + 20 cranial through a 4 French Cobra catheter. Note the very long-segment RVOT obstruction. (**b**) The 4 mm coronary stent was placed through a 4 F introducer sheath advanced to the right ventricle to allow for sidearm test injections—immediate result. (**c**) At 8 months of age (weight 5.3 kg), a further catheter was undertaken performing 30 RAO + cranial and lateral angiograms. Note the very good growth (post stenotic dilatation) of the branch pulmonary arteries. (**d**) Lateral angiogram after further stenting with a 6 mm biliary stent. The child underwent complete repair at 15 months, weighing 7.8 kg

a sum of distances added together to pay tribute to the curved nature of the RV outflow tract (Fig. 57.2). During stenting of the RVOT, an attempt should be made to avoid crossing the pulmonary valve, unless there is significant supravalvar stenosis.

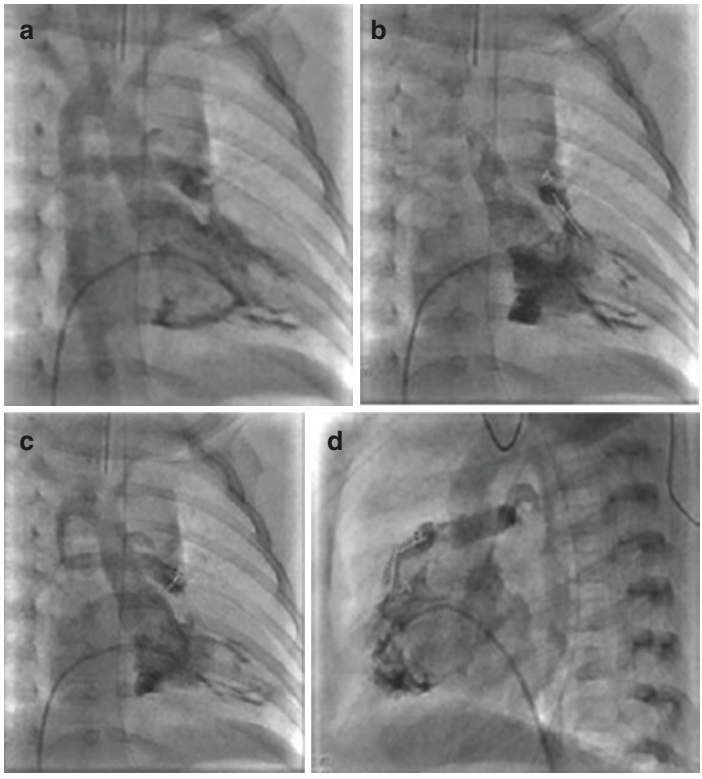

Fig. 57.2 Still frame RV angiograms in a 3-month-old child with complete AVSD and Fallot with significant co-morbidities. (**a**) RAO + cranial projection shows the anterior deviation of the outlet septum, a reasonably developed pulmonary valve annulus and decent branch pulmonary arteries. (**b**) The length of the RVOT is always underestimated using angiographic views. (**c**) Pulmonary valve and branch PA measurements are reliable. (**d**) In the lateral projection, the most reliable length measurement for the RVOT can be obtained by summation of at least two measurements, so as to allow for the curvature of the outflow tract. Always choose a stent slightly longer than the longest measurement taken—even those on ultrasound

57.3.5 Procedure

Following the initial RV angiogram (see above), the catheter is withdrawn to the IVC. Reference angiographic stills are selected and displayed in the room. Repeat measurements are being made from the angiograms.

The appropriate kit is selected and prepared before proceeding any further. In children weighing more than 2.5 kg where only short-term palliation is required, a 5 mm coronary stent is chosen (4.5 or 4 mm in kids weighing less). In those who require medium- to long-term palliation, a peripheral vascular or biliary stent such as the Cook Formula [13], the Omnilink or Genesis stent is chosen—which allow for later over-dilatation.

The stent chosen for implantation dictates the required delivery system. Departmental preference is for either 4 or 5 F Cook Flexor sheaths or for a 6 F short (55 cm) right Judkins Guide catheter (Cordis) (Fig. 57.3). All is prepared and is introduced over a 0.035″ wire placed in the SVC.

A 4/5 F right Judkins or Cobra catheter is inserted into the delivery catheter and is then used to enter the pulmonary arteries directly under pressure monitoring. Position is confirmed on hand test angio. The appropriate wire for the stent system is placed in the distal (right) pulmonary artery, and the delivery sheath or guide catheter is advanced from the IVC to the branch PA over the diagnostic catheter and delivery guidewire. Next the diagnostic catheter is removed over the wire and the stent is delivered. Angio test injections are used to confirm stent position prior to (hand) inflation of the balloon. The lateral *X*-ray projection is particularly useful for placing stents where it was decided to spare the pulmonary valve (Fig. 57.4). Hand inflation of the balloon for placement of the stent is almost always sufficient and allows for fine positional adjustments during placement. A further balloon inflation across the pulmonary valve should be performed so as to dilate the most commonly stenotic pulmonary valve. Next, the delivery sheath or guide catheter is advanced slightly to oppose the proximal part of the stent, so as to prevent dislodgement of the stent

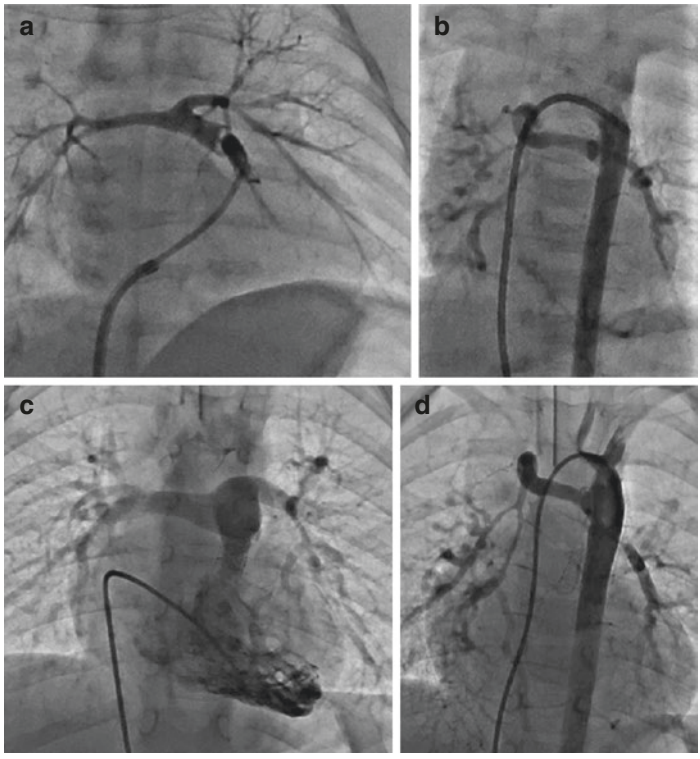

Fig. 57.3 Serial angiographic still frames in a child with severe Fallot and stenosed MAPCAs from a left arch. (**a**) After the initial angiogram, decision was taken to proceed to RVOT stenting. A 6 F right Judkins Guide catheter was placed in the right atrium, and the distal right ventricular outflow tract was intubated using a 4 F Judkins right catheter. Note the severe pulmonary valve stenosis and the appearance of the peripheral pulmonary arteries suggestive of Alagille syndrome (negative genetics). (**b**) AP angiogram depicting the left aortic arch and the stenosed MAPCAs to both lung fields—attempts at surgical recruitment are not warranted. A 5 mm coronary stent was implanted at 4 weeks of age (weight 3.9 kg) with good effect. (**c**) RV angiogram at 3.5 years of life (14 kg—saturations 84%) after further dilatation of the initial stent at 9 months (7.5 kg) and implantation of a 7 mm Cook Formula stent at 19 months (10 kg). The central PAs have grown very nicely, but there remain bilateral hilar stenoses, which were addressed by balloon angioplasty. (**d**) The repeat arch angiogram documents further progression of the long-segment stenoses within the MAPCAs. At 4.5 years of age, the child is now listed for complete surgical repair and preceding catheter occlusion of the MAPCAs

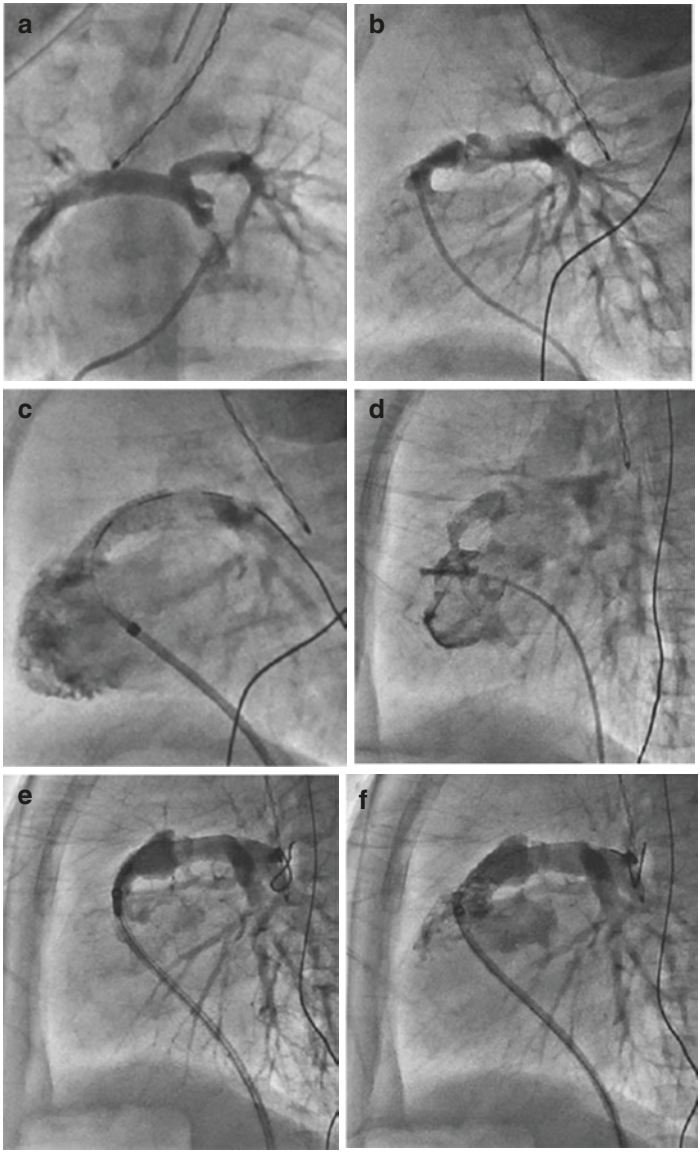
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Fig. 57.4 Sequence of angio stills in a 5-week-old boy (3.4 kg) with double outlet right ventricle (Fallot-type), complete AVSD and bilateral SVC with left SVC draining to an unroofed coronary sinus. (**a**) Four-chamber view documents tight infundibular and valvar pulmonary stenosis—well-developed central pulmonary arteries and peripheral arborization. (**b**) Lateral angiogram showing short infundibulum and significant anterior deviation of the outlet septum. (**c**) RVOT stent placement (5 × 20 mm coronary) via 4 F Flexor over 0.014" Thruway wire just crossing the pulmonary valve annulus. (**d**) Repeat lateral angio 4 months later (5.8 kg) shows proximal obstruction. (**e**) A further stent (6 mm Cook Formula) was placed proximally through a 5 F Flexor under repeat sidearm angio guidance. (**f**) Good final result after placement. Note the growth of the pulmonary valve annulus. Complete repair was performed at 13 months (9 kg)

whilst retrieving the deflated (negative suction!) delivery balloon. The delivery sheath/guide catheter is then withdrawn to the IVC, and further detailed cardiac ultrasound study is performed. This focuses on whether the proximal portion of the infundibulum is covered, assessment of ventricular function and exclusion of pericardial effusion.

RVOT stenting has evolved to be a valuable tool in patients with very complex anatomies bordering on pulmonary atresia with VSD (Fig. 57.5) and in our opinion is safer and more effective than PDA stenting in cases with duct-dependent pulmonary blood flow, as long as there is even a tiny residual right ventricular outflow tract with some anterograde flow (Fig. 57.6). Further, in our experience, it has become a low-risk effective initial palliation technique in selected univentricular patients with limited anterograde blood flow (Fig. 57.7).

57.3.6 Post Procedure Management

All patients receive antibiotics at induction of anaesthesia and two further doses after stent implantation. Prostaglandin infusions, when present, are stopped on placement of the stent. Patients who experience a rise in oxygen saturations of more than 20% are

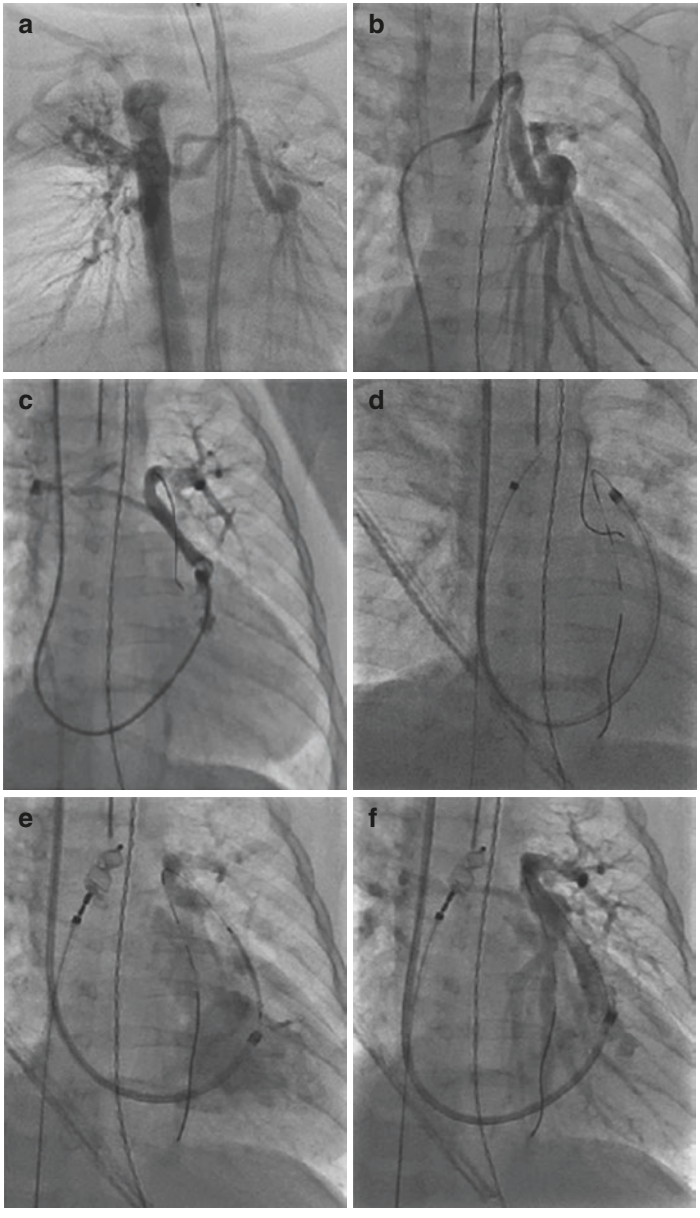
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Fig. 57.5 Still frames of RVOT stenting in a 5-month-old child with extreme tetralogy of Fallot and large MAPCAs to both lung fields. Weight 5.1 kg, on CPAP ventilation due to excessive pulmonary blood flow and airway compression by the MAPCAs. (**a**) Descending aortogram showing numerous collaterals to the right lung and at least two MAPCAs to the left lung. (**b**) Injection into the large MAPCA to the left lung does not show any significant stenosis and good peripheral arborization, suggesting early communication with the native left lung artery system. (**c**) Injection into a severely hypoplastic right ventricular outflow tract demonstrates a native confluent PA system with extreme hypoplasia of the right pulmonary artery. (**d**) During further simultaneous injections, it was confirmed that the left lung in fact had dual supply (native LPA and MAPCA) to all segments. The left MAPCA was test occluded. (**e**) Placement of a vascular plug to occlude the large left MAPCA and placement of a stent on the RVOT (crossing the hypoplastic pulmonary valve annulus) via a Flexor sheath over a coronary wire. (**f**) RVOT angiogram showing good flow to the entire LPA system and improved flow to the RPA. Subsequently the vascular plug was released. The patient weaned successfully from ventilation 5 days later. He is awaiting further catheter 6 months later (saturations 84%, weight 7.6 kg) with a view to dilate the stent further and ultimately to undergo recruitment of right MAPCAs and unifocalization to a restrictive valved conduit

commenced on twice daily diuretics. Patients are kept on intravenous fluid management for at least 12 h and are started on single-agent antiplatelet therapy once oral intake is re-established. If clinically stable and feeding well, patients are discharged home 48 h post procedure with frequent outpatient review.

Re-intervention is considered in cases where further delay to repair is desirable or in those who develop significant recurrent outflow tract obstruction either due to not entirely covering the proximal portion of the outflow tract, tissue ingrowth into the stent or somatic outgrowth.

57.3.7 Experience So Far

RVOT stenting is a complex procedure with a definite learning curve. Meticulous procedure planning and execution is essential. Early mortality rates of <2% compare favourably with either BT

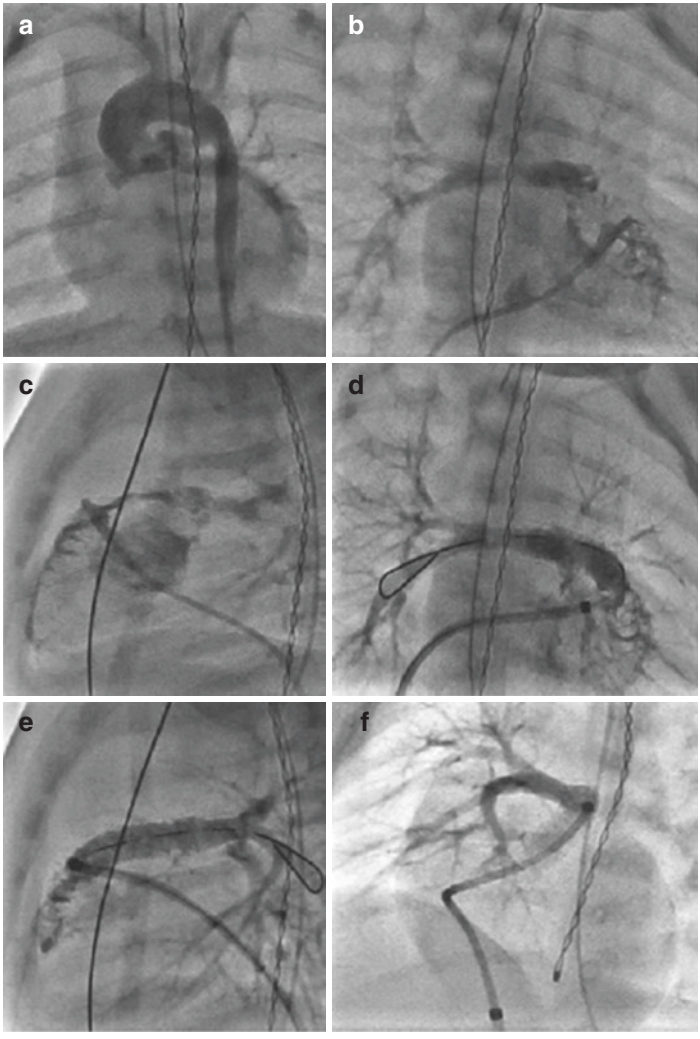
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Fig. 57.6 Sequence of angio still frames in a 2-week-old child who was initially thought to suffer from pulmonary atresia with a tortuous duct from the underside of the left aortic arch. (**a**) Retrograde aortogram in AP projection documents a left arch and a wide opening but rather tortuous duct to confluent PAs of reasonable size. (**b**) RV angio in 30 RAO + cranial documented some miniscule forward flow through the RVOT mainly to the right PA. (**c**) Lateral projection of the same angio. Decision was taken to stent the RVOT rather than to attempt stenting the wide and tortuous duct with the potential of acute instability and possible later branch PA stenosis. (**d**) RAO angio after 5 mm coronary stent placement through 4 F Flexor sheath over a 0.014" Thruway wire. Note the stent was placed across the pulmonary valve annulus but stops short of the bifurcation. (**e**) Lateral projection of the above. (**f**) Final angio in four-chamber angulation documents good flow to the right PA. The left PA was predominantly perfused through the patent duct, as confirmed on post-placement cardiac ultrasound. Prostin was stopped, and the child underwent complete repair at 6 months with transannular patch and surgical bifurcation plasty

shunt procedures or early Fallot repair. In as yet unpublished studies, we could document that RVOT stenting provides better pulmonary arterial growth (Fig. 57.8) compared to BT shunt and that time to complete repair was reduced.

57.4 Summary

RVOT stenting in the initial palliation of patients with tetralogy of Fallot is displacing BT shunt surgery. Catheter procedures are complex and have to be well planned, and executed, ideally by an established team. Strict adherence to the above step-by-step guide is likely to result in success and short procedure times.

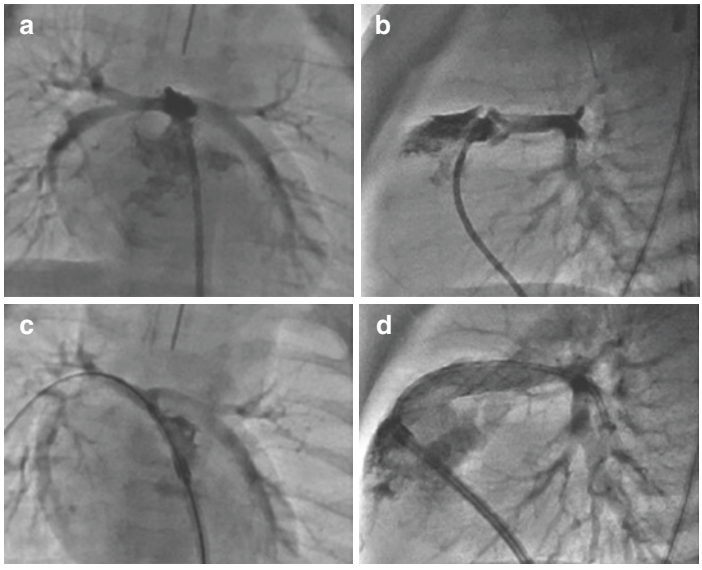
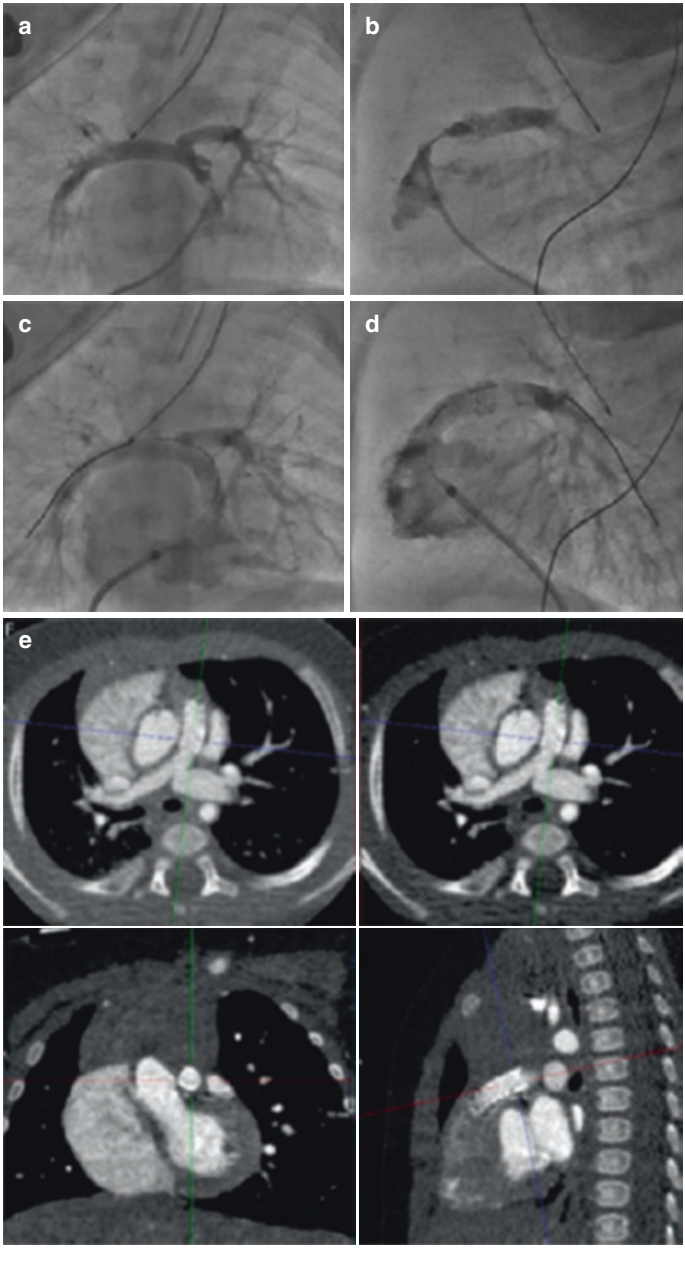


Fig. 57.7 Sequence of angio stills in a neonate with right atrial isomerism, unobstructed total anomalous pulmonary venous return, complete AVSD and severe valvar and supravalvular pulmonary stenosis. (**a**) AP projection with cranial tilt showing symmetric branch pulmonary arteries of a reasonable size. (**b**) Lateral angiogram documents wide open subpulmonary area and severe valvar and the supravalvular pulmonary stenosis. (**c**) Four-chamber projection documents hypoplasia of the main pulmonary artery up to the bifurcation. (**d**) RVOT stent was placed across the pulmonary valve and the main pulmonary artery up to the bifurcation. No further intervention was required until the age of 9 months (bilateral CP shunts, PA augmentation and repair of TAPVC)

Fig. 57.8 Image sequence in a neonate with severe Fallot undergoing RVOT stenting at 2.6 kg in weight. (**a**) Good-sized branch pulmonary arteries on four-chamber projection. (**b**) Tight right ventricular outflow tract obstruction on lateral projection. (**c**) Corresponding angio stills after placement of a 5 mm coronary stent (**d**) as above. (**e**) CT angio multiplanar reconstruction images of the branch pulmonary arteries and the stented right ventricular outflow tract prior to complete repair at 6 months of age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Part X

Imaging Techniques

58 The Use of 3D Rotational Angiography in Congenital Heart Disease

Gregor Krings

58.1 Introduction

3D rotational angiography (3DRA) is an angiographic technique with a computed tomography scan acquired during cardiac catheterization. Most angiographic catheterization systems offer a special acquisition mode in which a more or less 180° spin of the frontal plane around the patient is performed within 4–6 s during continuous contrast injection. This rotational angiography (RA) automatically is converted in a volume rendered 3D dataset within a few seconds and can be processed in the same way as a CTA or MRI. During postprocessing, different tools are available to rotate the 3D image, zoom in or out, use multiple clipping planes, scissoring, and adjustment of Hounsfield units (HU). When the region of interest is visualized, the condensed 3D image can be projected on the conventional angiography system to guide the intervention by synchronized 3D roadmapping. Imaging data of preprocedural CTA or MRI or a previous 3DRA can be imported, fused with the

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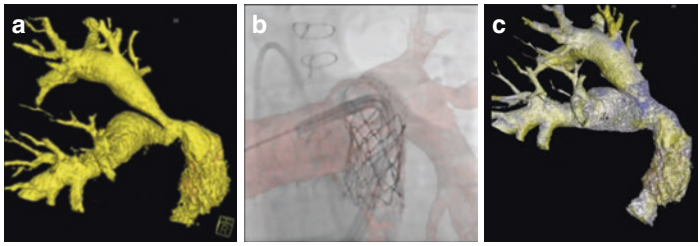

Fig. 58.1 (**a**) 3DRA in complex pulmonary artery bifurcation stenosis. (**b**) 3DRA guided Y-stenting procedure with stent-through-stent implantation. (**c**) exit 3DRA to visualize vessel integrity and final bifurcation geometry

current 3DRA or even used instead of a 3DRA after alignment with the anatomical landmarks of the patient. In interventions like stent placement or percutaneous valve implantation, 3DRA helps to guide the procedure with enhanced understanding of the topographic interactions and anatomic variability (Fig. 58.1).

58.2 Principle of 3D Rotational Angiography

The angiographic principle of 3DRA differs from a conventional angiography and from a CTA. Understanding of contrast injection timing and contrast distribution at the target region is crucial. If the blood flow through a region of interest (ROI) is compared to a train, then conventional angiography does film the train coming and going. With CTA the entire train is contrast loaded, runs along the entire track and is filmed after a full cycle. With 3DRA the train is slowed down and only the segments of interest are scanned at their first appearance. Spatial resolution is high and structures as pulmonary veins—potentially blocking the vision of the region of interest when stenting peripheral pulmonary artery stenosis—will not be scanned if not intended. Next to all cavities filled with contrast visualization in 3DRA includes bones, metallic structures like stents and airway. Dynamic information is present in the rotational angiography but not in the static 3DRA although experimental work proved to generate high resolution dynamic

4DRA. There are different ways to perform 3DRA as there are many ways to performing a conventional angiography, too. Multiple factors have influence on the spatial resolution in 3DRA as contrast dilution, location of injection, use of rapid pacing and breath hold. The image quality in 3DRA depends on the correct timing of the contrast injection, location(s) of injection, and the correct delay between injection and start of the scan. The need for rapid pacing is discussed controversially. However, little spatial resolution will occur when rotational angiography is done during spontaneous heart rhythm and breathing compared to a setup with rapid pacing and breath hold. Movement artifacts cannot be compensated by higher amount or concentration of contrast.

58.3 How to Start with 3DRA

The learning curve in 3DRA does involve the entire team at the cath suite including technicians, nurses, anesthesia staff, and interventionalists. Communication is crucial during 3DRA. Any kind of frustration linked to the "new workflow" will diminish the teams' willingness to participate and support the learning curve evidently linked to 3DRA. An application specialist from the 3DRA system as well as a proctor should be present during the first rotational angiographies. This will allow minimizing systematic errors and improving the 3DRA outcome. When 3DRA delivers suboptimal anatomic information in a series of patients, it will get difficult to justify the change in workflow compared to conventional angiography.

58.3.1 Role of the Anesthesiologic Team

Performing a rotational angiography has impact on the anesthesiologic team and their workflow. Ventilation tubes, perfusor lines, and monitoring cables should have a length of at least 250 cm and be bundled to avoid interaction with the rotating frontal plane. On biplane systems, the lateral plane has to be moved

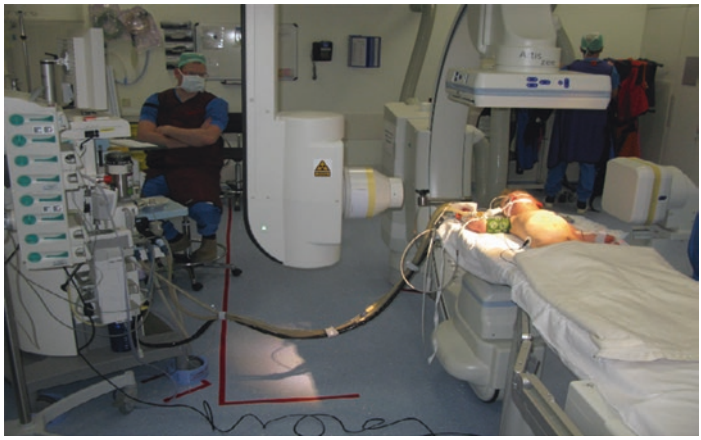

Fig. 58.2 Cables and tubes bundled, red tape on the floor to mark the corridor which has to be kept free for the angiographic system

away from the patient side to let the frontal plane rotate unrestricted. Thus the anesthesiologic hardware has to stay away from the reach of the rotating frontal plane as well as outside the corridor the lateral plane needs to move back and forth. Red tape on the floor is helpful to define the critical area which should not be blocked by any equipment during the entire procedure. Ventilator tubes, cables, NO units, and infusion pumps should be outside this corridor from the very beginning of the procedure (Fig. 58.2). Simple commands should be communicated to define the procedural steps in 3DRA and guarantee safety and success.

58.3.2 The 3DRA Workstation

Before performing any rotational angiography, the interventionalists should get familiar with the installed 3DRA workstation and its tools, capabilities but also limitations. Basic functions of the workstation include rotation of the 3D volume, zooming in and out, selecting a ROI and removing structures on the in- or outside, applying a preset for best visualization of contrasted structure,

adjusting Hounsfield units, safe the current status with bookmarks, work with cross sectional clipping planes, import CTA, MRI or 3DRA data, and backproject a 3DRA volume to the angiographic system for roadmapping.

58.3.3 Contrast Injection

Contrast (300 mg I/ml) should be diluted 6:4 with NaCl 0.9% and injected in the cavity proximal to the region of interest, or generally spoken the corresponding ventricle. Herewith contrast washout will be reduced to a minimum. Contrast injection into the main pulmonary artery trunc or bifurcation will result in washout in RPA and LPA, inhomogeneous contrast distribution, low density and low spatial resolution. Thus RV injection is preferable in general. Right atrial injection should be avoided in biventricular hemodynamic because of low flow contrast distribution and back flow into the caval veins. In single ventricle with stage I palliation and DKS contrast injection into the single ventricle during rapid pacing will guarantee crisp high detail imaging of the DKS, shunt, aorta, collaterals, and pulmonary arteries within one run. The amount of contrast can be estimated by two different approaches: 2 ml/kg total, distributed over 6 s scan time. A 3 kg newborn would receive 12 ml contrast (60% diluted) with 2 ml/s. Our preferred algorithm does differ and takes the ideal flow in ml/s into account related to the patient's weight following an experienced based curve (Fig. 58.3).

Each circulatory system receives contrast following this curve. In biventricular circulation RV contrast injection should start 2 s before for sufficient prefill of the pulmonary arteries and stop 3 s after scan start. LV contrast injection is initiated 1 s before scan start and continued the entire scan. Thus 5 s RV contrast injection and 6 s LV contrast are needed. RV contrast accumulates in the lung and remains longer in place which allows for terminating the RV contrast injection 2 s before scan stop. The short transition time of contrast in the aorta demands continuous contrast injection from 1 s before scan until the last scan moment, thus 6 s. In a

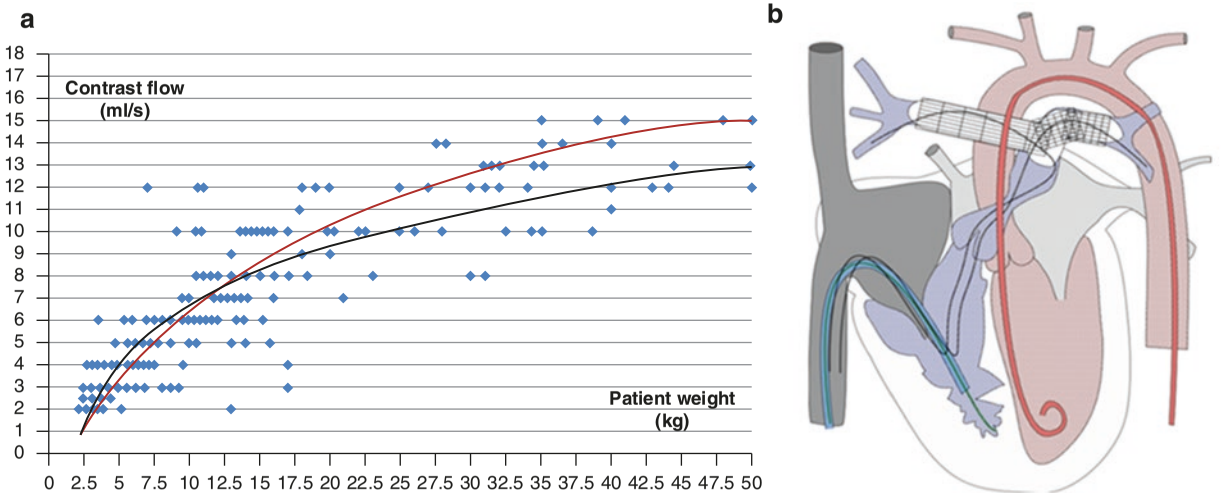

Fig. 58.3 (**a**) Relation between patient weight and contrast flow in 3DRA; the flow is multiplied with the injection time and used per chamber (RV or LV), contrast 40% diluted. (**b**) location of contrast injection (RV and LV) and rapid pacing, 4Fr pacing electrode advanced through long sheath in RV apex (5Fr or larger)

10 kg child, RV injection would be performed with 6 ml/s over a 5 s injection period (30 ml total), whereas LV injection would be done with 6 ml/s over a 6 s period with a 1 s delay (Fig. 58.4). Details for typical lesions are explained below.

When two locations for contrast injection are needed, a 1-in-2 Luerlock splitter can be connected two identical angiographic catheters. In our approach, we prefer manual contrast injection if more than one location is chosen for contrast filling for timing reasons as explained below. We typically connect the injector to the pigtail catheter placed in the left (or single) ventricle and use manual injection for all other locations (RV in biventricular, VCS and left jugular vein PCPC, VCI and VCS in TCPC) works fine. Double venous accesses can be avoided (one for pacing, one for contrast injection) when a long sheath (minimum 5Fr) is placed in the right ventricular apex with a 4 Fr bipolar pacing electrode being advanced out of the sheath tip. In 2 kg children, a 5Fr RV setup with a 10 ml RV hand injection works as well as a, 8–10 Fr setup in adults will parallel injection through two syringes 50 ml.

In PCPC and TCPC injection in VCS/VCI should be done 2 s before contrast filling of the single ventricle to take slow contrast progression in the cavopulmonary connection into account. In pulmonary venous interventions such as recanalization or stenting, the cavity proximal of the region of interest is the right or left pulmonary artery in which the contrast should be injected. The correct timing between contrast injection and strat of the 3DRA scan has to be delayed with the number of seconds estimated by performing a short contrast angiography in the pulmonary arteries and counting for the moment when contrast appears in the pulmonary veins. The typical contrast injection–scan delay will be around 5–7 s.

58.3.4 Rapid Pacing

Rapid pacing can be best performed in the RV apex via a long sheath enabling for contrast injection at the same location (Fig. 58.3). Atrial pacing often results in suboptimal myocardial contact with inconsistent capture. Transesophageal ventricular

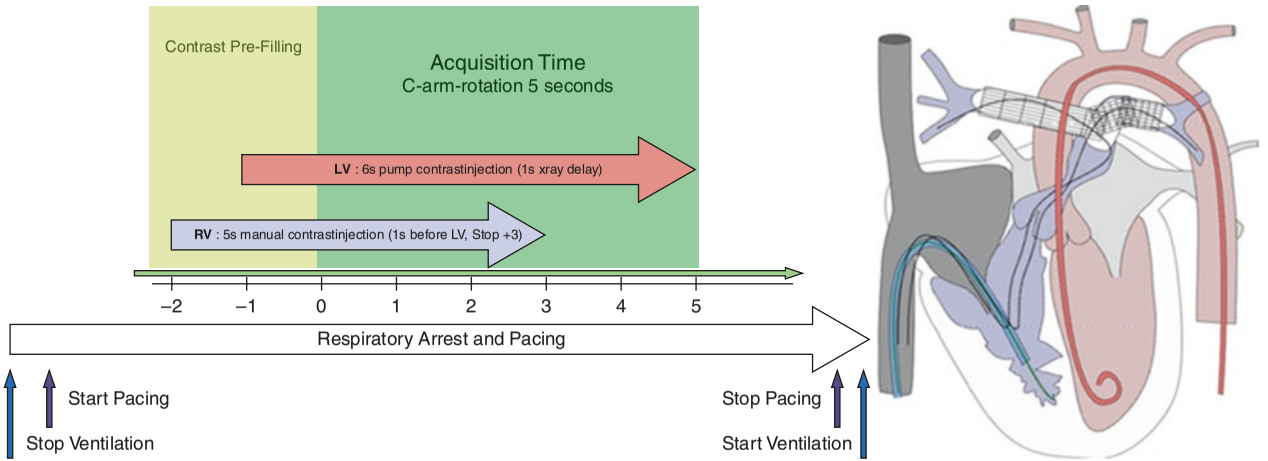

Fig. 58.4 Workflow sequence in 3DRA with biventricular contrast injection

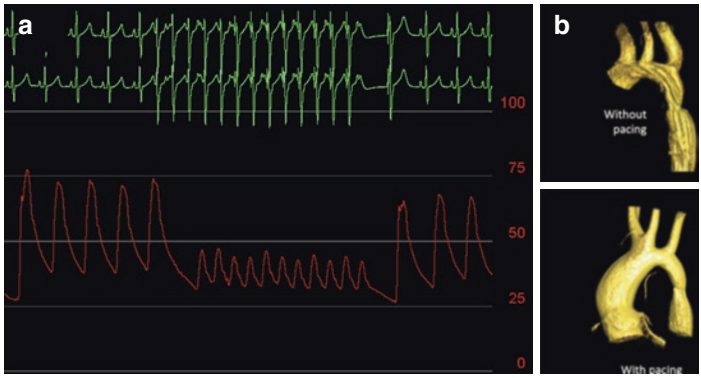

Fig. 58.5 (**a**) Heartrate and blood pressure during rapid RV apex pacing. (**b**) Low resolution volume rendering without enhances spatial resolution

pacing with a high output external pacer is a good alternative in small infants and Fontan patients where transvenous access doesn't allow for myocardial contact. In communicating with the anesthesiologic team, the rapid pacing test maneuver aims to detect the ideal frequency and output. Starting at 180/min the pacing frequency is increased stepwise until the systolic ABP is decreased by 40% (Fig. 58.5). When the capture is reliable and the ABP response is stable, this pacing test is stopped. When capture remains insufficient, it is recommended to check the pacemaker setting, cable connection, and electrode position. An inconsistent rhythm will result in inhomogeneous contrast distribution and low spatial resolution. In newborns and small children, advancing the electrode should be done with care due to the thin RV myocard and the risk of perforation. Bending the electrode 45° in between the distal tip and ring will make advancement into the RV apex less traumatic. Furthermore the bended electrode tip will prevent pushing the long sheath backwards through the tricuspid valve which finally would create a useless right atrial contrast injection.

When right ventricular rapid pacing is used, the entire team should be prepared to handle ventricular arrhythmias as fibrillation or Torsade de Point with an external defibrillator. In our

experience, sustained ventricular arrhythmia occurred in less than ten procedures out of 1600 3DRAs and was induced by rapid pacing in patients with significantly reduced RV function.

58.3.5 Isocenter and Collimation in 3DRA

In 3DRA, the frontal plane is centered to the patient's verbal spine in AP and to a midcardiac level in lateral. The amount of collimation (cranial and caudal) depends on the anatomical information needed. In PA VSD, the window of interest is kept open to scan for aortopulmonary collaterals from the vertebral and subclavian artery. In PCPC/TCPC, the window of interest is opened up even more in the cranial and caudal aspect to be able to scan veno-venous or veno-left atrial collaterals. In aortic arch and CoA, the window will be reduced to the arch/CoA region only. Shutters and lateral collimation cannot be applied during 3DRA since those would block the scan of the frontal when rotating to the lateral aspect.

58.3.6 Hounsfield Units and 3DRA

Technically 3DRA is comparable to a CTA available at the cathlab within one system. If the raw data in the rotational angiography and MIP planes look grey-in-grey, then postprocessing the volume rendered 3D dataset will not visualize anatomical figures sufficiently due to the lack of a good contrast ratio between the substrate and the surrounding. If the raw angiographic data look more black-and-white due to a high contrast ratio, then postprocessing and visualization of the target structure will be easy. The natural properties of a tissue and the contrast density during angiography determine at which specific range on the Hounsfield scale this tissue appears during X-ray (Fig. 58.6).

Coarctation stenting is an ideal indication to study the use of 3DRA because the aorta is a simple tube with only a few branches. When during rapid pacing and breath hold contrast is injected into the left ventricle than the entire aorta is filled homogeneously

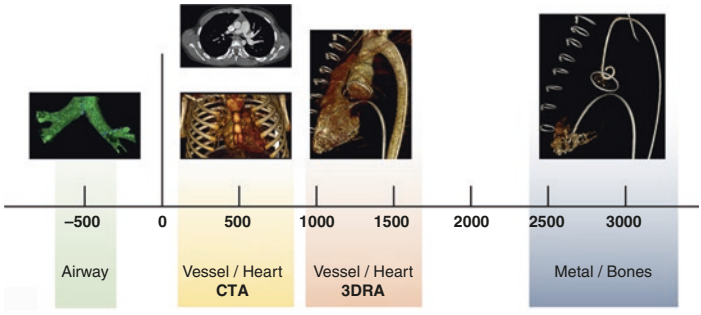

Fig. 58.6 Different tissues and their typical Hounsfield ranges in CTA versus 3DRA

with contrast and a high contrast ratio will allow for a quick postprocessing, understanding of the lesion, and use of a 3DRA overlay to guide the intervention. This is completely different when performing 3DRA in pulmonary artery interventions. The pulmonary arteries are not a simple tube but a complex double tree with delicate branches and capillary vessels. Pulmonary arteries during RA will have much more contrast distribution thus a much lower contrast ratio compared to the aorta. Simplistically the aorta is black-and-white, whereas pulmonary arteries are more grey-in-grey. Thus Hounsfield units representing the aorta are on a higher scale than those representing pulmonary arteries. Knowing this difference is crucial when using vendors presets to delineate specific tissues and even more when presets fail to do so.

Manual adjustment of the so-called "tables" should be trained to adapt the tissue of the 3D volume to its Hounsfield range. In the beginning, one should work with a single table and a single color to understand the relation between Hounsfield range and tissue representation (Fig. 58.7). When a table is setup like a ramp (one value with infinite range to the left or right), the structure represented by this Hounsfield range will appear solid (Fig. 58.7b–d). A trapezoid setup with a minimum and maximum cutoff value will create a hollow structure if both values are approximated correctly (Fig. 58.7a, e). This often is the best choice for roadmapping and reduces a structure to its outline. If a solid setup with a

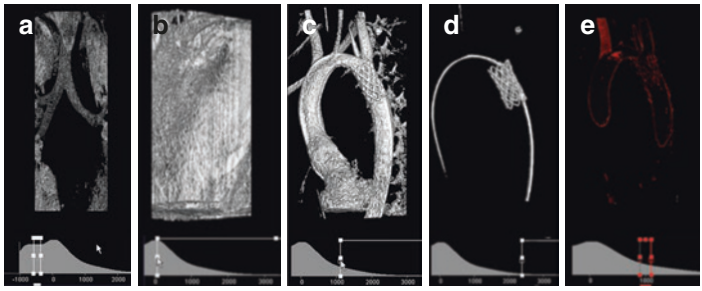

Fig. 58.7 3DRA in CoA stenting, contrast injection in LV under rapid pacing; same dataset and angulation with five different table settings on different ranges of the Hounsfield scale: (**a**) trapezoid −600 to −400, (**b**) ramp >50, (**c**) ramp >1000, (**d**) ramp >2300, (**e**) trapezoid 900–1100. Ramp: creates solid structures, Trapezoid: creates hollow figures

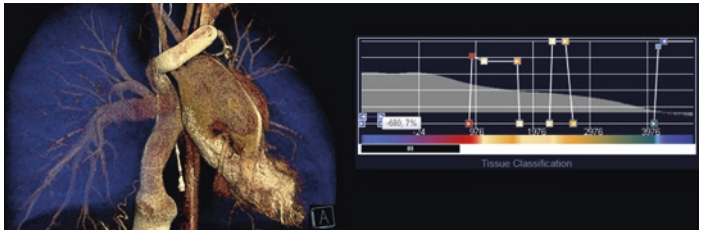

Fig. 58.8 DILV after DKS and TCPC: complex table setup to visualize tissue at specific Hounsfield ranges

ramp is used for 3D roadmapping, the closed surface of the structure (i.e., aortic arch) will block the vision of catheters, wires, and devices. Ramps can also be used as an endoscopic visualization to "fly through" stenotic vessels, complex outflow tract morphology, a VSD or to detect airway obstructions from the virtual inside of the lumen being compressed by a lusoric artery from outside.

Presets with multiple colors and various tables may seem attractive and beautiful but are useless for interventional purpose since adjustments are complex and difficult to understand (Fig. 58.8).

58.3.7 Postprocessing of a 3D Volume Dataset

The workflow of postprocessing a 3D volume differs from vendor to vendor. It is important to get familiar with the capabilities of the workstation being installed. Some vendors offer segmentation tools which identify anatomic structures based on their homogenous HU and sufficient contrast ratio. After measuring and editing, the results of the segmentation can be superimposed on the conventional angiographic system. However, the lower the contrast ratio compared to the surrounding tissue the less accurate the result of segmentation is. It is important to notice that segmented figures are non-dicom data which can differ in shape and dimension from the dicom raw data and the patients anatomy. Other systems avoid segmentation and offer scissoring tools to isolate target structures. Some vendors offer 3DRA systems with two separate channels for independent postprocessing with scissoring, Hounsfield units adjustment, etc. This feature allows to postprocess volume datasets with simultaneous multi location contrast injections, i.e., working with pulmonary arteries in channel A (lower HU) and the aorta in channel B (higher HU, Fig. 58.9a). When importing a preprocedural CTA, the volume rendering of the CTA can be done in channel A and a current 3DRA superimposed in channel B (Fig. 58.9b). Other options are the pre/post interventional comparison in channel A/B (Fig. 58.9c) or great arteries in channel A and the left bronchus compromised by aortic arch stenting in channel B (Fig. 58.9d).

58.3.8 Import CTA/CMR with Image Fusion

Fusion or *merge* describes the process when a CTA or MRA is imported to a 3DRA workstation and used with or instead of a 3DRA. The process of adjusting a 3D volume to the current patient anatomy is called registration. When CTA data are imported, then image fusion is based on the alignment of structures

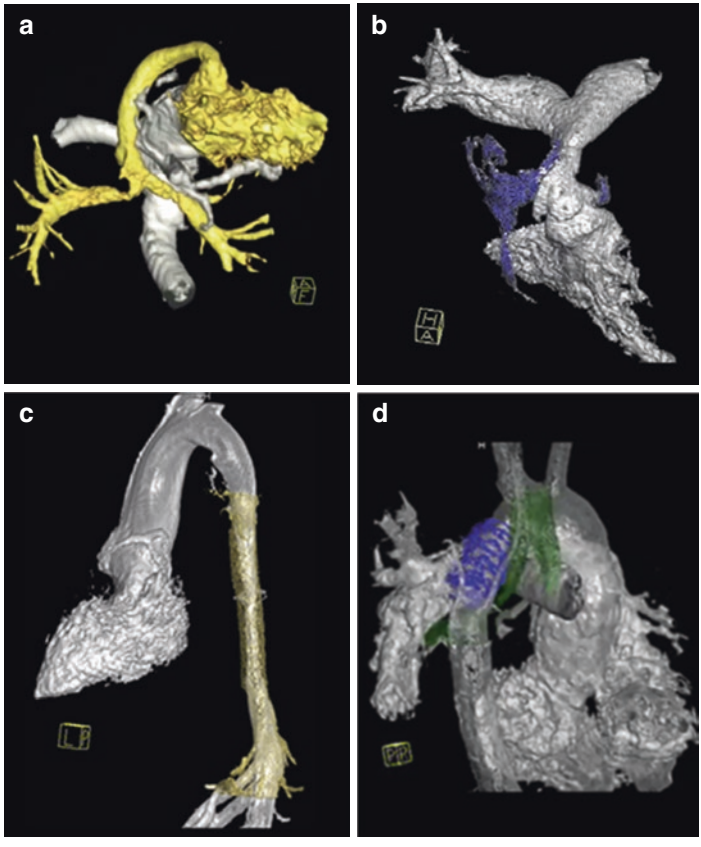

Fig. 58.9 Working with two independent 3D volume channels. (**a**) RV, Sano shunt and PAs (yellow), Norwood aorta (silver). (**b**) RVOT, RPA, and LPA (silver, CTA), aortic calcification (blue, 3DRA). (**c**) midaortic stenosis (silver), result after multiple stents (yellow). (**d**) left bronchus compression (green) after aortic arch stenting (silver blue)

with high HU as bones or metal. All workstations offer manual and automatic registration tools to perform this process. When CMR data are imported, registration cannot be performed based on high HU but has to be done manually by alignment of airway components from the CMR with the current patient airway

anatomy (LIT Fagan, CMR fusion). In selected cases, an intervention can be based exclusively on a CTA after registration (S. Goreczny, PDA closure, Philips VesselNav Lit). Registration can be challenging when body position and breathing status during CTA or MRA acquisition were different compared to the current patient position. Positioning the arms high or low does change the aortic arch geometry and make proper alignment during fusion imaging cumbersome. Once image fusion is performed, the merged dataset can be used for 3D roadmapping instead of or together with a regular 3DRA dataset.

58.3.9 Working with Clipping Planes

Clipping planes are virtual cross sections which can be applied to the 3D volume data in a frontal, lateral, or cranial orientation. Clipping planes are helpful to get a first idea of a region of interest without excessive scissoring. A frontal clipping plane for example can be moved as a vertical wall from anterior to posterior to make structures invisible being anterior from this plane. All workstations offer multiple clipping planes which can be angulated without restrictions. When preparing an anatomic region for roadmapping, a clipping plane can be set up as a virtual cut through the midline of an anatomic structure. In PPVI roadmapping for prestenting and valve implantation often is ideal from a left lateral view. When the stenotic MPA region is postprocessed as a hollow structure (trapezoid setting of the table) a left lateral cross section can be moved into the mid MPA position to virtually cut the hollow MPA in halve which will then result in an outline shape. With careful angulation the cross section can be centered at the MPA and be used for optimal overlay.

58.3.10 3D Roadmap

The use of a postprocessed 3D volume on the angiographic system for 3D roadmapping is a key feature in the 3DRA workflow.

In the 3DRA systems that are currently available, the 3D volume is projected to the frontal angiographic plane only and synchronized during angulation. The opacity or translucency of a superimposed 3D image can be adapted to allow identifying wires, catheters, and devices during intervention. The perfect match of the 3D image with the current anatomy should be checked before stent or device placement by short manual contrast injection through the delivery sheath and manual correction of the 3D volume in regard of the current anatomy. Up to now none of the available 3DRA systems is able to compensate the 3D roadmap for motion which inevitably occurs during every heart cycle and breathing but also when advancing stiff wires, catheters, or devices. Performing a 3DRA with stiff wires and long sheaths in place can minimize anatomic shift during guidance of the intervention for the price of a distorted non-native anatomy. Since the 3D roadmap will only appear in the frontal plane, it can be helpful to angulate the frontal plane into a lateral position to be able to use the 3D landmarks for interventional guidance.

58.3.11 Setup of the Rotational Angiographic System

It is recommended to ask the vendor of a specific 3DRA system to customize the setup with predefined programs related to patients' weight (3–5, 5–10, 10–30, above 30 kg), adapted filter and tube parameters to offer high spatial resolution with lowest radiation. The radiation dose of a well-adapted 3DRA system will result in 0.4–0.6 mSv or even less for one biventricular rotational angiography which is comparable to a biplane angiography of 10 s with 15 f/s or a state of the art CTA (literature : Stelt/Minderhoud dramatic dose reduction …). Some 3DRA systems offer 30 or 60 frames/s acquisition during rotational angiography. However, 30 f/s are sufficient and deliver enough spatial resolution in newborns as well as in adults. 60 f/s will unnecessarily double the radiation dose.

58.4 3DRA in Typical Lesions

58.4.1 Coarctation of the Aorta

Coarctation of the aorta is an ideal lesion to start with the implementation of the 3DRA workflow. Rapid pacing as well as ventilator stop is recommended as in all rotational angiographies although personal preferences may lead to a different workflow. LV contrast injection (40% diluted with NaCl 0.9%) should be performed with LV contrast injection timed 1 s before the scan starts (1 s X-ray delay, Fig. 58.10). In our experience, the visualization of coronary arteries, aortic valve cusps, and intercostal arteries does indicate high detail of the acquired image data. The goal of any kind of 3D imaging is the visualization of the CoA region, aortic arch hypoplasia ostium of the subclavian artery as a landmark for stent positioning. Imaging data acquired pre cath as CTA or CMR can be imported with image fusion/merge and superimposed for roadmapping as well as a 3DRA.

Studying the aortic arch and CoA from lateral but also cranial and posterior does offer complete understanding of the anatomy. A short manual contrast injection should be performed through the long delivery sheath prior to stenting in all kinds of 3D roadmapping to check for a perfect match of the 3D overlay with the current anatomy. Manual correction of the 3D overlay will allow for precise positioning based on the 3D image. Measurements of the vessel dimensions in the 3D data set tends to underestimate real proportions. Therefore proper inspection of the systolic diastolic aortic excursion during the rotational angiography is recommended and will help to find the adequate stent diameter. Depending on the excursions of the aortic wall and the prestenotic windkessel, a conventional biplane angiography can be performed to validate the 3D dimensions which are relevant for balloon or stent choice. For 3D roadmapping, a trapezoid table setup is helpful to convert the solid 3D volume into an outline figure. A left lateral clipping plane in a 60-degree angulation in the mid vessel portion often is ideal in a left aortic arch with the frontal plane moved into a 60-degree lateral position to enable the use of the 3D

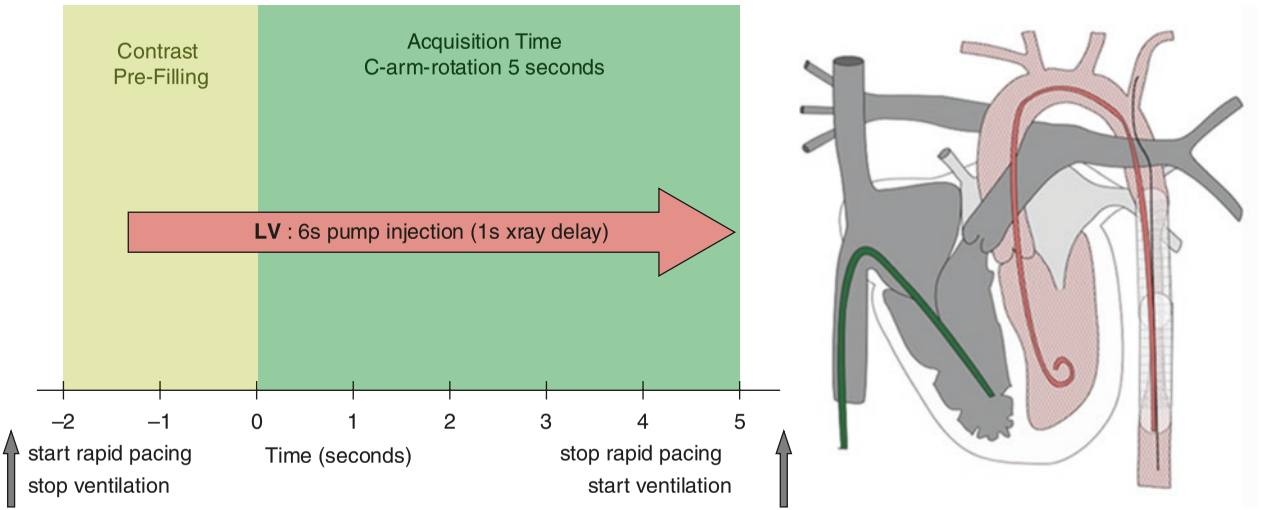

Fig. 58.10 3DRA in aortic lesions

roadmap in this plane. A post-stenting 3DRA with a collimated window of interest can be indicated to exclude early paravasation, dissection, or aneurysm formation (Fig. 58.11). A non-contrast 3DRA with collimation close to the stent margins can be used for high resolution 3D visualization to investigate stent fracture.

58.4.2 (Sub-)Atretic CoA

The workflow in (sub-) atretic CoA is more complex. Access from the right radial artery is used to place a pigtail catheter in the LV for pump injection of contrast during rapid RV pacing. With a second injection side contrast is injected manually and simultaneously via a long sheath in the highest aspect of the descending aorta with one 50 cc syringe. This dual location of contrast injection will visualize the entire aorta including the atretic segment and collateral arteries in a single 3DRA run. The radial artery side access is then used for antegrad perforation of the atretic segment and snaring of a perforation wire can be performed from the femoral side to establish a rail system radial-to-femoral with a long guide wire. Subsequently stent implantation can be done safely to using the rail system under 3D roadmapping (Fig. 58.12). A post-stenting 3DRA will allow to check for complete stent alignment with the aortic wall, aneurysm formation, or dissection.

58.4.3 RPA or LPA Stenting in TOF

MPA, RPA, or LPA stenoses are another typical indication for the use of 3DRA. Preprocedurally acquired CTA can be imported and used for fusion imaging. However, in peripheral multilevel PA stenosis (Alagille or Williams syndrome) postprocessing of the CTA dataset can be time consuming when pulmonary veins have to be scissored away to enable for a unrestricted view of the pulmonary arteries. When performing a rotational angiography contrast should be injected into the RV apex to fill the pulmonary arteries sufficiently with 2 s X-ray delay thus 2 s before the scan starts. The contrast injection can be stopped 5 s after injection

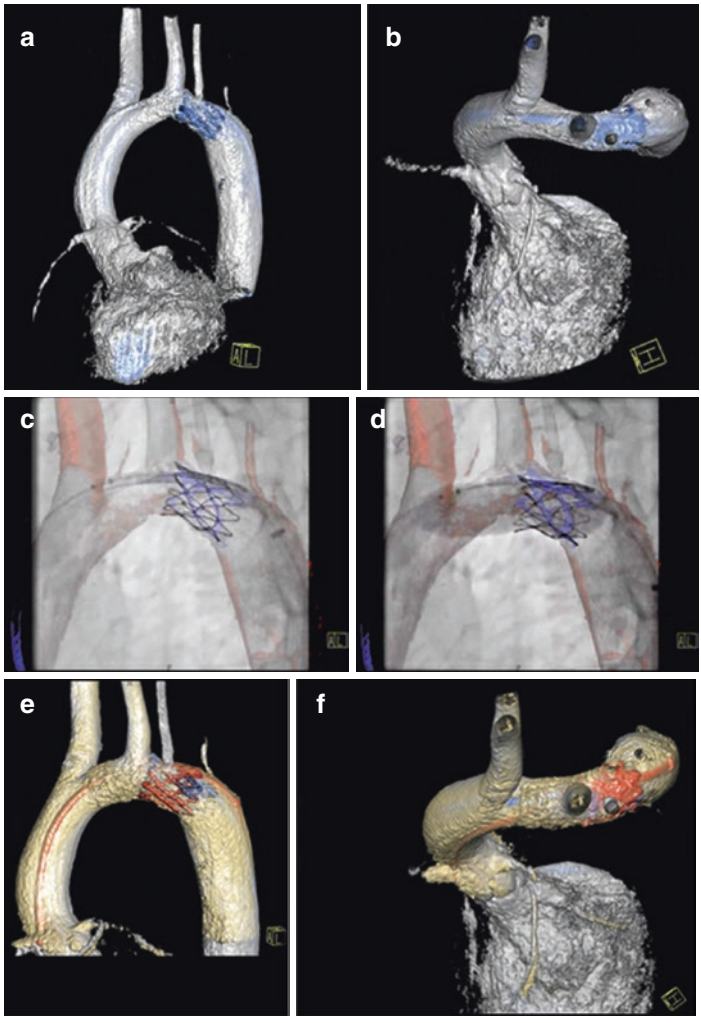

Fig. 58.11 Case: aortic arch hypoplasia after CoA stenting, 3DRA before aortic arch stenting (**a**, **b**), 3D roadmapping for stent positioning (**c**) and during implantation (**d**). 3DRA after aortic arch stenting and strut opening to left carotic artery (**e**, **f**)

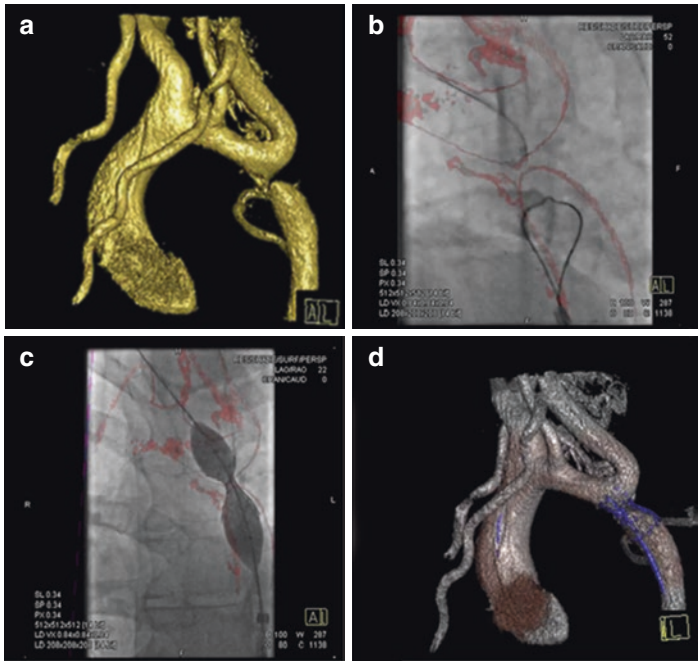

Fig. 58.12 3DRA in atretic CoA. (**a**) 3DRA with AoDesc (art fem) and LV apex contrast (art radialis dextra). (**b**) Snaring after antegrad perforation (sharp end of coronary wire). (**c**) Implantation covered CP stent. (**d**) Pre - post 3DRA overlay

thus 2 s before the scan ends. If in doubt of interaction with coronary arteries, a simultaneous LV or at least selective coronary injection should be performed with injection start 1 s after RV injection thus 1 s before the scan (image). If RPA or LPA stenting is intended in a postoperative situs after major aortic surgery (Ross, TAC correction, IAA, Yasui, Nikaido, Rastelli) then evaluation of coronary arteries and airway is recommended to avoid compression by implanting a stent in proximity to those structures. Balloon interrogation can be useful prior to stent implantation to identify critical diameters and strategies as oval stenting can be applied (Lit Krings et al.). In complex PA bifurcation stenosis involving the distal MPA and both branches 3D imaging is

extremely helpful to understand the offset of RPA and ostium, twisted branch anatomy, of set of upper and lower lobe branches and to apply techniques as Y-stenting or double/kissing balloon stenting (Lit Y Stent, Fig. 58.13). In patients with peripheral multilevel stenosis (Alagille), 3D imaging can shorten procedural and fluoroscopy time by guiding wire and balloon placement in different peripheral branches efficiently. Preparing a separate RPA and LPA 3D image for roadmapping can be useful with the contralateral PA side not being superimposed unnecessarily.

58.4.4 PPVI

The use of 3DRA during PPVI is based on a two-step workflow. First the entire anatomy is manually visualized either by importing a CTA or performing a 3DRA with RV apex and LV apex contrast injection during rapid pacing following the "Entire workflow" described in 6.4 for complex PA stent procedures. The CTA data or biventricular 3DRA will allow for identification of the anatomy in the region of the PPVI landing zone (calcification in grafted MPA versus large RVOT, coronary anatomy with risk for compression). In the second step, a 3DRA with balloon interrogation (30% contrast, 70% NaCl%) and simultaneous selective coronary injection (manually, pure contrast) can be very helpful to proof for coronary compression. Rapid pacing is not needed since the balloon does temporarily block the cardiac output. Maximal collimation should be applied close to the length of the balloon in the window of interest. In large RVOT balloon interrogation can be performed through a long sheath to facilitate contrast injection in the RVOT during balloon occlusion to demonstrate if the chosen balloon size is occlusive in the MPA (Fig. 58.14). The resulting 3D image will show the RVOT-balloon-coronary relation and allow to identify proximal and distal end of the landing zone, (sub-) total MPA occlusion, and possibly the presence of critical aneurysm formation in the MPA. In calcified conduits, the amount and distribution of calcification can be visualized by an optimized table setup with careful manual Hounsfield adjustment (Fig. 58.15).

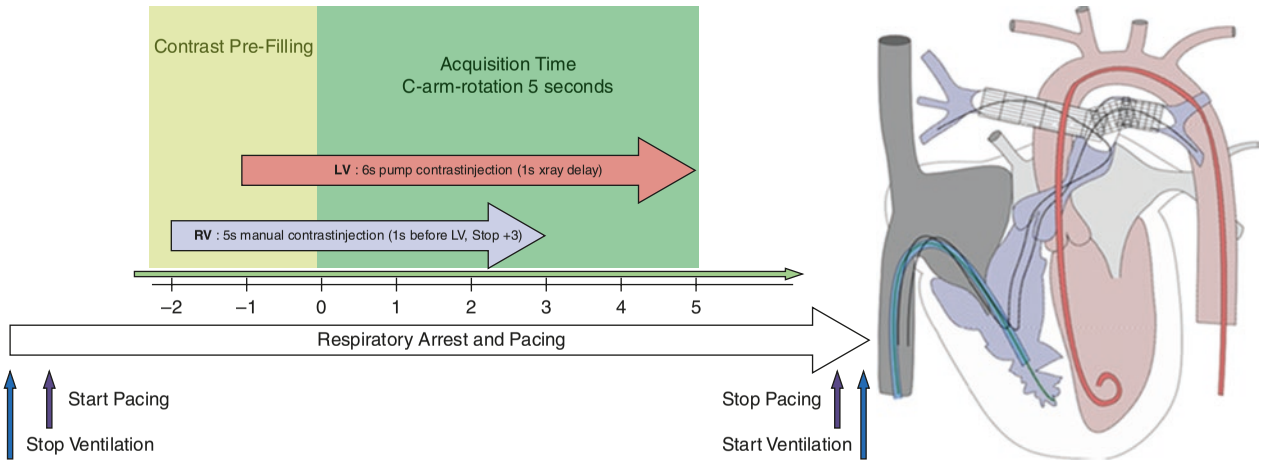

Fig. 58.13 Y-stenting of the PA bifurcation in distal MPA, RPA, and LPA stenosis. RV apex contrast injection (manually, long sheath, −2s to +3s related to scan) RV apex rapid pacing (4 Fr electrode though long sheath) LV with contrast injection (injector, −1 s to +5 s related to scan) ventilation and rapid pacing timing following symbolic markers

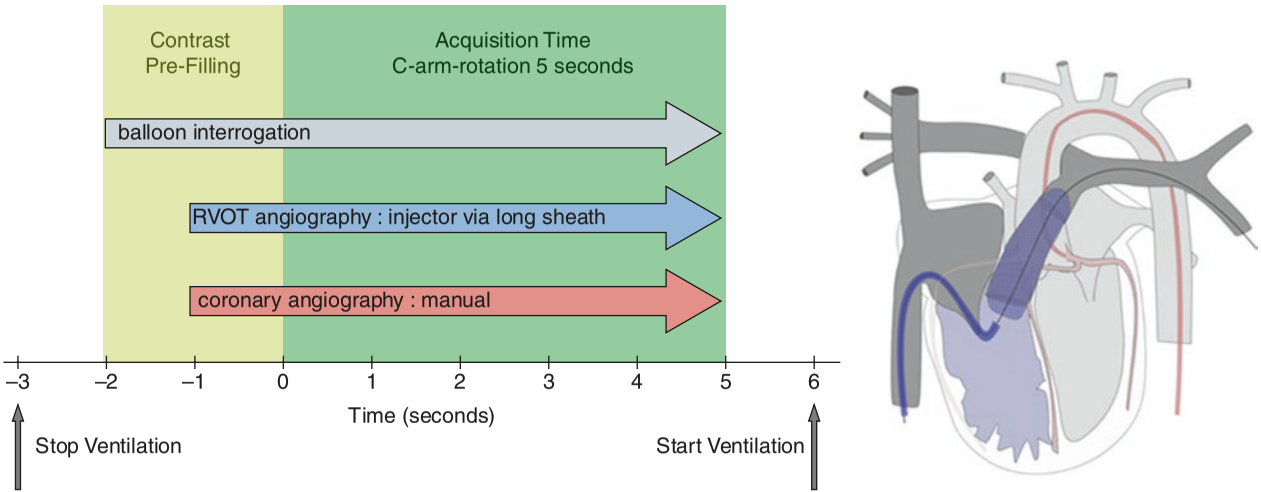

Fig. 58.14 3DRA workflow "balloon interrogation" no rapid pacing needed due to "0" output during balloon occlusion RVOT contrast injection (50 ml, 40% diluted through RVOT sheath) selective coronary angiography (pure contrast)

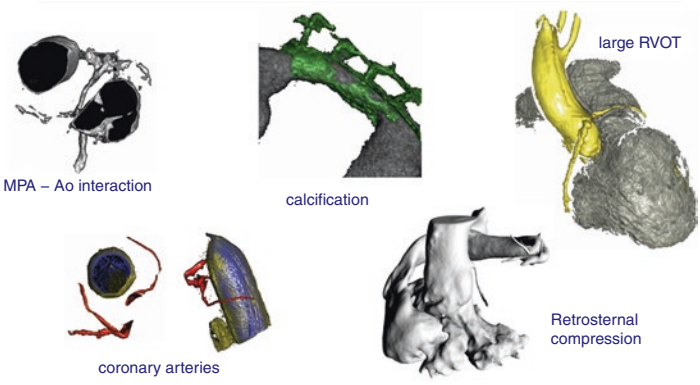

Fig. 58.15 Examples for characteristic interactions during 3DRA in PPVI as vessel-vessel, vessel-calcification, vessel-balloon, or vessel-bone

Patients with a combined volume and pressure loaded RV often indicate to be vulnerable for ventricular tachycardia during rapid pacing by showing lots of ventricular extrasystole and short VT runs during wire and catheter manipulation. In this case, RV rapid pacing can be performed but special attention should be put on post-pacing ventricular tachycardia, fibrillation, and Torsade de Point, possibly needing external defibrillation (Fig. 58.16).

58.4.5 Single Ventricle Stage I/Pre-Stage II

It is not the purpose of this 3DRA chapter to answer the question of the ongoing controversial discussion whether invasive catheterization prior to stage II palliation in single ventricle is useful or not. In critical patients after Norwood palliation low saturation can occur and delineating shunt or/and PA branch stenosis is crucial. This can be done by CTA or—when indication for cath is already made—during cath by performing 3DRA. It seems obsolete to perform the complex 3DRA workflow in a critical ill newborn. However, the experience of different center gives proof for the benefit of this approach especially in those fragile babies in

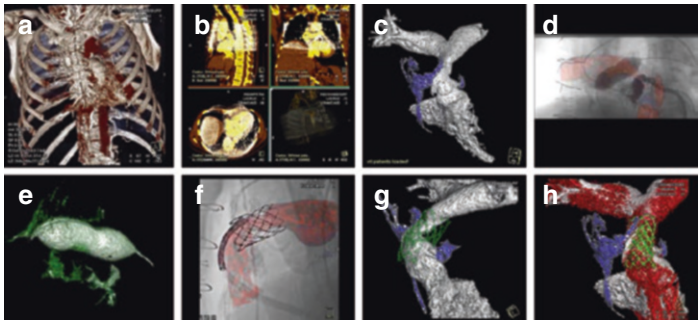

Fig. 58.16 3DRA workflow in PPVI, example with CTA imported on 3DRA workstation. (**a**) CTA postprocessing on 3DRA workstation. (**b**) Process of registration : CTA with 3DRA (balloon interrogation with LCA). (**c**) Visualization of severe para-aortic and homograft calcification (blue). (**d**) Balloon interrogation with LCA contrast. (**e**) Balloon interrogation postprocessed with LCA. (**f**) 3D guided prestenting (CP covered) and Melody valve implantation. (**g**) Post-PPVI 3DRA : pre-PPVI image **c**) superimposed with post-PPVI stent. (**h**) Post-PPVI 3DRA with vessel (red), calcification and stent

the early postoperative course. The reason is the all-in-one run characteristic of 3DRA when performing properly. This includes rapid esophageal or RV apex pacing (often up to 220–240/min, ideally through a 5 Fr blue long sheath with a 4 Fr pacing electrode advanced through the sheath) and additional contrast injection through a pigtail catheter in the aortic arch (Figs. 58.17 and 58.18).

58.4.6 Single Ventricle Stage II and III

3DRA in Stage II and III is complex due to timing, multi contrast location, and a complex postprocessing. Manual Hounsfield adaptation is crucial. Contrast should be applied at the locations described in Figs. 58.19 and 58.20. Pacing in TCPC can be achieved by esophageal approach. Special attention should be given to airway visualization since left bronchus compression can occur in LPA stenting especially in DILV with DKS anastomosis

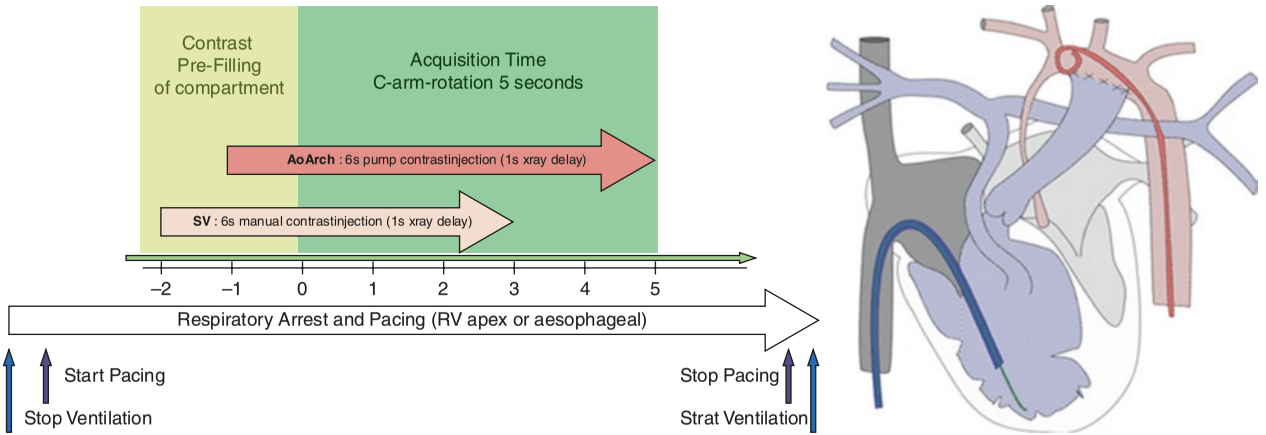

Fig. 58.17 3DRA workflow in Stage I in a 3 kg child: rapid pacing 240/min through 5Fr long sheath in RV apex contrast manually 2 ml/s = 12 ml total with 2 ml/s in RV apex contrast 2 ml/s = 12 ml total in AoArch with injector

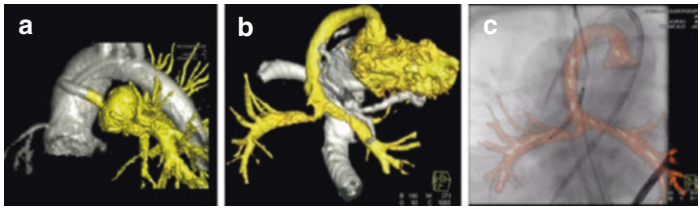

Fig. 58.18 Stage I cases 3DRA: (**a**) SV with central AP shunt, severe discrete distal anastomosis stenosis. (**b**) SV with Sano shunt, proximal RPA stenosis in caudal angulation. (**c**) Same newborn as (**b**), 3D roadmap with stent implantation

and in HLHS after Norwood procedure. Veno-atrial collateral vessels will be visible if the window of interest is adjusted correctly and the timing applied as demonstrated in the schematic drawing (Fig. 58.21).

58.5 Check List: 3DRA Step by Step

58.5.1 Before 3DRA

Anesthesia

- Position of all equipment outside the critical corridor?
- Defibrillator checked?
- ECG, ABP, and CO_2 monitoring faultless?
- Cables and ventilator tubes unaffected by frontal plane rotation?
- Patient arms outside the rotational area of the frontal plane?

Cath team

- Correct program chosen?
- Collimation adapted?
- Rapid pacing: check electrode position, detect threshold
- Perform rapid pacing, 180/min upwards until systolic ABP decrease to 60%

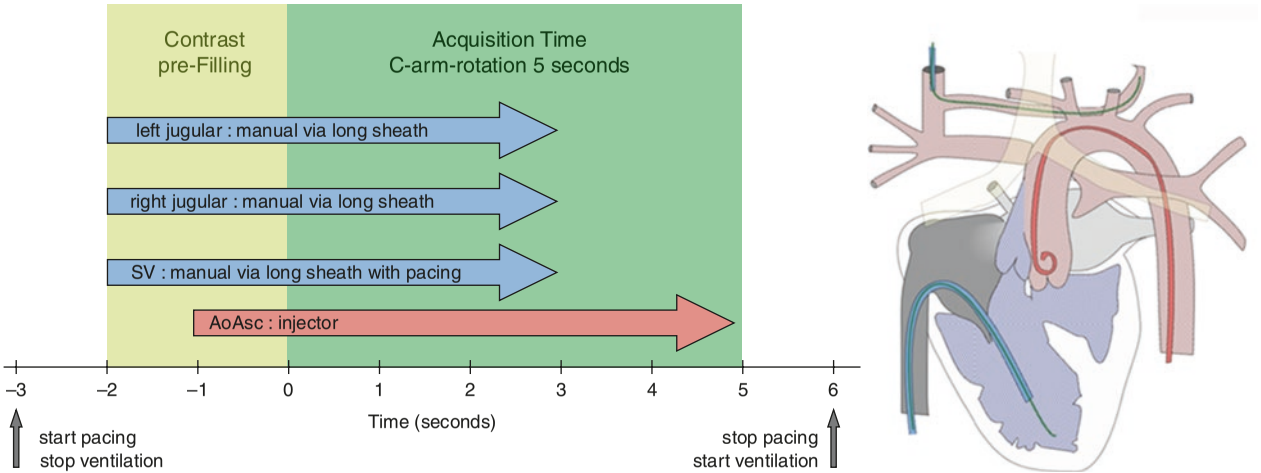

Fig. 58.19 3DRA workflow in Stage II/PCPC, calculation for 5 kg child contrast applied at four locations for best imaging results: left and right jugular vein manually, each 3 ml/s = 15 ml, SV manually 4 ml/s = 20 ml, AoAsc injector, 3 ml/s = 15 ml to visualize DKS/Norwood stenosis

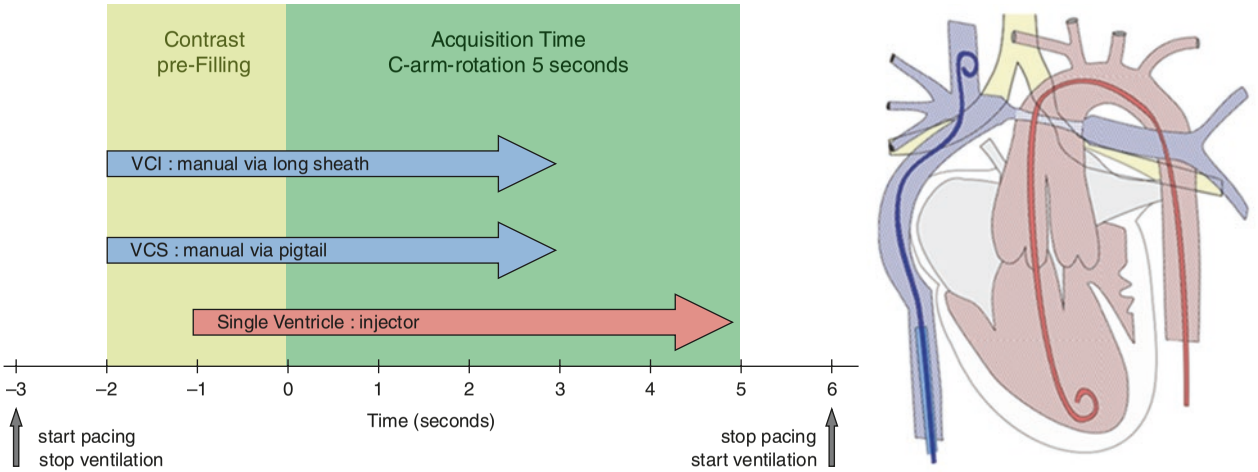

Fig. 58.20 3DRA workflow in Stage III/TCPC, calculation for 20 kg child contrast applied at three locations for best imaging results: superior caval vein (pigtail, 6 ml/s = 30 ml, manually) inferior caval vein (long sheath, pigtail, 6 ml/s = 30 ml, manually) SV (injector, 10 ml/s = 60 ml)

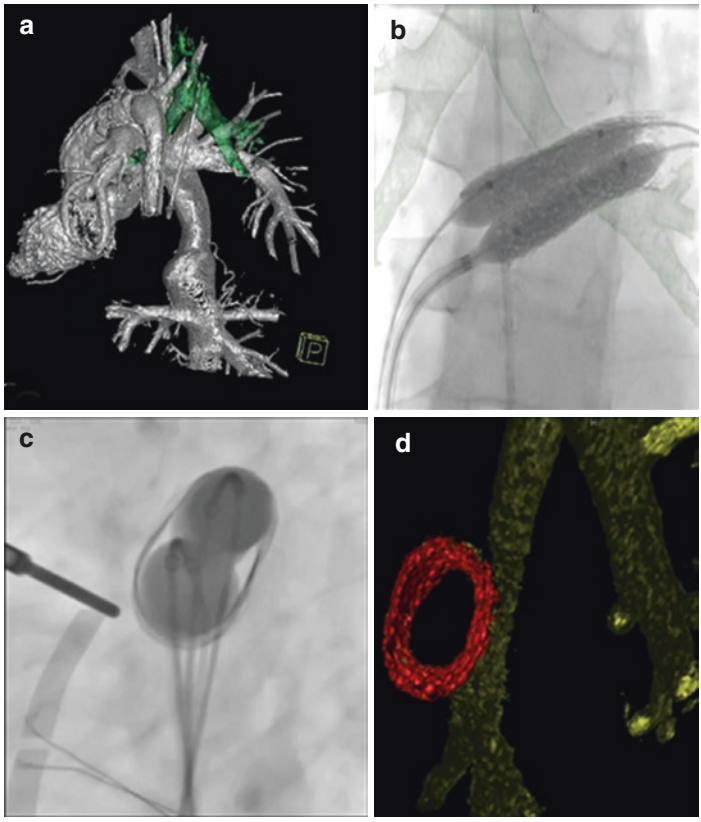

Fig. 58.21 3DRA in DILV, Stage III/TCPC with LPA stenosis. (**a**) Posterior view of TCPC with airway (green), left bronchus compression. (**b**) Oval stenting with double balloon on ev3 Mega LD 26 stent, AP view. (**c**) Lateral view. (**d**) 3DRA post oval stenting, oval shape prevents left bronchus compression

- Catheter position for contrast injection in the cavity proximal of the ROI?
- Contrast timing calculated (RV −2 s, LV −1 s, scan start at 0 s)?
- Contrast diluted to 60%, syringes ready ? all connections de-aired?

- Contrast injector active with setting checked?
- Contrast injection lines open?
- Injector and cine synchronized ? X-ray delay correct?
- Second capture test

Performing 3DRA

- The following commands are helpful:
  - Stop ventilation
  - Start rapid pacing (*wait a few beats to ensure stable capture)*
  - … 21, 22 start injection RV (manual), 23 start injection LV (injector) … 24 scan.

After 3DRA

- Check ECG, ABP, CO_2
- Withdraw pacing electrode
- Bring frontal and lateral plane back in position

58.6 List of Pitfalls

Low quality resolution and suboptimal volume rendering can be caused by:

- Contrast injection location at the region of interest and not in the proximal cavity
- Contrast injection started late (without 1 s prefill) or stopped to early
- High blood flow at region of interest not reduced due to slow rapid pacing
- Low blood flow at region of interest reduced too much due to very high rapid pacing
- Non-pacing capture during entire/partial scan time
- Low contrast concentration (below 50% dilution)
- Very high contrast concentration (scattering artifact)

Movement artifacts and scattering can be caused by:

- Patient moving during scan
- No breath holding which will result in low spatial resolution, unsharp vessel borders,
- Ascending aorta against RPA due to insufficient rapid pacing

High radiation dose

- Wrong program chosen
- Not enough frames acquired during pre-3DRA adjustment phase
- High contrast concentration
- Lots of metal in the scan circuit

Rhythm disorder

- AV block due to pacing electrode malposition
- Inconstant pacing due to instable electrode
- Sustained VT/VFib induced by rapid pacing in severely decreased RV function

CTA or CMR fusion/merge doesn't match current anatomy

- CTA performed with different patient position (arms low/up, body not straight)

Critical patients when indication for 3DRA has to be made with special care

- Williams syndrome with severe supravalvular aortic stenosis
- In patients with severely decreased RV function (CAVE VT/VFib/TdP)

59 Intracardiac Echocardiography

Jason H. Anderson and Allison K. Cabalka

59.1 Introduction

Intracardiac echocardiography (ICE) can be used for procedural guidance and device evaluation in many types of congenital and structural heart disease interventions. Miniaturization of phased-array probes, improvement in image quality, and ongoing development of three-dimensional capabilities have all contributed to ICE becoming a widely utilized ultrasound based imaging adjunct in the catheterization laboratory. As with all imaging modalities, there are advantages and disadvantages to consider when utilizing ICE. Established benefits include patient comfort (general anesthetic not typically needed for ICE as compared to TEE), capacity for single operator imaging, and superior near-field imaging of intracardiac structures without acoustic shadowing from extracardiac structures. The primary limitations are the need for an additional venous access site, limited depth of tissue

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penetration, single-use catheter design (cost implications can be alleviated by re-processing for sterile reuse), and the lack of a standardized approach to imaging technique. Specific training is often required to become confident with ICE.

59.2 Current ICE Systems

There are two types of ICE catheters, rotational and phased-array, available for commercial utilization. Rotational systems are not steerable with near-field imaging extending 5 cm circumferentially from the catheter producing cross-sectional images. They are cheaper and allow for adequate visualization of the atrial septum, which is the reason it has been predominately utilized for transseptal access in electrophysiology cases. The phased-array system is steerable and allows for far-field imaging, making it the more favored of the two catheter types among congenital interventionalists. Two phased-array systems are currently widely available: the AcuNav platform, compatible with the Acuson family of echo machines (Siemens-Acuson, Mountain View, CA) and the View Flex Xtra (Abbott Vascular, St Paul MN), compatible with ViewMate ultrasound consoles. Both systems are 90 cm in length with four-way tip steerability and 2D/pulse wave, Doppler and color Doppler capability. Longitudinal plane imaging only is standard for these systems. More robust three-dimensional ICE imaging systems are currently under development. The Acuson AcuNav ICE catheter is a newer 2D and 4D catheter that provides real-time three-dimensional ("4D") imaging. The 12.5 French catheter can only be used with the Acuson SC2000 platform provided specific software is installed. Currently the catheter is not able to be re-purposed so cost may be a significant barrier to widespread use. There is an advanced probe from Philips as well with 3D and multi-planar reconstruction and *x*-plane capabilities currently in development.

59.3 Description of Typical ICE Examination

The catheter is inserted into the femoral vein through an appropriately sized sheath (8 French or 10 French). Advancement to the right atrium is conducted under fluoroscopic guidance to reduce venous injury. Operators may utilize a moderate length sheath (30 cm) to ensure that the probe is advanced directly into the inferior vena cava rather than through the iliac vein. Once within the heart, a series of five different catheter positions allows for imaging of a majority of cardiac structures treated during transcatheter interventions.

- *Mid right atrium (RA):* In the mid RA in a fairly neutral position, the imaging palate is rotated toward the tricuspid valve for the standard "home view" (Fig. 59.1a). Clockwise rotation allows for visualization of the left ventricular outflow tract and aortic valve followed by the mitral valve and left

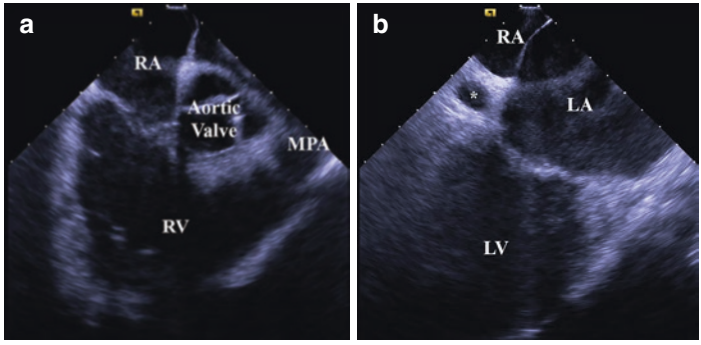

Fig. 59.1 ICE imaging from the mid RA. The imaging plane is directed at the tricuspid valve producing the home view. (**a**) This view results in an inflow-outflow view of the right ventricle with off-axis imaging of the aortic valve. Clockwise rotation through the cardiac structures will result in (**b**) a short axis view of the left atrium, mitral valve, and left ventricle. The coronary sinus is visualized in cross section (*asterisk*). *RA* right atrium, *RV* right ventricle, *MPA* main pulmonary artery, *LA* left atrium, *LV* left ventricle

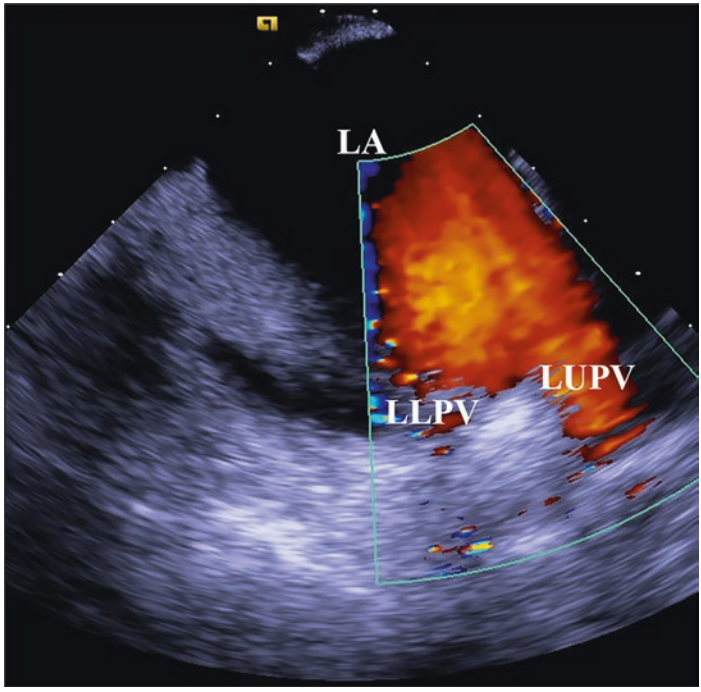

Fig. 59.2 Color flow Doppler image from the mid RA with slight posterior tilt opening up the ostia of both the left upper and left lower pulmonary veins as they return normally to the left atrium. *LA* left atrium, *LUPV* left upper pulmonary vein, *LLPV* left lower pulmonary vein

atrial appendage (Fig. 59.1b). A slight posterior tilt with advancement of the catheter demonstrates the left sided pulmonary veins (Fig. 59.2). Clockwise rotation is again performed to achieve a posterior sweep through the descending thoracic aorta, from left to right, to the right pulmonary veins (Fig. 59.3). Additional retroflexion may be necessary to visualize the right pulmonary veins in more detail. Advancing the probe cephalad will lead to visualization of the SVC and right pulmonary artery. The probe is then returned to the mid RA facing the aortic valve. Retroflexion and rotation of the

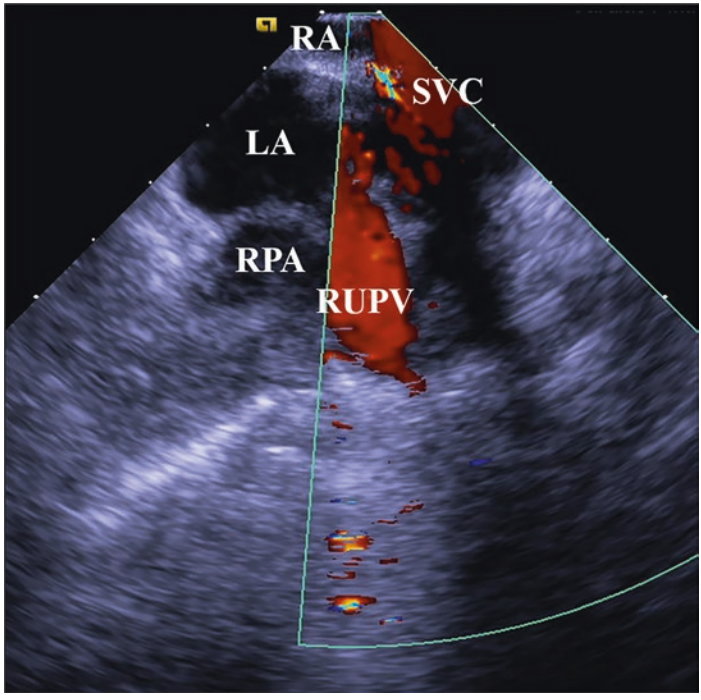

Fig. 59.3 Color flow Doppler image following advancement of the probe cephalad to the anastomosis of the SVC with the RA. The RUPV can be visualized entering the left atrium adjacent to the right pulmonary artery. *RA* right atrium, *LA* left atrium, *SVC* superior vena cava, *RUPV* right upper pulmonary vein, *RPA* right pulmonary artery

probe in this position provides detailed imaging of the atrial septum in a short axis and long axis plane (Fig. 59.4).

- *Low RA:* Withdrawal of the probe from the mid RA into the low RA provides additional imaging of the inferior and posterior portion of the atrial septum. Tilting maneuvers in this position can also provide a full sweep through the tricuspid valve leaflets.
- *Right ventricular (RV) inflow:* From the low RA position, anterior deflection is applied to the catheter with careful advancement just beyond the tricuspid valve. Clockwise

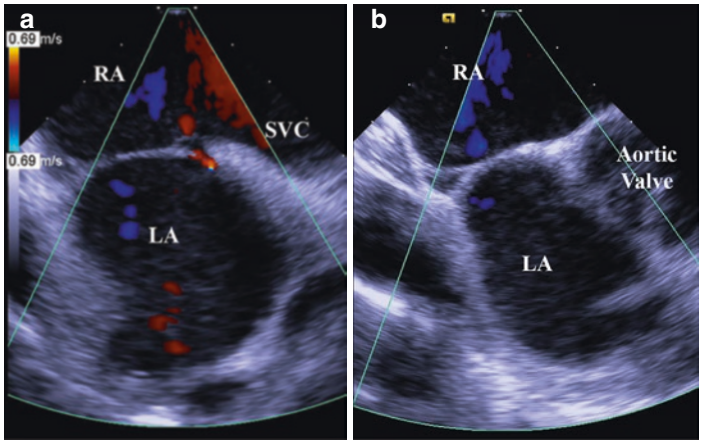

Fig. 59.4 Atrial septum visualized in (**a**) long axis and (**b**) short axis views. There is a small patent foramen ovale with left-to-right shunting. *RA* right atrium, *LA* left atrium, *SVC* superior vena cava

rotation with slow release of the tilt results in visualization of the left ventricle, interventricular septum, and mitral valve.

- *RV outflow:* From the RV inflow position, the probe may be advanced and rotated with full release of the anterior deflection achieving the RV outflow position. Alternatively, the maneuver can begin with the ICE catheter in the mid right atrium facing the tricuspid valve from the home view, and then rotating the probe 180°. The tip is then retroflexed and carefully advanced through the tricuspid valve into the mid right ventricle. Once the probe has passed the tricuspid valve, retroflexion can be relaxed, and the probe will settle into the subpulmonic position with the imaging plane directed toward the pulmonary valve.
- *Left atrium:* Advancement of the ICE catheter along a wire under fluoroscopic guidance through an atrial septal defect or transseptal puncture site results in catheter positioning immediately adjacent to the pulmonary veins, left atrial appendage, and mitral valve. A combination of tilting and left-right sweeping provides orthogonal angles to these structures.

59.4 Imaging During ASD Device Implantation

Evaluation of the ASD begins with the standard short axis and long axis views with imaging planes tailored to the anatomy at the time of closure. Rim deficiencies can be clearly identified (Fig. 59.5). ASD sizing is performed via static two-dimensional measurements and via stop-flow technique. Following device selection, ICE provides clear visualization of the left atrial disk of the ASD closure device as it comes into apposition with the atrial septum, ensuring appropriate position prior to delivery of the right atrial disk (Fig. 59.6). Following device deployment, a thorough evaluation of all septal rims should be completed. If the retroaortic rim is small or absent, the superior aspect of the left atrial disk may prolapse; this is easy to visualize via ICE imaging. The resolution of ICE also allows for visualization of the thin inferior septal tissue within the disks of the device to ensure adequate capture (Fig. 59.7). If it is not adequately visualized, a tug maneuver or partial retrieval and re-deployment of the right atrial disk can be performed. A post-implantation assessment via 2D and color flow Doppler for residual shunting concludes the intervention.

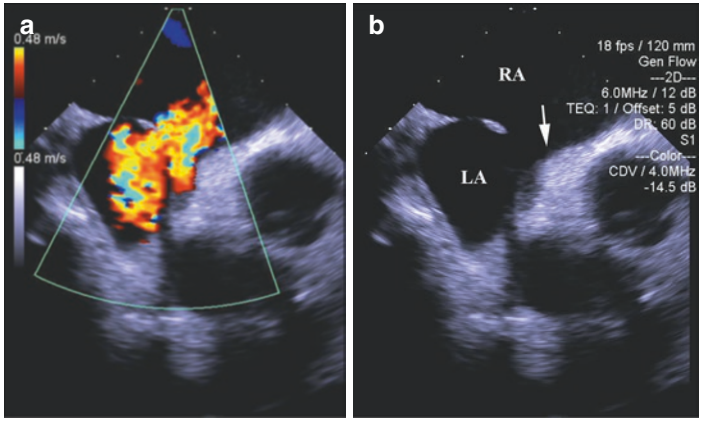

Fig. 59.5 Short axis ICE view of the atrial septum in the presence of a large secundum ASD. There is absence of the retroaortic rim (*arrow*). *RA* right atrium, *LA* left atrium

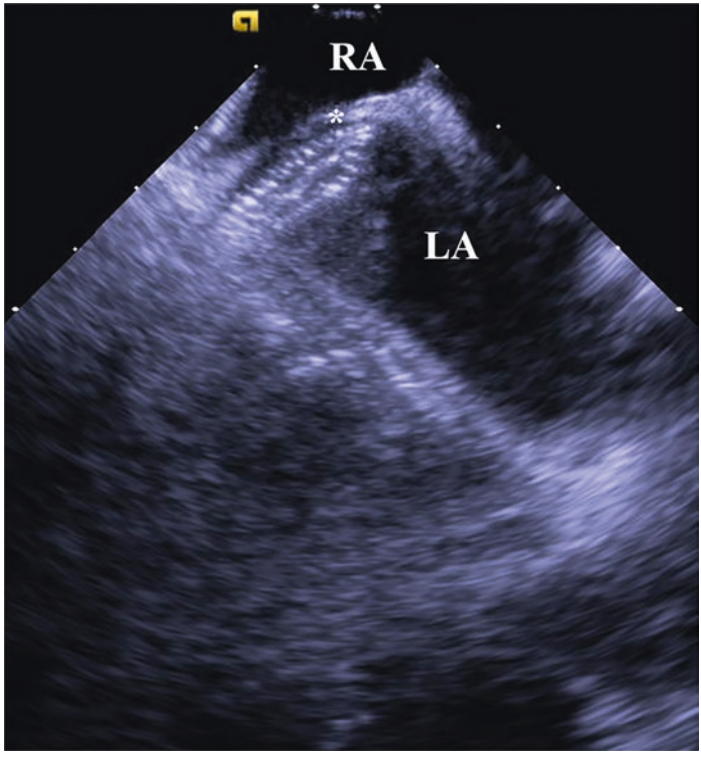

Fig. 59.6 Short axis view of the atrial septum during implantation of an Amplatzer Septal Occluder device. The left atrial disk is in apposition with atrial septum (*asterisk*) during deployment of the right atrial disk. *RA* right atrium, *LA* left atrium

59.5 Imaging During PFO Device Implantation

ICE guidance for PFO closure (Fig. 59.8) should include the following assessments: measurement of the PFO tunnel, excursion of the atrial septum (aneurysmal), and evaluation for additional fenestrations. To ensure full visualization for additional defects, a

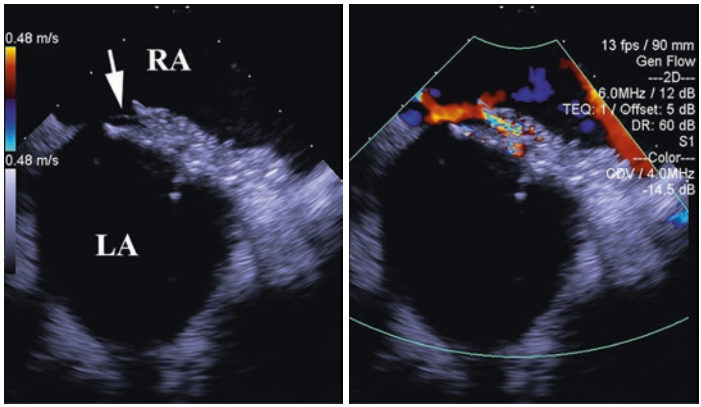

Fig. 59.7 Long axis ICE view of the atrial septum from the mid right atrium following implantation of a Gore Cardioform ASD Occluder. The inferior rim can be visualized (*arrow*) coursing between the left atrial and right atrial disks of the device consistent with adequate capture. *RA* right atrium, *LA* left atrium

central image in short axis of the atrial septum should be obtained followed by a left-right tilt of the imaging plane through the entire septum. A long axis of the atrial septum is then obtained with the same left-right maneuver documented, thus ensuring a full sweep of the atrial septum in two separate planes. Agitated saline injections with Valsalva maneuvers can be performed at baseline and immediately following device implantation to evaluate for right-to-left shunting (Fig. 59.9). Due to the position of the probe in the right atrium, the images may be obscured during administration of agitated saline for a bubble study. If this occurs, advancing the probe into the superior vena cava to evaluate the aorta for the presence of bubbles may provide additional confirmation of right-to-left shunt if needed. Balloon sizing of a PFO is rarely used. In patients with orthodeoxia syndrome, during spontaneous breathing, it is possible to document right-to-left shunt through a PFO (Fig. 59.10).

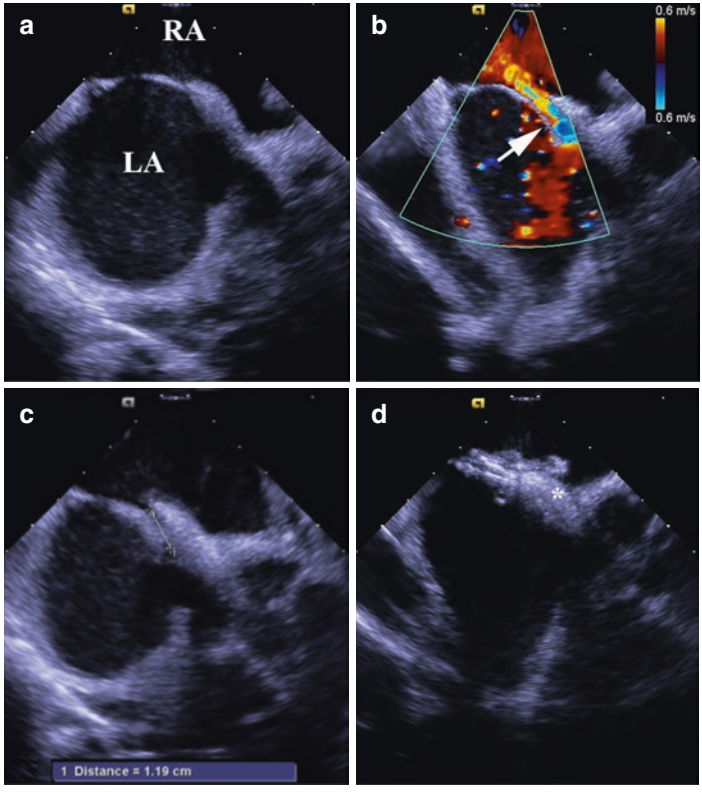

Fig. 59.8 Common images acquired during ICE guidance for PFO device closure. This assessment includes: (**a**) two-dimensional evaluation with sweep of the atrial septum, (**b**) color flow Doppler assessment of the atrial septum for additional fenestrations with a left-to-right shunt at baseline through the PFO (*arrow*), (**c**) measurement of tunnel length to determine optimal device selection, and (**d**) post device implantation assessment ensuring adequate capture of the septum secundum/lipomatous septum (*asterisk*) and inferior septal rim. *RA* right atrium, *LA* left atrium

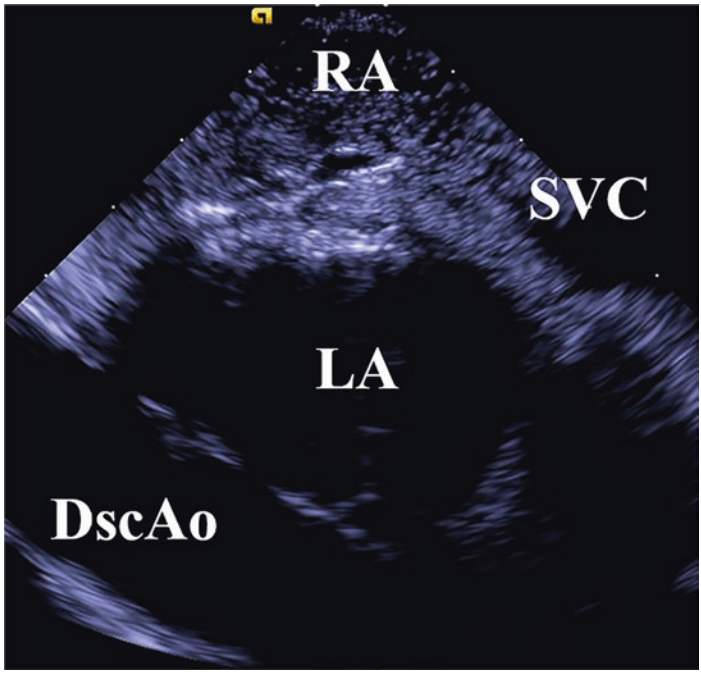

Fig. 59.9 Long axis view of the interatrial septum following PFO device closure utilizing a Gore Cardioform Septal Occluder device. Agitated saline injection has been performed with microbubbles present in the right atrium. There is no right-to-left shunting of microbubbles present. *RA* right atrium, *LA* left atrium, *SVC* superior vena cava, *DscAo* descending aorta

59.6 Imaging During Transseptal Puncture

Transseptal puncture can be guided by ICE with careful transition between long axis and short axis imaging planes. There is clear visualization of tenting of the atrial septum (Fig. 59.11a) prior to passage of the needle into the left atrium. Technically, this is slightly more challenging than with transesophageal echocardiography imaging where *x*-plane imaging provides simultaneous

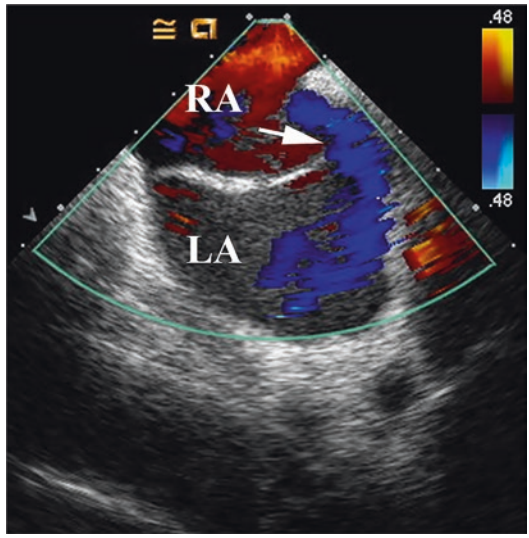

Fig. 59.10 Color flow Doppler assessment demonstrating marked leftward displacement of an aneurysmal atrial septum with right-to-left shunting in a patient with orthodeoxia syndrome. *RA* right atrium, *LA* left atrium

visualization of orthogonal planes. ICE can also guide subsequent interventions including dilation of the iatrogenic septal communication (Fig. 59.11b) or atrial stent implantation.

59.7 ICE Imaging for Evaluation of Valvular Abnormality

Intracardiac echo imaging is very useful to document the nature of presumed valve dysfunction, particularly in patients with bioprosthetic or transcatheter pulmonary valve replacements. ICE can assess leaflet thickness, mobility, and provide evidence into the mechanism of deterioration, thus supporting thrombus vs. vegetation (Fig. 59.12). This assessment may be difficult via transthoracic or transesophageal imaging due to tissue interference and artifact from the valve frame. ICE may also demonstrate

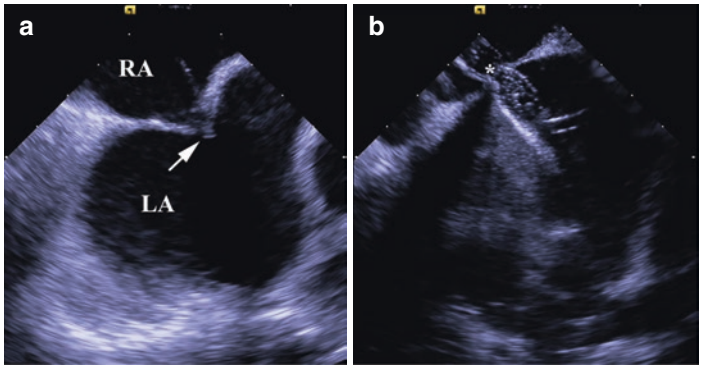

Fig. 59.11 (**a**) Long axis view of tenting of the atrial septum (*arrow*) just prior to transseptal puncture. (**b**) Balloon dilation of the atrial septum following transseptal puncture. There is clear visualization of the balloon straddling the atrial septum and resultant diameter achieved (*asterisk*). *RA* right atrium, *LA* left atrium

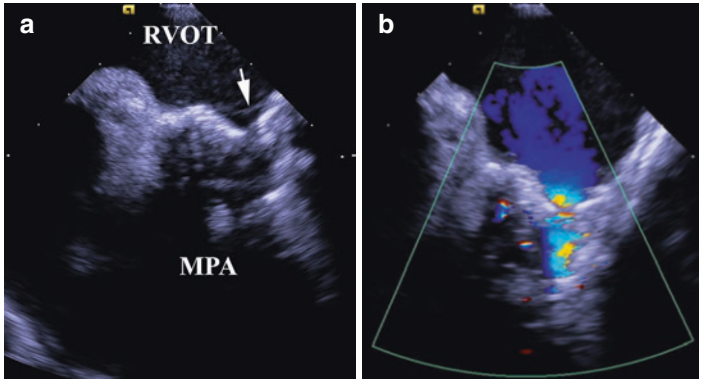

Fig. 59.12 ICE imaging of a bioprosthetic pulmonary valve 15 years after implantation. (**a**) There is severely limited leaflet excursion with the onset of ventricular systole (arrow) with the inferior leaflet fused in the closed position. (**b**) Color flow Doppler assessment demonstrates a small effective valve orifice with flow aliasing occurring at the leaflet tips. This constellation of findings is consistent with age-related degeneration of the valve. *RVOT* right ventricular outflow tract, *MPA* main pulmonary artery

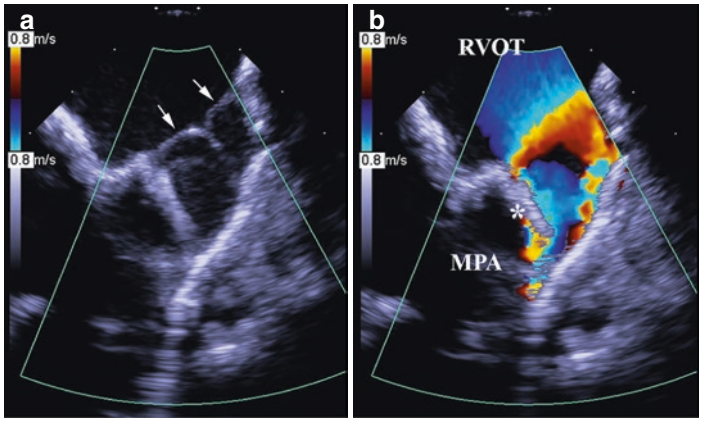

Fig. 59.13 ICE evaluation from the RVOT in ventricular (**a**) diastole and (**b**) systole. These images demonstrate normal bioprosthetic valve leaflet thickness and mobility with full coaptation (*arrows*). There is severe supra-valvar obstruction occurring between the valve struts of the bioprosthetic valve (*asterisk*) and hood of the main pulmonary artery. *RVOT* right ventricular outflow tract, *MPA* main pulmonary artery

that the valve leaflet function is normal with the primary issue being immediately sub-valvar or supra-valvar, altering the recommended intervention (Fig. 59.13).

59.8 Imaging During Transcatheter Valve Replacement

The role of ICE imaging in transcatheter valve implantation is primarily for assessment of the valve following implantation. The valve implantation procedure itself is predominately guided by fluoroscopic imaging in the current state of practice. ICE does allow for high-resolution imaging post valve implantation, thus serving as a baseline assessment for ongoing comparison (Fig. 59.14). This evaluation is comprised of visualization of leaflet size/mobility, valvular regurgitation, spectral Doppler assessment for valve gradient, and perivalvular leak. To date, ICE is most commonly utilized following transcatheter pulmonary valve

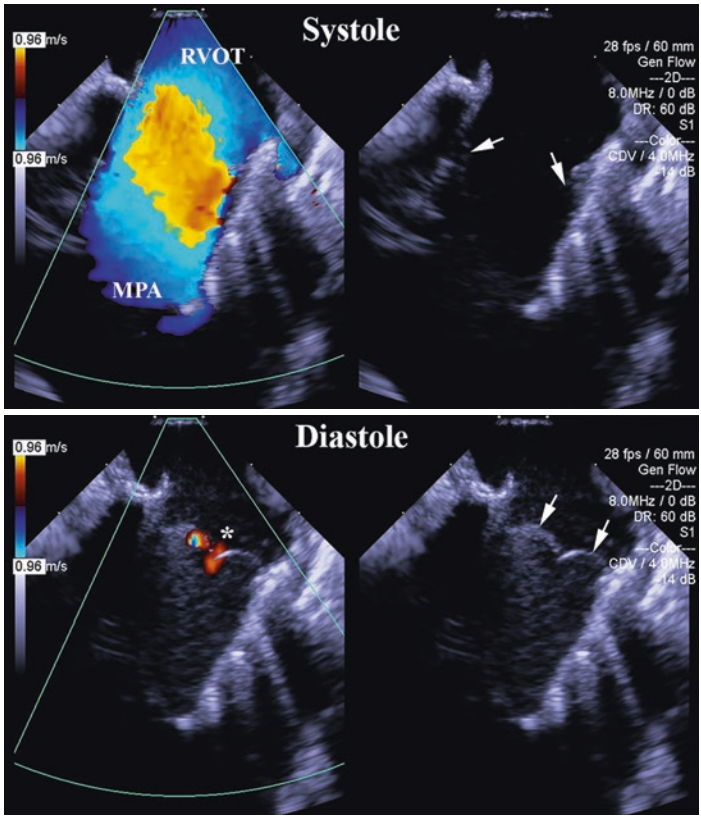

Fig. 59.14 ICE evaluation of a Melody valve implant following pulmonary valve-in-valve implantation. Two-dimensional and color flow Doppler assessment in ventricular systole (top panel) and diastole (bottom panel) demonstrate thin leaflets with full coaptation (*arrows*). There is trivial eccentric pulmonary valve regurgitation (*asterisk*) commonly observed following Melody valve implantation. *RV* right ventricle, *MPA* main pulmonary artery

replacement. This is in part due to the difficulty of imaging a pulmonary valve replacement via transthoracic and transesophageal echocardiography in certain scenarios, given the retrosternal location and orientation of the valve limiting the ability to obtain orthogonal angles for Doppler assessment. A similar evaluation can be conducted following transcatheter valve implantation for

all valve-in-valve and valve-in-ring implants, including tricuspid, mitral, and aortic. The tricuspid valve is imaged quite easily with the ICE probe from the mid right atrial view (Fig. 59.15). Imaging of the aortic and mitral valve implants is often best from the RV inflow position and LA position, respectively.

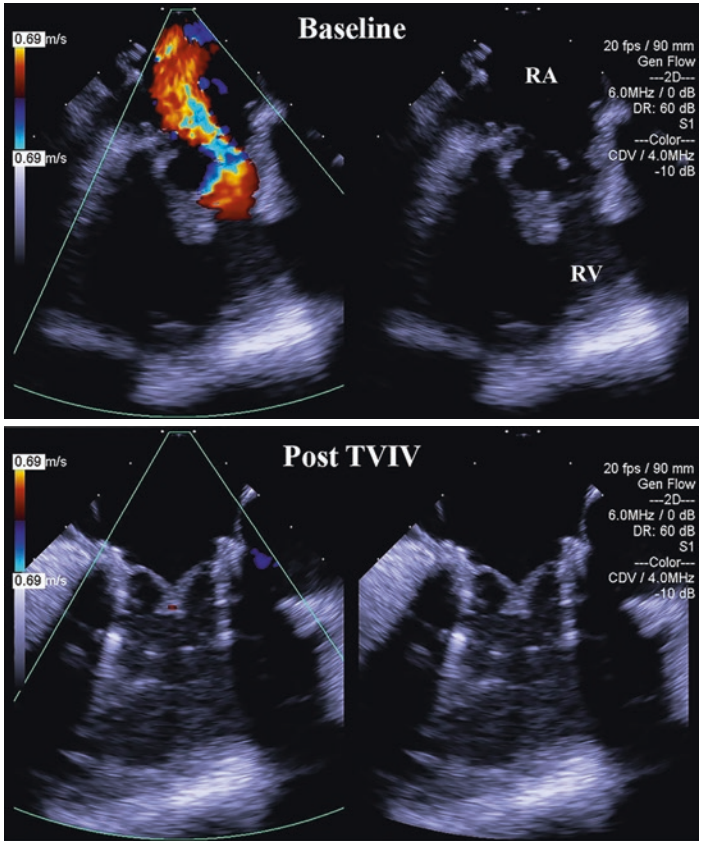

Fig. 59.15 ICE evaluation of a failed bioprosthetic tricuspid valve replacement. The medial leaflet is fused in the open position with resultant severe tricuspid valve regurgitation (top panel). A tricuspid valve-in-valve (TVIV) procedure was performed with implantation of a Sapien valve. The valve leaflets are thin with full coaptation and no regurgitation (bottom panel). *RA* right atrium, *RV* right ventricle

Further Reading

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60 Three-Dimensional Trans-oesophageal Echocardiography in Diagnosis and Transcatheter Treatment of Congenital Cardiac Defects

Carmelo Arcidiacono

60.1 Introduction

Trans-oesophageal echocardiography is widely established in most centers as a mandatory imaging modality for guidance of percutaneous closure of septal defects: coupled with fluoroscopy, it provides detailed and reliable information, enabling measurement of the defects and of their rims, visualization of devices during deployment, and evaluation of the results. Conventional Two-Dimensional TOE (2DTOE) has however some intrinsic limitations. Obtaining detailed information such as number, size, shape, and spatial relationships of multiple, multi-fenestrated or complex-shaped defects can often be challenging with two-dimensional imaging. 2DTOE guidance is also significantly

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limited in locating guidewires and catheters and assessing device position, particularly when multiple devices are used.

Real-Time Three-Dimensional Trans-oesophageal Echocardiography (3DTOE) is a rather recent innovation in echocardiographic imaging, based on miniaturized matrix array transducers, that allows three-dimensional imaging in real time without the need for multi-beat acquisition, and is particularly useful for guidance of percutaneous procedures. Real-time 3DTOE enables accurate assessment of multiple or complex defects (Figs. 60.1, 60.2, and 60.3) to confirm feasibility of transcatheter closure (Fig. 60.4) and guide the choice of the appropriate devices. During the procedure, it allows localization of guidewires (Figs. 60.5 and 60.6) and catheters when crossing the defects (Fig. 60.6), and monitoring of device deployment and release (Figs. 60.7 and 60.8). In cases

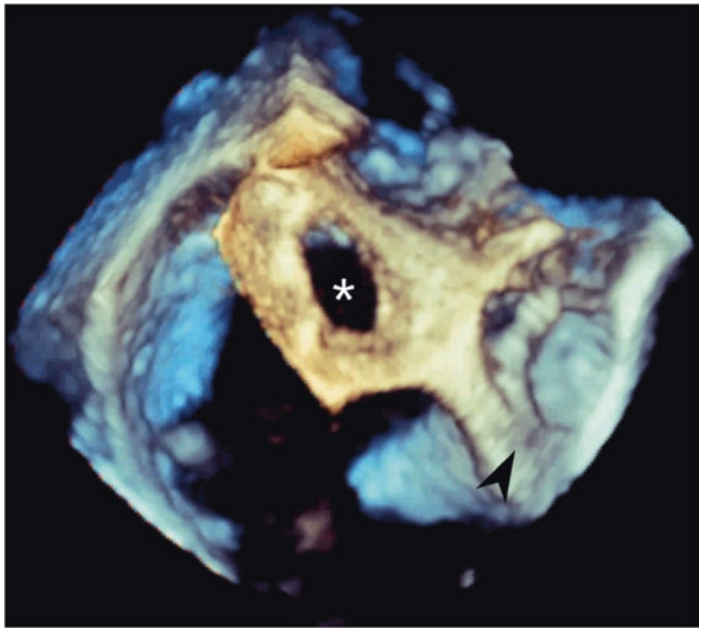

Fig. 60.1 3DTOE imaging in a case of an ostium secundum atrial septal defect (ASD). The defect is centrally located (asterisk) and the imaging mode can clearly show its relationship to the tricuspid and mitral valve annuli (lower left and upper right of the image, respectively) and with the aortic root (black arrowhead)

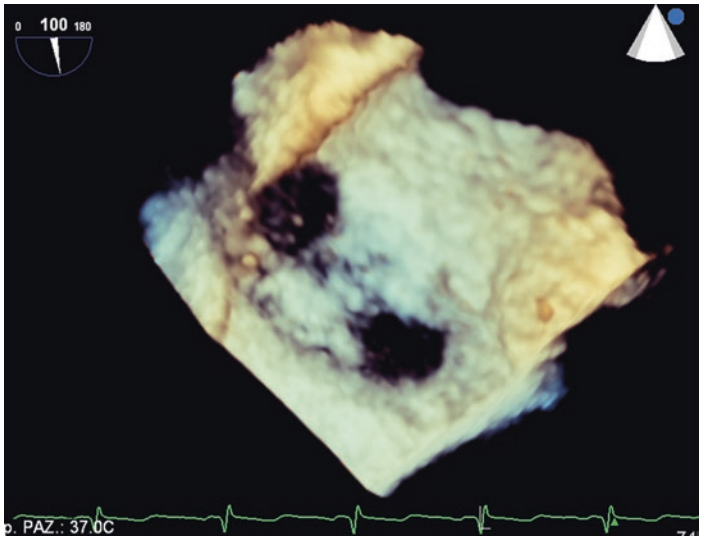

Fig. 60.2 3DTOE image of a case of multiple atrial septal defects. The view shows the left atrial side of the atrial septum. Two secundum-type ASDs of similar size are shown, both round shaped, separated by a large band of septal tissue

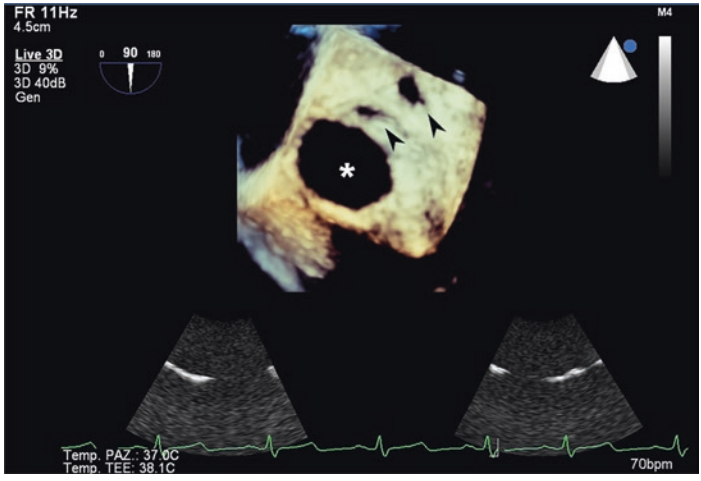

Fig. 60.3 3DTOE image of a case of multiple atrial septal defects. The view shows the left atrial side of the atrial septum. The main defect, indicated by an asterisk, is located in the middle portion of the oval fossa, while two small accessory defects are detectable on the posteroinferior portion of the septum (arrowheads)

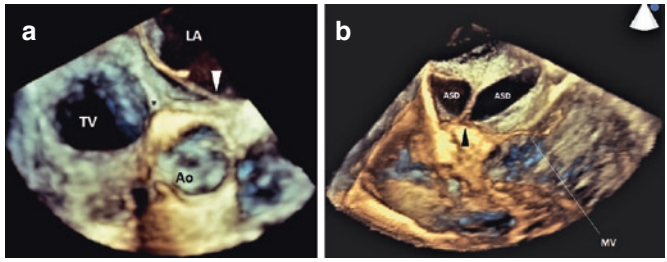

Fig. 60.4 3DTOE image of a malaligned type of secundum ASD, in which the flap valve of the oval fossa is multi-fenestrated and attached to the anterior left atrial wall via a thin fibrous strand. Panel (**a**) showing a view from above, clearly demonstrates the malalignment of the flap valve, with its thin attachment to the left atrial wall (white arrowhead), which is offset to the septum primum (asterisk). Panel (**b**) shows a view from the left atrial side, with two large fenestrations (ASD) in the oval fossa divided by a thin tissue strand (black arrowhead). After 3DTOE evaluation, device closure of the defect was deemed unfeasible. *TV* tricuspid valve, *MV* mitral valve, *Ao* aorta

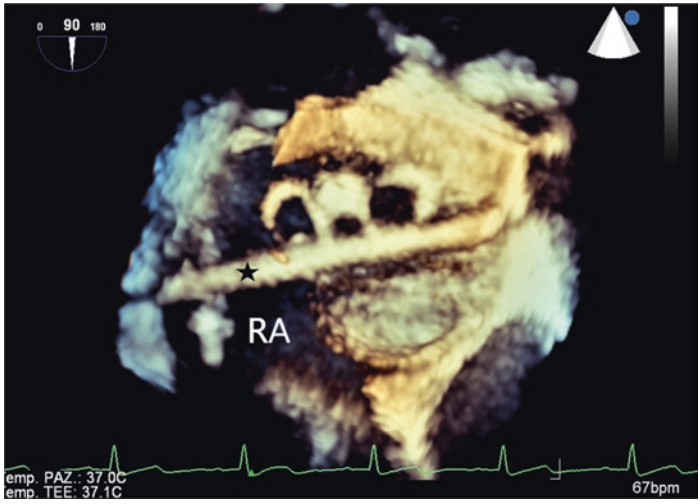

Fig. 60.5 A case of multi-fenestrated ASD. The catheter, indicated by a black star, is crossing the fenestration in the middle from the right atrium

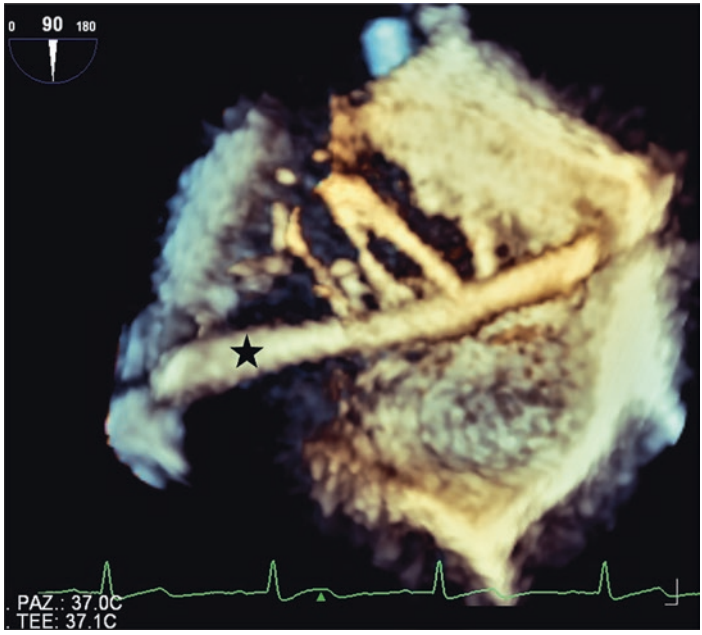

Fig. 60.6 The same case as in Fig. 60.5. When some tension is applied to the catheter, this stretches the strands of tissue separating the fenestrations, as clearly shown by the 3DTOE image

where multiple devices are used (Figs. 60.7, 60.9, and 60.10), 3DTOE provides information on their arrangement and relationship with surrounding structures (Figs. 60.7, 60.11, 60.12, 60.13, 60.14, and 60.15).

60.2 Optimization of Three-Dimensional Echocardiographic Images

The principles behind image generation on 3D echocardiography are the same as in conventional 2D echocardiography, but the results are presented in a pyramid-shaped volume of data instead of a two-dimensional sector.

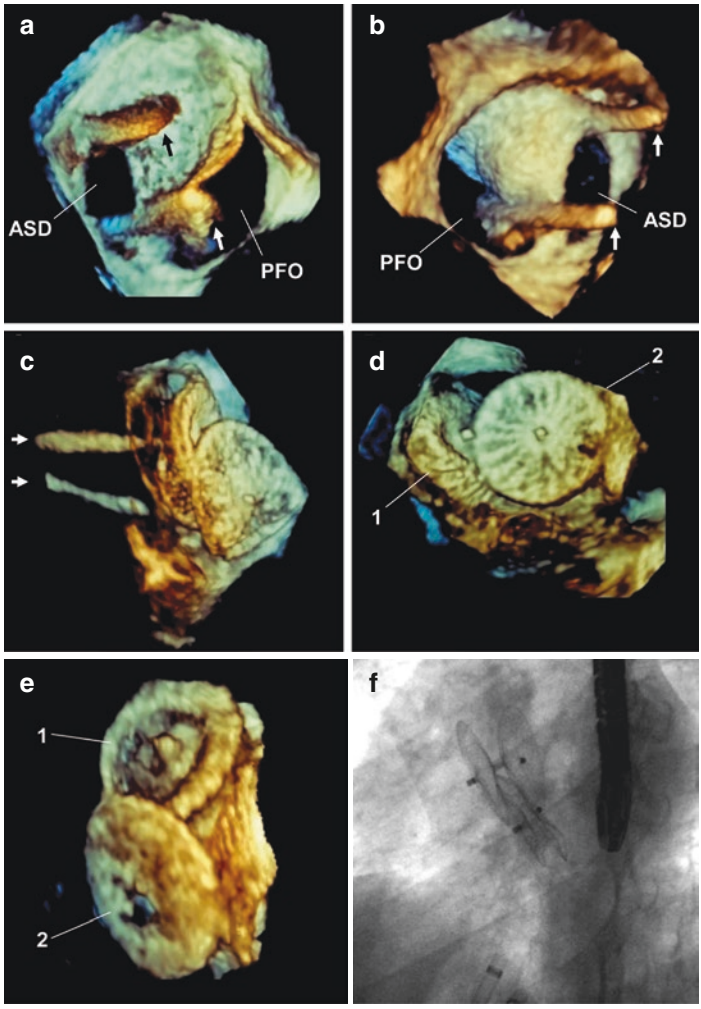

Fig. 60.7 Panels (**a**) and (**b**) show 3DTOE images the atrial septum, seen from the left and right side: a posterior secundum ASD is associated to an antero-superior patent foramen ovale with a very compliant and aneurysmal flap valve. The defects are both crossed by catheters (arrows). Panel (**c**) The two defects are closed with an ASD occluder device and with a PFO occluder device, respectively (angiogram on Panel f). The devices are still attached to the delivery wires (arrows). On Panel (**d**) and (**e**) the two devices have been released and are seen from the left and from the right atrial side, respectively. The different shape of the two devices is evident (PFO device: number 1; ASD device: number 2)

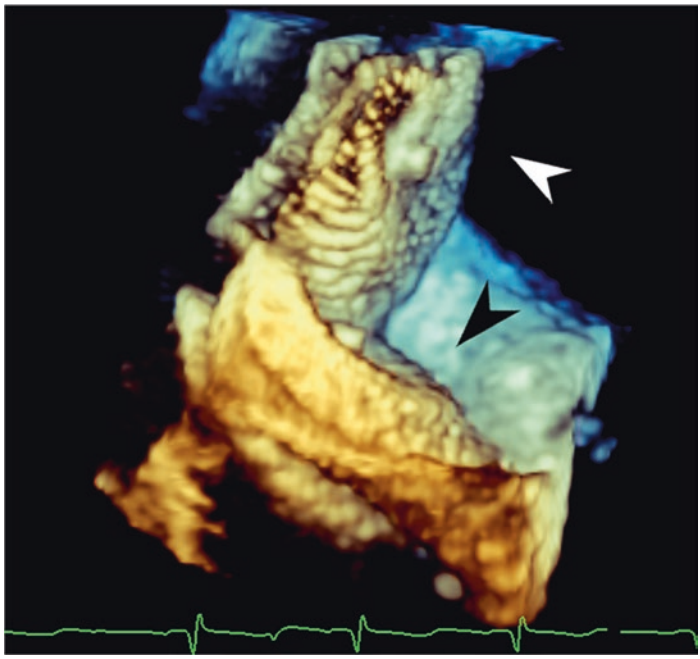

Fig. 60.8 The same case as in Fig. 60.2, after placement of two occluding devices (black and white arrowheads). Of note, the two devices are arranged at an almost orthogonal angle, reflecting the curved surface of the atrial septum

Keys to obtain good 3D echocardiographic imaging are:

1. **Appropriate probe selection**
   Matrix array probes allow different frequency ranges and, as usual, lower frequencies achieve better penetration but worse resolution. Commercially available matrix array probes come in adult and pediatric sizes, to fit patients weighing as low as 4 kg.
2. **Choice of the best transducer position and imaging view**: Depending on the heart structure to be studied, some views are more suited than others for 3D imaging. For example, atrial septum is best imaged on mid-esophageal 45°–90° probe orientation, ventricular septum on low-esophageal 0° and 120° setting.

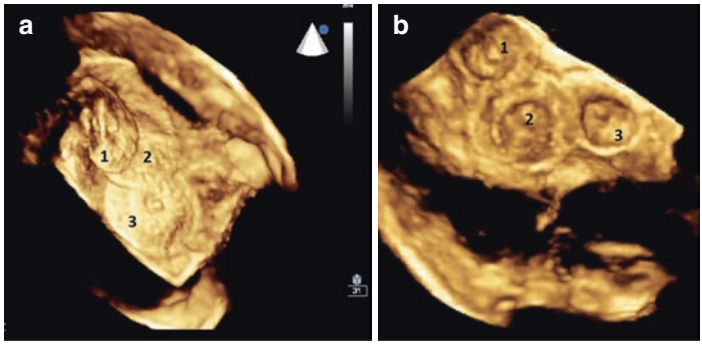

Fig. 60.9 3DTOE image of a case of multi-fenestrated ASD treated by placement of three ASD occluding devices. Panel (**a**) and (**b**) show the view from the left and right atrium, respectively. The devices are numbered from 1 to 3 starting by the one closest to the junction of the inferior vena cava. Note the partial overlapping of the three devices

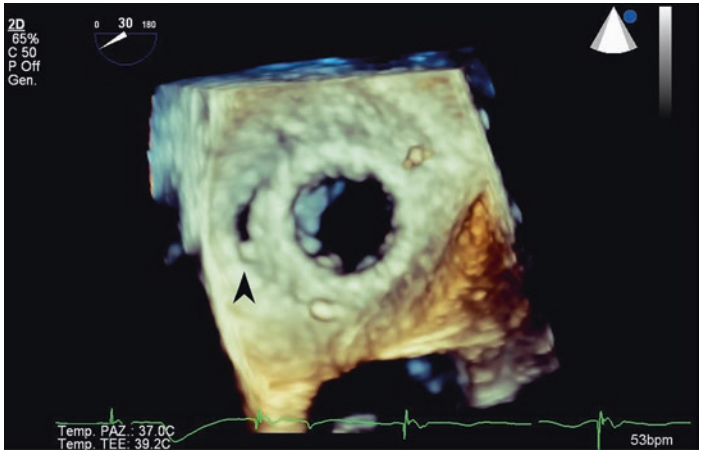

Fig. 60.10 Left atrial view of a case of para-valvular leak on a prosthetic mitral valve. The leak, pointed by a black arrowhead, has an elongated shape and is located towards the atrial septum

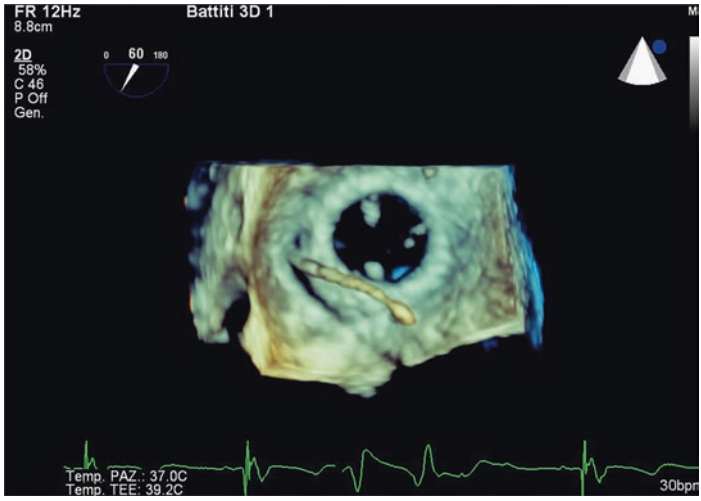

Fig. 60.11 The same case as in Fig. 60.10. A guide wire, as clearly shown by 3DTOE imaging, has crossed the leak

3. **Optimization of 2D images for 3D volume rendering**
   This is achieved by:
   (a) *obtaining the best signal-to-noise ratio*, i.e., ensuring all the structures of interest are well defined in the 2D image keeping background noise (or "haze") as low as possible. Use of harmonics and frequency settings, gain, compression, focus, and LGC adjustments is of great help for the purpose. Note that a not-so-good 2D image will result in an awful 3D image!
   (b) *Including all the structures and landmarks of interest* in the volume of data collected. Orthogonal biplane or steerable biplane visualization is useful to define the region of interest.
   (c) *Optimizing frame rate.* Frame rate is of crucial importance for imaging quality, especially in patients with higher heart rates. A narrower region of interest and a lower image density will result in a better frame rate. Wanting to improve frame rate without affecting image density and volume

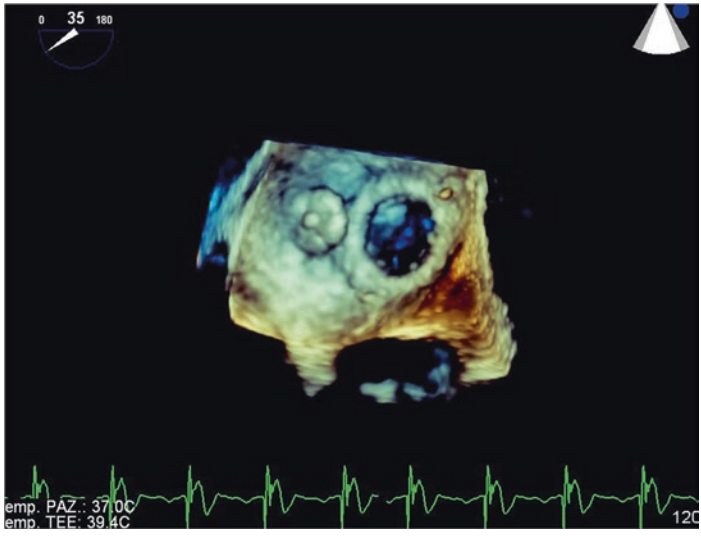

Fig. 60.12 Final check after placement of a muscular VSD device, obtaining occlusion of the para-valvular leak

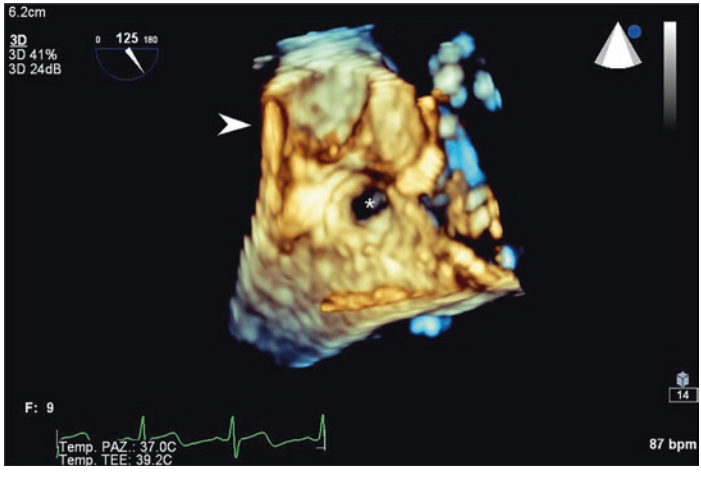

Fig. 60.13 3DTOE image of a case of perimembranous ventricular septal defect (VSD). The viewpoint is from the left ventricular outflow tract, and the defect, indicated by an asterisk, is clearly detected in close proximity to the aortic valve (white arrow)

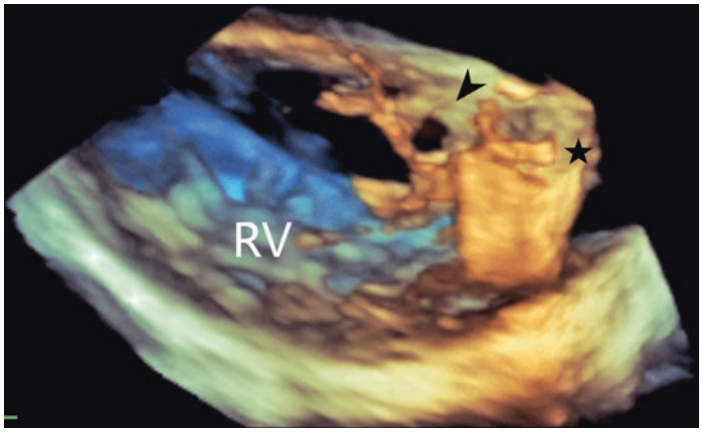

Fig. 60.14 A case of perimembranous VSD, where the defect, oval in shape (black arrowhead), is at some distance from the plane of the aortic valve (black star). Accessory fibrous tissue from the tricuspid valve apparatus is also evident

width usually implies using multi-beat acquisition with ECG gating.

4. **Adjustment of 3D image**

   Once the 3D volume is obtained, 2D gain, 3D gain, and compression significantly change the image:

   excessively high gain results in noisy and "foggy" images with loss of field depth; excessively low gain leads to loss of information and dropout artifacts («false defects»). Compression makes the image crispier and more defined. LGC faders also affect live-3D image. Other adjustments include image brightness, smoothness, and different volume rendering modalities (different presets).

5. **Moving, resizing, and cropping the 3D volume.**

   Modern 3D echocardiographic interfaces allow the operator to move the region of interest along two axis in real time, like a flashlight shining on the heart structures. The 3D box can also be resized along the two axis in real time by acting on specific controls, and cropped, using an orthogonal axial system or a freely steerable plane, to fit imaging needs.

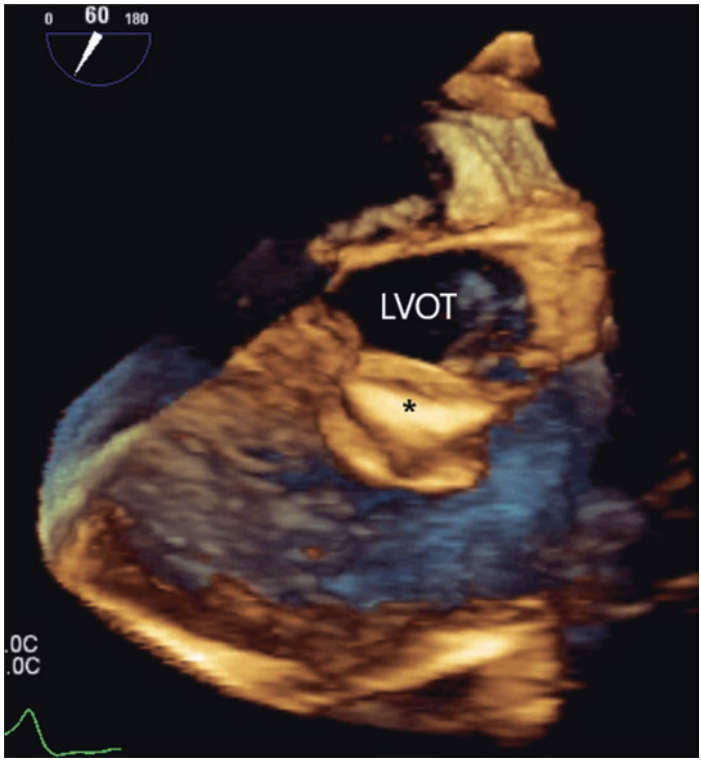

Fig. 60.15 Final check of the position of a muscular VSD occluding device after release. In this transverse 3DTOE view across the left ventricular outflow tract (LVOT), we can check the profile of the device (asterisk) and its relationship to the ventricular septum

6. **Color 3D settings**

While in older systems color 3D rendering was only possible on multi-beat volume acquisition, with modern interfaces it can also be obtained in real time. It must be noticed that this modality significantly affects the sector width, frame rate, and/or image resolution. Nontheless, it is very useful to locate small defects that are poorly imaged on 3D rendering (e.g., para-valvular leaks or small ventricular septal defects).

60.3 Conclusions

Three-dimensional trans-oesophageal echocardiography provides valuable information both in pre-procedural assessment of septal and valvar defects and in intra-procedural guidance of percutaneous device closure. Significant technologic improvements over the last few years have overcome many of its initial limits and have made it a standard adjunct of conventional 2D echocardiography.

Further Reading

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61

3D Mapping: Live Integration and Overlay of 3D Data from MRI and CT for Improved Guidance of Interventional Cardiac Therapy

Stephan Schubert and Felix Berger

61.1 Introduction

Multimodal picture integration has been developed for improved visualization of interventional procedure in congenital and structural heart disease. 3D rotational angiography (3DRA) was used for that purpose in the past, and a 3D dataset was than imposed as an overlay for fluoroscopy. But 3DRA includes a 180° turn of the

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Fig. 61.1 Work Flow for the use of 3D overlay with the use of VesselNavigator (Philips Healthcare): *A*. SEGMENTATION is done by importing a 3D dataset (DICOM) into the workstation (CT of a complete thoracic scan of the heart and vessels and thorax and 3D MRI of the whole heart sequence are optimal). The targeted vessels are selected with the mouse nearly automatically (aorta, pulmonary arteries, coronaries). *B*. PLANNING is then performed by using ring markers and small points for marking stenotic regions (landing zone) or landmarks (branches, coronaries). Additionally, the 3D dataset can be rotated in order to identify the best projection and visualization, which can be saved for at the workstation. *C*. REGISTRATION is performed, if the patient is lying on the table with the use of fluoroscopy or angiography and in the AP plane only. A difference of >30° is demanded in order to do the registration. This can be done from inside at the patients table or outside at the workstation. *D*. LIVE GUIDANCE will start after steps *A*–*C*, where also re-calibration of the accuracy of the overlay is possible during the whole examination

X-ray arm in order to generate a 3D dataset, including additional contrast application of 4–5 seconds, may need rapid pacing and preparation of the cath lab to allow X-ray arm movement.

VesselNavigator (Philips Healthcare) allows reuse of 3D vascular anatomical information from existing CT and MRI datasets as a 3D road map overlay on a live X-ray image (Fig. 61.1). Quality and success of interventional therapy may be improved by this 3D

dataset, and generation of this data can be done prior to the catheterization. Additionally with the use of MRI data, X-ray and contrast exposure can be reduced significantly in comparison to 3DRA.

In this chapter, we want to demonstrate the use of 3D overlay in different interventional procedures.

61.2 CASE: TPVI with Melody

Transcatheter pulmonary valve implantation (TPVI) was performed in a 13-year-old male patient after correction of Ebstein anomaly with tricuspid reconstruction, PCPC, and pulmonary valve replacement with an 18 mm Contegra® (Medtronic) conduit 2 years ago. Now a high-grade conduit stenosis was intended to be treated by TPVI.

After segmentation (MRI dataset) of the pulmonary artery and aorta, two rings were blue placed for marking the landing zone for stenting and pulmonary re-valvulation (Figs. 61.1 and 61.2). The

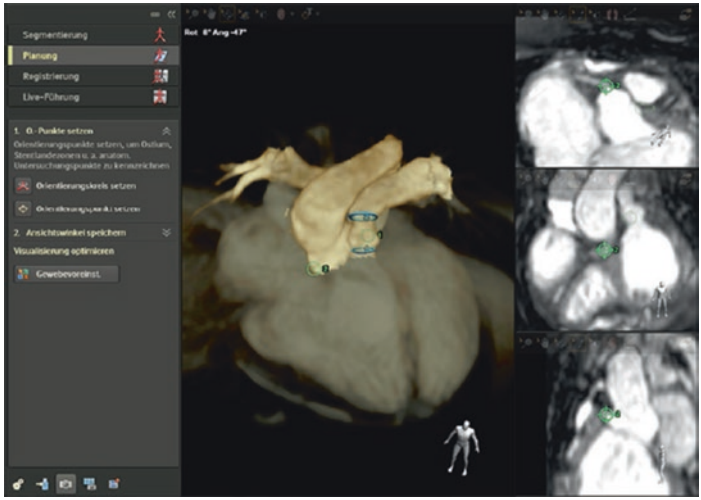

Fig. 61.2 Segmented vessel for TPVI in a 13-year-old patient with conduit stenosis in RAO 8° and cranial 47°. Two rings are marking the proximal and distal landing zone for stent placement. 1 and 2 are marking LCA and RCA

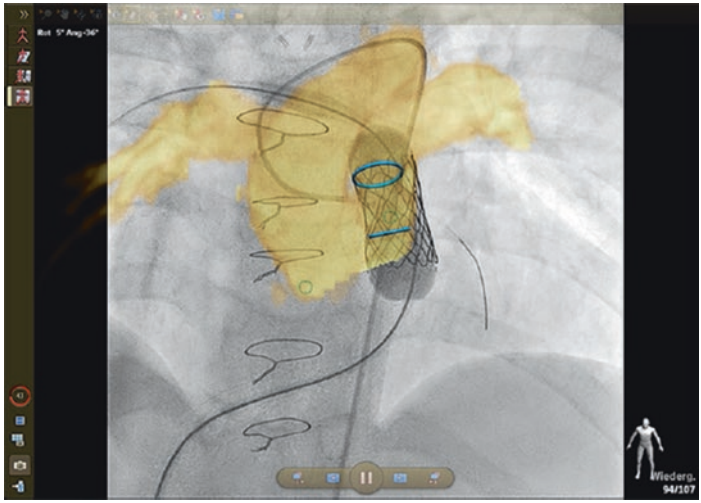

Fig. 61.3 Transcatheter Pulmonary valve (Melody® TPV, Medtronic) implantation in a pre-stented stenotic conduit with live guidance by MRI based image fusion (PA and Ao). Blue rings act as marker for the valve region. Left coronary artery is marked by a 0.014 inch wire

left and right coronary artery were also marked by green points (Fig. 61.2). Transcatheter pulmonary valve implantation was performed with the use of Melody® TPV (Medtronic) on a 18mm Ensemble (Fig. 61.3) and guided by the 3D overlay.

61.3 CASE: RPA Stenting

Right pulmonary artery (RPA) stenting was performed in an 8-year-old patient with pulmonary atresia with shunt palliation and right ventricular to PA conduit placement. Several surgical and catheter interventions had been performed, but RPA stenosis was reluctant to balloon dilatation and surgical patch reconstruction. Actual MRI showed a residual RPA stenosis with flow and size mismatch of left > right pulmonary arteries including supravalvular stenosis at distal conduit anastomosis (see Fig. 61.4).

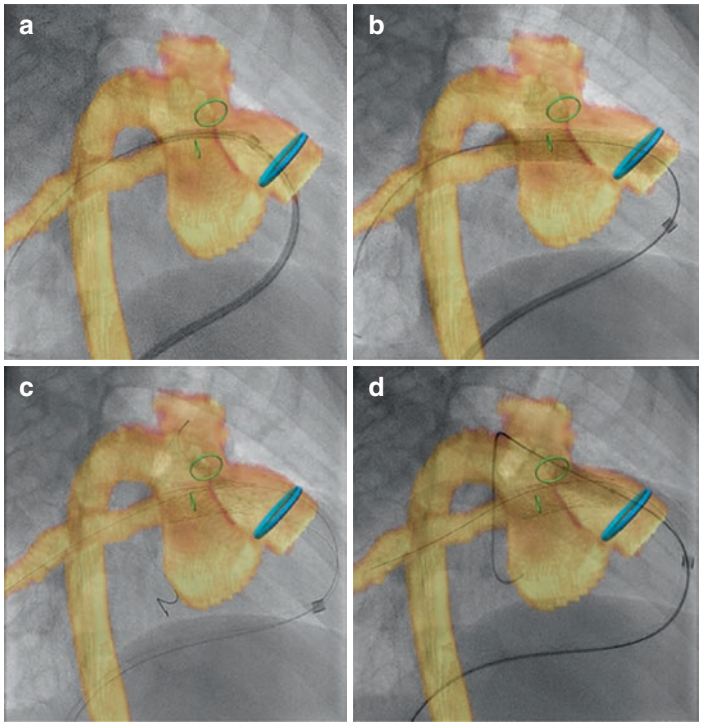

Fig. 61.4 3D overlay with pictures generated from MRI with segmented vessels (PA and Ao = yellow) during treatment of pulmonary branch stenosis. Green rings are marking the stenotic part in RPA and LPA ostium. (**a**, **b**) Stent implantation of a Mega LD (Ev3, 26 mm) on a 10 mm angioplasty balloon (Powerflex®, Cordis) with a use of a 9 Fr long sheath (Flexor, Cook™). (**c**, **d**) Re-opening of the LPA ostium after stent placement with the use of a 4 mm coronary balloon and 8 mm Powerflex® afterwards

61.4 CASE: CoA Stenting

A 35-year-old patient was treated for native and subatretic coarctation of the aorta (CoA). Vessel navigator was used after import of an external CT scan (see Figs. 61.5, 61.6, 61.7 and 61.8) and a covered stenting was performed.

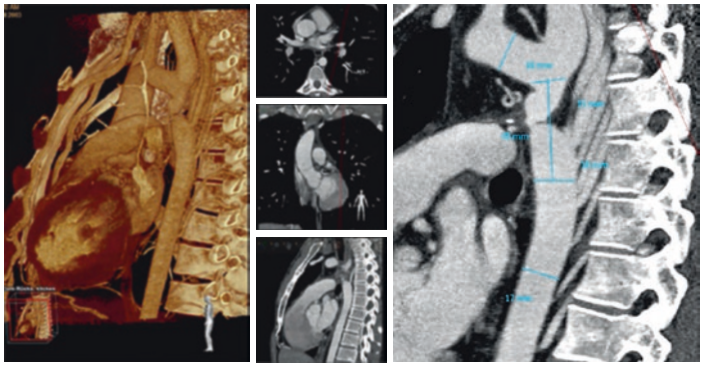

Fig. 61.5 CT data as used for planning of the procedure in this patient with native CoA imported into VesselNavigator (left). Numerous collateral vessels can be seen connecting from dorsally into the descending aorta. Measurements (right) were performed, and landing zone length (42 mm) and diameter (18 mm) of the stent were defined

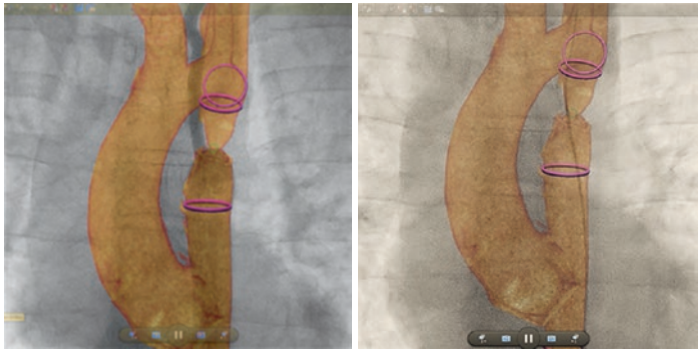

Fig. 61.6 Live guidance with the use of CT data was used. For calibration, two angiographies in the descending aorta were used (left) and in a second 40° RAO projection. Passage over the CoA into the left subclavian artery was then performed (right), and rings were placed in order to define the landing zone for stent placement

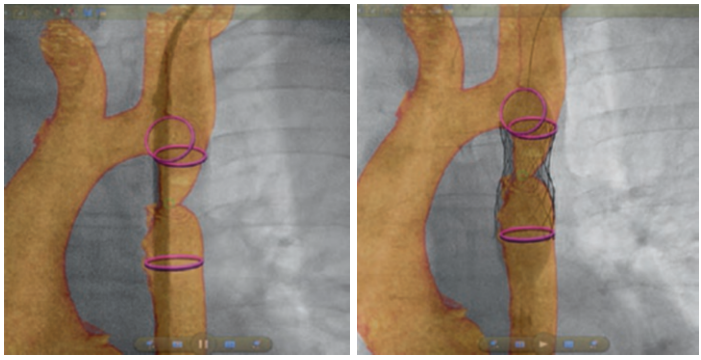

Fig. 61.7 Live guidance for 16 Fr-long sheath (Mullins™, Cook®) placement followed by implantation of a custom-made covered 10 zig CP Stent® (50mm lenth, NuMED) on a 22 mm Balloon-in-Balloon (BIB®, NuMED) catheter

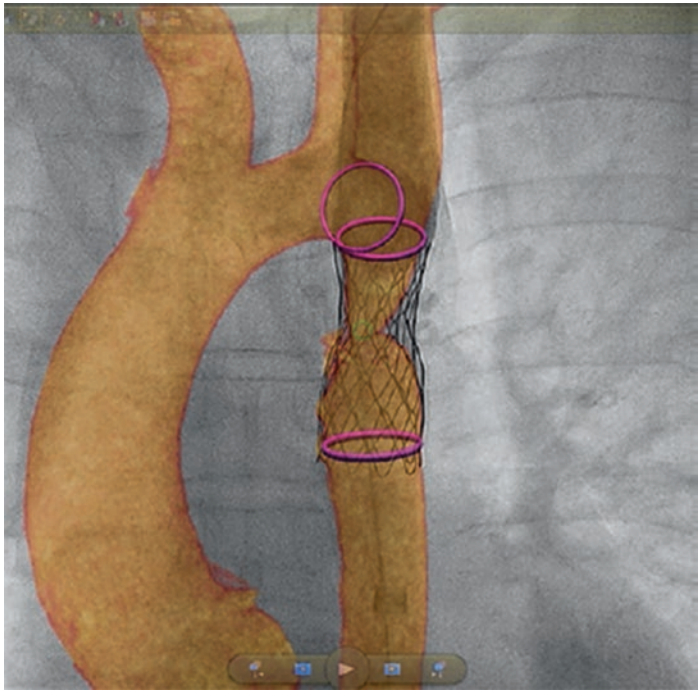

Fig. 61.8 Angiographic result after covered CP stent implantation into a native CoA with the use of overlay by VesselNavigator from CT data. No residual gradient was measured

62 3D Printing and Engineering Tools Relevant to Plan a Transcatheter Procedure

Elena Giulia Milano, Teodora Popa, Andrei-Mihai Iacob, and Silvia Schievano

62.1 Introduction

Over the past few decades, advanced in imaging technologies and computing power have been exponential, providing us with more and more detailed information about the structure and function of the heart and vasculature. This, along with general progress in medicine, biomedical research and device design, has allowed us to improve our diagnostic and treatment ability, exploring approaches and implementing solution to patient care not feasible before.

Despite the wealth of information provided by advanced cardiovascular imaging modalities, until 10–15 years ago, common cardiovascular practice relied on 2D flat screens to visualize

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complex 3D anatomical structures, thus relying on the expertise of trained cardiologists and surgeons to create 3D pictures in their mind, and evaluate/predict outcomes of different treatment options. This presented limitations related to perception, interpretation, communication and limited uptake of new technologies. In the past two decades, engineering tools such as 3D printing, extended reality and computational modelling have tried to overcome these limitations, and have been applied to cardiovascular problems, to support decision-making process, surgical and transcatheter planning, training and education, and overall to better comprehend the cardiovascular system function and dysfunction [1]. These methodologies can help solve complex problems and generate innovative solutions, by developing new device designs, and improving patient safety and procedural efficiency.

In this chapter, we aim to present an overview on how 3D printing, extended reality platforms and the most common computational engineering methodologies—finite element and computational fluid dynamics—are currently used to support percutaneous procedures in congenital heart disease (CHD), with examples from the scientific literature.

62.2 3D Printing

Patient-specific 3D printing is a fast growing technology that allows building of solid plastic replica of the patient anatomy by 'printing' successive layers of material. Input cardiovascular anatomies are provided by postprocessing of routine diagnostic 3D clinical images, in particular cardiovascular magnetic resonance (CMR) and computer tomography (CT) imaging. Case reports and small case series have suggested that 3D printed models can be used to plan surgical and percutaneous interventions by facilitating the decision-making process in complex cases [2–6]. Qualitative analysis of the models by means of satisfaction questionnaires has so far been used to evaluate their benefit. The largest multicentre prospective study aimed to evaluate the impact of 3D printing in planning 40 complex CHD surgeries, providing surgeons with a 3D printed model after a first

multidisciplinary discussion, showed a change in surgical strategy in 19/40 cases [3]. In catheterization procedures, meaningful clinical applications include:

1. Visualization of the size of the pathological structures in presence of rare congenital abnormalities
2. Three-dimensional visualization of intra-cardiac structure
3. Understanding of the spatial relationship of the great vessels in cases of complex CHD, particularly in post-surgical anatomies

Percutaneous pulmonary valve implantation. 3D printing has been first successfully employed for interventional planning to assess the right ventricular outflow tract in patients with pulmonary valve regurgitation, as a tool to aid clinicians in selecting patients eligible for percutaneous pulmonary valve implantation (PPVI) (Fig. 62.1) [7]. A crucial step in PPVI planning consists of identifying the adequate landing zone. This can be done with the help of 3D printed patient-specific models, with well-documented strengths and weaknesses. Known limitations of these models include the difficulty in obtaining adequate images to print the 3D model from the patient-specific CMR routinely acquired for PPVI assessment, as well as the innate differences between the features of native tissues and those of elastomeric materials.

Frame acquisition is usually done during the diastolic resting phase in order to benefit from the least cardiac motion, which could result in motion artefacts, and to facilitate inspection for cardiac defects. However, due to its failure to reliably mimic the

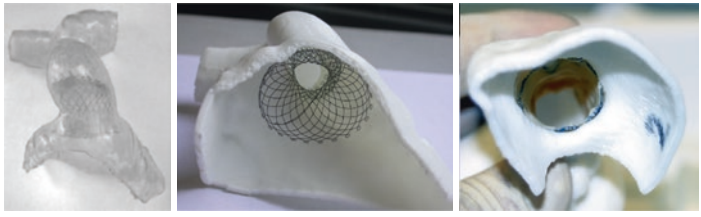

Fig. 62.1 Patient-specific realistic models printed in different plastic materials for PPVI planning and device testing

large deformation occurring between the RVOT and the pulmonary artery, the 3D printed diastolic model may underestimate the necessary implant dimensions.

Moreover, dysfunctional RVOT present great amount of movement and size variation throughout the cardiac cycle. Thus multidetector CT imaging has been employed more recently by some centres to produce multiphase rapid prototyping models and compliant 3D printed models to test in vitro feasibility of the percutaneous procedure and assure a safe landing zone for the device [6, 8]

Pulmonary arteries and pulmonary veins. 3D printed mock circulatory models have been produced to study the haemodynamic of the pulmonary vascular tree, in particular to study the differential split flow at the pulmonary bifurcation and to test different clinical scenarios by increasing the pulmonary vascular resistance [9] (Fig. 62.2). The evaluation of the pulmonary vascular tree poses major technical challenges in performing percutaneous intervention. The procedure requires extensive planning and intraprocedural precise visualization of the stenotic pulmonary branches; however, the pulmonary vascular tree is difficult to visualize in the catheterization laboratory with conventional planar angiography. New approaches for 3D visualization of medical data such as 3D printing and extended reality experiences have

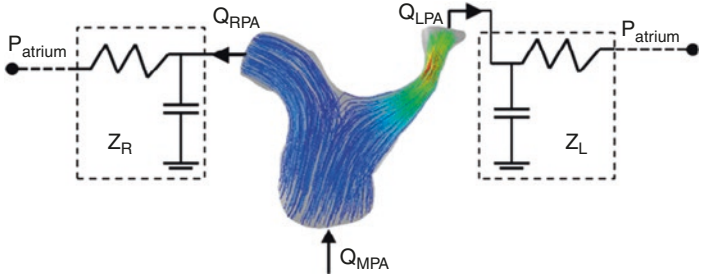

Fig. 62.2 Simulation of flow (Q) split in the left and right pulmonary artery (LPA and RPA) using computational fluid dynamics (CFD) and lumped parameter networks to represent the peripheral left and right pulmonary circulation to the left atrium. P pressure, Z impedance

been used as preoperative planning tools. In a proof of concept study, a 3D printed pulmonary artery model has been successfully employed in order to better select the target lesions, avoid vessel injury caused by oversized balloons, provide more complete revascularization, and decrease the volume of contrast medium [10]. The use of 3D printed models has also been reported in planning complex percutaneous procedures on pulmonary vein baffle in a case of TGA post-atrial switch with Mustard procedure and obstructed pulmonary vein baffle [11].

Major aorto-pulmonary collaterals. Planning of intervention and surgery in patients with major aorto-pulmonary collateral arteries (MAPCAs) is challenging as the anatomy of the collateral vessels is often complex and unique to each patient. To deliver successful embolization coiling via catheter, a 3D map of the collateral pathways and adjacent structure needs to be evaluated prior to entering the cath lab. 3D printed models of these small vessel structures built from CT scans can help reduce bypass, anaesthesia and fluoroscopy time, thus decreasing complications and improving outcomes [12, 13].

Atrial septal defects and ventricular septal defects. Atrial and ventricular septal defects are some of the most common CHD and can present either isolated or in combination with complex cardiac anomalies. When large and haemodynamically relevant, treatment is indicated either via surgical closure or by inserting a percutaneous closure device, nowadays considered a safe alternative. Simple ASD closure is usually guided by trans-oesophageal echocardiography and rarely requires advanced pre-procedural planning. However in complex cases, 3D printing of the anatomical structures surrounding the defect, from 3D echocardiography, CMR or CT guides the selection of the most adequate treatment technique and allows better understanding of the positioning of the device within the neighbouring structures, due to the high variability in morphology, location and presence of adjacent structures [14] (Fig. 62.3).

In particular, patient-specific 3D printed models have been used to introduce stenting of the superior vena cava-right atrium junction in superior sinus venosus atrial septal defects, which are commonly associated with partial anomalous pulmonary venous

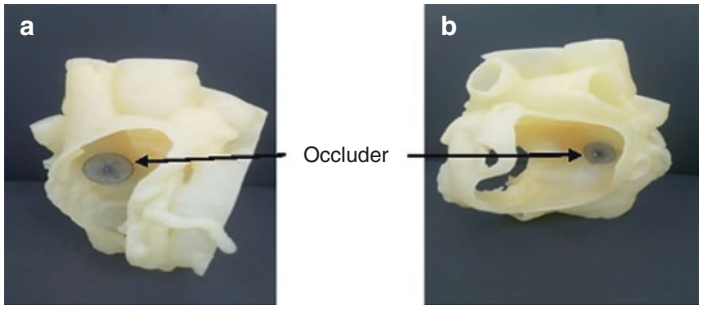

Fig. 62.3 3D printed model of a patient with multiple ASDs. (**a**, **b**) Illustrate the status after occluder deployment in the 3D printing model [14]

drainage. Patients who present with low complexity of this pathology are surgically corrected with excellent results. However, for those high complexity patients with co-morbidities who cannot tolerate surgery, meticulous planning through 3D modelling can guide the percutaneous intervention [15, 16].

3D printing has also proven greatly beneficial in the navigation of the occluder device and the optimization of patch sizing. In the case of atrial septal defects, the dimensions of the surrounding rim play an important role in the selection of occluder devices, since misplacement can lead to complications. 3D printed models allow for occluder device sizing, preoperative evaluation of the defect and in some cases can serve as the subject for an occlusion trial, in order to prevent an unnecessary transcatheter closure.

A 3D printed model of ventricular septal defect can aid in crossing the defect in cases of congenital muscular ventricular septal defects. It can also prove useful in bench testing occluder device selection and successful in vivo deployment in the case of post-myocardial infarction ventricular septal defects. In these cases, although rare, percutaneous closure devices are preferred over surgical repair, due to the latter's high mortality in the context of a myocardial infarction [17].

Patent ductus arteriosus. Both in paediatric and adult cases, patent ductus arteriosus may present a wide anatomical variability. A small case series described the use of 3D printed hollow models

in procedural planning of PDA closure supporting the device selection and shortening the fluoroscopic and total procedural times in transcatheter PDA closure [18].

Aortic Coarctation. Aortic coarctation stenting is a treatment option in patients with recoarctation after initial surgical repair or adult native coarctation. Depending on the aortic arch anatomy, 3D printing models may be useful to select the best stent landing zone in complex aortic coarctation and to better appreciate the spatial relationship with the left subclavian artery or in cases of aortic arch hypoplasia (Fig. 62.4) [5]. 3D printed models have been used to simulate the endovascular procedure under fluoroscopy and identify the best device size in case of complex aortic coarctation in order to predict the risk of infolding of large stent grafts [19].

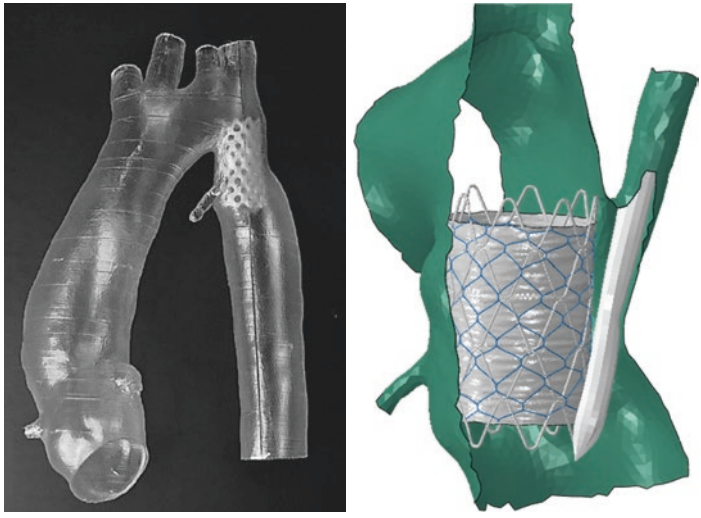

Fig. 62.4 3D printed model of a patient with previously stended aortic coarctation (left panel), and in the finite element model (right panel) to simulate covered stenting deployment and spatial relationship with subclavian artery, in order to avoid aneurysm rupture and subclavian artery obstruction during balloon inflation

62.3 Extended Realities

Recent technological advanced have enabled clinicians to use advanced 3D visualization tools such as extended realities, thanks to the development of high resolution display technology with relatively low costs and a very user friendly interface. Extended reality technologies have found several applications in medicine, ranging from telemedicine and education to emergency response, patient point of care, rehabilitation, procedural planning and intraprocedural visualization. Extended reality encompasses two main different experiences: virtual reality (VR) and augmented reality (AR). VR consists of a fully synthetic environment that replaces the user's auditory and visual fields, while AR interferes to a minimal level with the normal field of vision by presenting an annotated 'window-on-the-world'. The AR interface has very little interference with the user field of view and experience since it is activated on demand only when needed by the user and does not replace the user surroundings allowing the clinician to continue with their activity (surgery, cath intervention, etc.). In the medical setting, this translates to presenting relevant graphics, 3D anatomical models, patient information and reference data alongside the physical surroundings of the physician, giving the user full control over both the virtual and real-time scenarios.

The use of VR setup in congenital cardiology has been described in planning complex surgical procedures such as the biventricular repair of double outlet right ventricle with uncommitted ventricular septal defect [20] (Fig. 62.5). In the setting of interventional procedures, the use of AR with holograms created

Fig. 62.5 VR platform showing manipulation of a complex case of DORV with ultrasound like project (left panel), a stented pulmonary artery (central panel) and the baffle pathway of a DORV case (right) for surgical planning

from 3D echocardiography or angiography has been reported to guide structural valve disease intervention such as transcatheter aortic valve implantation [21–23].

62.4 Computational Modelling

Computational models have been extensively developed to investigate cardiovascular problems and predict clinical outcomes, thus enriching the information provided by clinical advanced imaging to improve understanding of pathophysiology, support surgical as well as interventional planning, and develop new device solutions [24, 25]. Patient-specific computational tools, combining clinical imaging and numerical methods with individual patient data to build realistic simulations, foster precision medicine, particularly relevant in CHD. Translation of these computational technologies—mainly finite element (FE) and computational fluid dynamics (CFD) analyses—into routine clinical practice depends on large-scale testing and validation, which remain a major challenge for CHD studies [26]. However, in the last decade, patient-specific computational models have become increasingly realistic, taking into account anatomical variability, implantation site data and specific pathophysiologic conditions, thus raising the interest of regulatory agencies and medical device industry, and gaining wider clinical acceptance.

Finite Element (FE) analysis is used to define how a structure deforms under given loading conditions and how structures interact with each other, by defining the relationship between stress and strain, force and deformation. The core principle of FE is that of reducing a complex problem into a number of small, finite parts, which are assembled together and interconnected by nodes. The deformation of these finite parts (elements) affects the behaviour of adjacent elements, resulting in a local approximate solution for the initial problem. The overall behaviour of the structure is defined by a global approximate solution from all the local approximate solutions of the FE analysis. The end output of FE models is a detailed visualization of the stresses and deformations affecting the structures and their distribution [27–29].

Computational fluid dynamics (CFD) examines and quantifies fluid flow patterns and behaviour by utilizing different computational techniques and physical properties, such as temperature, velocity, density, pressure and viscosity. In a closed system, the physical properties of a fluid (mass, energy and momentum) are stable constants, which allow CFD to provide valuable hemodynamic parameters for the clinical assessment of heart performance, the diagnosis of heart dysfunction and the comparison between different treatments.

FE and CFD can be combined together to simulate fluid-structure interaction (FSI), a more advanced numerical technique where fluid flow and tissue mechanics are coupled to mimic more realistically the cardiovascular function. For example, FSI is used to reproduce blood circulation in compliant vessels, or the pumping action of the ventricle resulting in blood flowing through the valve leaflets opening and closing during the cardiac cycle. FSI cardiovascular models are numerically highly complex and, therefore, mainly confined to the engineering development domain, and less mature than FE and CFD methods for use in clinical practice.

Patient-specific computational methodologies have been adopted in several examples of procedural planning for CHD transcatheter procedures, as they allow to investigate and quantify clinically relevant risks associated to the percutaneous intervention.

For example, as already explained in the 3D printing paragraph above, successful PPVI requires accurate pre-procedural patient evaluation to minimize risks of device dislodgment, arterial dissection, coronary artery compression or other adjacent structure interference and injury. All these PPVI-related adverse events have been studied and assessed using FE models [30, 31] (Fig. 62.6). The methodology has been used not only to support PPVI planning in specific patients, but also to study PPVI stent mechanical behaviour, design new devices and enhance safety in the compassionate use of prototype devices [8, 32–34].

Surgical or percutaneous treatment of aortic coarctation (CoA), despite successful in the short term, results in late hypertension, which has been linked to several factors such as aortic arch geometry, attenuated baroreceptor reflex sensitivity or

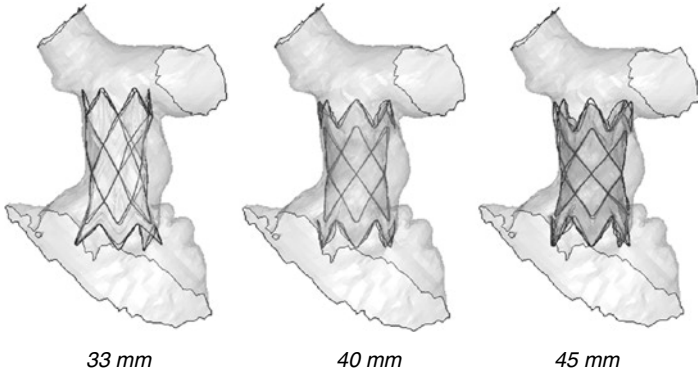

Fig. 62.6 Example of PPVI simulation of different stent sizes in the same patient-specific anatomical reconstruction

persistent abnormalities of central aortic biomechanics. Because of this, virtually reproducing the percutaneous procedure through patient-specific numerical models might improve our knowledge on the effects of the procedure on the surrounding anatomy. While CFD studies based on postoperative imaging can only be used for ex post quantifications of the fluid dynamic effects of the CoA treatment, FE analysis based on preoperative imaging allows for ex ante predictions, which support further procedure planning [35].

In order to accurately estimate the impact of CoA stenting on aortic post-stenting biomechanics and geometry, the FE analysis must take into account some key modelling aspects. First, since the aorta is never unloaded throughout the cardiac cycle, the anatomical configuration reconstructed from CT images should be consistent with the pressure loads and stresses acting on it. This is possible by computing the corresponding prestressed field. Second, in the FE simulation of the stenting of a severe aortic obstruction the quality of aortic wall elements could deteriorate due to their massive circumferential stretching, thus hampering further progress of the simulation [36, 37].

Advanced CFD methodologies for cardiovascular disease applications have recently been applied for the development of

the first successful clinical trial and subsequent FDA approved simulation platform in 2014 (HeartFlow Inc.) which is based on the use of patient CT imaging datasets and CFD analysis based fraction flow reserve to evaluate the significance of coronary artery stenosis in ischemic heart disease noninvasively [38–40].

62.5 Discussion

Three-dimensional modelling has become an important tool to support clinical decision-making in selected cases. In complex cases, the collaboration between clinicians and biomedical engineers can better address clinical problems and improve the confidence of the operator and the success of complex procedures. Modelling and simulation can provide the clinician with quantitative information before entering the cath lab and improve the understanding of the patient-specific haemodynamic.

Machine learning algorithms that will improve the segmentation of the cardiac and extra-cardiac structures will reduce the processing time and improve the accessibility and integration of these biomedical tools into the clinical workflow.

Mostly in congenital heart disease, where the anatomy is often unique for each individual patient, the use of 3D modelling and computational modelling is also supporting the development of new devices and also new materials by the use of bioprinting.

Despite many case studies and small case series report the use of engineering tools in supporting interventional procedures, these techniques are not widely available yet; this is mainly due to the initial costs, for instance related to the 3D printing, the relatively long processing time required to create a 3D model or complete a CFD simulation, but also the need of a close integration of the biomedical engineering team into the hospital setting. Moreover, although modelling techniques have been now available for decades, their use and their benefits still remain limited to case reports and small case series and there is still the need of large prospective clinical trials to support their use on larger scale.

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63 Development of a Quality Improvement Culture in the Congenital Cardiac Catheterization Laboratory

Brian P. Quinn, Priscila Cevallos, and Lisa Bergersen

63.1 Introduction

Cardiac catheterization for pediatric patients with congenital heart disease (CHD) has undergone rapid expansion of both its diagnostic and interventional capabilities over the past several decades. Ongoing technological advancements, augmenting the capabilities and safety of catheterization treatment options, contribute to an increased life expectancy and enhanced quality of life in children with CHD. Continuous efforts to increase the quality and safety of cardiac catheterization procedures through quality improvement projects have shown the potential to further improve patient care, especially in areas of risk for patient harm and radiation safety [1–4]. The continued evolution of interventional cardiology for CHD necessitates the development and implementation of quality tools specific to the needs of practitioners and pediatric patients.

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63.2 What Is Quality?

Traditionally, quality in healthcare focused on outcome measures but has since grown to encompass a much broader meaning. Quality efforts previously emphasized processes for *quality assurance*, which measure compliance with recommended standards of care. The quality assurance (QA) process gears towards individual practitioner performance and behavior, and was found to propagate a defensive and punitive culture. QA processes have largely been replaced with methodology focused on continuous quality improvement (CQI). This modernized approach builds upon traditional QA methods while proactively improving systems by analyzing patterns of care and using data to identify areas for improvement.

63.3 Quality in Healthcare

As the healthcare industry began to adopt a CQI approach to the processes, structures, and outcomes of medical care, quality improvement has become an expected component of delivering high-value patient care [5]. While CQI application in healthcare can be difficult and time-intensive, its implementation has resulted in multiple areas of improvement, such as resource utilization and, above all, patient safety. The increasing utilization of the CQI approach in healthcare has fostered a culture in which optimal quality of care provided to patients is achievable.

The Institute of Medicine (IOM) has put forward a framework for quality assessment for the healthcare system with six aims [6]:

- **Safe:** Avoid harm to patients from the care that is intended to help them
- **Effective:** Provide services based on scientific knowledge to all who could benefit and refrain from providing services to those not likely to benefit (i.e., avoid underuse and misuse of services, respectively)

- **Patient-centered:** Provide care that is respectful of and responsive to individual patient preferences, needs, and values and ensure that patient values guide all clinical decisions
- **Timely:** Reduce waits and harmful delays for both those who receive and those who give care
- **Efficient:** Avoid waste, including waste of equipment, supplies, ideas, and energy
- **Equitable:** Provide care that does not vary in quality due to personal characteristics such as gender, ethnicity, geographic location, and socioeconomic status

Quality frameworks, such as the one developed by the IOM, have increased our understanding of quality measures and their value in healthcare.

63.3.1 Requirements for Institutions

In the United States, the federal government has mandated the adoption of the CQI process in the healthcare industry. Governmental and accreditation bodies, such as the Joint Commission on Accreditation of Healthcare Organizations (JCAHO), now require CQI programs as part of licensing and regulation. In addition, per the Accreditation Council for Graduate Medical Education (ACGME), physicians in training are required to complete quality improvement projects prior to the completion of residency.

While required quality monitoring in the cardiac catheterization laboratory can vary regionally in the United States, professional organizations such as the American College of Cardiology (ACC), the American Heart Association (AHA), and the Society of Cardiovascular Angiography and Interventions (SCAI) have produced guidelines and expert consensus documents recommending CQI programs for enhancing cardiovascular care [7, 8]. To date, these programs have facilitated measurable success in improving care for this complex population of patients.

63.3.2 Importance in Pediatric Interventional Cardiology

Pediatric cardiac catheterization encompasses many complex procedures that are highly dependent on a sophisticated organization system, coordinated efforts of multiple individuals working as a team, as well as a high level of technical proficiency. While essential for diagnosis and treatment, pediatric cardiac catheterization remains among the highest-risk procedures for potential complications due to hemodynamic vulnerabilities of young infants and small children, technical demands on operators, and procedural diversity within the field [2, 3, 9]. The occurrence of an adverse event during a catheterization procedure can be as high as 1 in 4 depending on patient acuity and procedural complexity [9]. Some events not successfully managed at the time of identification can result in downstream patient harm, such as requiring unplanned surgery, permanent disability, or death.

Due to the complexity of the field, the increasing pressure for cost-efficiency, and demands for quality and public reporting of morbidity and mortality rates, efforts to adhere to the highest quality standards available in the cardiac catheterization laboratory have become a high priority for hospital management, physicians, and patients [5].

For these reasons, interventional cardiologists must continuously evaluate their practices and work with internal and external multidisciplinary teams to establish rigorous strategies to ensure that the highest quality of patient care is provided.

63.4 Methods for QI Success

Due to regional variations in institutional and practitioner practices, no single quality improvement model is universally appropriate. However, the basic principles for all CQI models, as applied to cardiac catheterization laboratories, include several key factors: a problem, a measure, an intervention, and an analysis.

Once a problem is identified, either by a healthcare team or an individual, that stands to benefit from a QI initiative, a multidisciplinary QI team is encouraged to meet to discuss and determine a valid quality indicator for this problem. Appropriate quality measures are systematically trackable over time by the institution and, ideally, can be compared to regional and national benchmarks. Next, the multidisciplinary team establishes an intervention(s) that can be feasibly made within their healthcare delivery system to target the problem. It is important to establish aims that are *time-specific*, *targeted*, and *measurable*. A timeline for data collection should be determined, this is then followed by the identification of an individual or group who will take responsibility for the execution of the QI plan. A method for testing the proposed changes through action-oriented learning can then be conducted. Initially, it is encouraged to maintain data collection on a smaller scale to analyze the impact of the QI intervention. After successful implementation of changes, the QI process can be expanded or started anew with additional changes.

63.4.1 Identifying a Problem

There are three overarching domains that build the framework by which cardiac catheterization laboratory physicians and staff can measure, review, and improve quality to enhance patient care [10]. These domains are structural, procedural, and outcomes driven.

63.4.1.1 Structural Domain: Is the Care Provided to Patients Appropriate?

The *Structural Domain* is concerned with ensuring that clinicians are prepared to adequately perform procedures and that systems are in place to provide clinicians with impartial assessments and opportunities for didactic feedback [8]. This includes creating a space to routinely review complex clinical cases and ensure adherence to standards of care.

63.4.1.2 Process Domain: Are the Parts/Steps in the System Performing as Planned?

The *Process Domain* is focused on monitoring patient care processes. These include direct patient care, system, guidelines, cost, and utilization processes (Table 63.1).

63.4.1.3 Outcomes Domain: How Does the System Impact the Values of Patients, Along with Their Health and Wellbeing?

The *Outcomes Domain* monitors outcomes and generates data sharing and reports for the purpose of quality improvement (Table 63.2).

Table 63.1 Examples of process domains

Direct patient care	Systems related	Guidelines related	Cost and utilization
Structured handoff to post-procedural service	Critical contingency plans	Standardized clinical assessment and management plans	Availability and quality of inventory
Management of complications	Generation and completion of reports	Pre-procedure checklists and patient verification process	Appropriate use of post-procedural resources and capacity management

Table 63.2 Examples of outcomes domains

Monitoring of outcomes		Data sharing and reporting
Risk-adjusted complication rates		Internal aggregation of case- and operator-specific data
Radiation exposure		Multicenter outcome reporting and collaboration

63.4.2 Defining Quality

63.4.2.1 Procedural Quality

Procedural quality takes into consideration the success or failure of the intervention(s) performed, the quality of the data ascertained at the time of the catheterization, and the presence of an adverse event—all of which have the ability to positively or negatively impact the observed outcome for a given case. In congenital cardiac catheterization, there is an innate heterogeneity among the procedures performed and the patient population, making the assessment of quality and performance challenging.

As an example, there continues to be a strong cultural change in the field to strive for radiation doses as low as reasonably achievable. While doing this, an individual operator must be cognizant to not compromise efficacy of the procedure nor the diagnostic yield. Keeping the patient's best interests first will help guide our therapies so that we can achieve radiation best practices without compromising the integrity of the catheterization and avoid unnecessary consequences.

Furthermore, there exists certain established radiation dose standards for common interventions, serving as balancing measures to compare one's performance. Using these reported outcomes for comparison allows a local cardiac catheterization lab and/or individual operator to judge their performance relative to others. While the use of these reported standards is an important tool, one size does not fit all, and care must be individualized for each specific patient. Therefore, local institutions can consider the use of these external benchmarks to assess performance relative to reported standards in the industry, all the while, ensuring that the patient-specific goals of the catheterization were still met.

Why Have a Process to Assess Performance Issues?

Providing optimal patient care is at the forefront of every pediatric and cardiac care center worldwide. Goals of meeting and exceeding standards of care through quality practices and policies is imperative. Additionally, it is important to understand that performance evaluation starts at the institutional level, as all cath lab

staff involved in patient care directly impact the quality of the care provided, not just at the operator level. Through public and/or aggregate comparative performance reporting, processes lacking in optimal performance can be quickly identified and adjusted to ensure that appropriate standards of care are met.

63.4.2.2 Procedural Checklist

The cardiac catheterization lab is a unique environment distinct from cardiac operating rooms and therefore should have its own set of best practices guidelines. It is an area of high patient throughput and provides elective, urgent, and emergent treatments. Physicians and staff should be properly trained and equipped to handle increasing patient complexity and emerging technology.

Given the unique nature of the cath lab environment, with high patient throughput including both elective cases and urgent/emergent unscheduled cases, there is inherent risk where human error can be introduced into the system. Building upon established practices in other industries, such as aviation and construction, procedural checklists can be used to combat these risks, minimizing the chance of human error. Checklists have been shown to reduce risk and improve efficiency and are being increasingly adopted in the practice of medicine.

Procedural checklists can improve safety and efficacy of a given procedure by empowering a multidisciplinary team approach to patient care and improving communication among providers. Checklists may identify pre-procedural and pre-anesthetic risk factors for poor outcome, with a focus of reducing morbidity and mortality. Ensuring a universal protocol is in place for pre-procedure patient verification and implementation of a "time out" procedure prior to the start of the case will help meet these goals. These checklists can also facilitate handoffs among services and have the ability to reduce unnecessary costs in care. Lastly, ensuring these standards of care are met can reduce the chance of litigation should there be a poor procedural outcome.

63.4.3 Promoting QI in the Workplace

A culture of quality improvement in the cardiac cath lab is founded on the engagement of all staff in the environment. Through the formation of a team-based approach, active members of the team will empower others, focusing on methods of improvement rather than an emphasis on quality assurance. Furthermore, providing data relevant to current practice trends can be helpful in motivating staff to achieve these goals.

63.4.3.1 Creation of a Quality Improvement Team

To successfully conduct quality improvement projects in the catheterization lab, it is helpful to establish a small, multidisciplinary team to oversee its execution. QI efforts are most effective when the team is comprised of individuals offering differing perspectives on the delivery of care, such as technical staff, nursing, and cardiologists [8].

63.4.3.2 Best Practices

Some examples of best practices for pediatric cardiac catheterizations are detailed below.

Pre-procedure:

- Incorporate the use of risk prediction methodologies to anticipate patient complexity and the need for post-procedural resources [4].
- Utilize pre-procedural checklists to minimize the chance of a preventable event and mitigate risk for morbidity and mortality.

Intra-procedure:

- Time Out: Confirm the right patient is identified along with correct procedure(s) (including necessary equipment needed), and an appropriate informed consent has been completed. Mitigate risk by verifying the presence of allergies, any complicating medical condition(s), or presence of any other patient or procedure related risk factor that may result in patient harm.

Post-procedure:
- Sign out: Following the procedure, communicate the important findings, complications or concerns, radiation dose used, and monitoring plan needed for post-procedural care.
- Conduct formal handoffs between the procedural attending and referring physician with completed procedural notes following case completion.
- Utilize a reliable and efficient system to monitor for procedural-related complications.

63.4.3.3 "Key Conferences" for Quality Improvement

"Key Conferences" serve as an essential tool to link current practices with best practices, fostering interdisciplinary collaboration and process improvement. Not only are they required by the Joint Commission to assess operator performance for Ongoing Professional Performance Evaluations (OPPEs) using metrics such as the standardized adverse event ratio (SAER) using the Congenital Heart Adjustment for Risk Method (CHARM), but they are also helpful in maintaining CME credit [2]. Conferences may also be required by the ACGME if an institution operates a fellowship training program in the United States.

"Key Conferences" facilitate practice improvement, continuing medical education, and professional development. To be successful, "key conferences" are encouraged to be regular, inclusive, non-punitive, and focused on practice improvement.

The three most common "Key Conferences" conducted to improve quality in the cath lab include Cath Lab Morbidity and Mortality Conferences, Case Review Conferences, and Cath Lab Educational Conferences.

Invasive Cardiology Morbidity and Mortality (Cath Lab M&M) Conferences

These meetings are separate from the clinical cardiology M&M and include an open review and evaluation of all cath lab complications and in-hospital events following any invasive cardiovascular procedure.

Why Have Cath Lab M&M?

Cath lab M&Ms are essential to achieving meaningful practice improvement in the cath lab. These non-punitive conferences serve as a vehicle for process improvement via collaboration, education, and feedback. Cath lab M&Ms offer an opportunity for staff to review adverse events that have occurred among peers in a colloquial setting, while engaging multiple key stakeholders: physicians, allied health, and other disciplines.

How to Make It Happen

Prior to the cath lab M&M, a quality officer (physician, PA, or designated cath lab staff member) identifies all cases with complications that occurred during the review period. Case identification is unbiased and comprehensive. All cases resulting in death within 30 days of the procedure are reviewed during the next cath lab M&M. Additionally, all major complications, as defined by ACCF, SCAI, and/or state reporting requirements, are also reviewed. Other complications may also be prospectively selected for review as aligned with specific process and quality improvement initiatives.

These meetings typically occur at least quarterly with mandatory cath lab staff attendance. The meeting environment should feel safe and transparent to allow for critical review of events as a means for performance improvement. Consideration should be made to include other multidisciplinary staff such as non-interventional physicians, nurses, and/or other allied health personnel, especially for events involving specific complications or other departments (i.e., anesthesia-related complications, intensive care unit, etc.). The responsible attending for each presented case must be in attendance when the case is reviewed and a sign-in sheet for participating staff is encouraged.

A Case Review Form is typically filled out during these meetings. These forms include an action plan and/or response to the complication and can be tailored to each institution. A mechanism to track complications (i.e., spreadsheet or database) and case review forms can be created and maintained for record keeping.

Case Review Conferences

Case Reviews consist of an open review of a random sample of cath lab cases, including diagnostic and interventional cases.

Why Have Case Review?

Different from the review of cases for which an adverse event occurred, case review meetings provide a formal process for assuring that indications for invasive procedures and intra-procedure decision-making conforms to standards of practice. They offer a great opportunity for education and further performance improvement by reviewing all relevant data collected at the time of the catheterization. This venue allows for an open discussion of pertinent decisions made during a case and provides an educational opportunity for providers to share lessons from past mistakes and successes, encouraging a culture of continued performance improvement.

How to Make It Happen

A randomly selected group of cases should be made available for review by a designated physician, cath lab manager, or quality officer and presented for review at a scheduled frequency (weekly, monthly, or quarterly). All cases are reviewed openly in a group setting with the responsible physician in attendance, followed by a discussion. Educational pearls and progress of cases discussed should be tracked and shared among the group.

Catheterization Laboratory Educational Conferences

Cath Conferences are primarily formal educational events that occur on a consistent basis and serve as an opportunity to discuss complex patient management.

Why Have Cath Conferences?

Catheterization Conferences provide a forum for continued professional development for faculty and fellows. The goal is to stimulate discussion and clinical perspectives. Therefore, the

attendance by non-cath lab physicians, especially cardiac surgeons, are encouraged to attend. These conferences can also help meet ACGME core curriculum requirements for fellows.

How to Make It Happen

Typically, the Cath Lab Director or Fellowship Program Director is responsible for holding these recurring events. The fellowship core curriculum may be used to structure a calendar of topics, with individual meetings being run by alternating fellows. A sign-in sheet can be maintained to catalogue attendance, and CME credit may be provided to encourage participation.

During these conferences, a fellow presents all clinical, imaging, and catheterization data for a selected patient and provides a summary of existing literature and/or review on the topic. A discussion of actual patient management and other strategies for optimal patient management follows.

63.5 Final Considerations

Over the past decade there have been numerous advancements in the field of congenital cardiac catheterization, including novel therapies and new technologies along with a refined understanding of complex cardiac physiology, with the goal of improving care in this vulnerable pediatric population. As the healthcare industry continues to evolve, with a current focus of rewarding value and quality, QI initiatives have become paramount in the success of an institution. The initiatives provide a framework for continued performance improvement and outcome assessment methodology in an ever-changing field. An interventionalist who prioritizes quality-based research will be adept at providing meaningful care to this population and play an integral role in expanding infrastructure necessary to meet the needs of the evolving healthcare delivery system.

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64 Quality Improvement Tools and Risk Mitigation in the Congenital Cardiac Catheterization Laboratory

Brian P. Quinn, Priscila Cevallos, and Lisa Bergersen

64.1 Introduction

Quality improvement (QI) methodology in the congenital cardiac catheterization laboratory is essential to providing high-quality patient-centered care to this complex population of patients. Through focused analysis of adverse events and near misses, along with risk mitigation strategies, patient safety in the laboratory can be optimized. With the development of innovative therapies and the shift from diagnostic cases to more complex interventions and procedures, the importance of implementing QI strategies and multi-center collaboration is paramount to leading a successful catheterization laboratory.

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64.2 Quality Improvement Tools

Successful quality improvement efforts require change, whether they are in workflow practices, technical performance, practitioner behavior or system-level adaptations. In order to continuously improve patient care, it is essential for a practitioner, department, or hospital to be able to develop, test, and implement changes [1].

Determining what changes to pursue to ensure practice improvement is a common challenge for QI teams. It is often helpful to develop a "change concept," combining QI concepts with specific QI methodology to help generate ideas for change [1].

The Model for Improvement, developed by Associates in Process Improvement [1], established three fundamental questions for a QI team to address to accelerate improvements [1]:

1. What are we trying to accomplish?
2. How will we know that a change is an improvement?
3. What change can we make that will result in improvement?

One of the most commonly used QI methodologies is known as the Plan, Do, Study, Act (PDSA) cycle (Fig. 64.1) [1]. The PDSA cycle tests changes in real settings to determine if the change is an improvement. The PDSA cycle allows for rapid assessments of the interventions pursued, whereby (1) a new change is proposed (Plan), (2) changes are implemented (Do), (3) effects are measured or described (Study), and (4) the process is reviewed, upgraded, and repeated (Act).

Typically, several different changes are repeatedly tested through iterative PDSA cycles as the team continues to learn and make systematic improvements. PDSA cycles are often scaled up to larger samples when improvements are shown.

64.2.1 Selecting and Implementing QI Interventions

There are many established QI methodologies for selecting, testing, and implementing QI interventions.

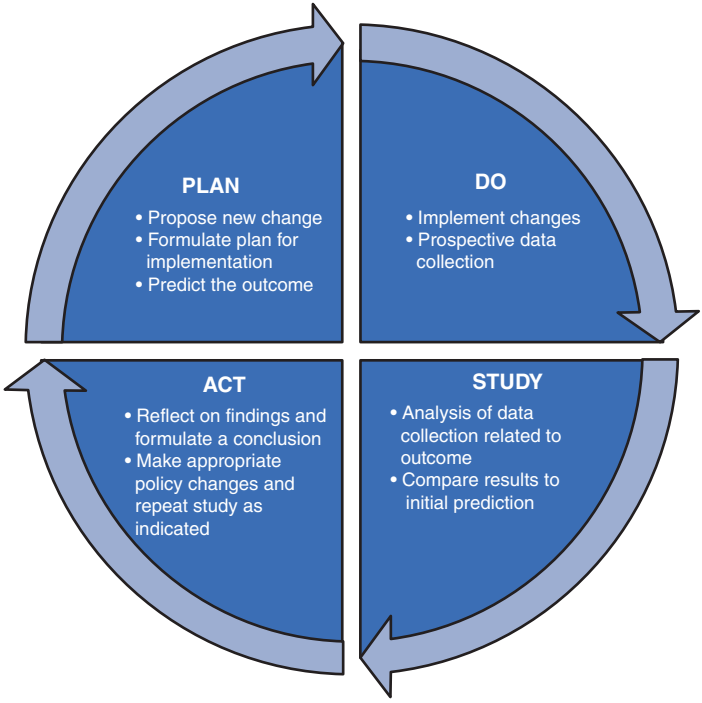

Fig. 64.1 PDSA (**P**lan, **D**o, **S**tudy, **A**ct) cycle

- *Flowchart*
  It is helpful to break down and understand a process in current operation as an important first step towards brainstorming ideas for improvement.

  A flowchart or "process map" is a visual representation of sequential steps in a process, making it especially useful in the early phases of improvement work. To create a flowchart, each step in a process is determined, indicating the points where decisions are made that lead to further branching of steps.

  Such flowcharts enable a shared understanding of current operations that can help teams identify problems, focus discussions, and unify goals.

- *Cause and Effect Diagrams*
  Once a problem has been identified, QI teams often struggle to define testable changes that may improve the process [1]. A cause and effect diagram can be used in such cases to graphically represent the many causes contributing to a certain effect or outcome, highlighting areas for improvement.
- *Key Driver Diagrams*
  The key driver diagram, created by the quality improvement team during the design phases of their project, provides a framework for the proposed aim, key factors (or drivers) necessary for improvement, and potential change strategies. The diagram identifies the primary and corresponding secondary drivers that contribute to the achievement of a project aim and establishes testable measures for each secondary driver. Multidisciplinary teams, with unique expertise of the problem, strengthen the development of key driver diagrams.
- *Failure Modes and Effects Analysis*
  Failure Models and Effects Analysis (FMEA) is a tool used to conduct a comprehensive and proactive analysis of a new or modified process in which harm to patients or staff may occur [2]. In a FMEA, a team predicts and records where, how, why, and to what extent a system under review might fail, and works together to devise improvements to prevent such failures. This tool is useful in evaluating and preventing possible failures by adjusting processes proactively rather than reacting to adverse events as they occur.

64.2.2 Data Collection

It is important to collect adequate data to determine the impact and success of the quality improvement project. Optimal data collection will be appropriate for the specific aim of the project, comprehensive for the proposed project specifications, and feasible in regard to data measurement, access, and consolidation with personnel and financial constrictions in mind.

The determination of what measures to collect will typically be guided by expert opinion and published literature. A common mistake with data selection includes collecting too many variables, leading to a data burden and ambiguous interpretation of data definitions. This mistake can be overcome by appropriately selecting data measures of importance and clearly outlining all variable definitions or using standardized definitions. It is preferable that data collection occurs prospectively on every case eligible for inclusion in the project.

It is often useful to understand the data collection burdens that the project specifications demand prior to the start of data collection and to determine if adequate resources are in place to appropriately manage these demands. Creating a culture of continuous improvement in the cath lab will lead to a more engaged staff dedicated to identifying and implementing sustainable changes that improve patient care.

64.2.3 Monitoring of Progress

After data collection has commenced, it is important to monitor outcomes regularly to improve upon QI methodologies being utilized and to determine the impact of the project. Establishing processes to analyze and display the ongoing data collection allows for close monitoring of progress.

It is often helpful to review data by subgroups, such as by procedure type or patient characteristics. Identification and review of outliers can reveal unusual behavior and areas to target for improvement. Some common ways used to display data graphically for quality improvement projects include histograms, run charts, control charts, and scatter diagrams.

64.2.4 Comparing Outcomes

The comparison of outcomes against established benchmarks is an important aspect of a quality improvement project. Evaluating local results is important; however, it is essential to determine

how these results compare to peer institutions and avoid misinterpretation of one's performance. In some countries, national registries can be a good source of reference information. However, many large-scale clinical databases are designed specifically to compare results of a specific treatment or condition and to provide data for ongoing and future research.

64.3 Multi-center Collaboration

Cardiac catheterization in congenital heart disease is characterized by groupings of complex heterogeneous procedures performed infrequently, making it difficult for individual care centers to achieve an appropriate sample size and generalizable methodology for QI metric development.

To combat this problem, a handful of collaborative cardiac catheterization QI projects have been established, such as Improving Pediatric and Adult Congenital Treatment (IMPACT), Congenital Cardiac Interventional Study Consortium (CCISC), and the Congenital Cardiac Catheterization Project on Outcomes (C3PO) registries. This has enabled large-scale data collection and implementation of important multi-institutional efforts to develop risk prediction and adjustment methodology, as well as identify best practices and areas requiring improvement [3–6].

64.4 Examples of QI Initiatives

64.4.1 Radiation Exposure

Single institutional efforts over the past decade and the field's focus on quality improvement have cultivated an awareness around radiation safety and reducing patient and operator radiation exposure. Radiation reduction efforts are critical in the pediatric population due to the ongoing physical developments of children and a potential of cumulative radiation exposure over a patient's lifetime. With the development of innovative technologies and the shift from diagnostic cases and isolated interventions

to complex interventions performed in the cardiac catheterization laboratory, the importance of radiation dose reduction is magnified. In the past decade, published reports emerged on single- and multi-center quality improvement efforts to reduce radiation exposure leading to the creation of ideal doses for common procedures and methodology to allow for radiation reduction and monitoring.

64.4.1.1 Establishing a Radiation QI Project

Designating a radiation QI champion at a local center can promote radiation safety practices and accelerate radiation-related QI efforts throughout the department. Such QI initiatives typically begin by setting a measurable radiation safety aim. Examples of measurable aims may include outcomes such as reducing patient and/or staff radiation exposure. Additionally, process aims may target improvements in areas such as recording radiation dosages in the medical record, completion of follow-up protocols to identify radiation-related skin injuries, and compliance with staff dosimeter monitoring.

Examples of metrics that may be used to evaluate patient dose include:

- Percent reduction in dose per year by all cases
- Percent reduction in dose quarter by radiation exposure category
- Reduction in the number of cases which exceed a specified threshold value over time
- Maintaining radiation doses within a range of reference values

64.4.1.2 Data Collection

Once the aim of the project is agreed upon, the metric determined radiation data can be analyzed based on a reported measure. There are numerous measurements of radiation exposure including: air kerma, peak skin dose, dose area product (DAP), and/or effective dose. Given the heterogeneity in patient sizes in congenital cardiac catheterization and the direct relationship of patient size to

dose received, we advocate for the use of DAP adjusted for weight (DAP/kg) as the optimal measure for exposure reporting. An additional advantage of using this metric is the growing published literature available for comparison.

In addition to the specific radiation measure, other important radiation-related data should be determined and collected. These include patient and procedural information which should be linked to existing cath reporting systems. Accessibility to the data elements and ease of data extraction is essential for efficient and timely reporting and review of performance both in real time and longitudinally.

Once data has been collected and audited for missing fields and/or outliers, analysis of the initial observation period can commence. This observation period should be long enough to contain a sufficient number of patients but short enough to allow QI efforts to take place using recent experience feedback, usually monthly or quarterly intervals depending on the metric.

64.4.1.3 Data Analysis

Reports in the literature have commonly focused on selected and isolated procedure types among a heterogeneous practice experience. To broaden the available population for outcome assessment, radiation exposure categories (REC) have been created by stratifying procedure types into three categories of expected radiation exposure: low, medium, or high [7]. This facilitates the assessment of performance of rarely performed procedures by aggregating the experience with similar procedures for the outcome radiation exposure. This improves analytic capabilities and allows for more frequent assessment of radiation outcomes. Further, aggregating case types (Table 64.1) into groups of similar expected radiation exposure and summarizing the outcomes by REC allows individual institutions, providers, and collaboratives to perform meaningful comparisons, while accounting for case mix differences.

Figures can be generated to depict the median and/or average radiation dose observed over the observation period. Aggregating data into the established REC (Table 64.1) allows the interpretation of meaningful trends and identification of areas where perfor-

Table 64.1 Radiation exposure categories

Low radiation exposure	Medium radiation exposure	High radiation exposure
Biopsy	Proximal pulmonary angioplasty or stent	Mitral valvotomy + intervention[a]
ASD or PFO closure	VSD device closure + intervention[a]	TPV implantation
PDA device or coil closure	RVOT dilation/stent	≥2 vessel proximal or distal angioplasty or stent
Vasodilator testing	ASD or PFO closure + intervention[a]	Coil systemic pulmonary collateral + intervention[a]
Atrial septostomy	Venous collateral closure	Aortic valvotomy + intervention[a]
Pulmonary valvotomy	Distal pulmonary angioplasty or stent	RVOT dilation/stent and ≥2 vessel proximal or distal pulmonary angioplasty or stent
Biopsy + CA	Aorta dilation/stent + intervention[a]	TPV implantation and PA Intervention[a]
PDA stent placement	Atrial needle transeptal puncture	≥2 vessel proximal or distal pulmonary angioplasty or stent + intervention[a]
Diagnostic catheterization	Atrial Septostomy + intervention[a]	Pulmonary vein dilation or stent
Fenestration device closure	Coil systemic pulmonary collateral	TPV implantation + intervention[a]
Aortic valvotomy	Proximal R and L pulmonary angioplasty	Pulmonary vein dilation or stent + intervention[a]
Aorta dilation and or stent	Proximal or distal pulmonary angioplasty or stent + intervention[a]	
Pulmonary valvotomy + intervention[a]	Atretic valve perforation	
	Atrial septum stent placement	
	Fenestration device closure + intervention[a]	
	RVOT dilation or stent + proximal pulmonary angioplasty or stent	

AS aortic stenosis, *ASD* atrial septal defect, *CA* coronary angiography, *L* left, *PDA* patent ductus arteriosus, *PFO* patent foramen ovale, *PS* pulmonary stenosis, *R* right, *RVOT* right ventricular outflow tract, *TPV* transcatheter pulmonary valve placement, *VSD* ventricular septal defect

[a]Intervention defined as additional angioplasty and/or stent placement, valvuloplasty, transeptal needle puncture, or coiling of systemic or venous collateral vessel. Case types ordered in observed radiation exposure for each category

mance may be less than optimal. Furthermore, data can be stratified by age group or other patient and procedural characteristics. These iterations allow for consideration of targeted areas for improvement, and frequently have the benefit of resulting in global improvements across the entire population treated at an institution.

64.4.1.4 Multi-center Collaboration

With the evolving understanding of the importance of radiation reduction, collaborative efforts and large data registries such as Improving Pediatric and Adult Congenital Treatments (IMPACT) and Cardiac Congenital Catheterization Project on Outcomes (C3PO) have made it possible to compare outcomes through a standardized methodology, providing summarized data on common procedures along with established procedure risk groups for radiation exposure [7, 8]. These efforts have allowed for the comparison of outcomes among centers and operators.

An example of a large-scale quality improvement project focusing on radiation reduction for congenital heart disease was spearheaded at Boston Children's Hospital with fifteen participating institutions nationwide [9]. Providers from all participating institutions came together aiming to reduce patient radiation exposure in the catheterization laboratory by improving individual performance and establishing new performance expectations. The QI initiatives were conducted using established PDSA methodology and included the creation of a key driver diagram to highlight the goals of the project and changes for execution (Fig. 64.2).

Through structured radiation- and patient-related data collection and the use of key elements within the key driver diagram, the collaborative was able to achieve their initial goal of 10% radiation dose reduction over a 3-year time period, exceeding that goal with a 30% median radiation dose decrease for all procedures (Fig. 64.3). Dose reduction was achieved through initiatives such as the sharing of best practices and operator techniques, regular webinars, use of reporting tools to allow longitudinal radiation reporting, identification of procedural specific areas requiring improvement, and creation of institutional radiation safety com-

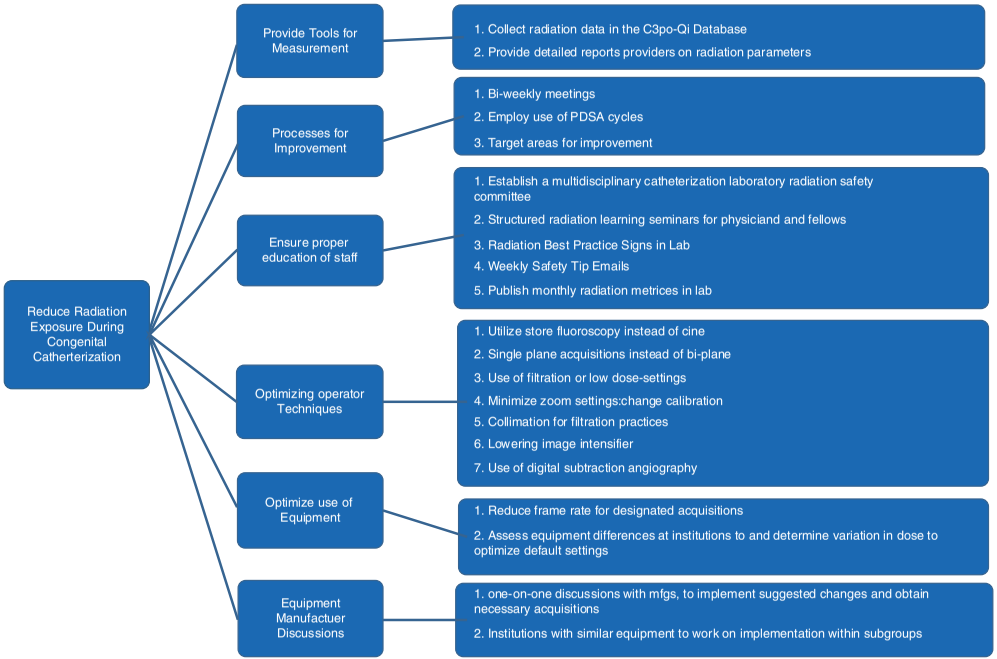

Fig. 64.2 The C3PO-QI key driver diagram used in the implementation of the QI initiative to reduce radiation exposure in the catheterization laboratory serves as a proposed outline of the strategies incorporated to reach the project's goals

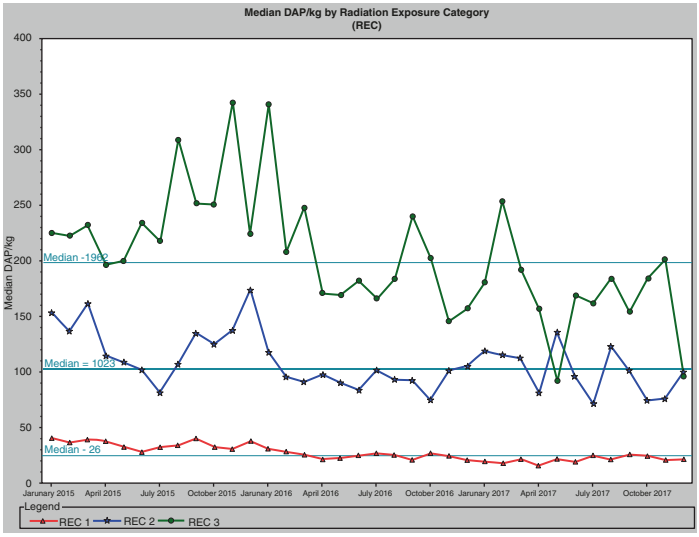

Fig. 64.3 Median DAP/kg by each radiation exposure category over the observed time period

mittees. QI efforts were further tailored to individual institutional environments and specific needs.

64.4.2 Adverse Events

Adverse events during pediatric cardiac catheterization can cause substantial morbidity and mortality. Complications due to a broad range of events can lead to downstream harm resulting in increased resource expenditure and sustained deterioration in a patient's clinical status, including potential permanent impairment. Thus far, many efforts have focused on determining patient and procedural factors associated with AE occurrence and developing measurement tools. These efforts have identified important determinants of risk including age, procedure complexity, and hemodynamic vulnerability [6, 10, 11].

64.4.2.1 Risk Adjustment Methodology

Over the past decade, there have been substantial advancements in the understanding of risk factors for adverse events associated with pediatric cardiac catheterization procedures. The Congenital Cardiac Catheterization Project on Outcomes (C3PO) was the first effort to standardize adverse event nomenclature (Table 64.2) in the field of congenital heart disease [12]. This led to the development of a model for risk adjustment for adverse outcomes using a combination of patient and procedural variables, the Congenital Heart Adjustment for Risk Method (CHARM) [10]. Empirically driven procedure type risk categories were generated to allow groupings of heterogeneous procedures in categories of similar risk. In addition, the final risk adjustment model included patient risk factors, specifically hemodynamic vulnerability and age, which improved the risk adjustment methodology.

Since its inception in 2010, CHARM methodology has been used to measure physician performance following event occurrence and has been supported by the National Quality Forum (metric endorsed in 2014) as the first quality metric in pediatric interventional cardiology. The CHARM model has made the equitable comparison of adverse event rates among providers and institutions possible via a standardized adverse events ratio (SAER) calculation.

Following the success of the CHARM model, the IMPACT registry developed a risk-standardization model for major adverse event rates [11]. The outcome chosen for the model was Major Adverse Events (MAE), equivalent to level 4 and 5 severity classification events, whereas CHARM was built on the outcome "serious" adverse events as defined by any level 3, 4, and/or 5 event. There were similarities in the foundation of the two models such as: the creation of procedure type risk categories using empiric and expert opinion-based methodology, identified markers for hemodynamic vulnerability, and assessment of additional patient-level risk factors. The IMPACT risk adjusted MAE rate reporting methodology has contributed another credible metric for comparison of complication rates in pediatric interventional and congenital heart disease therapy.

Table 64.2 Definitions for adverse event severity

Severity level	Definition	Examples
1–None	No harm, no change in condition, may have required monitoring to assess for potential change in condition with no intervention indicated	Balloon rupture
		Equipment problem
2–Minor	Transient change in condition, not life-threatening, condition returns to baseline, required monitoring, required minor intervention such as holding a medication, or obtaining laboratory test	Groin hematoma
		Self-resolving arrhythmia
3–Moderate	Transient change in condition may be life-threatening if not treated, condition returns to baseline, required monitoring, required intervention such as reversal agent, additional medication, transfer to the intensive care unit for monitoring, or moderate transcatheter intervention to correct condition	Unstable arrhythmia with preserved blood pressure requiring intervention
		Vascular damage not life-threatening but requiring intervention
4–Major	Change in condition, life-threatening if not treated, change in condition may be permanent, may have required an intensive care unit admission or emergent readmit to hospital, may have required invasive monitoring, required interventions such as electrical cardioversion or unanticipated intubation or required major invasive procedures or transcatheter interventions to correct condition	Event requiring cardiopulmonary resuscitation
		Event leading to surgery or repeat catheterization
		Stroke
5–Catastrophic	Any death, and emergent surgery or heart lung bypass support (ECMO) to prevent death with failure to wean from bypass support	Event resulting in death

64.4.2.2 Procedure Risk Classification

Since the development of CHARM, there have been numerous advancements in the field of pediatric catheterization including novel procedures, advancements in technology and techniques, as well as a better understanding the most important patient determinants of risk. The second iteration of CHARM improves upon and modernizes established risk adjustment methodology by identifying events at the case level, yielding improved procedure risk classification, and the expansion from four stratified categories in CHARM to five risk categories in CHARM II for consideration (Table 64.3).

64.4.2.3 Hemodynamic Vulnerability

Hemodynamic vulnerability of a patient is an important consideration when developing a risk adjustment and/or risk prediction methodology and serves as a strong independent determinant of risk. In addition to accounting for contemporary procedures, determination of hemodynamic risk has been improved upon in CHARM II with a new weighted score rather than simple presence of hemodynamic variables of vulnerability. CHARM II includes the most predictive hemodynamic variables, now validated in multiple data sets, stratified when appropriate by single or biventricular circulation and including: systemic arterial saturation, mixed venous saturation, pulmonary artery pressure, systemic ventricle end-diastolic pressure (EDP), and ratio of pulmonary to systemic blood flow (Qp:Qs) (Table 64.4).

64.4.2.4 Risk Prediction

Advancements in risk prediction included the development of a Catheterization RISk Score for Pediatrics (CRISP) [6] developed by the Congenital Cardiac Interventional Study Consortium (CCISC) registry participants in 2007. This prediction tool provides a pre-catheterization risk scoring system that can be applied to individual pediatric patients undergoing cardiac catheterization procedures to determine risk of an adverse event based on anticipated procedure type and patient characteristics. The CRISP calculator has been made available online and serves as a widely

Table 64.3 CHARM II procedural risk categories

	Risk category 1	Risk category 2	Risk category 3	Risk category 4	Risk category 5
Diagnostic case		Diagnostic 1 month—<1 year Diagnostic ≥1 year	Diagnostic <1 month		
Valvuloplasty	Pulmonary valvotomy ≥1 month	Pulmonary valvotomy + procedure	Pulmonary valvotomy <1 month		Aortic valvotomy ± procedure, <1month Aortic valvotomy ± procedure, ≥1 month Mitral valvotomy Atretic valve perforation and/or valvotomy
Device or coil closure	Fontan fenestration or baffle leak device closure	ASD or PFO closure Venous collateral occlusion PDA closure	Fontan fenestration or baffle leak device closure + procedure Systemic pulmonary collateral closure ± procedure	ASD or PFO closure + procedure	VSD closure

Balloon angioplasty and/or stent placement			Pulmonary artery (1 vessel) Pulmonary artery (1 vessel) + RVOT conduit dilation/ stent Aorta (coarctation) dilation and/or stent	Pulmonary artery (1 vessel) + procedure Pulmonary artery (≥2 vessels) Pulmonary vein dilation and/or stent RVOT conduit dilation and/or stent PDA dilation and/or stent	Pulmonary artery (≥2 vessels) + RVOT and/or other procedure Aorta (coarctation) dilation and/or stent + procedure
Other	Endomyocardial biopsy Endomyocardial biopsy with coronary angiography		Atrial septostomy		Atrial septostomy + procedure TPV implantation ± procedure Atrial septum static dilation and/or stent placement

Table 64.4 Hemodynamic vulnerability score

Hemodynamic indicator	Qualifier	Weighted Score (0–2)
Systemic arterial saturation	BiV: <95%	1
	SV: <78%	2
Mixed venous saturation	BiV: <60%	1
	SV: <50%	1
Pulmonary artery pressure	BiV: ≥45 mmHg	2
	SV: ≥17 mmHg	2
Systemic ventricle EDP	≥18 mmHg	1
Qp:Qs	>1.5	1

BiV biventricular, *SV* single ventricle, *EDP* end-diastolic pressure

popular tool to predict procedural risk and used by operators and for patient counseling. Future improvements in risk prediction can be anticipated based on the recent enhancements integrated in CHARM II, specifically the new procedure risk categorization and a better understanding of hemodynamic risk.

64.4.2.5 Procedural Efficacy

As the field evolves and considers additional outcome assessment tools, it will be important to consider metrics that measure procedural efficacy. Rather than simply measuring complications, metric developers are considering a composite measure for comparison which takes into consideration achieving procedural goals while not experiencing an adverse event during a procedure. For example, a metric for a balloon valvuloplasty procedure would consider both the change in gradient achieved with the intervention, as well as any complication occurrence. Such a metric would account for an important limitation of measuring just complications, and capture missed opportunities such as a valvuloplasty procedure which is ineffective at reducing the valve gradient but does not result in an adverse event, perhaps due to risk aversion. Work on this type of metric is currently being piloted but may be an important tool to assess institutional outcomes and operator performance in the future.

64.4.2.6 Resiliency

Fortunately, major adverse events in pediatric cardiac catheterization only occur in 1–2% of cases and rarely result in permanent injury. But when a life-threatening event occurs, there may be a critical metric to consider with a significant opportunity to improve clinical care and patient outcomes. Consider the occurrence of identical events with different outcomes impacted by the resiliency of the system to manage the event effectively and efficiently. For example, a device may embolize during a procedure and successful retrieval and management of the embolized device may result in no harm to the patient, whereas failure to successfully retrieve a device may result in further adverse events, need for cardiac surgery, and/or possible end organ damage. Also, consider a cardiac arrest with an expedited resuscitation with all required resources compared to a prolonged and ineffective resuscitation due to delays in resources or ineffective management. A successful metric for "resiliency" will take into consideration two different outcomes following a primary event (Fig. 64.4) with a

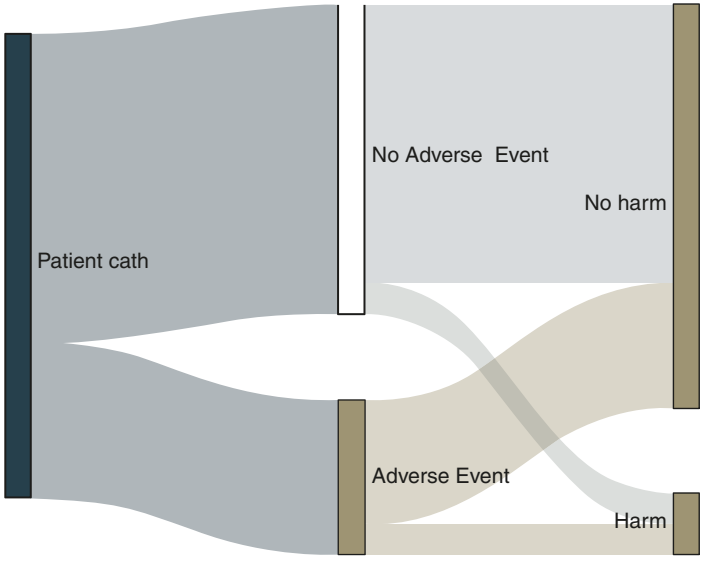

Fig. 64.4 Clinical outcomes determined by the presence of patient harm post procedure

potential to strengthen a system when opportunities for improvement can be identified. Through future identification of modifiable factors that improve resiliency, a cardiac catheterization team will be empowered to anticipate hazards in advance, make plans to mitigate risk, strengthen responses of the system, and improve appropriate access to resources.

64.5 Final Considerations

With an increasing focus of providing quality care within the field over the past decade, many successful strides have been made to promote a QI culture with the aim of providing outcome assessment methodology, radiation safety best practices, and risk mitigation strategies within the congenital cardiac catheterization laboratory. By implementing appropriate QI methodology and enhancing collaboration with large-scale dataset analysis, the goals of achieving optimal patient care and safety are possible. Although many influential advancements have occurred, as outlined above, more work needs to be done to ensure optimal delivery of care and a continued focus on patient-centered outcomes without sacrificing procedural efficacy.

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65 Hemodynamic Formulae, Calculations, and Charts

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65.1 Body Surface Area Formulae (BSA)

Mosteller [1]:

$$\text{BSA}\ \left(\text{m}^2\right) = \sqrt{\frac{\text{height}\ \left(\text{cm}\right) \times \text{weight}\ \left(\text{kg}\right)}{3{,}600}}$$

Dubois and Dubois [2]*:*

$$\text{BSA}\ \left(\text{m}^2\right) = 0.007184 \times \text{height}\ \left(\text{cm}\right)^{0.725} \times \text{weight}\ \left(\text{kg}\right)^{0.425}$$

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65.2 Derived Hemodynamic Data

65.2.1 Cardiac Output

$$SV = EDV - ESV$$

$$CO = SV \times HR$$

$$EF = SV / EDV$$

where SV = stroke volume, EDV and ESV = end-diastolic and end-systolic volumes, CO = cardiac output, HR = heart rate, and EF = ejection fraction.

65.2.2 The Fick Equation

$$Q\ (\mathrm{l/min}) = \frac{\mathrm{VO_2\ (ml\ O_2/min)}}{\mathrm{arterial\ O_2\ content - venous\ O_2\ content\ (ml\ O_2/l)}}$$

where ***Q*** is cardiac output expressed in liters per minute (l/min) and VO2 is the oxygen consumption in ml O2/min.

The denominator of the Fick equation is the arteriovenous oxygen content difference (a − v O_2 diff) and is expressed as ml O_2/l of blood.

$$\mathrm{Oxygen\ capacity\ (ml\ O_2/l)} = \mathrm{Hgb\ (g/l)} \times 1.39\ \mathrm{(ml\ O_2/g\ of\ Hgb)}$$

The oxygen content of the blood is the amount of oxygen in that specific sample (either arterial or venous) and can be estimated by the following formula:

$$\mathrm{C_aO_2\ (ml\ O_2/l)} = \mathrm{Oxygen\ capacity\ (ml\ O_2/l)} \times \mathrm{arterial\ oxygen\ saturation\ (\%)}$$

$$\mathrm{C_vO_2\ (ml\ O_2/l)} = \mathrm{Oxygen\ capacity\ (ml\ O_2/l)} \times \mathrm{venous\ oxygen\ saturation\ (\%)}$$

If the patient is breathing enriched oxygen ($F_I O_2 > 30\%$), the amount of dissolved oxygen must be accounted for in the flow equation. The solubility coefficient of oxygen in plasma is 0.00003 O_2 ml/ml plasma/mmHg O_2 tension or 0.032/l of plasma.

$$\mathrm{C_aO_2} = \text{Oxygen capacity} \times \text{arterial oxygen saturation } (\%) + 0.032 \times \mathrm{P_aO_2}\,(\mathrm{mmHg})$$

$$\mathrm{C_vO_2} = \text{Oxygen capacity} \times \text{venous oxygen saturation } (\%) + 0.032 \times \mathrm{P_vO_2}\,(\mathrm{mmHg})$$

65.2.3 Assessment of Flows and the Q_p:Q_s Ratio

Flow calculations are based on the Fick principle and can be applied to both pulmonary (Q_p) and systemic blood flows (Q_s).

Q_p can be estimated by the following equation:

$$Q_p = \frac{\mathrm{VO_2}}{\text{pulmonary venous } \mathrm{O_2} \text{ content} - \text{pulmonary arterial } \mathrm{O_2} \text{ content}} \text{ or}$$

$$Q_p = \frac{\mathrm{VO_2}\,(\text{ml } \mathrm{O_2}/\min)}{(\mathrm{PV\ sat} - \mathrm{PA\ sat}) \times 1.39 \times \mathrm{Hgb}\,(\mathrm{g/l})}$$

where PV is pulmonary vein and PA is pulmonary artery saturation.

Similarly, Q_s is estimated as

$$Q_s = \frac{\mathrm{VO_2}}{\text{systemic arterial } \mathrm{O_2} \text{ content} - \text{mixed venous } \mathrm{O_2} \text{ content}} \text{ or}$$

$$Q_s = \frac{\mathrm{VO_2}\,(\text{ml } \mathrm{O_2}/\min)}{(\mathrm{Ao\ sat} - \mathrm{MV\ sat}) \times 1.39 \times \mathrm{Hgb}\,(\mathrm{g/l})}$$

where Ao is aortic and MV is mixed venous saturation.

Finally, effective pulmonary blood flow (Q_{ep}) is the amount of deoxygenated blood that is pumped to the lungs.

$$Q_p = \frac{VO_2}{\left(\text{pulmonary venous } O_2 \text{ content} - \text{mixed venous } O_2 \text{ content}\right)} \text{ or}$$

$$Q_{ep} = \frac{VO_2\left(\text{ml/min}\right)}{\left(\text{PV sat} - \text{MV sat}\right) \times 1.39 \times \text{Hgb}\left(\text{g/l}\right)}$$

$$\text{Mixed venous saturation} = \frac{\left(3 \times \text{SVC sat} + \text{IVC sat}\right)}{4} \text{ or}$$

$$= \frac{\text{SVC sat} - \left(\text{SVC sat} - \text{IVC sat}\right)}{4}$$

$$Q_p : Q_s = \frac{\left(\text{Ao sat} - \text{MV sat}\right)}{\left(\text{PV sat} - \text{PA sat}\right)}$$

where Ao is the aortic saturation, MV is the mixed venous saturation, and PV and PA saturations are the pulmonary vein and artery, respectively. SVC is superior caval and IVC inferior caval vein saturations.

65.2.4 Oxygen Transport

Global oxygen delivery (DO_2), also known as systemic oxygen transport (SOT):

$DO_2 = Q_s \times C_aO_2$ expressed in ml/min.

The oxygen extraction ratio (O_2ER):

$$O_2ER = \frac{VO_2}{DO_2}.$$

65.2.5 Resistance (Wood Units)

Pulmonary vascular resistance:

$$\mathrm{PVR} = \frac{(\mathrm{mPAP} - \mathrm{mLAP})}{Q_{\mathrm{p}}}$$

where PVR = pulmonary vascular resistance, mPAP = mean pulmonary artery pressure, mLAP = mean left atrium pressure (alternatively, pulmonary vein or PCWP may be used), and Q_{p} = pulmonary blood flow.

Similarly, systemic vascular resistance can be calculated as follows:

$$\mathrm{SVR} = \frac{(\mathrm{mAoP} - \mathrm{mRAP})}{Q_{\mathrm{s}}}$$

where SVR = systemic vascular resistance, mAoP = mean arterial pressure, mRAP = mean right atrial pressure, and Q_{s} = systemic blood flow.

Wood units × 80 = dyne − sec − cm − ^{5}.

Normal values:

PVRI: 1–3 Wood units × m^2 or 80–240 dyn × s × cm − ^{5} × m^2.

SVRI: 15–30 Wood units × m^2 or 800–1600 dyn × s × cm − ^{5} × m^2.

65.2.6 Oxygen Consumption per Body Surface Area (ml/min/m²) by Gender, Age, and Heart Rate [3, 4]

Oxygen consumption (assumed values):

- Infant <3 months is ~130 ml/min/m^2.
- 2–5 years ~150–200 ml/min/m^2.
- Adolescents ~120–180 ml/min/m^2.
- Adult females ~100 ml/min/m^2.
- Adult males ~110–120 ml/min/m^2.
- 1–2 years ~200 ml/min/m^2.

65.2.6.1 Male Patients

	Heart rate (bpm)												
Age	50	60	70	80	90	100	110	120	130	140	150	160	170
3	155	159	163	167	171	175	178	182	186	190			
4	149	152	156	160	163	168	171	175	179	182	186		
6	141	144	148	151	155	159	162	167	171	174	178	181	
8	136	141	144	148	152	156	159	163	167	171	175	178	
10	130	134	139	142	146	149	153	157	160	165	169	172	176
12	128	132	136	140	144	147	151	155	158	162	167	170	174
14	127	130	134	137	142	146	149	153	157	160	165	169	172
16	125	129	132	136	141	144	148	152	155	159	162	167	
18	124	127	131	135	139	143	147	150	154	157	161	166	
20	123	126	130	134	137	142	145	149	153	156	160	165	
25	120	124	127	131	135	139	143	147	150	154	157		
30	118	122	125	129	133	136	141	145	148	152	155		
35	116	120	124	127	131	135	139	143	147	150			
40	115	119	122	126	130	133	137	141	145	149			

65.2.6.2 Female Patients

	Heart rate (bpm)												
Age	50	60	70	80	90	100	110	120	130	140	150	160	170
3	150	153	157	161	165	169	172	176	180	183			
4	141	145	149	152	156	159	163	168	171	175	179		
6	130	134	137	142	146	149	153	156	160	165	168	172	
8	125	129	133	136	141	144	148	152	155	159	163	167	
10	118	122	125	129	133	136	141	144	148	152	155	159	163
12	115	119	122	126	130	133	137	141	145	149	152	156	160
14	112	116	120	123	127	131	134	133	143	146	150	153	157
16	109	114	118	121	125	128	132	136	140	144	148	151	
18	107	111	116	119	123	127	130	134	137	142	146	149	
20	106	109	114	118	121	125	128	132	136	140	144	148	
25	102	106	109	114	118	121	125	128	132	136	140		
30	99	103	106	110	115	118	122	125	129	133	136		
35	97	100	104	107	111	116	119	123	127	130			
40	94	98	102	105	109	112	117	121	124	128			

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Correction to: Melody Valve Implantation in Pulmonary Position

Gianfranco Butera, Simon Thomas MacDonald, Massimo Chessa, and Philipp Bonhoeffer

Correction to:
Gianfranco Butera (Ed) ***Cardiac Catheterization for Congenital Heart Disease: From Fetal Life to Adulthood***
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This book was inadvertently published with the incorrect Author name. This has now been amended throughout the book from Simon Mac Donald to Simon Thomas MacDonald.

The updated version of the book can be found at https://doi.org/10.1007/978-3-030-69856-0_42

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