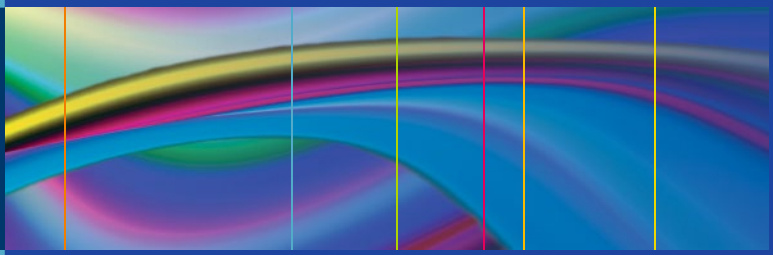
Gianfranco Butera · Massimo Chessa
Andreas Eicken · John Thomson *Editors*



Cardiac Catheterization for Congenital Heart Disease

From Fetal Life to Adulthood

Forewords by
Shakeel Qureshi and Mario Carminati

Springer

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Editors
Gianfranco Butera
Pediatric and Adult Congenital Heart Center
IRCCS Policlinico San Donato
Milano
Italy

Massimo Chessa
Pediatric and Adult Congenital Heart Center
IRCCS Policlinico San Donato
Milano
Italy

Andreas Eicken
Department of Pediatric Cardiology and Congenital Heart Disease
German Heart Center, Technical University of Munich
Munich
Germany

John Thomson
Pediatric Cardiology
Yorkshire Heart Centre
Leeds
UK

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Foreword

There has been a revolution in cardiac catheterization and interventional techniques in congenital heart disease in the last three decades and, in particular, in the 1990s and the early part of this century. There is always concern amongst clinicians that the pace of developments is likely to slow down. However, fortunately, there does not seem to be any slowing in the progress of developments of newer interventional techniques. Therefore, there has been a need for a book dealing with practical aspects of cardiac catheterization in congenital heart disease. Such books are not only aimed for trainees and interventionists in early parts of their careers, but they are also invaluable to experienced interventionists, helping them to keep abreast of developments. The Editors of this handbook on *Cardiac catheterization for Congenital Heart Disease: From Fetal life to Adulthood* are to be commended for their efforts and for addressing this important task.

This book, composed of 47 chapters, will attract a large audience from all over the world, thanks to the involvement of some of the leading authorities in our specialty. The interventional techniques from the fetus to the adult with congenital heart

disease are covered. All the technical chapters follow a similar format, dealing with important practical steps that each interventionist needs to know. These include the indications for interventions, any pre-procedure assessment, step-by-step technical information, as well as practical tips and tricks. Issues such as informed consent represent important steps in the performance of interventional procedures, and these aspects are not forgotten in this book. There are new interventional balloons and devices appearing regularly, and attempts are made to provide up-to-date information on these. Interventionists need detailed step-by-step information about access into the heart, and so usual and unusual accesses are dealt with. Inevitably with the interventional techniques there will be a spectrum of complications, and so these and many retrieval techniques are addressed.

The editors have produced a much-needed book at the correct time. It will act as a quick practical guide to all the interventionists. I have no doubt whatsoever that it will become a crucial requirement as a stock item for all the departments and libraries.

Evelina Children's Hospital, Shakeel A. Qureshi
London, UK

Foreword

It is my great pleasure to write a foreword for this book on transcatheter interventions of congenital heart defects. The volume follows "how-to" format, as a result of a worldwide cooperation of many international experts, who put together their knowledge and expertise.

As the chief of Pediatric and Adult Congenital Cardiology Department at Policlinico San Donato IRCCS, I have been involved for many years in the fascinating world of catheter interventions in congenital heart defects. During this period I had the pleasure to be the mentor of Gianfranco Butera and Massimo Chessa, who shared with me the enthusiasm in taking part of continuous and rapidly evolving process of a variety of new techniques in this field. They should be congratulated, together with Andreas Eicken and John Thomson, as Editors of this book, for the excellent work they did. This book can really become a day by day companion of

all young fellows and trainees around the world. I am convinced that also seniors and already expert interventionalists will find it useful in the daily practice.

Pediatric Cardiology Unit,
IRCCS Policlinico San Donato,
Milan, Italy

Mario Carminati

Preface

The population of patients recognized with congenital heart disease has dramatically increased over the past years as a consequence of significant advances in diagnosis and the improvement in surgical and transcatheter techniques.

Actually, the number of adults with congenital heart disease in the United States is almost the same as that of children with these anomalies. Additionally, the evolutions in both device technology as well as non-invasive imaging technology have allowed the safe and effective catheter-based treatment of various congenital or post-surgical residual cardiac defects. Furthermore, these patients have a quicker recovery and a shorter hospital stay, and a better quality of life.

On the other hand, these techniques require not only a specific training, but also the necessity of a tight collaboration between pediatric and adult interventionist as well as with a trained echo-cardiographer and anesthesiologist.

Actually, interventional cardiology of subjects with congenital heart disease is a well-established field on its own and, programmatically, is a fundamental component of any center providing care for these patients.

Excellence in the field of interventional cardiology is obtained with passion, competence, hard study and attention to details.

This book is the result of the collaboration of more than 30 world renowned experts from 4 continents and gives plenty of details, tips and tricks on more than 40 different topics.

It provides a practical guide to the large majority of procedures performed during daily life, and its step-by-step approach will be a precious tool during hard times in the catheterization laboratory.

Almost everything is in the book, to the reader the effort to put in hard work and passion!

Milan, Italy — Gianfranco Butera
Milan, Italy — Massimo Chessa
Munich, Germany — Andreas Eicken
Leeds, UK — John Thomson

Contents

Part I
General Issues

Chapter 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 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

M. Witsenburg
Departments of Congenital and Pediatric Cardiology, Thoraxcentre, Erasmus MC, 2040, 3000 CA Rotterdam, The Netherlands
e-mail: m.witsenburg@erasmusmc.nl

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informed appropriately. The combination of the duty to inform and the agreement of the patient with the treatment plan is called informed consent.

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. 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.

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 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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Chapter 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

G. Isgrò, MD • M. Ranucci, MD, FESC (✉)
Department of Cardiothoracic and Vascular Anaesthesia and Intensive Care, IRCCS Policlinico San Donato, San Donato Milanese, Milan, Italy
e-mail: cardioanestesia@virgilio.it

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arteriosus closure, or STENT implantation), during which it is essential to keep the patient deeply relaxed to permit the precise deployment of the device.

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

Table 2.1 Suggested anaesthetic drugs

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

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.

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).

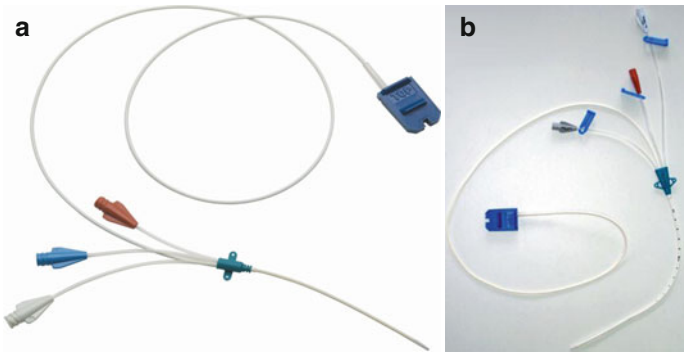

Fig. 2.1 (**a**, **b**) PediaSat catheter (paediatric) (**a**) and PreSep catheter (adult) (**b**) (Courtesy of Edwards Lifesciences)

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. 2.2 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 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

Table 2.2 Suggested emergency drugs

Glucose 33 %
Atropine
Epinephrine
Norepinephrine
Dopamine
Isoproterenol
Diazepam
Calcium chloride or calcium gluconate
Sodium Bicarbonate
Lidocaine
Corticosteroids
Aminophylline

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).

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Chapter 3
Antibiotics and Anticoagulation

Luciane Piazza

In the catheterization laboratory, antibiotics are used to prevent bacterial endocarditis. Despite advances in diagnosis, antimicrobial therapy, surgical techniques and the management of complications, infective endocarditis (IE) is still a serious problem with high rates of mortality and morbidity. Theoretically endocarditis can be prevented using prophylactic antibiotics. Therefore, since 1955, IE antibiotic prophylaxis strategies have been in place for patients with cardiac disease potentially at risk of IE following bacteraemia during an invasive procedure.

It is important to recall two concepts:

(a) Prophylaxis—The administration of an antimicrobial substance in the periprocedural period in the absence of clinical infection to prevent an infectious complication
(b) Bacteraemia—The presence of bacteria within the bloodstream with or without clinical signs or symptoms of infection

L. Piazza
Pediatric and Congenital Heart Disease, Policlinico San Donato IRCCS, Via Morandi 30, San Donato Milanese, Milan 20097, Italy
e-mail: lppiazza@hotmail.com

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In the catheterization laboratory, specific interventions to prevent infections consist of measures to prevent bacterial ingress into the patient and antibiotic prophylaxis.

3.1 Infection Control Measures

- It consists of infection-control measures and specific techniques. The most important recommendations for sterility are (a):

  1. For physicians (Table 3.1)
  2. The laboratory environment (Table 3.2)
  3. For patients (Table 3.3)

Table 3.1 Recommendation for physicians

Hand washing	The most important procedure to prevent infection. For a minimum of 2–3 min, thoroughly scrub the hands and distal arms using an antiseptic agent. In some units it is acceptable to use volatile antiseptic solution in subsequent cases after an initial thorough wash to prevent skin irritation and dermal abrasion
Body protection	Each laboratory should have a specific policy on caps, masks and gowns and gloves. As a minimum, sterile gowns and gloves should always be worn by operators in contact with the patient and the scrub trolley

Table 3.2 Environmental catheterization of the laboratory

Clothes prevention	Appropriate laboratory-specific clothes and shoes are necessary
Cleaning of the room	The laboratory must be cleaned in accordance with the institutional requirements. In particular policies should be in place for blood spillage; notifiable infections, e.g. MRSA; and other potentially infectious situations, e.g. a patient with an active viral illness such as HIV or hepatitis B where specific cleaning of the laboratory may be required
Air vents	Well-serviced and efficient ventilation is a mandatory requirement. Local regulations should be in place

Table 3.3 Recommendation for patient preparation

Hair removal	Remove hair at the access site using clipper or depilatory on the day of the procedure or the day before
Skin cleaning	A 2 % chlorhexidine with alcohol is the most effective antiseptic agent but can be substituted with povidone-iodine. Take care in neonates and premature infants because of increased risk of skin irritation or even skin burns and absorption (b)
Drapes	As in an operating theatre, nonporous drapes are necessary to cover the working area. Drapes should be large enough and should contain an appropriately sized adhesive part attached to the skin to be attached around the area involved in the intervention

3.2 Antibiotic Prophylaxis

Prophylactic intravenous antibiotic should be routinely administered to high-risk patients (Tables 3.4 and 3.5) undergoing cardiac catheterization. The practical issues related to prophylaxis are the following:

- The choice of the antimicrobial agent
- The timing of the first administrated dose
- The duration of the prophylactic regimen

Most catheterizing cardiologists do not give antibiotic prophylaxis for standard diagnostic catheterization. In some units, antibiotics can be given in case of prolonged or complicated procedures.

However, other clinical conditions may be the causes of bacteraemia. For this reason, the physician has to consider each situation in order to better identify the most adequate indication for prophylaxis. For example, different conditions could be used for preterms, neonates and immunocompromised patients.

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 mouths 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 because of immunosuppression

Table 3.5 Antibiotic prophylaxis: dose and regimen

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

3.3 Anticoagulation in Catheterization Laboratory

Heparin is frequently used during cardiac catheterization to prevent thrombosis and thrombus formation. The most common access points for cardiac catheterization are the femoral artery and femoral vein, although the jugular vein and brachial or radial arteries are used on occasion.

In all interventional procedures, heparin is also used as a thromboprophylactic agent. Heparin has the advantage over many other anticoagulants that it has a short half-life and is easily reversible. It has also been used for many decades so there is considerable therapeutic experience.

Table 3.6 Type of procedure and heparin dosage

Type of procedure	Heparin dosage in bolus
Diagnostic venous catheterization	50 UI/kg
Diagnostic arterial catheterization	50–100 UI/kg
Venous and arterial diagnostic catheterization	50–100 UI/kg
Interventional catheterization	100 UI/kg

Table 3.7 Reversal of heparin therapy

Time since last heparin dose (min)	Protamine dose in mg (per 100 U Heparin received by the patient)
<30	1.0
30–60	0.5–0.75
60–120	0.375–0.5
>120	0.25–0.375

The effect of a particular dose must be evaluated in order to reduce excessive bleeding (Table 3.6). It can be difficult to predict the therapeutic level in an individual patient, because there is a high degree of variability in response. Another inconvenient issue is that of heparin-induced thrombocytopenia. This is an immune-mediated adverse reaction that occurs in 3 % of patients exposed to heparin for ≥ 5 days.

It is necessary and relatively quick and easy to measure the heparin level during the procedure using the activated clotting time (ACT) and/or partial thromboplastin time (APTT). Usually the ACT can be done after 30–60 min of bolus heparin. It can be repeated if in any doubt (bleeding or suspected thrombotic formation).

Intravenous protamine can be used to acutely reverse the effect of heparin (Table 3.7).

The maximum dose of protamine is 50 mg. Infusion rate of 10 mg/ml solution should not exceed 5 mg/min.

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

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Chapter 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

J.L. Zunzunegui
Pediatric Cardiology Unit, Gregorio Marañon Hospital,
O'Donell 48-50, Madrid 28009, Spain
e-mail: zunzu69@hotmail.com

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(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 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.

X-rays that reach the target are converted into electrons once again by interacting with the input phosphor and photocathode; 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, aorto-pulmonary 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 1,570 mosm/kg H_2O, while IOCM have an osmolality similar to serum at 290 mosm/kg H_2O.

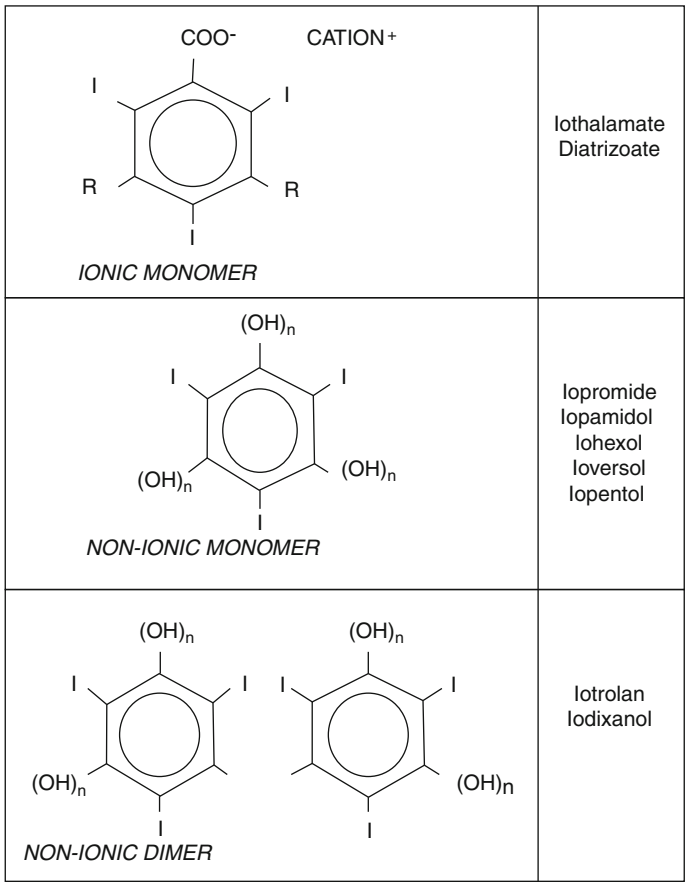

Fig. 4.1 Chemical structure of iodinated contrast agents and examples of contrast media

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.

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 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.

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].

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Chapter 5
Angiography: Radiation Exposure and Standard Projections

Federico Gutierrez-Larraya, Angel Sanchez-Recalde, and Enrique Balbacid-Domingo

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.

F. Gutierrez-Larraya (✉) • A. Sanchez-Recalde • E. Balbacid-Domingo
Department of Pediatric Cardiology, Hospital Universitario La Paz, Paseo de la Castellana 261, Madrid 28046, Spain
e-mail: flarraya@yahoo.es; asrecalde@hotmail.com; enriquebalbacid@hotmail.com

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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 misrepair 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 1,717 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 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).

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
Fis.: 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
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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

b

Protocolo de examen

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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  5sDR-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
Fis.: 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
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Fig. 5.1 (continued)

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 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

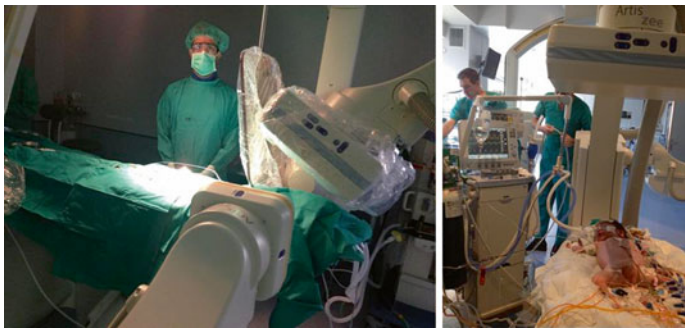


Fig. 5.2 Pediatric patients are often very sick and need ventilator, pumps, GE tubes, etc. Patient should be close to flat detector or intensifier; operator should use shields, curtains, glasses, collars, and apron. All these cautions make often impossible to achieve convenient angulations or biplane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

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

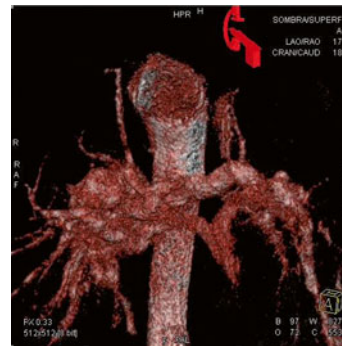


Fig. 5.3 3D reconstruction from an angiographic computed tomography (ACT) acquired and processed during the catheterization. Best C-arm angulation can be predicted upon figure movement. Aortogram from an infant with pulmonary atresia and ventricular septal defect with multiple aortopulmonary collaterals

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 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 210[a], 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

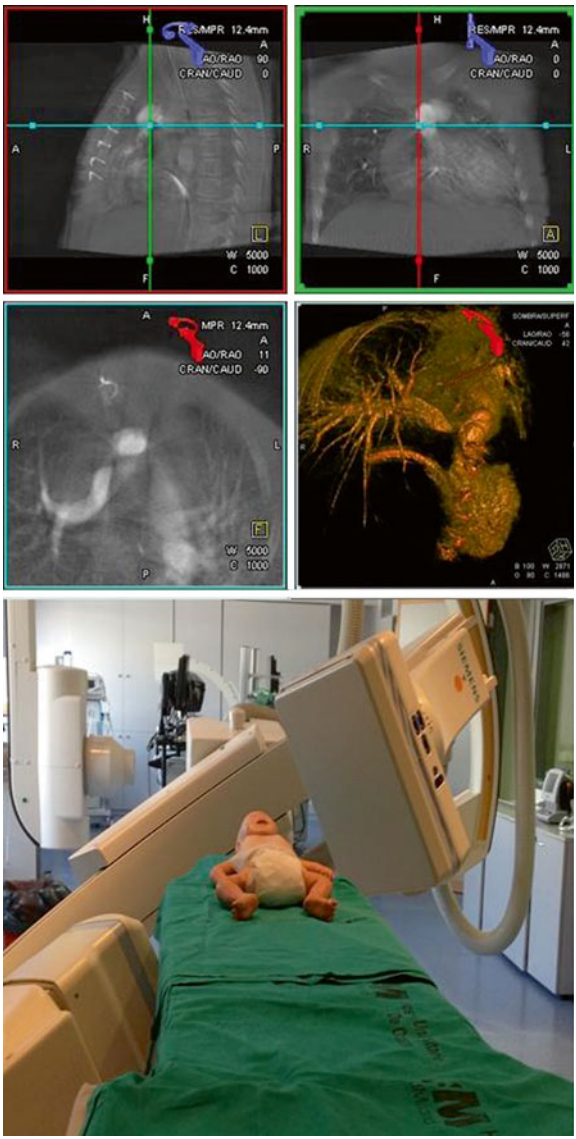
Specific lesions	Angles, plane A	Angles, plane B
Pulmonary stenosis	0° + 30° cranial	Left lateral
RVOT-MPA (sitting up)	10° LAO + 40° cranial	Left lateral
Long axial for LPA biplane	30° RAO	60° LAO + 30° cranial
LPA long axis		60° + 20° cranial
ASD	30° LAO + 30° cranial	
PA bifurcation and branches	30° caudal + 10° RAO	20° caudal

(continued)

Table 5.1 (continued)

Specific lesions	Angles, plane A	Angles, plane B
Left ventricular outflow tract obstruction	RAO	Long-axis oblique
Aortic coarctation	0°/shallow RAO/ shallow LAO	Left LAT/long axis oblique
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	

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


RES/MPR 12.4mm
A
AO/RAO 90
CRAN/CAUD 0
W 5000
C 1000
RES/MPR 12.4mm
A
AO/RAO 0
CRAN/CAUD 0
W 5000
C 1000
MPR 12.4mm
A
AO/RAO 11
CRAN/CAUD -90
W 5000
C 1000

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

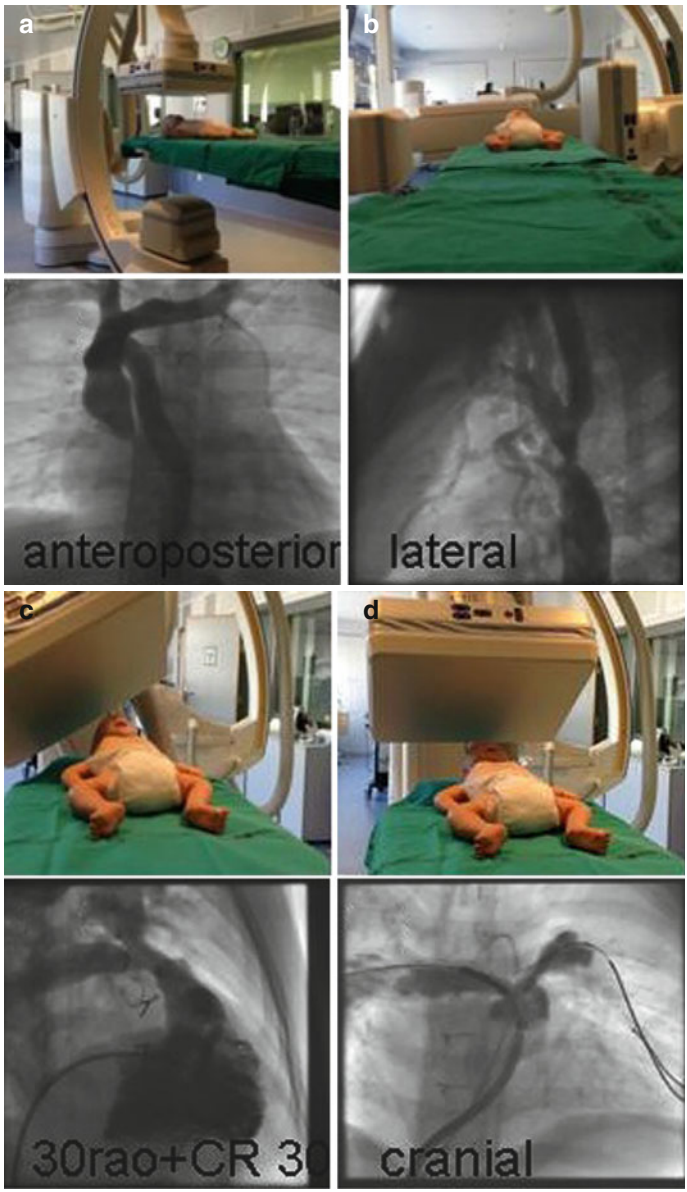
5.7.1 Secundum Atrial Septal Defect and Fenestrated Fontan

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° 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

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


a
b
anteroposterior
lateral
c
d
30rao+CR 3
cranial

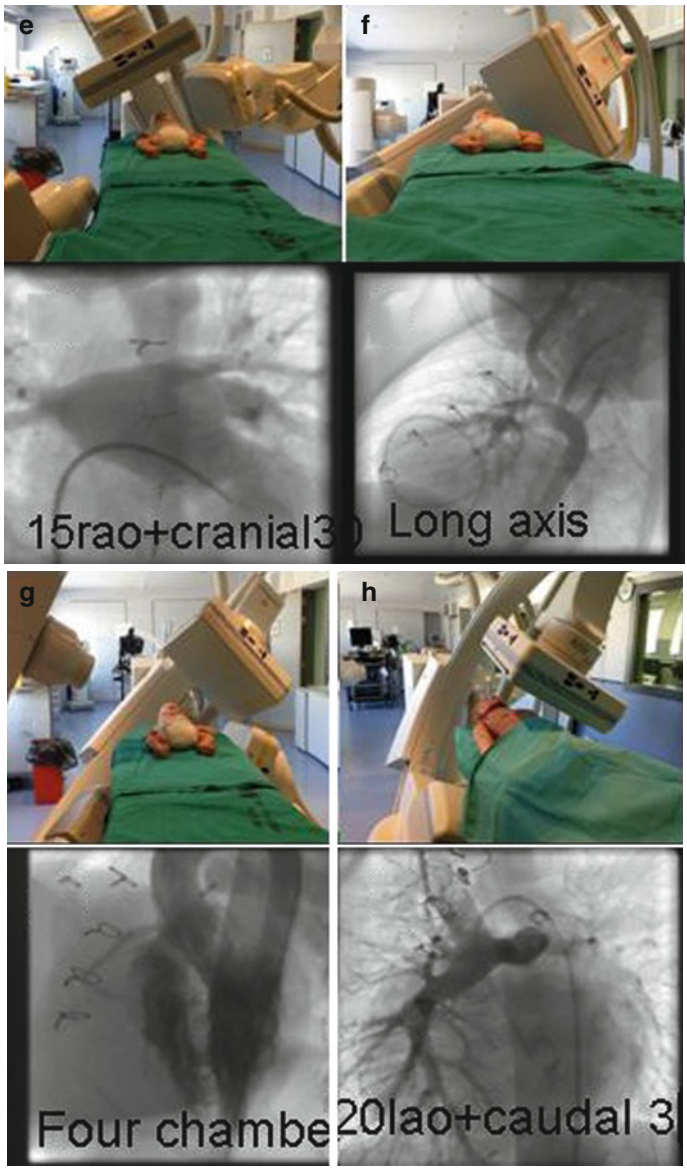


Fig. 5.5 (continued)

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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Chapter 6
Catheters and Wires

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 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.

A. Koleśnik (✉) • G. Brzezińska-Rajszys
Cardiovascular Interventions Laboratory,
The Children's Memorial Health Institute,
Al. Dzieci Polskich 20, Warsaw 04-730, Poland
e-mail: adam.kolesnik@wum.edu.pl; gbrzezinska@hoga.pl

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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 the 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-tipped catheters have a CO_2 inflatable balloon at their tips. This balloon is supposed to allow free floating with the blood stream and prevent tangling between the chordae tendineae in the cardiac chambers. Other catheters have hydrophilic coating that makes them slippery and facilitates 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.

Pigtail catheters have their tip shaped in a shape suggested by their name (Figs. 6.1a and 6.2). They are thin walled especially at their distal ends, and this makes them soft but susceptible to kinking. Thus, pigtail catheters have to be advanced and withdrawn with the guidewire inside them. While passing through the arterial valves retrogradely, the loop of the guidewire should precede the tip of the catheter to prevent kinking in the valvar sinuses. Side holes are placed proximal to the curved tip. This should be kept in mind when positioning the catheter for contrast infusion or pressure measurement.

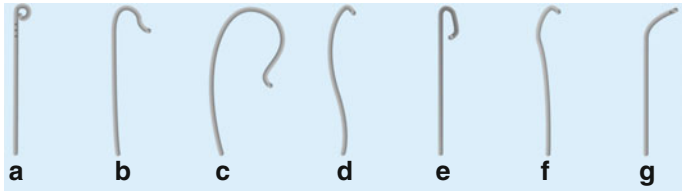

Fig. 6.1 Shapes of selected torque-controlled catheters (see text). (**a**) Pigtail catheter, (**b**) Amplatz left coronary catheter, (**c**) Amplatz right coronary catheter, (**d**) internal mammary catheter, (**e**) Judkins left coronary catheter catheter, (**f**) judkins right coronary catheter, (**g**) multipurpose catheter

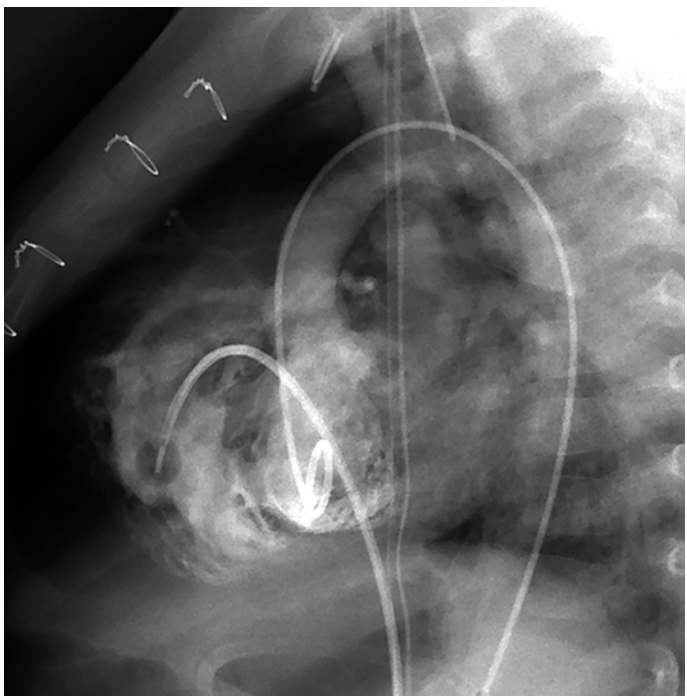

Fig. 6.2 Left ventriculography (retrograde approach) with pigtail catheter (Cordis, Miami Lakes, FL) – lateral projection. Multiple ventricular septal defects in a 2-year-old patient after pulmonary artery banding and surgical repair of aortic arch hypoplasia and coarctation

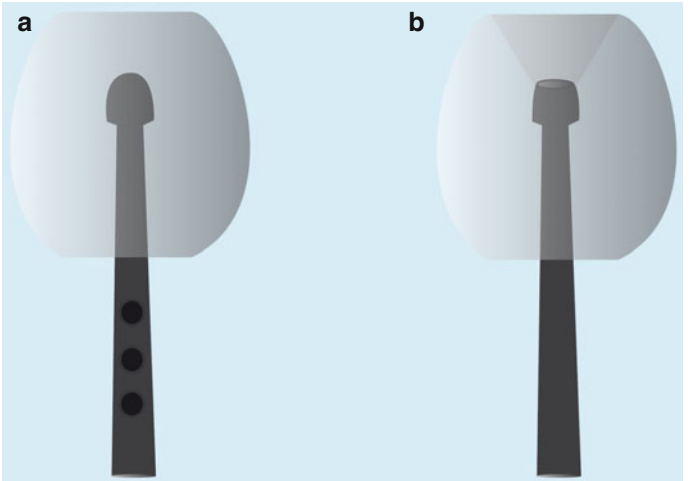

Fig. 6.3 Tips of floating catheters. (**a**) Berman angiography balloon catheter. (**b**) Pulmonary wedge balloon catheter

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. Therefore, they are not described in this chapter.

Berman angiographic catheter is a balloon-tipped catheter without the end hole (Fig. 6.3a). 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. Usually Berman catheter is used to access the right ventricle and the pulmonary arteries (Fig. 6.4). However, in the presence of interatrial or interventricular communications, it can be used to catheterize the left heart structures as well (Fig. 6.5). Antegrade approach to the

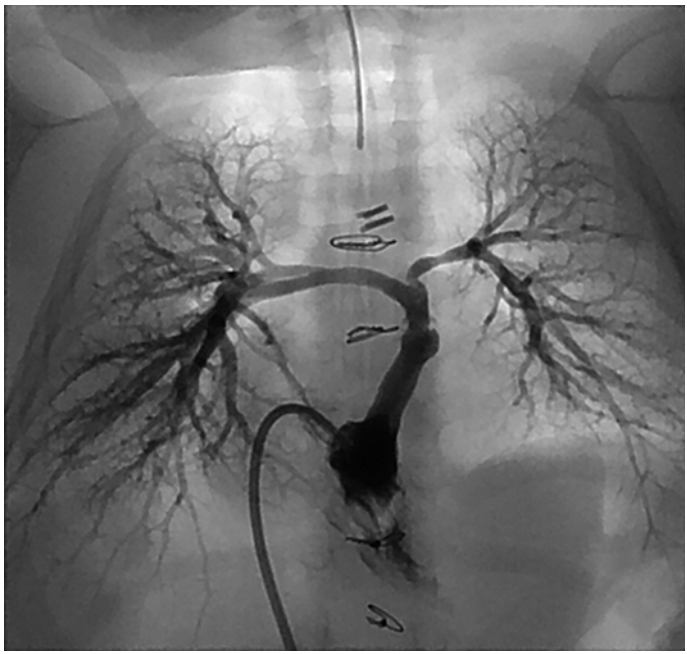

Fig. 6.4 Right ventriculography (antegrade approach) with Berman angiography balloon catheter (Arrow International Inc., Reading, PA) – cranially tilted frontal projection. Patient with hypoplastic right heart syndrome (weight 4 kg) after central shunt due to pulmonary artery hypoplasia

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. Antegrade balloon occlusion aortography with 35° caudal angulation is used to visualize coronary arteries in cases of transposition of the great arteries (Fig. 6.6). Balloon occlusion

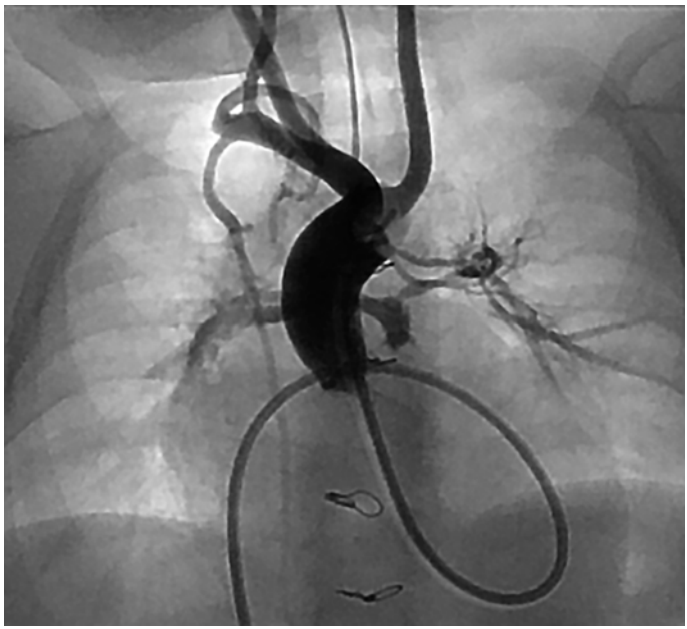

Fig. 6.5 Aortography (antegrade approach) with Berman angiography balloon catheter (Arrow International Inc., Reading, PA) – cranially tilted frontal projection. The patient presented in Fig. 6.4

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 (Fig. 6.7). 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.

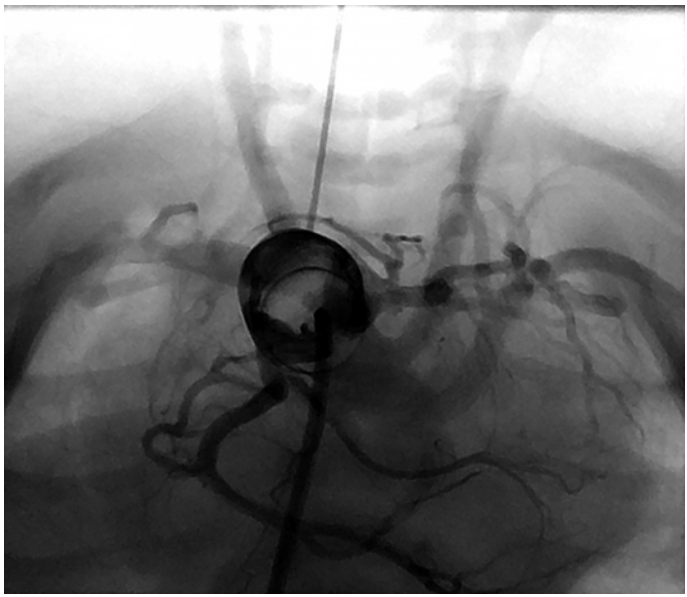

Fig. 6.6 Balloon occlusion ascending aortography (antegrade approach) with Berman angiography balloon catheter (Arrow International Inc., Reading, PA) – caudally tilted frontal projection. Anatomy of the coronary arteries (origin of the left circumflex artery from the right coronary artery) in a 2-day-old patient with transposition of the great arteries

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. 6.3b). 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

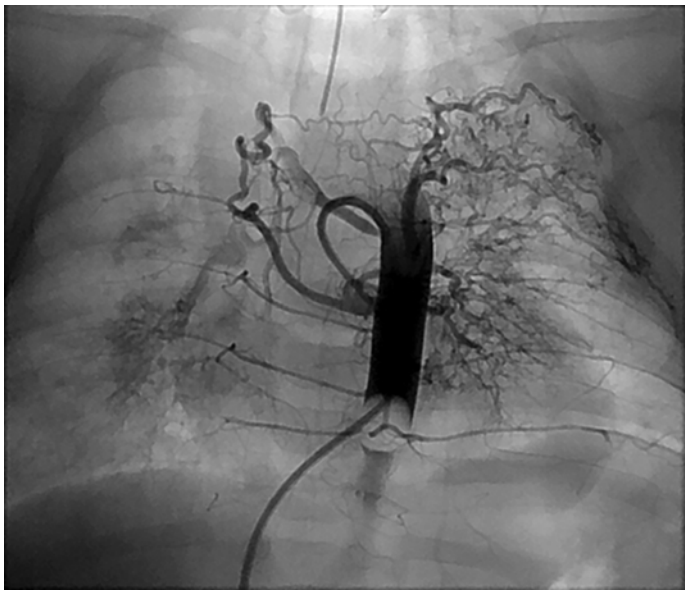

Fig. 6.7 Balloon occlusion descending aortography (antegrade approach) with Berman angiography catheter (Arrow International Inc., Reading, PA) – posterior-anterior projection. Aortopulmonary collaterals in a 6-month-old patient with double inlet left ventricle after pulmonary artery banding

vein, one can measure the pressure in the pulmonary arterial bed, based on exactly 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: balloon valvuloplasty, pulmonary arteries 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 flushing 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 used for selective angiography, pressure measurement, and guidewire placement.

Selected curved catheters are presented in Fig. 6.1. 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 (Figs. 6.1b–e, 6.8, 6.9, 6.10, and 6.11). 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

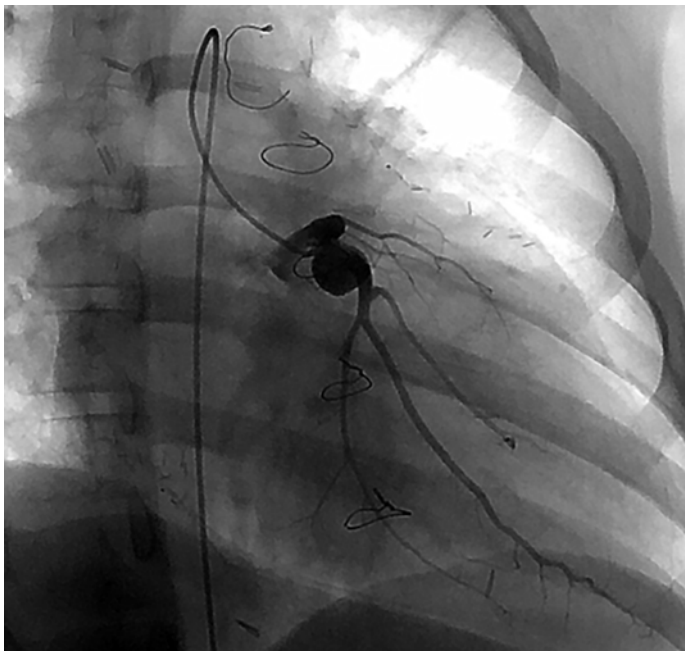

Fig. 6.8 Left coronarography (right anterior oblique projection) with Judkins left coronary catheter (Cordis, Miami Lakes, FL). Details of left coronary artery anatomy (aneurysms, interruption, stenosis) in a 10-year-old patient with Kawasaki disease after bilateral internal mammary arteries/coronary arteries bypass.

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 (Fig. 6.10). Furthermore, it appears to be the best catheter to enter the right ventricular outflow tract, in cases of extreme pulmonary valve stenosis and pulmonary atresia (Fig. 6.11). In crossing the restrictive interatrial communication in the hypoplastic left heart syndrome, it is worth

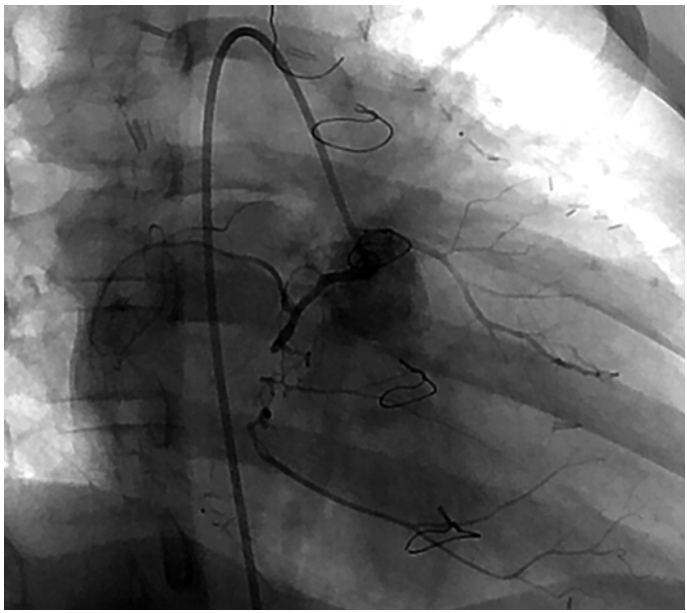

Fig. 6.9 Right coronarography (right anterior oblique projection) with Judkins right coronary catheter (Cordis, Miami Lakes, FL). Details of right coronary artery anatomy (interruption, stenosis) in the patient presented in Fig. 6.8

considering the use of JR catheter inserted over the guidewire to the superior vena cava and gently withdrawing it with the tip directed toward the interatrial septum. Prior to aortic valvuloplasty for neonatal critical aortic stenosis, the JR catheter is the best one for searching the orifice between the stenotic valvar leaflets (Fig. 6.12). JR catheter is also very useful in crossing the interventricular septal defects which is the important step in their interventional closure.

- *Internal mammary catheters* with their C-shaped tips can be used to enter vessels having origins at acute angles. Their

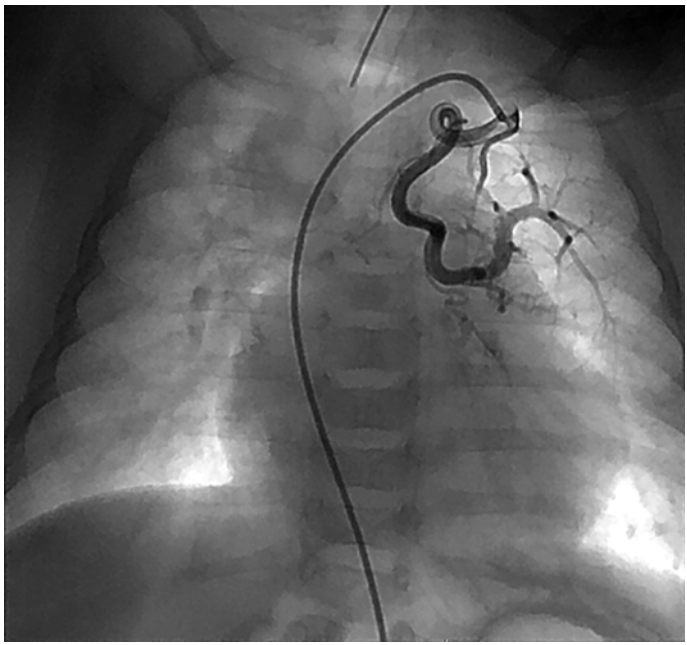

Fig. 6.10 Selective angiography of aortopulmonary collateral artery arising from the left subclavian artery with Judkins right coronary catheter (Cordis, Miami Lakes, FL) – posterior-anterior projection

applications include selective catheterization of Blalock-Taussig shunts and collateral vessels (Fig. 6.13).

- *Multipurpose catheters* have their distal ends curved at an obtuse angle (Fig. 6.1g). 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

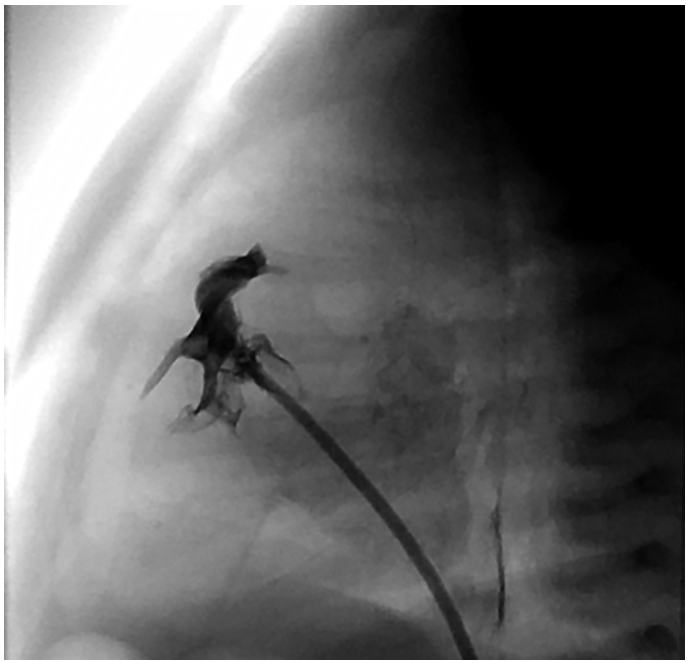

Fig. 6.11 Right ventricle outflow tract angiography (lateral projection) with Judkins right coronary catheter (Cordis, Miami Lakes, FL). Right ventricle outflow tract anatomy and catheter position before radiofrequency valvotomy for pulmonary atresia in a 2-day-old patient

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

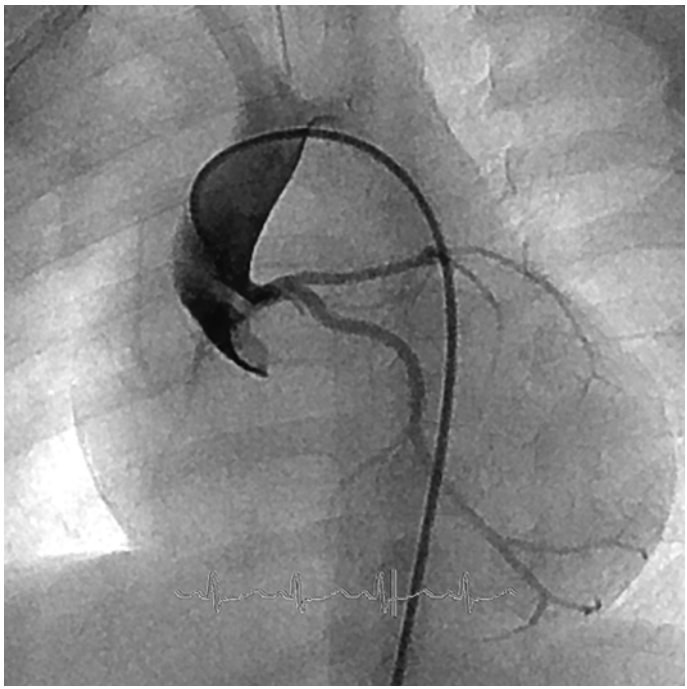

Fig. 6.12 Ascending aortography (retrograde approach) with Judkins right coronary catheter (Cordis, Miami Lakes, FL) – left anterior oblique projection. Catheter positioned in front of the opening of critically stenosed aortic valve in a 3-day-old patient allowing for easy insertion of the guidewire to the left ventricle and balloon valvuloplasty

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-size vessels. Once

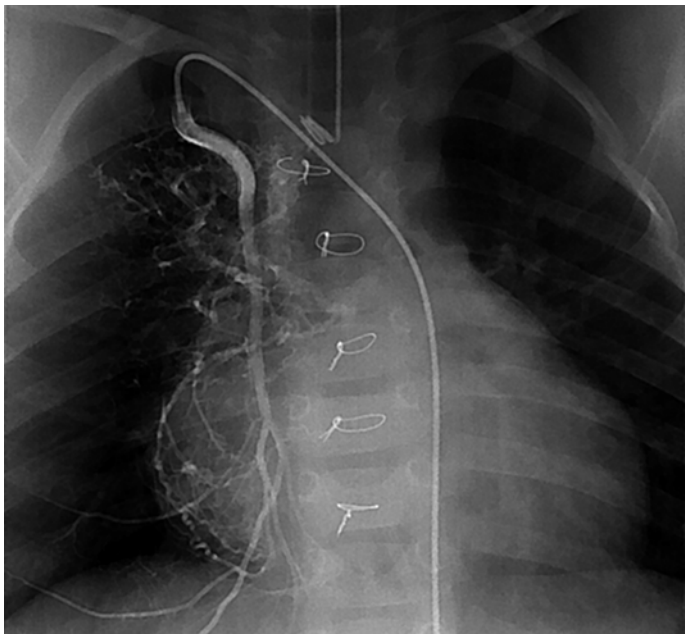

Fig. 6.13 Selective angiography of the right internal thoracic artery (posterior-anterior projection) with right internal mammary catheter (Cordis, Miami Lakes, FL). Small systemic-to-pulmonary collaterals in a 2-year-old patient with hypoplastic left heart syndrome after Norwood-Sano operation

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, that 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, catheterization for idiopathic pulmonary hypertension angiography is not a standard component, and the pulmonary wedge catheter is an obvious choice. Should right ventriculography or pulmonary angiography be planned, the Berman floating angiography catheter and multipurpose catheter are reasonable choices. 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, bi-plane 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.

6.2 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 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 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.

6.2.1 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, pre-shaped 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.

6.3 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 the vascular or cardiac wall. The soft tip can adapt to a vessel shape and 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, what 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 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. 6.14). 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.
3. Hydrophilic coating: it helps to cross tortuous vessels and narrow stenoses. In spite of the 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

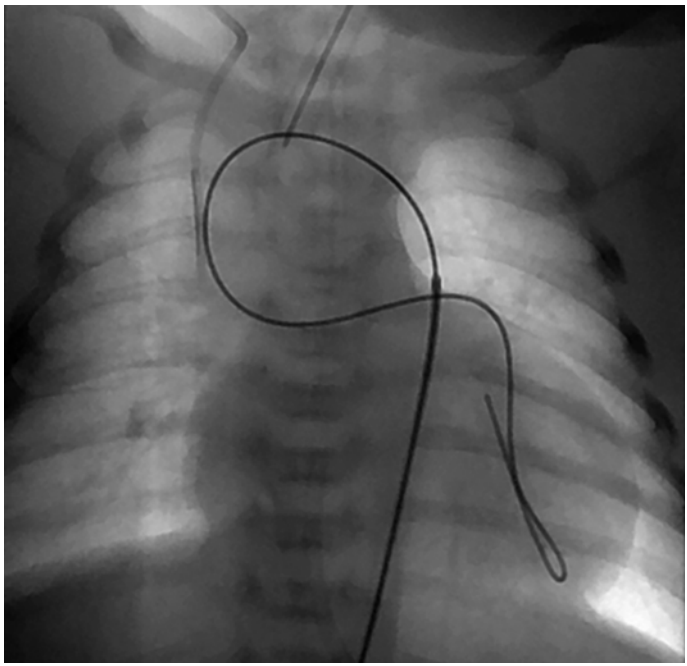

Fig. 6.14 Guidewire (Cook Inc., Bloomington, IL) position before stent placement to critical left pulmonary artery stenosis in a 3-week-old patient with complex congenital heart defect and duct-dependent pulmonary circulation after modified right Blalock-Taussig shunt. Guidewire introduced retrogradely 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

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.

6.4 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, 4 French introducer sheath can accommodate 4 French catheter. 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.

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. Most of them can be recurved using hot steam or air to meet the needs of particular procedures. Mullins sheath is a long, curved sheath with multiple diagnostic and interventional applications. High flexibility and ability to pass through especially tortuous vessel without kinking or collapsing are the features of long Super Arrow-Flex sheath. Details of long sheaths usage are presented in chapters devoted to specific procedures.

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 manipulations impossible. The valves with a screw-tightened hub (Tuohy type) can be regulated according to the needs of the operator. Detachable back-bleed valves with side ports can also be removed once the device is in the sheath and obturates its lumen, so that the control over the equipment is easier and more effective.

Suggested Reading

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Chapter 7
Balloons

Caroline Ovaert

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

C. Ovaert
Pediatric Cardiology, Timone enfants – Assistance Publique Hopitaux de Marseille (AP-HM), 264 Rue St Pierre, Marseille 13385, France
e-mail: caroline.ovaert@gmail.com

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DOI 10.1007/978-88-470-5681-7_7,

lesions. When performing 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 manoeuvre) 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 haemodynamic 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. 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) [1]. This also means that, in large balloons, lower pressure is needed when compared to small balloons, to generate identical wall tension and, as a correlate, clinical efficacy. Four categories of balloons, according to pressure characteristics, are currently available in paediatric and congenital heart catheterization: low-pressure, medium-pressure, high-pressure and ultra-high-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. In compliant balloons, the diameter may increase, especially in the areas of the balloon facing less resistance from the surrounding structures. 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. 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 paediatric 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 haemodynamic 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 paediatric 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 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. Figure 7.1 shows four different balloons used in different locations and clinical indications.

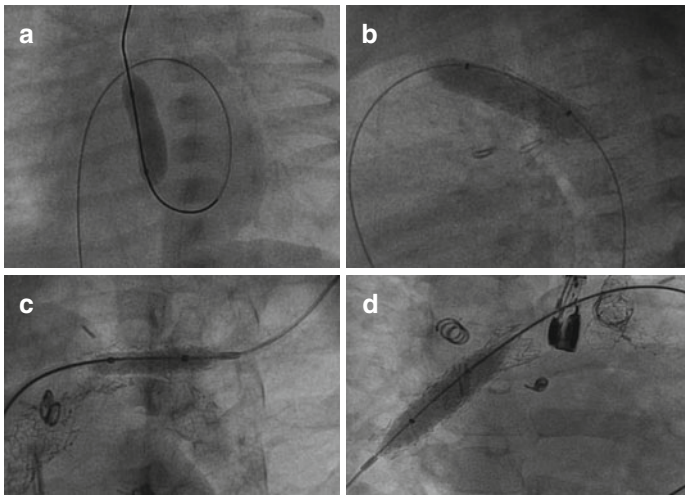


Fig. 7.1 Fluoroscopic views of four different dilation balloon catheters. (**a**) Tyshak II® (NuMED) 6 mm for dilation of neonatal critical aortic stenosis. (**b**) Z-MED™ (NuMED) 6 mm balloon for re-dilation of a stent in ductal position. (**c**) Ultra-thin™ SDS (Boston Scientific) 6 mm for dilation of a side branch, through a stent cell. (**d**) Conquest® (Bard) 5 mm balloon for dilation of in-stent stenosis. Note the large shoulders of the Conquest® balloon

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

Balloons can be divided in categories according to the maximal pressure they can sustain: low-pressure, medium-pressure, high-pressure and ultra-high-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 balloons.

Table 7.1 Non-exhaustive list of some of the frequently used balloons in Europe, classified according to pressure characteristics

	Name	Company	Diameter (mm)	Profile (Fr)	NP (atm)	RBP (atm)	Wire
Low-pressure balloons	Tyshak Mini®	NuMED	4–10	3–4	3–4.5	3.5–6	.014″
	Tyshak II®	NuMED	4–12	4–6	3–4.5	3.5–6	.021–.035″
Medium- and high-pressure balloons	Opta™ Pro	Cordis	3–10	5–8		10	.035″
	Opta™ Pro	Cordis	12	7		6	.035″
	Ultra-thin™ SDS	Boston Scientific	4–10	5–7		12	.035″
	Z-MED II™	NuMED	4–10	5–7	6	13–15	.025–.035″
	Z-MED II-X™	NuMED	8–30	7–16	2–6	3–15	.035″
	Powerflex™	Cordis	4–12	5–8		8–15	.035″
	Advance® 35LP	Cook	3–12	5–7	5–10	8–15	.035″
	Mullins-X™	NuMED	12–25	9–16		9–14	.035″
	Cristal Balloon	Balt	2–40	5–10	3–10 bar		.014–038″
Ultra-high-pressure balloons	Conquest®	Bard	5–12	6–8	8	20–30	.035″
	Atlas® Gold	Bard	12–26	7–12	4–6	12–18	.035″

The name of the company, the diameter range (in millimetres), the profile in French (Fr), the nominal pressure (NP) and rated burst pressure (RBP) in atmospheres (atm) and the maximum wire diameter in inches are mentioned

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 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 *ultra-high-pressure balloons* are more recent in the paediatric cardiology field. They are completely noncompliant balloons, made of ultra high molecular weight polyethylene (UHMWPE). This very resistant fabric supports very high pres-

sures, 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 ultra-high-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. Other bladed balloons are on the market but their experience in paediatric or congenital interventions is scarce. The rationale is to create 4 controlled tears in a thickened intima and media, without overdilating the vessel, avoiding by there a noncontrolled deeper tear which puts the vessel at risk for aneurismal dilation or rupture (Fig. 7.2). 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 [2, 3].

The Boston Scientific Cutting Balloon® comes in diameters between 2 and 8 mm and has a burst pressure of 8 atm. The 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 indeflator, in order to allow proper opening and refolding of the blades. Balloon dilation with

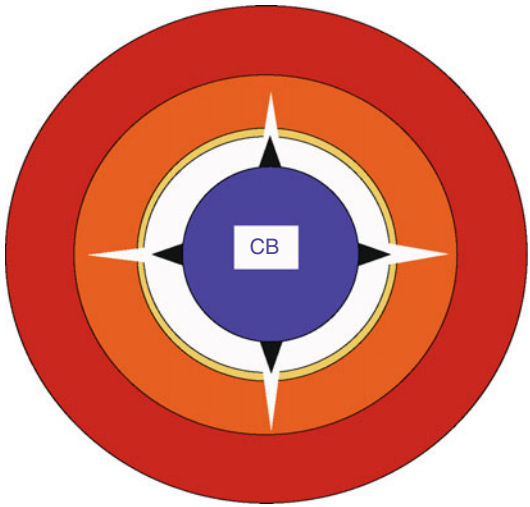


Fig. 7.2 Schematic representation of how the cutting balloon (*CB*) works. The 4 blades (in *black*) make controlled tears in the internal layers of the vessel

a cutting balloon may be followed by angioplasty with a standard compliant balloon or by stent insertion.

7.2.4 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. A small balloon (inner balloon) is inside a larger balloon (outer balloon). 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

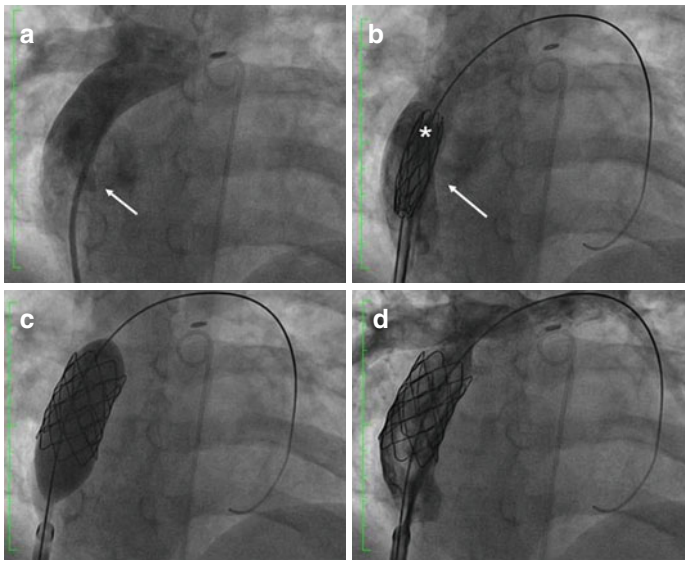


Fig. 7.3 Covered stent insertion using a BIB® balloon (NuMED) for closure of fenestration in an extracardiac fenestrated conduit. (**a**) The white arrow shows the contrast through the fenestration. (**b**) The inner balloon of the BIB® balloon catheter (*white star*) is inflated. The stent is in the appropriate position. (**c**) The outer balloon of the BIB® balloon catheter is inflated. (**d**) Perfect location of the stent with complete closure of the fenestration

inflation which is very helpful for stent placement in 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. 7.3).

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

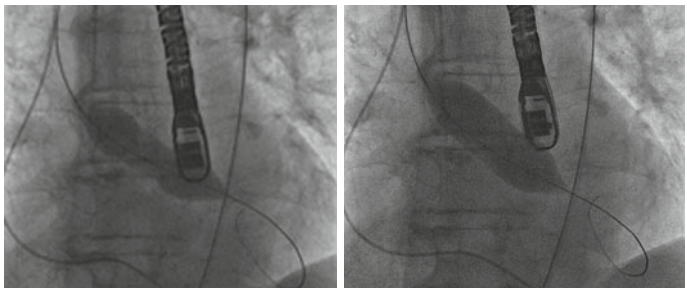


Fig. 7.4 Nucleus™ balloon (NuMED) used for aortic valve dilation. *Left panel*: the balloon is incompletely inflated and has the dumbbell morphology; the narrowest part is at the level of the aortic annulus. *Right panel*: complete inflation with loss of the dumbbell shape

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. 7.4).

7.3 Septostomy Balloons

Septostomy balloon catheters are specifically designed to cross the foramen ovale and to perform the Rashkind atrial septostomy manoeuvre. The balloons are round and noncompliant. The currently most used balloon catheters are the Edwards 5 F atrioseptostomy catheter (Edwards Lifesciences Corporation, CA, USA) and the NuMED Z-5™ atrioseptostomy catheter (NuMED Inc., NY, USA). The Edwards 5 F balloon catheter has a single lumen for inflation of the balloon. The catheter is rather stiff and has a curve at the end to facilitate crossing of the foramen ovale. The balloon takes larger volumes of contrast (4 cc) than the other atrioseptostomy balloons which is useful

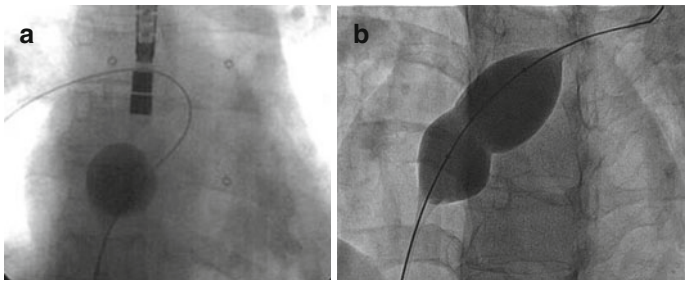


Fig. 7.5 Sizing balloons for atrial septal defect closure. (**a**) The pull-through technique. The balloon is circular and is pulled through the defect. (**b**) Static occlusion of the atrial defect with a soft compliant balloon. The waist on the balloon is obvious (stretched diameter)

when a large atrial septal defect is needed. The Z-5 atrioseptostomy balloon catheter is a 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 2 cc balloon, 5 F catheter), the smallest being particularly useful in small preterm babies.

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 (Fig. 7.5). The first method is the 'pull-through' technique. 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 defect will be considered as the stretched 'atrial

septal defect diameter'. The other method is the 'static' method, 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 manoeuvre 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 trough 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 manoeuvres 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 haemodynamic 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 manoeuvre 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 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 reinflations and deflations, to extract the balloon catheter. Withdrawal into the sheath is sometimes difficult. In those cases, gentle reinflation 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. Using 2 balloons may also be useful in smaller patients, to reduce the sheath size. Insertion of 2 smaller balloons in 2 separate veins will require smaller sheaths than 1 large balloon through 1 access vein. Additional benefit may be the haemodynamic 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 haemodynamic 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 [2].

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. 7.6). 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. 7.6). 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

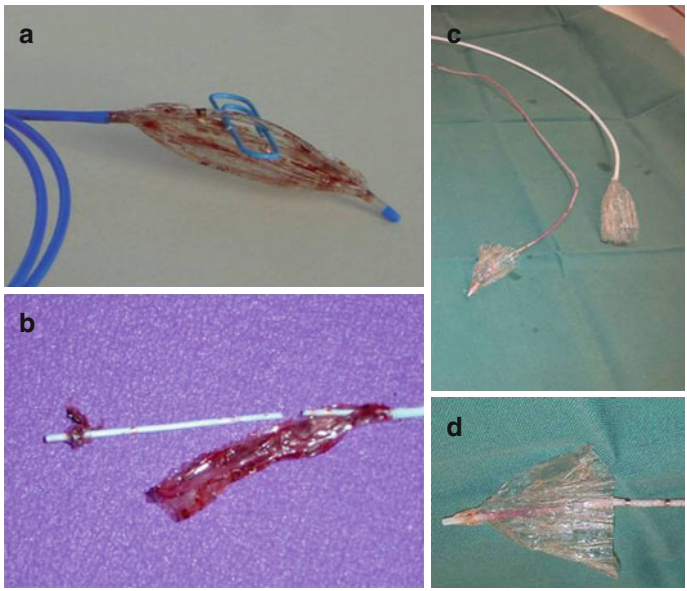
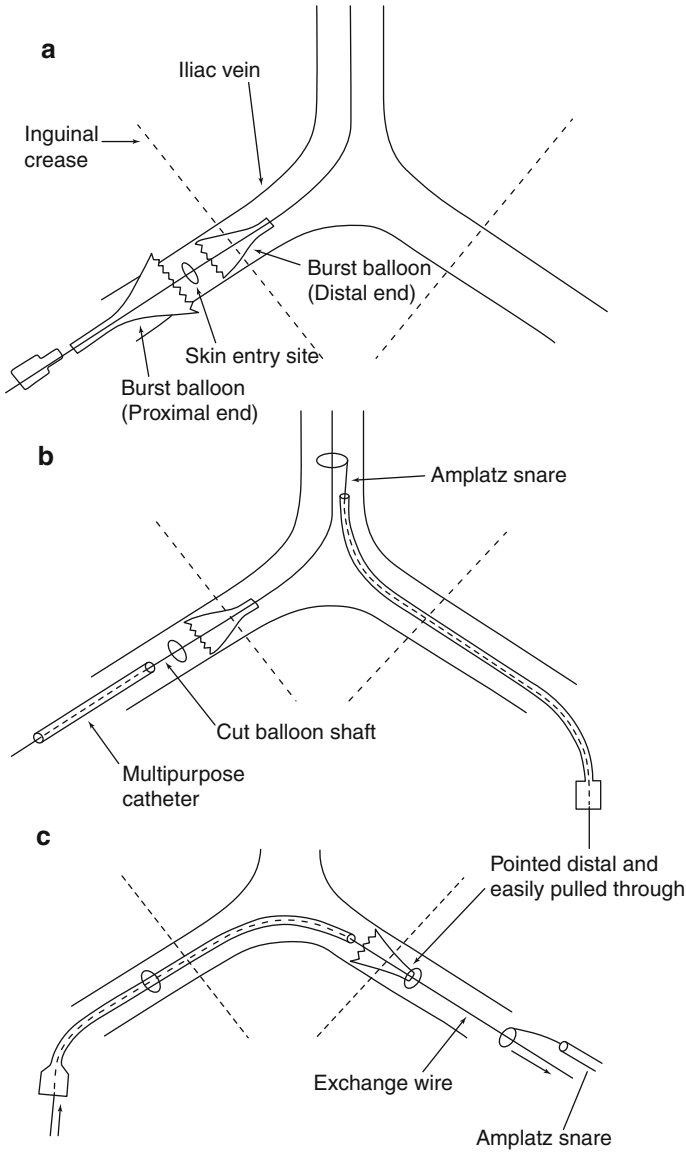


Fig. 7.6 Images of ruptured balloons. (**a**) Longitudinal tear. The *blue paper clip* shows the balloon shaft exposed by the longitudinal tear (Courtesy Dr Joseph De Giovanni, Birmingham, UK). (**b**) Circumferential tear, close to the distal end of the balloon. The shaft has been cut to retrieve the distal part (Courtesy Dr Joseph De Giovanni, Birmingham, UK). (**c**, **d**) Circumferential rupture of a balloon during a percutaneous pulmonary valve implantation. The rupture of the balloon is associated with a shaft rupture. The distal part remained on the internal shaft (**d**), the proximal part on the external shaft (Courtesy Prof R. Berger, Groningen, Netherlands)

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 7.7 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


a
Iliac vein
Inguinal crease
Burst balloon (Distal end)
Skin entry site
Burst balloon (Proximal end)
b
Amplatz snare
Cut balloon shaft
Multipurpose catheter
c
Pointed distal and easily pulled through
Exchange wire
Amplatz snare

obstructing the communication between the balloon and the balloon catheter lumen. In these cases, when haemodynamic 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. 7.7 Figures showing the steps for retrieval of a transversely ruptured balloon. (**a**) Wire position has to be conserved. (**b**) The wire is snared distally, using the contralateral 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 towards the contralateral vein. (**c**) The distal end of the balloon will easily be pulled through the contralateral sheath (Courtesy Dr Jo De Giovanni, Birmingham, UK)

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Chapter 8
Stents

Eric Rosenthal and Sebastian Goreczny

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.

Stent implantation prevented the immediate elastic recoil, allowed the vessel to be dilated only to the required diameter

E. Rosenthal (✉) • S. Goreczny
Paediatric Cardiology, Evelina London Children's Hospital,
St Thomas' Hospital, Lambeth Palace Road, London SE1 7EH, UK
e-mail: eric.rosenthal@gstt.nhs.uk

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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 co-operation between surgeons and interventionalists. Covered stents can seal aneurysms and fistulae 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 picking 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) Post surgical
   (b) Native
2. Pulmonary vein stenosis
   (a) Post surgical repair of TAPVD
   (b) Native
3. Aorta and branches
   (a) Native coarctation
   (b) Recoarctation
      (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) Post surgical (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

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

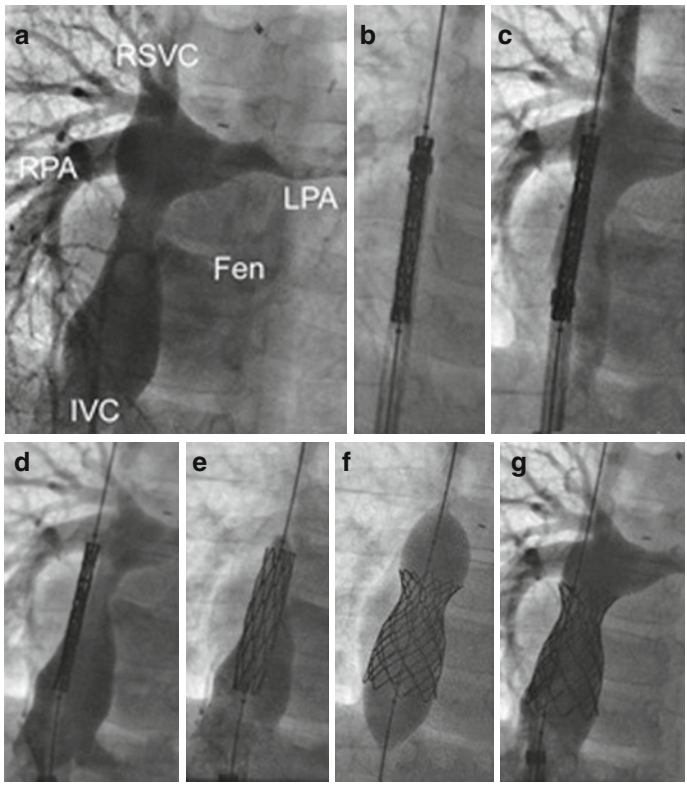

Fig. 8.1 Covered stent (Premounted Cheatham Platinum on a balloon-in-balloon) implantation into a lateral tunnel of an 8-year-old boy with exercise-induced cyanosis and protein-losing enteropathy. Angiogram in (**a**) shows a tunnel stenosis before the branch pulmonary arteries and a patent fenestration. 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	N	O	7–9	12–20	4	0.018	Self-expandable	Neonate and infant lesions
Zilver 518	N	C	6–10	20–80	5–7	0.018	Self-expandable	Neonate and infant lesions
Palmaz Blue	Cc	C	4–7 (12)	12–24	4–5 5	0.014 0.018	Aviator plus Slalom	Neonate and infant lesions
Genesis medium	Ss	C	4–8 (12)	12–24	5–6 6	0.018 0.035	Slalom Opta Pro Unmounted	Neonate and infant lesions
Formula 535	Ss	O	5–8 (20)	12–30	5–6	0.035	High-pressure balloon catheter	PAs, atrial septum, RVOT
Genesis large	Ss	C	5–10 (12)	19–79	5–6 6–7	0.018 0.035	Slalom Opta Pro Unmounted	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
Visi-Pro	Ss	O	5–10 (14)	12–57	6–7	0.035	High-pressure balloon catheter	PAs, atrial septum, RVOT
Valeo Lifestent	Ss	O	6–10 (20)	18–56	5–6	0.035	High-pressure balloon catheter	PAs, atrial septum, RVOT
Genesis XD	Ss	C	10–12 (18)	19–59	8	Depending on the balloon catheter	Unmounted	Pulmonary arteries
Double Strut LD	Ss	O	9–12 (18)	16–76	8	Depending on the balloon catheter	Unmounted	Pulmonary arteries
Mega LD	Ss	O	9–12 (18)	16–36	9	Depending on the balloon catheter	Unmounted	Pulmonary arteries
Maxi LD	Ss	O	12 (26)	16–36	11	Depending on the balloon catheter	Unmounted	Coarctation pulmonary arteries, veins

Advanta V12	Ss	O	12–16 (22)	29–61	9–11	0.035	High-pressure balloon catheter	Coarctation pulmonary arteries, veins s
Covered Cheatham Platinum	Pi	C	12–24 (26) (Up to 30 mm for 10 Zig)	16–45	12–14	0.035	Unmounted or premounted on BIB catheter	Coarctation pulmonary arteries, veins
Cheatham Platinum 8 Zig	Pi	C	12–24 (26) (Up to 30 mm for 10 Zig)	16–45	10–12	0.035	Unmounted or premounted on BIB catheter	Coarctation pulmonary arteries, veins
Andrastent XL	Cc	H	14–25	13–57	8–9	Depending on the balloon catheter	Unmounted	Coarctation pulmonary arteries, veins
Andrastent XXL	Cc	H	20–32	17–57	10–11	Depending on the balloon catheter	Unmounted	Coarctation pulmonary arteries, veins

Potential diameter from reported experience – not confirmed by the manufacturer
C closed, *O* open, *N* nitinol, *H* hybrid, *Ss* stainless steel, *Cc* chromium cobalt, *Pi* Platinum-iridium

  - 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!)
  - 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

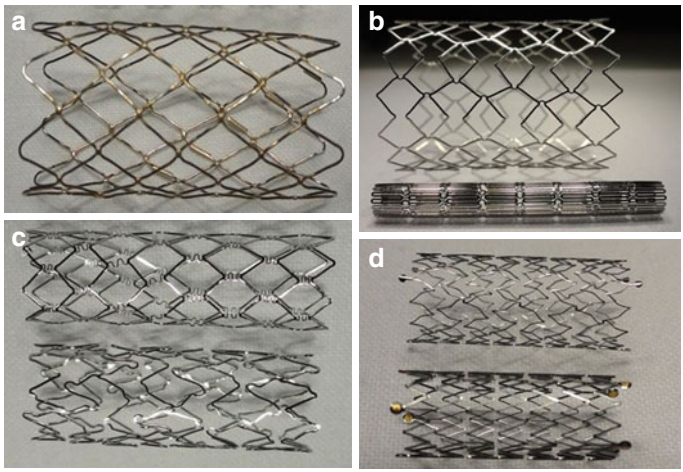

Fig. 8.2 Large balloon-expandable stent in (**a**) expanded closed-cell (Cheatham Platinum) stent and (**b**) expanded and unexpanded hybrid cell (Andramed) stent. Medium-sized balloon-expandable stents in (**c**) closed cell (Palmaz) above and open cell (Valeo) below. Medium-sized self-expanding stents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

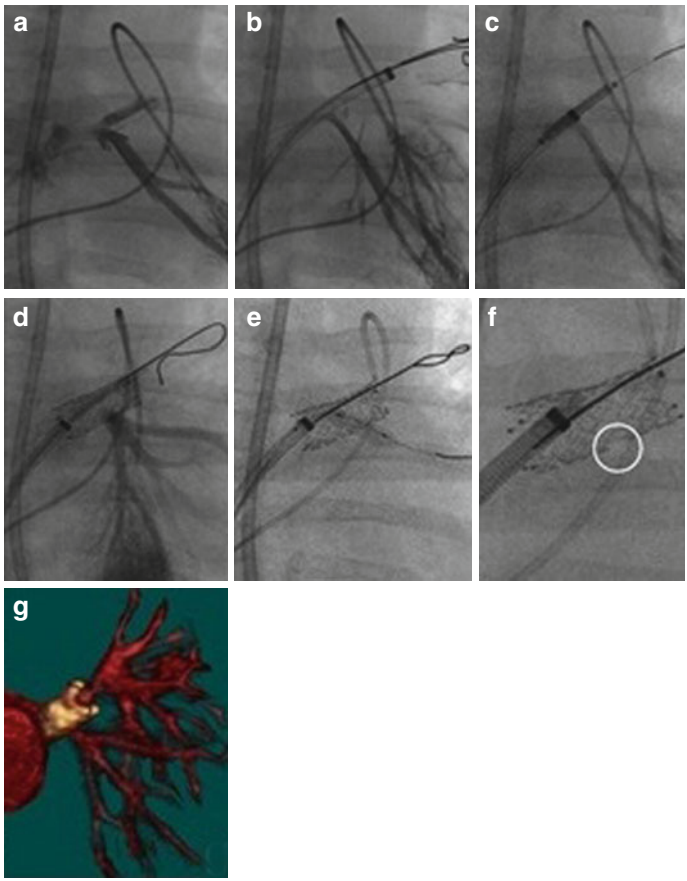

Fig. 8.3 Premounted Visi-Pro stent implantation into a native common left pulmonary vein stenosis after two previous cutting balloon dilations in a 15-month-old boy. Tight stenosis in (**a**) shown by pulmonary artery wedge injection. Long sheath advanced over two guide wires into upper lobe branch and 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**)

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 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.
- 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 for high-risk 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 fact, 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.
- 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 (Fig. 8.4) 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

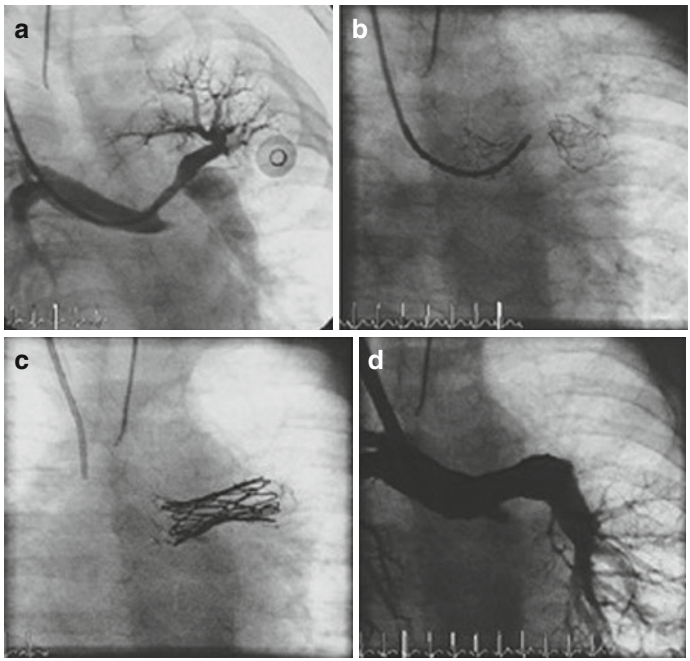

Fig. 8.4 Left pulmonary artery stenosis in a 5-year-old accessed via an internal jugular vein approach through a Glenn shunt (**a**). Palmaz Genesis medium stent fractured after approximately 6 months (**b**). A Cheatham Platinum stent is placed across the two ends of the fractured stent with good flow to the left lower lobe but reduced flow to the upper lobe (**c**, **d**)

adjacent structures (coronary arteries during RVOT stenting, left main bronchus after Norwood surgery (Fig. 8.5)).

- 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.*

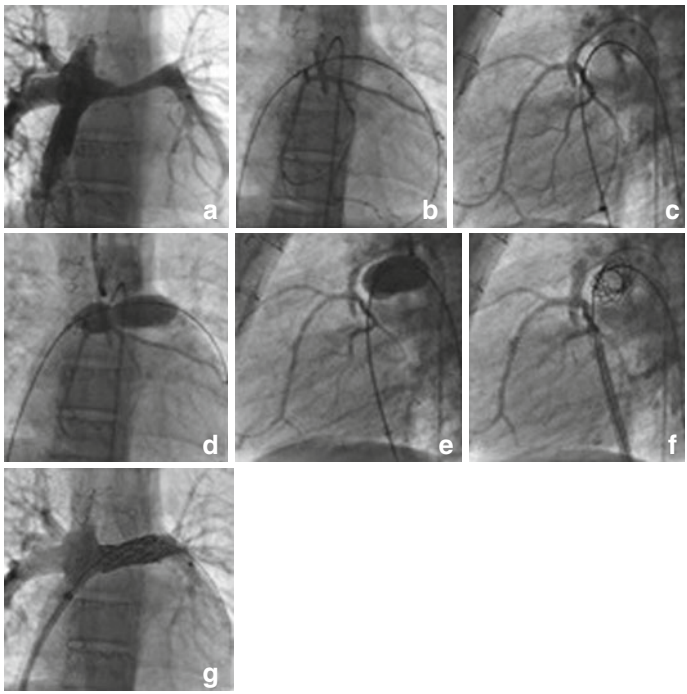

Fig. 8.5 Stent implantation into left pulmonary artery after stage III Norwood procedure in a 17-year-old boy (**a**). MRI scanning raised concerns of proximity of both the left main bronchus and native aorta to the stenosed segment confirmed on angiography in AP and lateral projections with the guide 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**)

- 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 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 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 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.

- 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 haemodynamically 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. 8.4).

- 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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Chapter 9
Hemodynamics: Pressures and Flows

Juan Pablo Sandoval Jones and Lee Benson

9.1 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.

J.P.S. Jones, MD • L. Benson MD, FRCPC, FACC, FSCAI (✉)
Division of Cardiology, The Department of Pediatrics,
The Hospital for Sick Children,
University of Toronto School of Medicine,
555 University Ave, Toronto, ON M5G 1X8, Canada
e-mail: juanpablo.sandovaljones@sickkids.ca; lee.benson@sickkids.ca

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9.2 Pressure Evaluation and Waveforms (Table 9.1)

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 applica-

Table 9.1 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

tion 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.

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 overdamped. 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.

9.3 The Pressures

9.3.1 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.

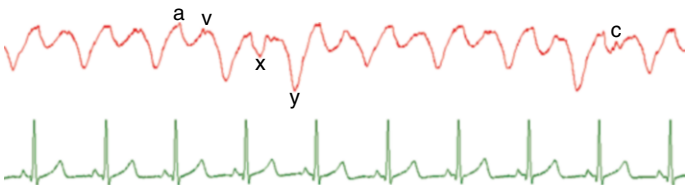

Fig. 9.1 *Upper panel*: venous pressure tracing from the right atrium denoting timing of the various wave components. The *a* wave is generated from the pressure rise following atrial contraction and occurs following the P wave on the ECG. The positive deflection following the nadir of the *x* descent is the *v* wave and is generated by passive venous filling of the atrium while the tricuspid valve is closed. *Lower panel*: the ECG

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. 9.1). The *a* wave is generated from the pressure rise following atrial contraction and occurs following the P wave on the ECG by approximately 80 msec 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 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.

9.3.2 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

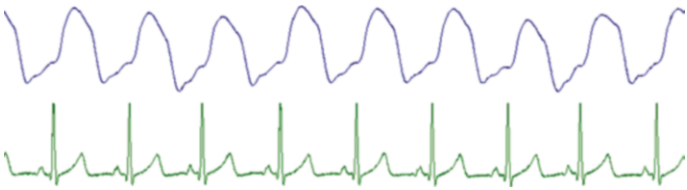

Fig. 9.2 *Upper panel*: typical appearance of the pressure wave generated by the right ventricle. Note the contour contrasted to that of the right ventricle in the presence of valvar stenosis as shown in Fig. 9.3. *Lower panel*: the ECG

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. 9.2). 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.

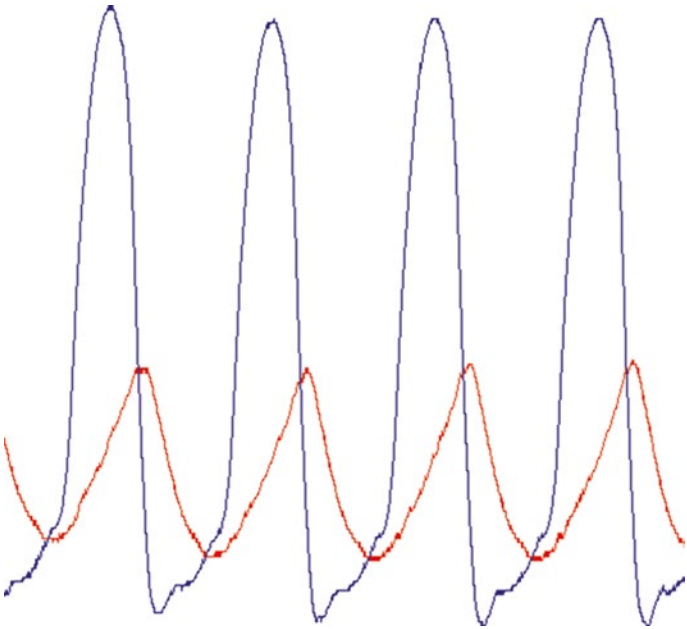

Fig. 9.3 A right ventricular trace in the presence of pulmonary valve stenosis. Note the triangular appearance contrasted to that in Fig. 9.2. Pressure transfer to the pulmonary artery is delayed and lower than that of the right ventricle due to the restricted outlet

9.3.3 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. 9.3). 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).

9.3.4 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 waveform 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.

9.3.5 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.

9.3.6 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. 9.4). 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,

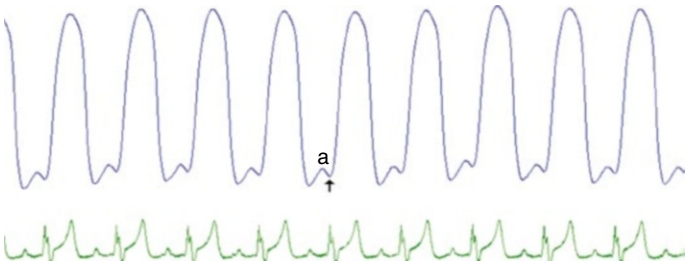

Fig. 9.4 *Upper panel*: typical left ventricular trace, note the trajectory compared to that of the right ventricle in Fig. 9.3. The *arrow* defines the post A wave end-diastolic pressure. *Lower panel*: the ECG

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.

9.3.7 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

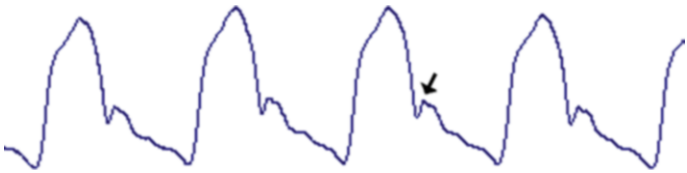

Fig. 9.5 A normal aortic pressure trace obtained from the ascending aorta. The waveform has a rapid upstroke and a clear dicrotic notch due to closure of the aortic valve as pressure decays (*arrow*)

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. 9.5). 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.

9.4 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.

9.4.1 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

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

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:

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

$$C_vO_2\ \left(\text{ml}O_2/\text{l}\right) = \text{Oxygen capacity}\left(\text{ml}O_2/\text{l}\right) \times \text{venous oxygen saturation}\,(\%)$$

For example, if the Hgb was 140 g/l, venous saturation 70 %, the oxygen capacity would be $140\ \text{g/l} \times 1.39 = 194.6\ \text{ml}\ O_2/\text{l}$. The oxygen content would be $.70 \times 194.6 = 136.22\ \text{ml}\ O_2/\text{l}$.

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

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

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

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 [3]. 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 [6]. 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 [2] 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.

9.4.2 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.

9.4.3 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.

9.4.4 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.

9.5 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:

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

Similarly Q_s is estimated as

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

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

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

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 [1]. 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.

9.6 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.

9.7 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

$$\text{Oxygen capacity} = \text{Hgb}(\text{g}/\text{l}) \times 1.39 \times 100 = 139\,\text{ml}\,\text{O}_2/\text{l}$$

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

$$\text{PA} = 139 \times 80\% = 111.2\,\text{ml}\,\text{O}_2/\text{l}$$

$$\text{PV} = 139 \times 95\% = 132.05\,\text{ml}\,\text{O}_2/\text{l}$$

And the pulmonary AV O_2 difference:

$$\text{PV} - \text{PA} = 132.05 - 111.20 = 20.85\,\text{ml}\,\text{O}_2/\text{l}$$

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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Chapter 10
Psychological Aspects in Congenital Heart Disease: A Lifelong Perspective

Edward Callus, Cristina Farè, and Emilia Quadri

10.1 Introduction

In order to explore this topic, the divergence in the expectations regarding the education and counseling that should be provided to parents of children with congenital heart disease (CHD) in both the prenatal and neonatal period by cardiologists [1] will be described. This will be followed by the description of the psychological functioning of patients with congenital heart disease from childhood to adulthood [2, 3].

E. Callus (✉) • C. Farè • E. Quadri
Pediatric and Adult Congenital Heart Disease Centre,
IRCCS Policlinico San Donato,
Piazza Edmondo Malan 1, San Donato Milanese,
Milan 20097, Italy
e-mail: edward.callus@grupposandonato.it;
emilia.quadri@grupposandonato.it

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10.2 The Importance of Information Provided by the Cardiologist in the Prenatal and Neonatal Phase

- In the literature on this topic, it is outlined that there is a discrepancy when it comes to the importance and relevance on which information needs to be given in the prenatal phase and the neonatal period between the cardiologists and the parents of children with CHD, although there was also an agreement on what needs to be known between the two groups.
- It could be possible that an increase in prenatal diagnosis of CHD could be connected to a rise in decisions to terminate pregnancy, and therefore it becomes particularly crucial to know what and how much the parents want to know and how much they want to communicate.
- Interestingly, the factors considered in deciding whether to continue the pregnancy of a known fetus with CHD differed according to the two groups (parents and cardiologists). Whereas both groups considered the quality of life of the child and the survival into adulthood, for the cardiologist group, the deciding factors were also a potential for neurodevelopmental delay and the severity of the CHD. On the other hand, the factors which also lead to deciding against pregnancy termination of the parents were moral/religious beliefs and survival into birth.
- During the prenatal phase, both physicians and the parents felt that parents should be able to name the heart condition and understand it, what is to be expected in the first hours of life, survival probability at birth, childhood and adulthood comorbidities, whether there were possibilities of not intervening medically after birth, and how to obtain additional emotional/psychological support.
- During this phase, the parents ranked the following issues as being of higher importance than the cardiologist: number of

lifetime surgeries needed, potential need for transplant, exercise limitations, and potential for child to have his or her own baby with CHD.

- In the neonatal phase, both groups agreed that the parents should be able to name the heart lesion, identify who their cardiologist and pediatrician are, the discharge medications, comorbidities, when to seek medical attention, and cardiopulmonary resuscitation training.
- On the other hand, the issues which the parents ranked as being more important than the cardiologist though they would be in the neonatal phase were why the child has CHD, why he or she needs surgery, ability to explain the CHD to a doctor, and follow-up needed.

10.3 Psychosocial Adjustment and Quality of Life in Children and Adolescents with Congenital Heart Disease

- In a literature review conducted in 2009, the psychological aspects in children and adolescents with CHD relating to outcomes after open heart surgery were considered [3]. The review took into consideration studies assessing psychological adjustment (assessed by semi-structured interviews, proxy reports administered to parents and teachers, and self-reports) and quality of life.
- When it comes to long-term psychological adjustment as assessed by the parents, it was reported that psychological maladjustment was present in a considerable proportion of these children and that this proportion is comparable with other children with chronic illnesses (who, when compared with the children in the general population, have a twofold higher risk of having a psychiatric diagnosis).

- Parents also report the operated children as displaying behavior which is outside the normative range and as having more psychological difficulties when compared to the controls (except for children with surgically corrected transposition of the great arteries).
- On the contrary, the proxy reports done by teachers reported that psychological functioning and behavior of CHD children were similar to the ones of healthy controls, which could either mean that these difficulties are more difficult to detect in a school setting or that parental anxieties influence the way these parents perceive their children.
- There are fewer self-report studies on the psychological adjustment in children after cardiopulmonary bypass surgery, which do not report differences between children with operated heart defects when compared to reference group, contrasting the proxy report studies on parents. Two studies on samples which included a large percentage of adolescents reported a significant degree of self-reported behavioral problems, suggesting that there might be difficulty in puberty for these patients possibly linked to increasing academic demands.
- Also when it comes to quality of life, impairments tend to be reported more by the parents than the patients themselves.
- Although current data regarding risk factors for psychological functioning and quality of life in children with CHD is insufficient and conflicting, the cardiac diagnosis in itself does not seem to be a risk factor for psychological maladjustment after cardiac surgery.
- One child-specific risk factor which has been consistently found is developmental delay; however, this applies to proxy reports done by the parents and are not confirmed by self-report studies.
- The actual parents of children with operated heart defects might directly impact the children's long-term quality of life and psychological adjustment; however, more research is needed on this aspect.

10.4 Psychological Functioning in Adults with Congenital Heart Disease

- In a current review, the focus was on the psychological functioning of grown-up CHD patients with a special attention to the link between medical and other variables and the psychological functioning.
- In some studies in the review, it was underlined that the CHD population has similar or even better psychological functioning than the healthy reference population – possibly because they have acquired a higher sense of coherence, due to the constant dealing with physical limitations and difficulties. The types of medical care available in different countries could also impact these patients' well-being.
- It could also be that denial and high achievement motivation could have influenced self-report data.
- Levels of anxiety and depression are similar to acquired cardiac populations – which is probably the best comparison available.
- It is important to note however that in a few studies where in-depth clinical psychological and psychiatric interviews were utilized, patients who were assumed to be well adjusted reveal to experience psychological and psychiatric difficulties and to be undertreated.
- Most of the studies available indicated that psychological functioning in this population is not related to medical variables such as diagnosis and physical status. The variables that result as being predictors of psychological distress reported by this population pertain to their subjective experience and result as being:
  - Loneliness
  - Fear of negative evaluation
  - Imposed limits
  - Low capacity for physical exercise
  - Perceived health status

10.5 Tips and Considerations on How to Approach Families and Patients

- These families and patients have to deal with many issues which often begin during the prenatal phase and which have an impact on how the developmental tasks are dealt with. Pediatric cardiologists often find themselves having to communicate difficult news which entails having to deal with strong emotions, and the integration of psychologists in the medical team can provide the necessary support in order to handle particularly difficult situations and to provide psychological support to the families and the patients when required.
- It is also particularly useful to have a connection with nonprofit associations which provide peer to peer support. The parents might be relieved to be able to meet adults with the conditions their children have and also the adolescents and adults can gain great benefits from meeting people with similar conditions and participating in events created by these associations.
- It is indicated that the parents would like to have more information than the cardiologists think that is necessary on a variety of topics. It is possible that there is a tendency of the cardiologists to believe that it is important not to give excessive information which could worry and overwhelm parents. On the other hand, it seems that parents seem to be willing to know more about their situation to prepare psychologically and also to make adequate decisions.
- It is therefore important for the cardiologists to customize the relationship with the parents and the patients, according to the situation the parents are in and the age of the patients, their wishes about the amount of information they would like to receive, and also their psychological and religious necessities.
- When handling the families, it is important to bear in mind that the parents and the patients themselves often have a

completely different approach to the illness. For example, someone born with a chronic illness is somehow used to that condition and for them it is a situation of "normality." For the parents, this is very hard to conceive – and this is supported by the divergence in data when it comes to the psychosocial adjustment and quality of life in children and adolescents with CHD.

- When it comes to adolescents and adults, it is important not to assume that a cardiac condition and severity of disease are automatically correlated to psychological distress, which is more likely to be caused by factors of a psychological nature and a low capacity of physical exercise.
- Finally, it would be ideal for medical and nursing staff to dedicate some time for training in communication skills and also for them to have moments of support in order to prevent burnout symptoms from emerging.

10.6 Conclusions

In conclusion it is essential for cardiologists to spend some time to enquire about how much information parents and also the patients themselves when they are older would like to know and to also consider their psychological needs and belief systems in order to be able to provide an adequate and efficient communication.

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Part II
Vascular Access

Chapter 11
The Usual Vascular Access

Jochen Weil

For cardiac catheterization, the most frequently used access to the heart is via the femoral artery and/or vein.

Other ways of access are possible or necessary, for instance, if the pulmonary arteries can only be reached via the SVC in patients after a Glenn shunt or in patients having an occlusion of the femoral vessels due to previous catheterizations.

The principles of getting access are explained using the femoral vessels.

Thereafter alternative ways of access are mentioned which are the access of:

- Internal jugular vein
- Subclavian vein
- Umbilical vein and artery
- Radial artery

J. Weil
Department of Paediatric Cardiology/Congenital Heart Diseases, University Heart Centre, Martinistr. 52, Hamburg D-20246, Germany
e-mail: j.weil@uke.de

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11.1 Femoral Venous and Arterial Access

11.1.1 Positioning and Landmark

Especially in infants and small children, a small elevation of the bottom, e.g. with a napkin, helps to expose the femoral vessels. The legs should be fixed in a strait and slightly outwards rotated position.

The landmark to be sought is the inguinal ligament which runs between the superior iliac spine and the pubic tubercle (Fig. 11.1).

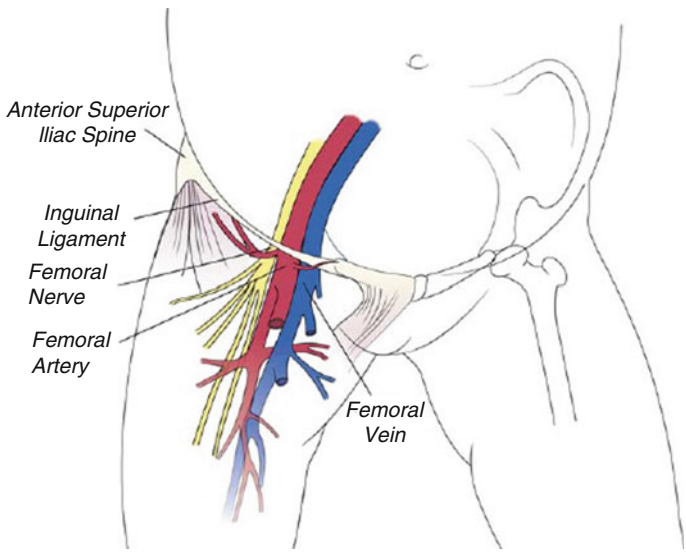

Fig. 11.1 Landmarks of femoral artery and vein access (From Bergersen [1], with permission)

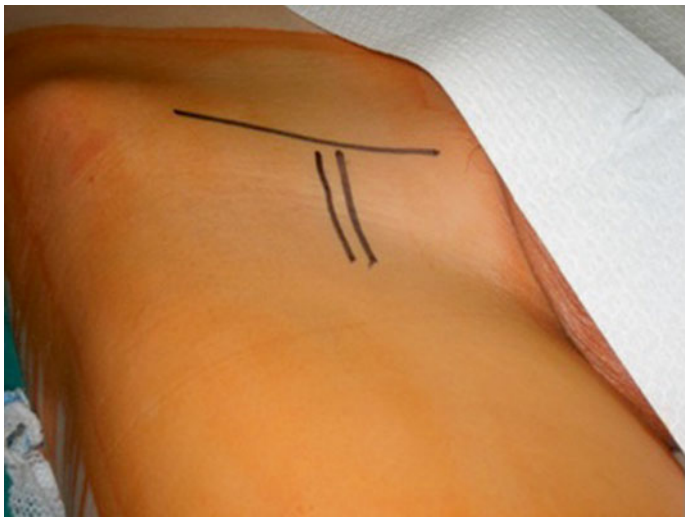

Fig. 11.2 Marking of the landmarks for femoral artery and vein access

The pulse of the femoral artery should be sought. The femoral vein runs closely medial to the femoral artery. A useful landmark for this area is the inguinal crease which is located about 1 cm below the inguinal ligament in normal weight children and adolescents, but can be misleading in overweight patients [1, 2].

The femoral pulses should be palpated in both groins to feel whether there is a difference in the quality of pulses. If a pulse is blunted on one side due, e.g. vessel stenosis, the puncture of the contralateral vessel should be preferred.

Marking of the vessels on both sides with a pen is done by many operators before covering the child. These markings help to identify the vessels when the landmarks are more difficult to see when the patient is draped (Fig. 11.2).

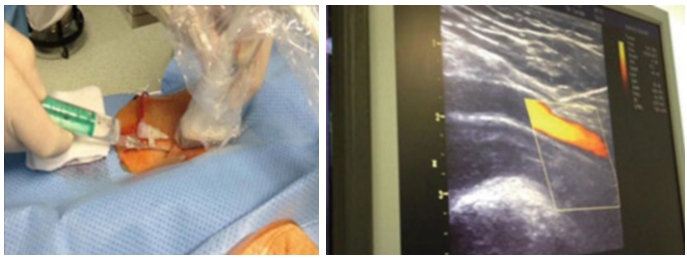

Fig. 11.3 Ultrasound-guided femoral vessel access using a linear probe. The pulsating flow of the femoral artery and the needle directing towards the femoral artery are seen

Delineation of the vessels by 2D ultrasound and Doppler facilitates the puncture of the vessels (Fig. 11.3). By this method, a stenosis or an occlusion of the vessel can be detected. Furthermore, the passage of the needle can be directed under vision into the respective vessel.

11.1.2 Technique

- It is useful always to prepare both sides of the groin.
- For local anaesthesia, infiltrate a small amount subcutaneously. Always withdraw on the hub of the needle to be sure that you are not injecting into the vessel.
- Large amounts of lidocaine will distort the underlying vessel. Be aware that the feeling of pain is mainly coming from the skin!

11.1.3 Femoral Venous Access

- Usually venous access prior to the arterial one.
- Palpate femoral artery. The femoral vein lies directly medial to the artery.

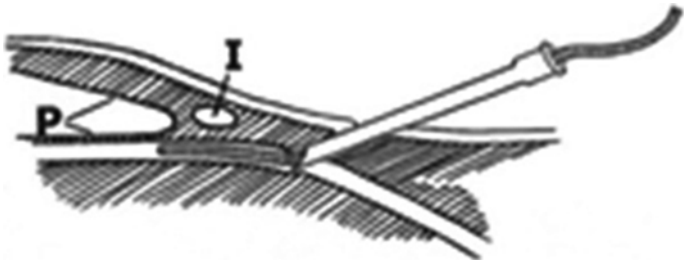

Fig. 11.4 Introduction of guide wire and sheath into femoral vessel below the inguinal ligament (*I*) far away from the peritoneal sac (*P*) (From Mullins [2], with permission)

- Introduce the needle into the skin just below the inguinal crease.
- Angle between needle and skin about 45°.
- Advance the needle in short 1–2 mm jabs and wait for the backflow into the needle.
- If advanced too far, withdraw the needle slowly with or without a negative suction on an attached syringe.
- Having backflow into the needle, hold the needle with the right hand and insert the guide wire into the needle and then into the vessel. The guide wire should get into the vessel smoothly without applying force (Fig. 11.4).
- Having the wire in the vessel, make a tiny cut with a blade and insert the sheath with the introducer under a mild rotation into the vein. Thereafter remove the dilator and wire and check appropriate sheath position by withdrawing blood through the side arm of the sheath.

Hints

- Withdraw guide wire and needle when the wire cannot be advanced easily. Very likely the vessel is not hit properly.
- When the guide wire is very likely in the vessel, but advancement is not possible, change the needle to a small short cannula and try to aspirate blood by withdrawing the cannula and then re-advance the guide wire.

- Consider a careful injection of a small amount of contrast medium if there is suspicion of a more distal occlusion of the vessel by, e.g. thrombosis due to previous catheterization [1].

11.1.4 Femoral Arterial

It is done similar as described for the venous access. When the needle hits the artery, there is a brisk and pulsating backflow. A syringe to aspirate the blood is not needed.

11.1.5 Complication of Femoral Access

- Retroperitoneal bleeding if the femoral vessels are punctured above the inguinal ligament [3]!

11.2 Internal Jugular Vein Access

11.2.1 Positioning and Landmarks

- Extend the head backwards by putting a towel underneath the shoulder.
- Turn the head to the contralateral side.
- Identify the landmark which is the triangle formed by the sternal and the clavicular head of the sternocleidomastoid muscle and the clavicle. The internal jugular vein lies lateral to the carotid artery which pulse can be felt (Fig. 11.5).

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

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

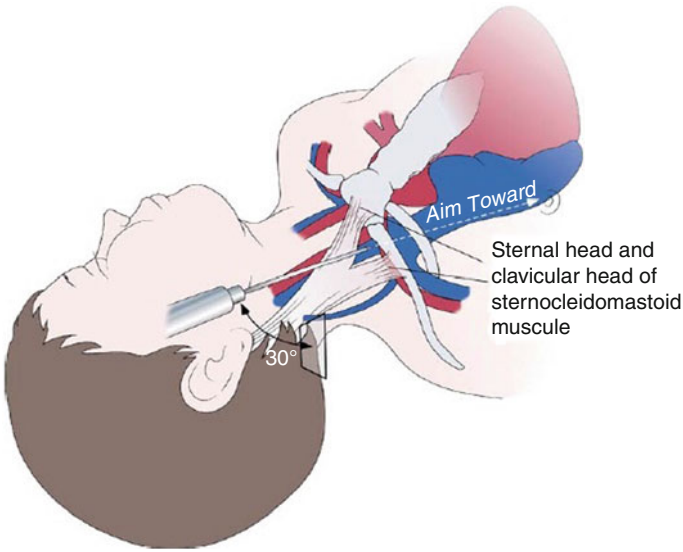

Fig. 11.5 Landmarks of the internal jugular vein access using the anterior approach (From Bergersen [1], with permission)

11.2.2 Ultrasound-Guided Approach

- Clear delineation of carotid artery and internal jugular vein is possible.
- When applying slight external compression the vein, collapses while the artery does not.

11.2.3 Anterior Approach

- The entry site of the needle is lateral to the carotid artery in the middle of the triangle formed by the carotid artery, the mandible and the sternocleidomastoid muscle.
- The degree of the needle to the skin is about 45° aiming to the ipsilateral nipple.

11.2.4 Posterior Approach

- The needle is inserted at the border of the sternocleidomastoid muscle midway between the angle of the mandible and the clavicle.
- The needle has an angle of 30° to the skin and is directed towards the sternal notch.

11.2.5 Central Approach

- Enter the site with the needle at the border of the sternocleidomastoid muscle midway between the angle of the mandible and the clavicle.
- Needle has an angle of 30° to the skin and is directed towards the sternal notch.

11.3 Subclavian Vein Access

11.3.1 Positioning and Landmarks

- Entry is possible on both sides. Some operators prefer the on left side, since the course of the wire and catheter is easier to the heart.
- The arm on the ipsilateral side should be straight downwards along the thorax.
- Small elevation of the shoulders by a towel and head turned to the contralateral side.
- Landmarks are the clavicle and the suprasternal notch.

The subclavian vessels are situated underneath the clavicle. The subclavian artery runs closely cephalad to the subclavian artery (Fig. 11.6).

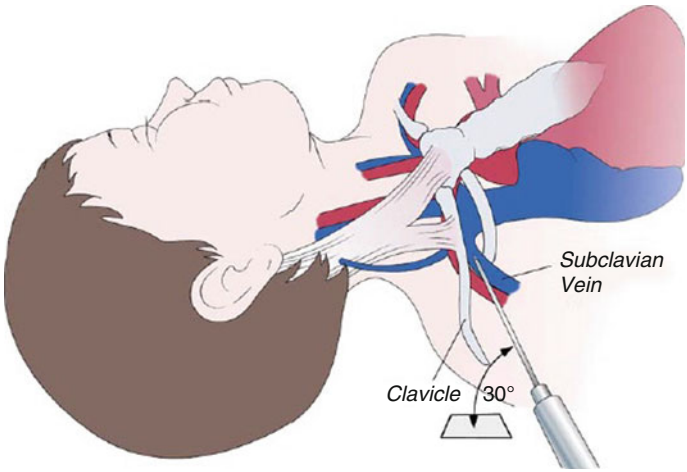

Fig. 11.6 Landmarks of the subclavian vein access (From Bergersen [1], with permission)

11.3.2 Technique

- Needle entry is at the lateral third of the clavicle about 2 cm below the clavicle aiming to the suprasternal notch.
- Advance of the needle under continuous aspiration with an attached syringe.
- When free blood flow is visible, advance a long flexible guide wire to be sure that you are is in the venous structures (SVC, RA) or in PA in the Glenn circulation.

11.3.3 Complications and Avoidance

- Puncture of subclavian artery and subsequent haematothorax. Avoid medial entry of the needle; otherwise the site of the puncture cannot be compressed against the first rib.

- Pneumothorax
  The apex of the lungs is situated under the first rib. The needle should not go too deep, rather aiming with an angle of about 30° towards the suprasternal notch. All these landmarks must be identified before draping the patient [1].

11.4 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″ guide wire.

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 guide wire.

11.4.1 Problems

- Umbilical vein
  The guide wire and catheter are difficult to manoeuvre into the ductus venosus because you are the 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 guide wire 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 guide wire which has a stiff shoulder.

11.5 Radial Artery Access

11.5.1 Positioning and Landmarks

- The arm is abducted 90° and positioned on an arm support.
- Slightly elevate the wrist by a cotton swab and fixate the hand.
- The landmarks are the distal ends of the radial and ulnar bone and the radial pulse.

11.5.2 Technique

- Insert the needle superficially with a 30° angle. No syringe attached. Advance until jerks of pulsating blood will flow.

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Chapter 12
Unusual Access

Stephan Schubert and Felix Berger

12.1 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 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.

S. Schubert (✉) • F. Berger
Department of Congenital Heart Disease/Pediatric Cardiology,
German Heart Center Berlin, Augustenburger Platz 1,
Berlin 13353, Germany
e-mail: sschubert@dhzb.de; berger@dhzb.de

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12.2 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. 12.1.

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

12.3 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 approach after sonographic visualization of the liver structure and especially liver veins [1].

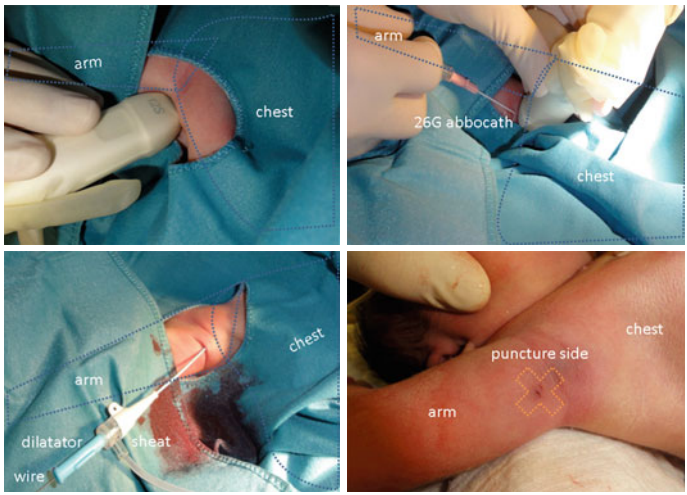

Fig. 12.1 Ultrasound-guided puncture: Right axillary arterial access with ultrasound-guided puncture for ductal stenting of a complex ductus-dependent defect in a 3-day-old infant with a BW of 2.9 kg. A 4.5–11.5 MHz ultrasound probe (GE Healthcare, Wauwatosa, USA) was used to visualize the axillary artery, 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

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. 12.2).

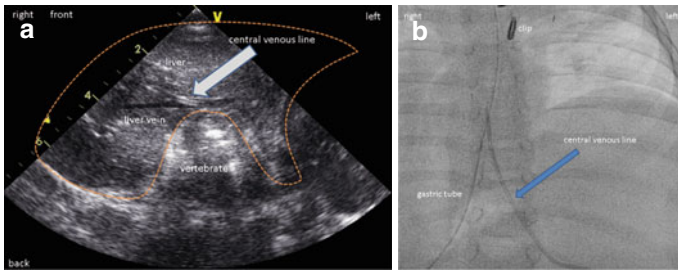

Fig. 12.2 Transhepatic central venous line placement: ultrasound-guided puncture by a left paravertebral and median percutaneous approach due to inverse abdominal situs with dextrocardia in a patient with caval thrombosis. (**a**) Sonographic picture after transhepatic placement of the central venous line. Visualization of liver veins and liver tissue is essential for percutaneous approach. (**b**) Fluoroscopic picture of central venous line with a transhepatic approach

12.4 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. 12.4). 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. 12.3 and 12.4).

12.5 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 control 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. 12.5a–c).

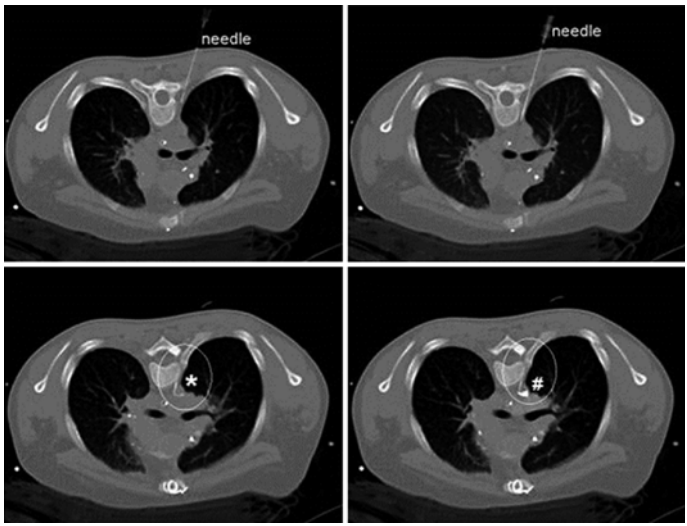

Fig. 12.3 CT scan of the initial paravertebral approach (patient in prone position), with the needle passing through the paravertebral space. (**b**) Puncture of the paravertebral vein with a needle on the right side. (**c**) Introduction of a guide wire (*) into the vertebral vein. (**d**) Insertion 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])

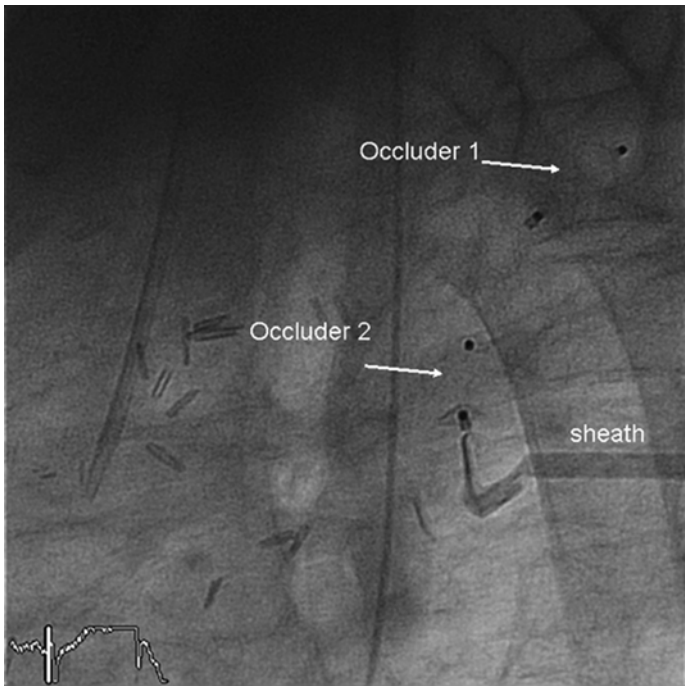

Fig. 12.4 Angiogram in lateral projection without contrast medium: two Amplatzer vascular plugs in situ. One occluder was placed in the inflow to the collateral; one was placed in the collateral itself. Access of the sheath through the vertebral vein is documented via paravertebral access (From Schubert et al. [3])

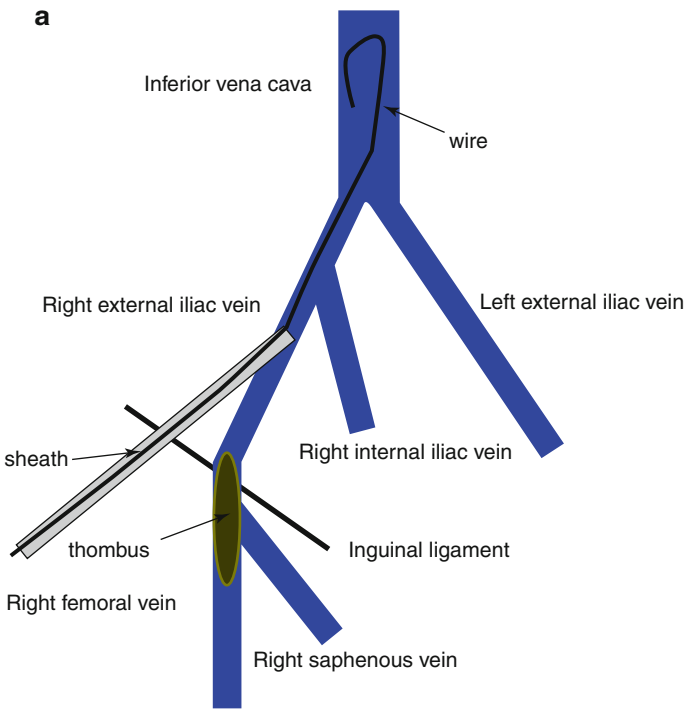

Fig. 12.5 (**a**) Iliac and femoral vessel area around the inguinal ligament. Venous puncture and sheath implantation can be performed via the right iliac vein under sonographic control. (**b**) Example of an angiographic and fluoroscopic picture in a 12-year-old girl with multiple vascular punctures 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

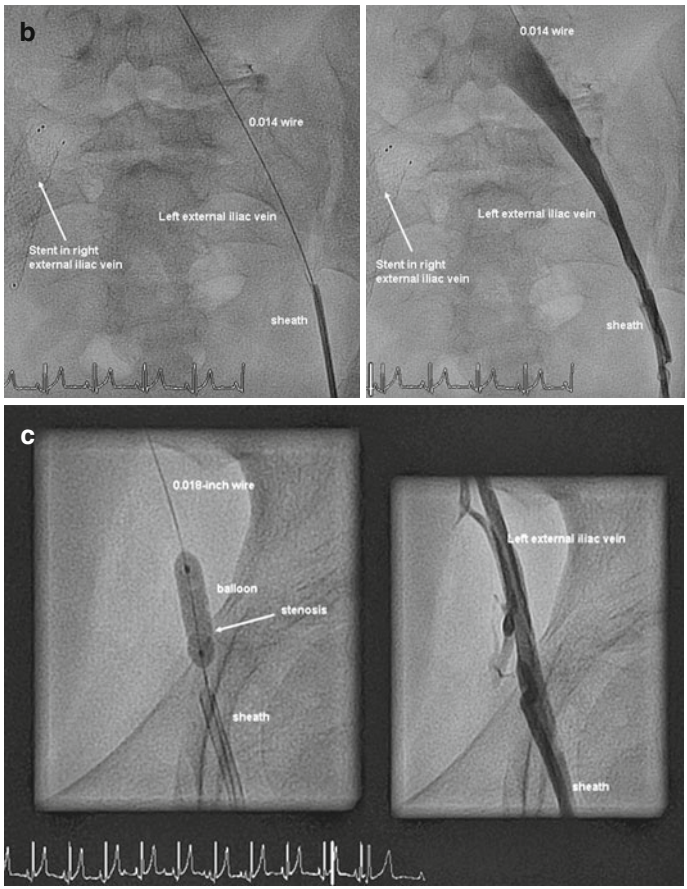

Fig. 12.5 (continued)

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Chapter 13
Hemostasis

Zakhia S. Saliba, Sami G. Slaba, and Elie B. Sawan

13.1 Hemostasis

Vascular hemostasis after catheterization is crucial for a successful procedure. The hemostasis stops external and internal bleeding without compromising vessel patency after sheath withdrawal.

Several factors like anticoagulant medications, the ratio of sheath size to vessel size, several attempts to obtain the vascular access (poorly anesthetized and not well-immobilized patients), multiple cardiac surgery or repeated catheterization using the

Z.S. Saliba (✉)
Department of Pediatric Cardiology, Hôtel-Dieu de France Hospital, Alfred Naccache Blvd, Ashrafieh, Beirut, Lebanon
e-mail: zakhiasaliba67@yahoo.fr

S.G. Slaba
Department of Radiology, Hôtel-Dieu de France Hospital, Alfred Naccache Blvd, Ashrafieh, Beirut, Lebanon
e-mail: sslaba@hotmail.com

E.B. Sawan
Department of Pediatric Cardiac Surgery, Hôtel-Dieu de France Hospital, Alfred Naccache Blvd, Ashrafieh, Beirut, Lebanon
e-mail: elie_sawan@hotmail.com

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same vascular access site, bleeding disorders, polyglobulia, and the complexity and length of exposure may affect hemostasis.

Direct percutaneous puncture of femoral vessels using the Seldinger method is the most popular method and access site for congenital invasive procedures at any age. This is due to the relatively larger vessels in the groin and the better accessibility to the cardiac structures and great vessels. The point and the method used in accessing the vessels implicate the type of hemostatic maneuver.

13.1.1 Manual Compression (MC)

MC remains the "gold standard" in achieving hemostasis of a femoral arteriotomy or venotomy. It is effective and well tolerated in the setting of diagnostic procedures and procedures that do not use anticoagulation.

MC does not require costly devices, has a good safety profile (with a very long experience spanning over 50 years), has a short learning curve, does not leave foreign bodies at the arteriotomy site, and is therefore still regarded as the favorite method for controlling bleeding in the majority of hospitals around the world.

With MC, the sheath is removed immediately after a diagnostic procedure, but its removal may be delayed (often 2–4 h) after an interventional procedure to allow the activated clotting time to decrease to <170 s. Before sheath withdrawal, the peripheral IV line patency should be checked to ensure that it is usable for IV medication if needed. While retrieving the sheath, it is first flushed with saline, and the skin is supported to minimize traction and vessel damage [1].

The distal immobilization bandage should be taken off during MC to avoid "tourniquet effect" on the venous access.

Vital signs are monitored and a pulse oximeter is placed distally on the limb to detect arterial pulsation. The lower extremity

coloration should be continuously inspected during compression (the distal limb perfusion should not be compromised). It is important to remember that even if the access vessel is venous, MC may compromise arterial circulation.

As the sheath is removed, firm manual pressure with a sterile dressing is placed over the femoral artery or vein, typically a very few mm above the skin entry site (arterial access) or below (venous access).

Firm compression to overcome intravascular pressure, without compromising the oximeter pulsation signal, should be held for 10 min, followed by a slightly less firm pressure for 2–5 min and then light pressure while applying a pressure dressing. Pressure should be maintained longer for larger sheath sizes and in the setting of anticoagulation. MC should be maintained until the patient is awake from anesthesia because agitation may cause rebleeding.

If bleeding persists, MC is maintained for an additional 15 min. If bleeding does not stop after 30 min and the ACT exceeds 200 s, consider protamine sulfate administration for heparin neutralization. The dose of protamine should, ideally, be calculated according to the elapsed time since the heparin was given.

The protamine doses to neutralize 100 units of heparin are:

- <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).

If, despite all the previous measures, hemostasis is not achieved, a cutdown of the vessel and direct surgical restoration or simple ligation (venous access) may be used, but this is a rare occurrence in small children.

Once hemostasis is successful, bed rest is recommended for 6–8 h. In infants and younger children, immobilization is difficult to obtain and not mandatory.

The elastic dressing should not be very tight and should be left for 12–24 h. The puncture site and lower extremity vascularization are 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.

13.1.2 Vascular Closure Devices (VCD)

Device-based hemostasis became the current standard practice in many hospitals, especially in adults. The primary goal of VCD was to improve patients' comfort and to reduce the time to hemostasis, as long as the safety profile was comparable to MC.

Currently, 10–15 % of all catheter-based procedures performed in adults utilize a VCD for femoral access site hemostasis. However, the use of these devices is not widespread in pediatrics.

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

13.1.2.1 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)

Compression devices are machines that replace humans with mechanical compression, either by using an inflatable bubble (FemoStop) or a clamp (ClampEase).

13.1.2.2 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].

13.1.3 Hemostasis in Non-femoral Access Sites

13.1.3.1 Jugular Access

The use of the internal jugular access in congenital heart procedures is common. In general, even when large introducers are used, simple MC successfully achieves hemostasis after sheath withdrawal for 5–10 min. Neck compression should not compromise the carotid artery vascularization.

13.1.3.2 Umbilical Access

In newborns of 3–5 days of life, umbilical arteries and vein are available for vascular access. The umbilical vein can accommodate relatively large catheters and facilitates access to the heart. Less frequently, an umbilical artery can be cannulated and used for aortic and left ventricular catheterization. Catheters and guidewires are withdrawn at the end of the procedure, and hemostasis is achieved by a simple light compression of the umbilicus for 5–15 min. If bleeding persists, 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.

13.1.3.3 The Transhepatic Access

It has become an important alternative for diagnostic and interventional procedures in patients with obliteration of the typical access sites. It allows introducing large sheaths and provides relatively direct access to the atrial septum, left atrium, and pulmonary veins.

Transhepatic access is possible in patients as small as 3.1 kg but is mostly used in patients weighing more than 10 kg. At the end of catheterization and in order to reduce the risk of significant hemorrhage, the parenchymal sheath tract is obliterated with Gelfoam plugs. If the ACT exceeds 200 s, 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. Then, the catheter is removed, the dilator is replaced, and the sheath is slowly withdrawn from the hepatic vein into the liver parenchyma.

A very small volume (1–2 ml) of dye contrast is injected to ensure that the sheath is no longer within the hepatic vein. 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. 13.1). The sheath is then completely withdrawn and the puncture site covered with sterile dressing.

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].

13.1.3.4 Surgical Vascular Access

When the Seldinger method fails or when larger introducer sheaths are used, a surgical access may be preferred. Sheaths are inserted through the artery or vein and secured with two 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 after 2 weeks).

13.1.3.5 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.

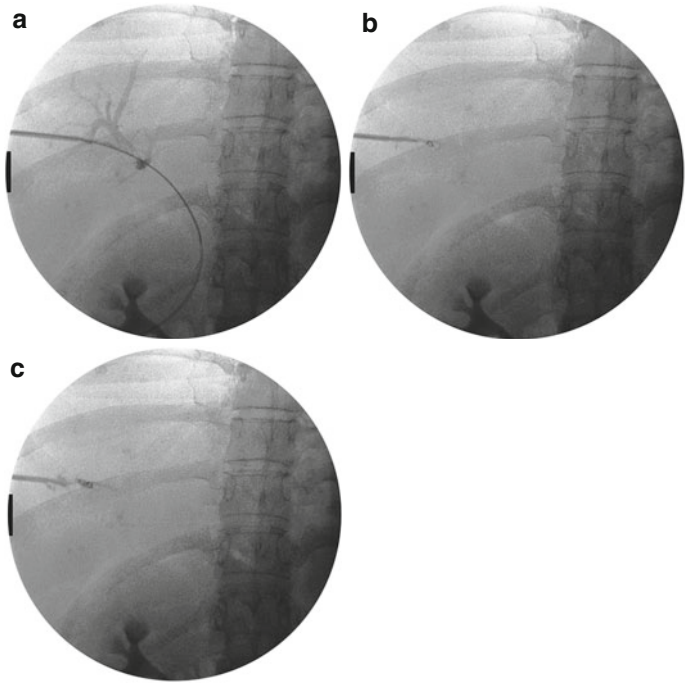

Fig. 13.1 Hemostasis after transhepatic catheterization: (**a**) A small volume (1–2 ml) of the contrast is injected to visualize the hepatic veins. (**b**) An MReye Embolization Coil (0.035 in. × 4 cm × 3 mm) is advanced to the end of the sheath and deployed between the hepatic vein and the liver capsule. (**c**) The intraparenchymal position of the sheath tip is documented by contrast injection

13.1.3.6 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.

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Chapter 14
Access Complications and Management

Zakhia S. Saliba, Georges S. Tabet, and Chady C. Kallassy

14.1 Access Complications and Management

Vascular access site (VAS) complications may cause significant morbidity and their incidences are variable depending on the definition of complications and covariates.

Children's iatrogenic vascular injuries are complicated by specific characteristics of pediatric anatomy and physiology. Unlike adults, children are not expected to have calcified arteries that could fracture under the needle pressure, advanced

Z.S. Saliba (✉)
Department of Pediatric Cardiology, Hôtel-Dieu de France Hospital, Alfred Naccache Blvd, Ashrafieh, Beirut, Lebanon
e-mail: zakhiasaliba67@yahoo.fr

G.S. Tabet
Department of Thoracic and Vascular Surgery, Hôtel-Dieu de France Hospital, Alfred Naccache Blvd, Ashrafieh, Beirut, Lebanon
e-mail: georgesstabet@gmail.com

C.C. Kallassy
Department of Angiology, St. Joseph Hospital, Raymond & Aïda Najjar St., Dora, Beirut, Lebanon
e-mail: chady.kallassy@gmail.com

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stages of atherosclerosis, or arterial occlusive disease that would complicate vascular repair.

But, children have thicker subcutaneous fat making compression less effective. They could be less compliant with postoperative motion and ambulation restrictive recommendations, and they will not express abnormal thigh tenderness.

Younger arteries are also more fragile and prone to vasospasm or sub-adventitial laceration, if over-dilated. Moreover, multiple sites of access may be required in structural heart diseases, while repeat interventions and multiple cardiac surgeries may result in incremental loss of sites. Thus, every effort should be made for preservation.

To anticipate and/or manage these complications, it is crucial to understand the predisposing factors, which can be schematically divided in three categories:

1. Operator-related factors such as experience and the inability/ability of using ultrasound for access guiding
2. Patient's conditions including younger age, anticoagulant medication, bleeding disorders, polyglobulia, connective tissue disorders, multiple cardiac surgery, or repeated catheterization using the same VAS
3. Procedural factors like the use of larger sheath in smaller patient, not well-immobilized patient, complex and long exposure, unplanned access, simultaneous venous and arterial ipsilateral femoral access, 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.

14.1.1 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. 14.1).
- 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.

14.1.2 Complications Linked to the VAS and Techniques

14.1.2.1 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,

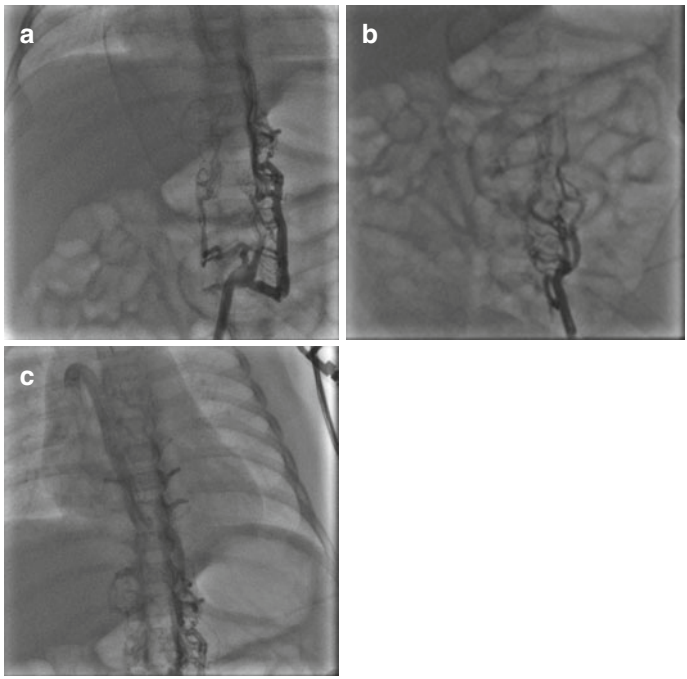

Fig. 14.1 Bilateral occlusion of the femoral veins in a 12-month-old child: dye injection in both veins (**a**, **b**) shows the contrast drainage to the right heart via a paravertebral collateral network to the right azygous vein and the right superior vena cava (**c**)

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. 14.1), 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

A direct communication between an artery and a vein that occurs when they are punctured once the sheath is removed is usually caused by multiple access attempts, punctures above or below proper site level, and impaired clotting.

It can be asymptomatic and result in bruit and/or thrill at VAS and in swollen and tender extremities. Distal arterial insufficiency and/or deep venous thrombosis can cause limb ischemia and congestive heart failure.

Some arteriovenous fistulas resolve spontaneously without intervention. Others require ultrasound-guided compression but surgical repair is rarely needed especially in children.

14.1.2.2 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].

14.1.2.3 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.

14.1.2.4 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.

14.1.2.5 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].

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Chapter 15
Transseptal Access

Chad Kliger and Carlos E. Ruiz

15.1 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 growing number of congenital heart patients reaching adulthood, the operator needs skills to safely perform a TS puncture.

C. Kliger, MD, MS
Department of Cardiothoracic Surgery, Division of Structural and Congenital Heart Disease Lenox Hill Heart and Vascular Institute – North Shore/LIJ Health System, 130 East 77th Street, 4th Floor Black Hall, New York, NY 10075, USA
e-mail: ckliger@nshs.edu

C.E. Ruiz, MD, PhD (✉)
Department of Cardiothoracic Surgery, Division of Structural and Congenital Heart Disease Lenox Hill Heart and Vascular Institute – North Shore/LIJ Health System, 130 East 77th Street, 9th Floor Black Hall, New York, NY 10075, USA
e-mail: cruiz@nshs.edu

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15.2 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, anteroinferiorly 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.

15.3 Transseptal Technique

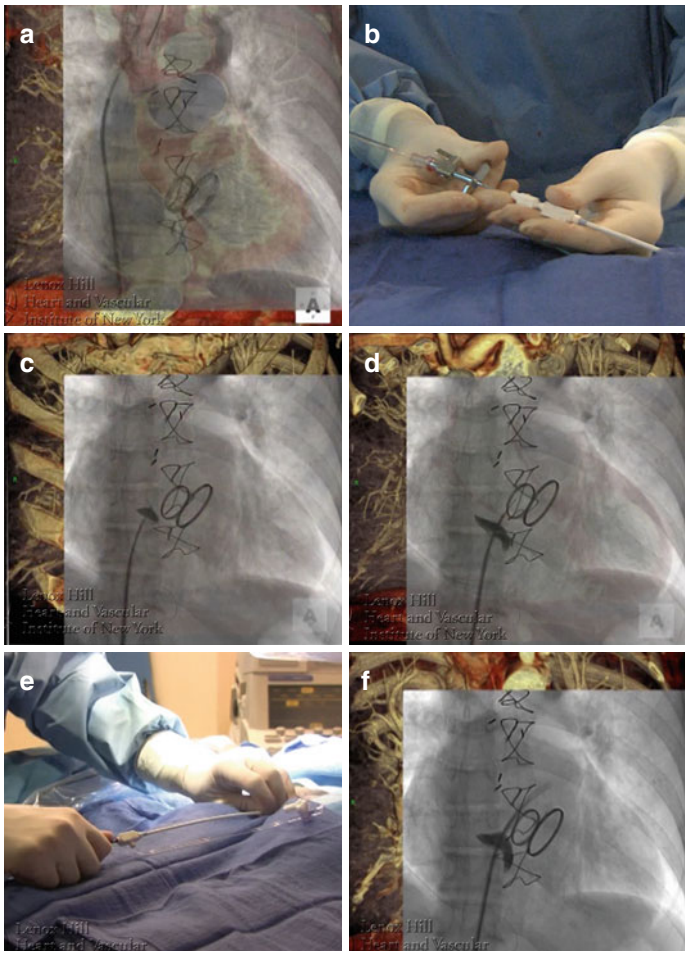
The technique of *TS puncture* has a multitude of steps with variations made based on operator preference and patient-related factors (Fig. 15.1). First, a 0.032 or 0.035 in. J-tipped 135 cm guidewire is advanced to the SVC and a 8 F 62 cm-long sheath (SL or Mullins) placed. The guidewire is removed, 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. 15.2). 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.

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 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.

Fig. 15.1 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

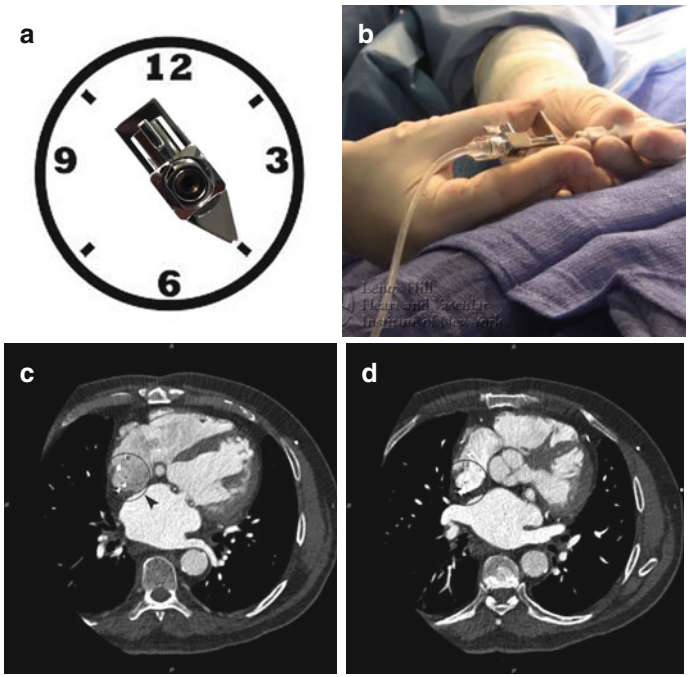


Fig. 15.2 Transseptal needle position. (**a**) The transseptal (TS) needle indicator arrow dictates the location of the needle tip. With the patient on a horizontal plane (3/9 o'clock), the sheath with sideport and indicator arrow are pointed in the same direction to the 4–5 o'clock location (45° from 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

15.4 Knowledge of Imaging

15.4.1 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 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° [3] (Fig. 15.3). 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 halfway 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.

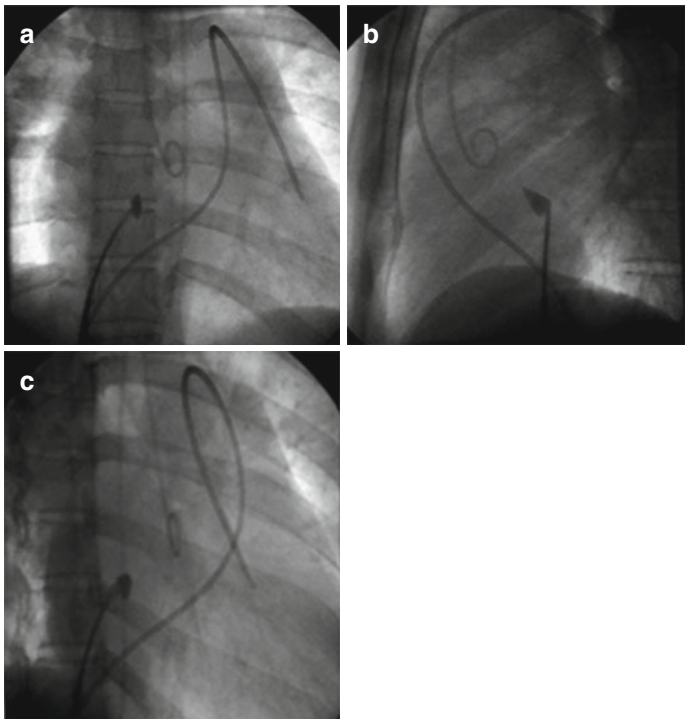


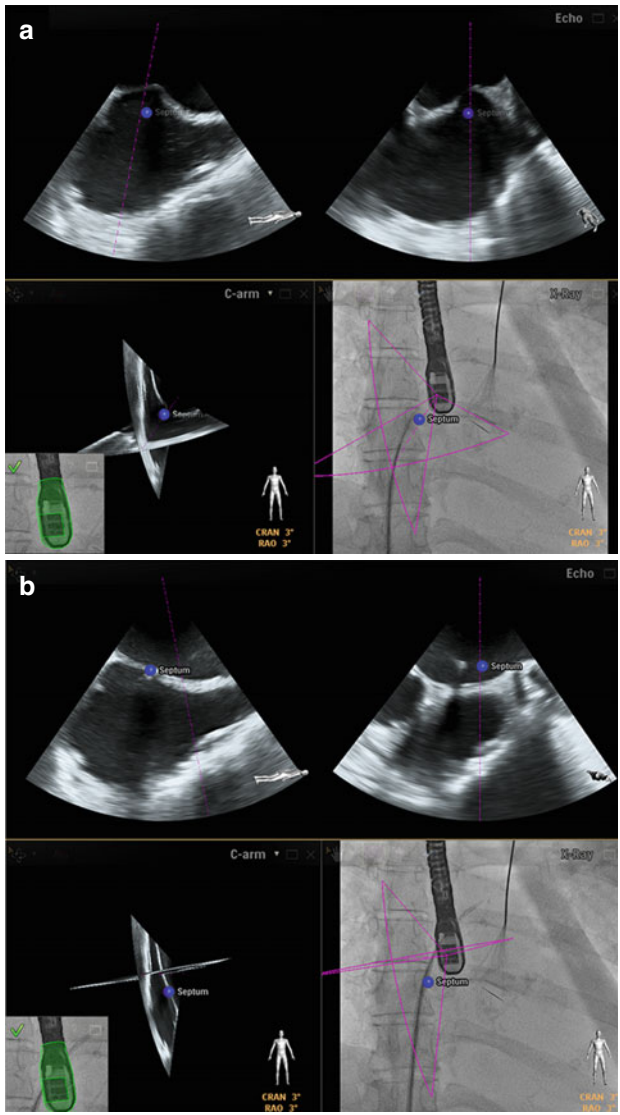
Fig. 15.3 Fluoroscopy for transseptal puncture. (**a**) In the anteroposterior (AP) view, the pigtail can be noted placed into the noncoronary aortic valve cusp, identifying the posterior border of the aortic wall and aortic root. A balloon-tipped catheter is also visualized within the right ventricular 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

15.4.2 Echocardiography

Nonetheless, 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, originally via transthoracic and more recently through transesophageal (TEE) and intracardiac (ICE) echocardiography. *3D TEE* enables even more comprehensive imaging of the heart, using either volumetric or multiplane 2D imaging [4, 5]. 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 clear definition of all intracardiac catheters, the IAS, septal tenting prior to puncture, and bubble visualization in the LA confirming needle position [6].

15.5 Advanced Imaging and Site-Specific Puncture

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. 15.4).


a
Echo
C-arm
X-Ray
Septum
CRAN 3°
RAO 3°
b
Echo
C-arm
X-Ray
Septum
CRAN 3°
RAO 3°

15.6 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 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. 15.5a, c, d).

Fig. 15.4 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

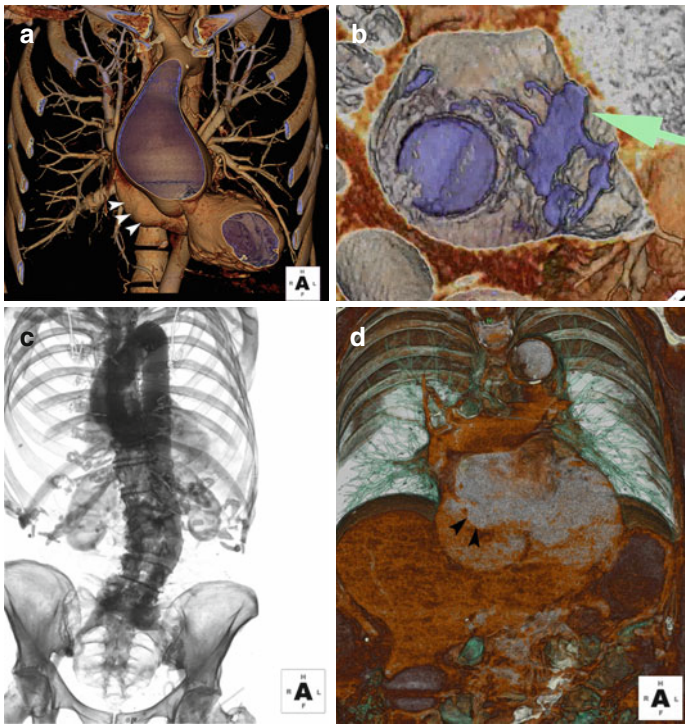


Fig. 15.5 High-risk transseptal anatomy identified by CTA. (**a**) Extracardiac distortion by an ascending aortic aneurysm can cause bulging of the antero-superior aspect of the interatrial septum (IAS) (*white arrowheads*). (**b**) Extensive calcification (*green arrow*) of the IAS can be visualized 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*)

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 [7]. 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 [8, 9] (Fig. 15.5b). 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.

15.7 Alternative Approaches and Advanced Techniques

Difficulties at engaging the FO may be related to many of the anatomical variations described [10]. 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* [11]. 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 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 [12, 13].

This can be achieved by using a dedicated RF TS system (Baylis Medical, Montreal, Canada) 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.

Finally, increasing numbers of patients presenting for TS cardiac catheterization have undergone previous percutaneous ASD or PFO closure [14–16]. 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 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.

15.8 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.

15.9 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 % [17–19]. 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.

15.10 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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Part III
Fetal Procedures

Chapter 16
Fetal Interventions

Carlos A.C. Pedra and Simone Fontes Pedra

16.1 Introduction and Clinical Scenarios

With evolving catheter and imaging technologies and better patient selection, fetal interventions have become an important therapeutic modality in last 10–15 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

C.A.C. Pedra, MD, PhD (✉)
Catheterization Laboratory for Congenital Heart Disease, Instituto Dante Pazzanese de Cardiologia and Hospital do Coração, Av Dr Dante Pazzanese 500 14 andar, Sao Paulo, SP 04012-180, Brazil
e-mail: cacpedra@uol.com.br

S.F. Pedra, MD, PhD
Echocardiography Laboratory for Congenital Heart Disease, Instituto Dante Pazzanese de Cardiologia and Hospital do Coração, Av Dr Dante Pazzanese 500 14 andar, Sao Paulo, SP 04012-180, Brazil
e-mail: sfpedra@uol.com.br

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scenarios, a prenatal intervention may remodel cardiac morphology and function and result in improved pre- and postnatal outcomes, including an increased likelihood of achieving a biventricular (BV) circulation [1–3]. In this chapter we review the indications, techniques, and current results of fetal cardiac interventions.

16.2 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. 16.1).
2. HLHS and intact or highly restrictive IAS [1–3]: A prenatal echocardiographic diagnosis of HLHS with either an intact IAS or a tiny ($\leq$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

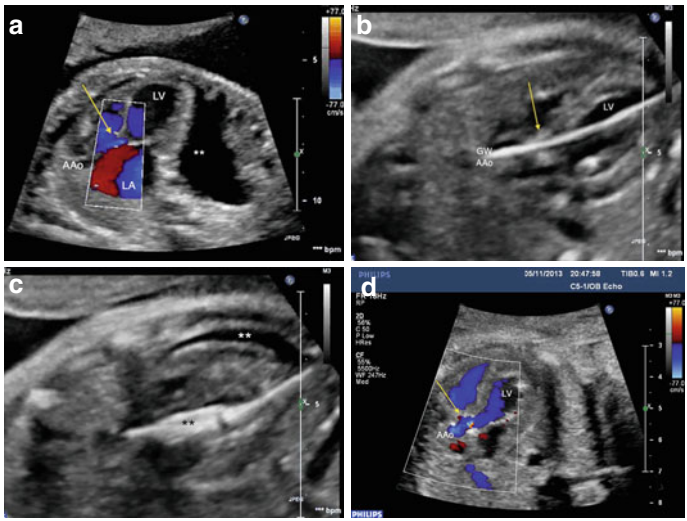

Fig. 16.1 Critical aortic stenosis in a 26 weeks' gestational age hydropic fetus. (**a**) Pre-intervention echocardiographic assessment. Color flow mapping across the aortic valve (indicated by an *arrow*) showing tiny forward flow (*blue color*). The *red color* displays the flow into the right ventricle 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 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 contrata across the aortic valve). *Abbreviations*: *LA* left atrium, *LV* left ventricle, *AAo* ascending aorta, *GW* guidewire

ASD creation are ideally performed between 29 and 32 weeks' gestation in order to endure until term (Fig. 16.2).

3. PA/IVS or CPS/IVS and evolving HRHS [1–3]: Patients should have a prenatal echocardiographic diagnosis of PA/

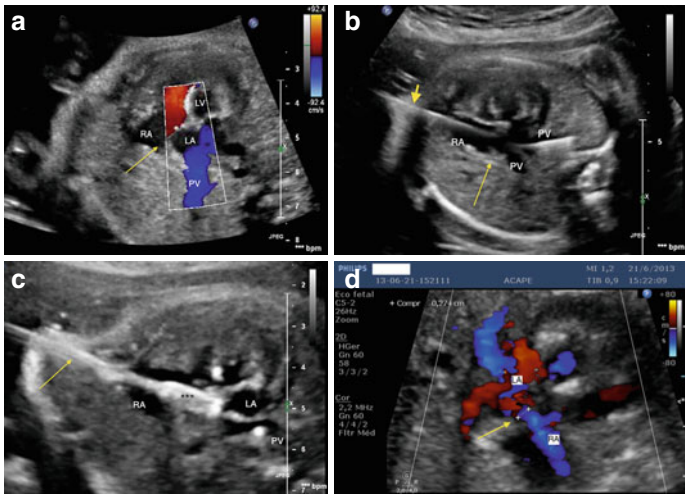

Fig. 16.2 Fetal atrial septostomy in a 29 weeks' gestational age fetus with established hypoplastic left heart syndrome (**a**). Pre-intervention echocardiographic assessment. The LA is conspicuously dilated and the interatrial septum (indicated by an *arrow*) is almost intact. There is flow reversal 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*). *Abbreviations*: *LA* left atrium, *RA* right atrium, *PV* pulmonary vein, *LV* left ventricle

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;

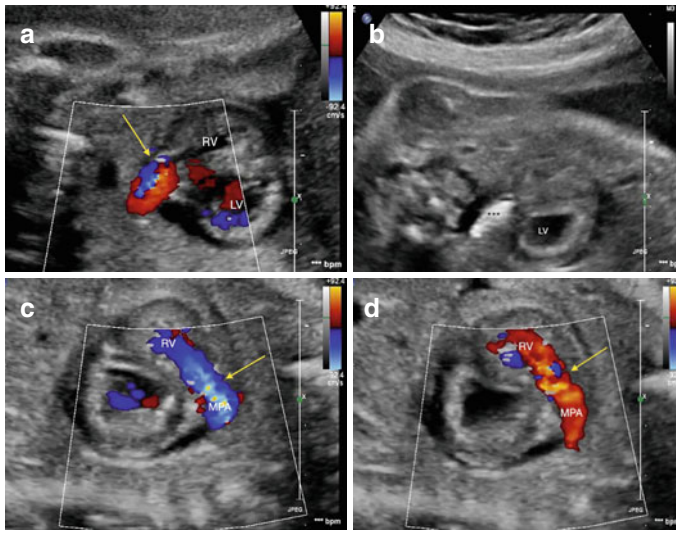

Fig. 16.3 Critical pulmonary valve stenosis in a 28 weeks' gestational age fetus. (**a**) Pre-intervention echocardiographic assessment. Color flow mapping across the pulmonary valve (indicated by an *arrow*) shows tiny forward flow (in *blue color*). The *red color* depicts the retrograde flow across 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. *Abbreviations*: *RV* right ventricle, *LV* left ventricle, *MPA* main pulmonary artery

left-to-right shunting across a patent ductus arteriosus (PDA); and right heart hypoplasia, with a tricuspid valve (TV) annulus Z-score below ≤ 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. 16.3).

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.

16.3 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.

16.4 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. 16.4). Under continuous two-dimensional ultrasound guidance, a 15-cm-long 17–18-gauge 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. 16.4). A premarked 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 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. 16.4). 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

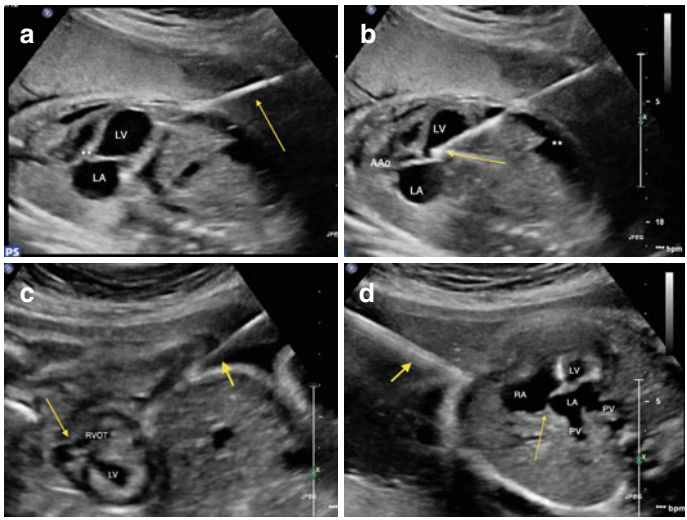

Fig. 16.4 Proper needle course, angulation, and positioning during the fetal cardiac procedures. (**a**) During fetal aortic valvuloplasty the tip of the needle (indicated by an *arrow*) is aimed at the aortic valve (marked with *asterisks*) in an imaginary line across the left ventricular apex toward 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*). *Abbreviations*: *LV* left ventricle, *LA* left atrium, *RV* right ventricle, *RA* right atrium, *AAo* ascending aorta, *RVOT* right ventricular outflow tract, *PV* pulmonary vein

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. 16.1 and 16.3). 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. 16.2 and 16.4). The same needle is used to perforate the IAS to gain access to the LA (Fig. 16.2). 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.

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 mcg/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. 16.1).

16.5 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.

16.6 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.

16.7 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.

16.8 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. 16.1 and 16.3). We have considered post-procedural AR and PI as a marker of effective dilatation of the aortic and pulmonary valves (Fig. 16.3). 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. 16.2) associated with reduction in LA size and improvement in the pulmonary vein Doppler pattern (Fig. 16.5). The ASD size is determined by measuring the width of the color jet (vena contracta) (Fig. 16.4).

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,

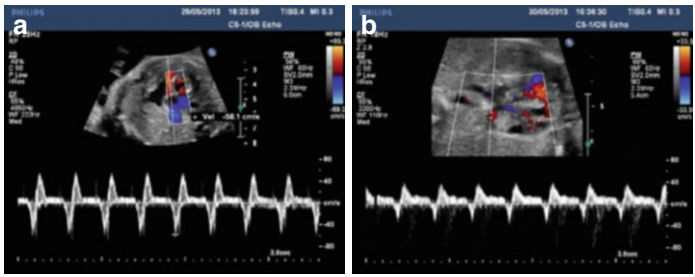

Fig. 16.5 Doppler tracing of the pulmonary veins pre- and post-fetal atrial septostomy in a fetus with hypoplastic left heart syndrome and almost intact interatrial septum. (**a**) Pre-intervention assessment. Bidirectional flow in the pulmonary vein with high reversal flow velocity (58 cm/s) during 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)

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 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. 16.6). We have employed a staged strategy for such patients with fetal aortic valvuloplasty followed by a neonatal hybrid procedure ± balloon aortic valvuloplasty. This approach works as a bridge to LV overhaul and BV repair later in

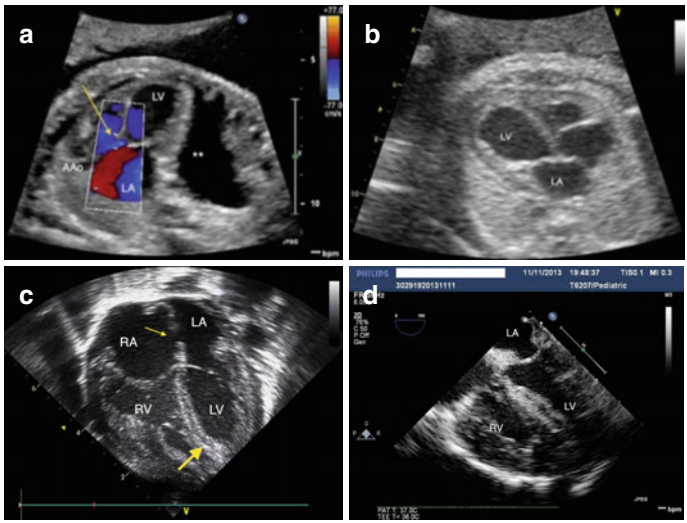

Fig. 16.6 Staged rehabilitation of the left ventricle in a fetus with critical aortic stenosis (same patient as in Fig. 16.1). (**a**) Pre-fetal intervention echocardiographic assessment at 29 weeks' gestational age. Color flow mapping across the aortic valve (indicated by an *arrow*) showing tiny forward 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 significant improvement in the LV size (LV length Z-score −1.2). *Abbreviations*: *RA* right atrium, *LA* left atrium, *RV* right ventricle, *LV* left ventricle, *AAo* ascending aorta

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].

Fetuses with critical AS, severe MR, and gigantic LA have a somber prognosis no matter what is performed in the pre- or postnatal periods. This condition is commonly associated with either fetal loss or prematurity. In our experience, only one out of four patients with this condition survived with a BV circulation and normal LV function after an initial hybrid procedure and LV overhaul at 9 months [2].

Fetuses with HLHS who underwent 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.

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. In our experience with six fetuses, technical failure was observed in the first case performed. One is still in utero. The remaining four patients showed significant growth of the RV structures achieving an eventual BV circulation after initial neonatal palliation with pulmonary valvuloplasty and ductal stenting [2].

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Part IV
Step-by-Step Procedures: Valve Dilatation

Chapter 17
Aortic Valvular Stenosis

Xiangbin Pan

17.1 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)

X. Pan
Department of Cardiovascular Surgery, National Center for Cardiovascular Disease, Fuwai Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, 167 Beilishi Road, Xi Cheng District, Beijing 100037, China
e-mail: xiangbin428@hotmail.com

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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 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 to one side. The rest lie to 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.

17.2 Clinical Scenarios

17.2.1 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.

17.2.2 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.

17.3 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.

17.4 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.

17.5 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.

17.6 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. 17.1). 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. 17.2).
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.

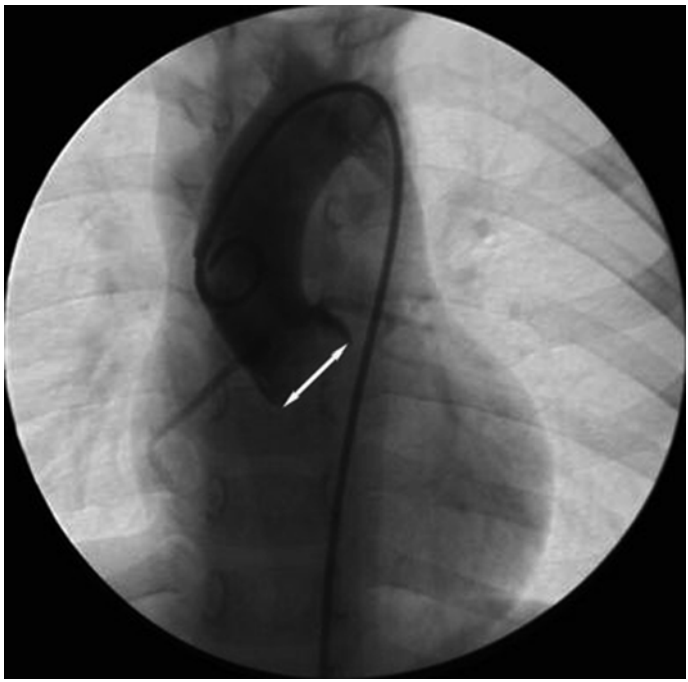

Fig. 17.1 The aortic valve annulus is measured by aortography in posterior-anterior projection. The *double-headed arrow* indicates the aortic annulus diameter between the hinge points of the valve leaflets

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.

5. Techniques of valvuloplasty: The techniques of valvuloplasty include single- and double-balloon valvuloplasty.

   5.1. Single-balloon valvuloplasty technique: With the help of an angiographic catheter (Judkins right, multipurpose

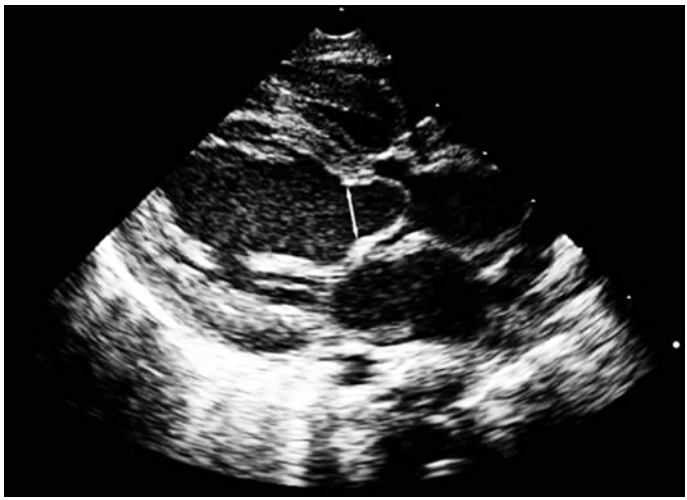

Fig. 17.2 Echocardiographic measurement of the aortic valve annulus between the hinge points of the valve leaflets (*double-headed arrows*) in the parasternal long-axis view

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″ extrastiff guidewire in adolescents) that will be used for the angioplasty. The wire is placed in the LV apex along the septum, anterior to the mitral valve chordal apparatus.

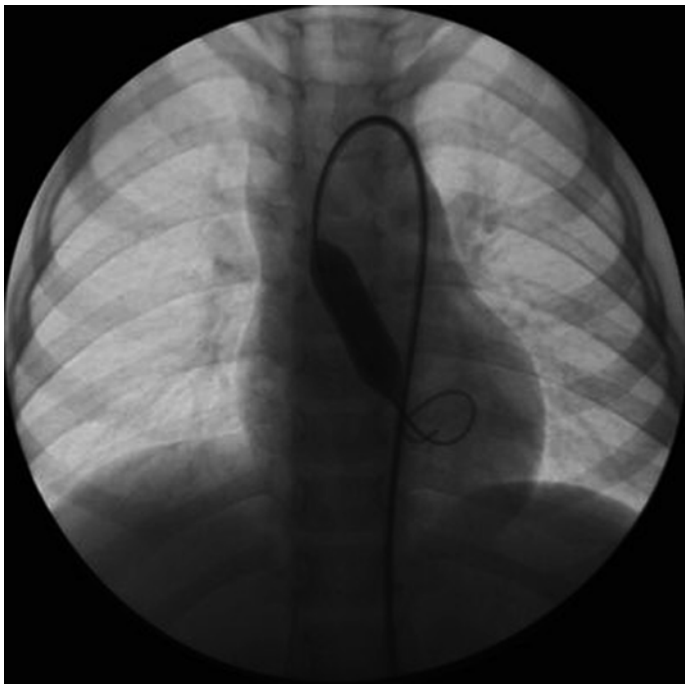

Fig. 17.3 Single-balloon technique in posterior-anterior projections

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. 17.3). 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

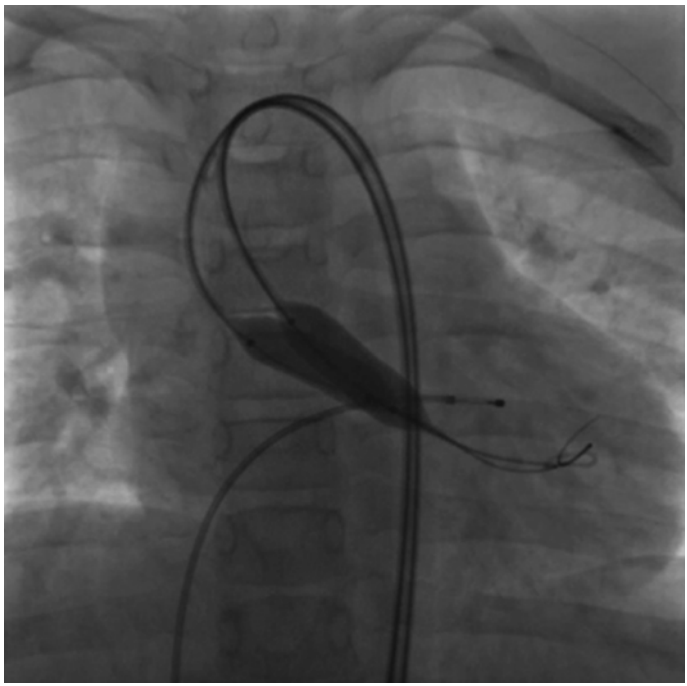

Fig. 17.4 Double-balloon technique in posterior-anterior projections

impulses per minute during the balloon inflation. Ultimately, such pacing can increase the success rate of the procedure.

5.2. 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. 17.4). 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 balloon. 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 $\leq$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.

17.7 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 in., the 0.018 in. 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.

17.8 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.

17.9 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.

17.10 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.

17.11 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 complication 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.

17.12 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 (Fig. 17.5). 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, no need for temporary pacemaker, cardiac compression under direct vision, and extracorporeal circulation, if needed. Hybrid balloon valvuloplasty appears to be an effective, simple, and safe procedure for low-weight infants with severe congenital valvular AS.

17.13 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

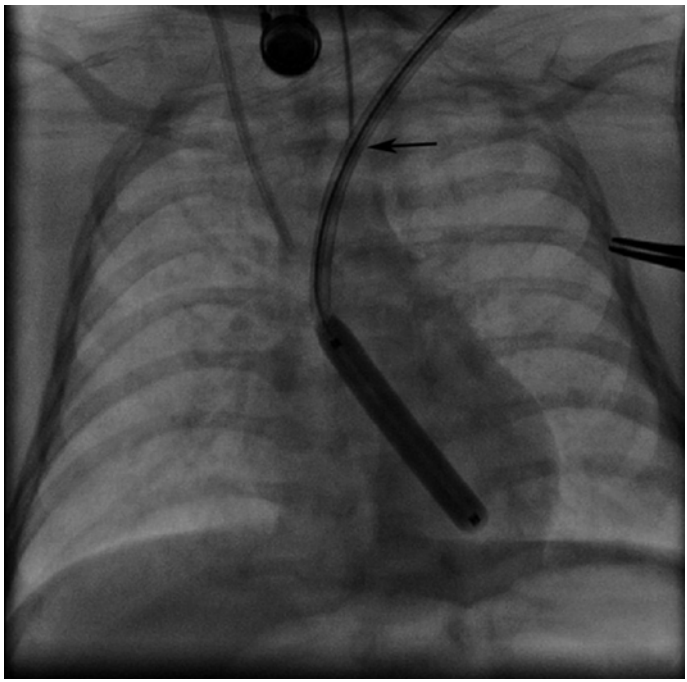

Fig. 17.5 Patients underwent tracheal intubation under general anesthesia on supine position in the hybrid operation room. After median sternotomy, the wire and balloon advanced into the aorta by an arterial sheath which was inserted into the aorta (the *arrow* pointed to the arterial sheath)

of the femoral artery or vein, the dorsalis pedis artery pulse should be taken regularly.

17.14 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 month, 3 months, 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 [2] found that after initial percutaneous balloon valvuloplasty, 65 % percent 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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Chapter 18
Pulmonary Valve Stenosis

Tingliang Liu and Wei Gao

18.1 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.

T. Liu, MD • W. Gao, MD (✉)
Department of Cardiology, Shanghai Children's Medical Center,
No. 1678 Dongfang Road, Shanghai 200127, China
e-mail: liutingliang@hotmail.com; davidgao1963@163.com

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The obstruction is mainly related to thickened, myxomatous immobile pulmonary valve cusps and valve ring hypoplasia.

18.2 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.

18.3 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.

18.4 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.

18.5 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.

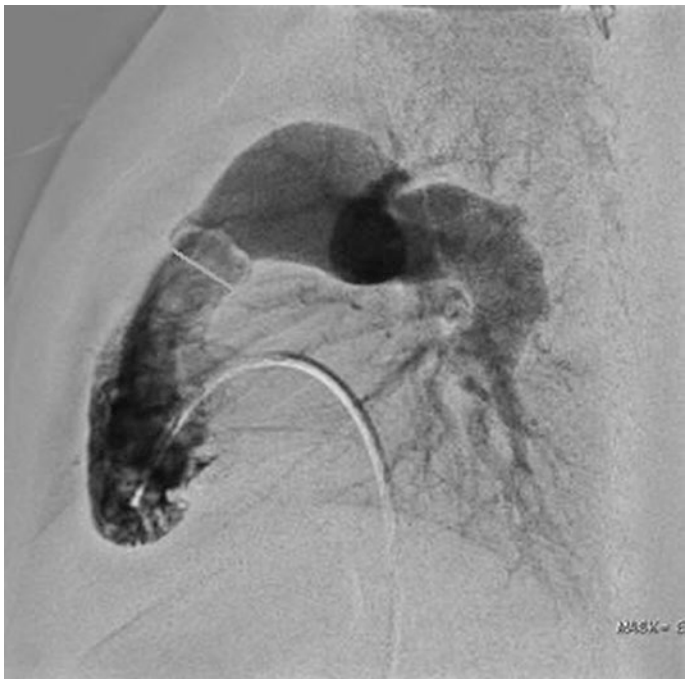

Fig. 18.1 Right ventriculogram in left lateral view showing pulmonary valve stenosis; thickened, domed pulmonary valve; and poststenotic dilatation of the main pulmonary artery. The white line shows the length of the pulmonary valve annulus

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. 18.1).

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 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. 18.2). The inflation pressure is gradually increased up to the manufacturer's recommendation (Fig. 18.3), 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 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.

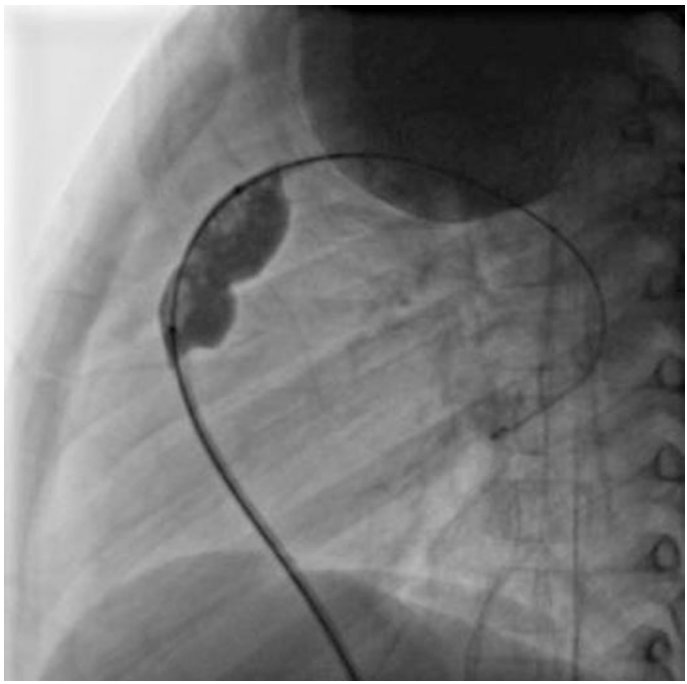

Fig. 18.2 Left lateral view of a partially inflated balloon catheter positioned across the pulmonary valve. As the balloon is inflated, a waist appears at the site of the pulmonary valve

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?

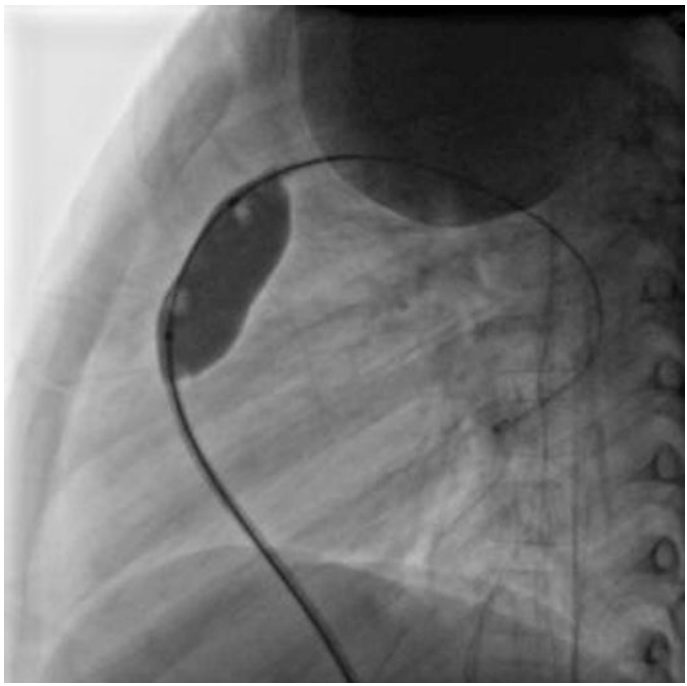

Fig. 18.3 Left lateral view of the balloon waist completely disappeared

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.

18.6 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. 18.4). 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.

18.7 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.

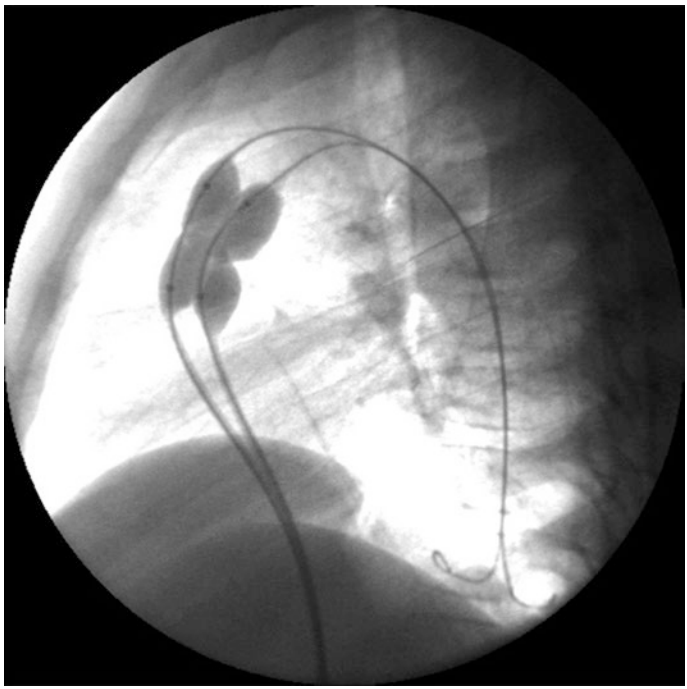

Fig. 18.4 Left lateral view of two balloon catheters positioned across the pulmonary valve showing waisting of balloons produced by the stenotic valve

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 stenosis is higher after balloon dilation. But the prevalence of pulmonary insufficiency and ventricular arrhythmias is higher after surgery.

18.8 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.

18.9 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.

18.10 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. ß-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.

18.11 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.

18.12 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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Chapter 19
Pulmonary Atresia and Intact Ventricular Septum

Zaheer Ahmad and Mazeni Alwi

19.1 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 muscle 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 [1].

Z. Ahmad
Department of Paediatric Cardiology, Southampton University Hospital NHS Foundation Trust, Tremona Road, Southampton, SO16 6YD, UK
e-mail: drzaheer@yahoo.com

M. Alwi (✉)
Department of Paediatric Cardiology, Institut Jantung Negara, 145, Jalan Tun Razak, 50400 Kuala Lumpur, Malaysia
e-mail: mazeni@ijn.com.my

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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 a modified Blalock-Taussig (BT) shunt on account of small RV cavity size [2, 3].

19.2 Anatomic Description

The principal anatomic lesion in PAIVS completely obstructs 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 the RV cavity by overgrowth of the muscles. However in the rare cases with associated severe Ebstein's anomaly, the RV (and RVOT) is thin-walled and markedly dilated (Fig. 19.1c). RV-coronary connections are a peculiar association in PAIVS, seen most often in those with diminutive RV cavity.

19.2.1 Major Anatomic Subtypes

(i) *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

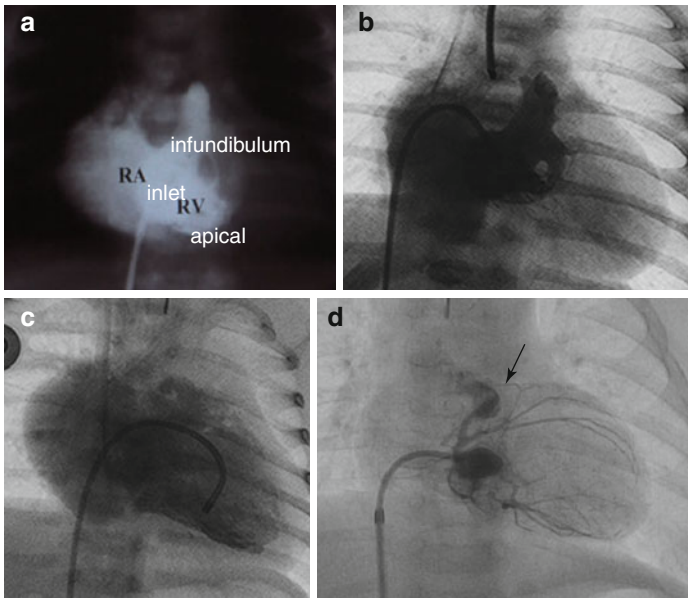


Fig. 19.1 Angiographic morphology of the RV in PAIVS. (**a**) Mild RV cavitary hypoplasia with well-developed infundibulum, inlet and apical parts and 'tripartite' RV. There is moderate TR and the RA is dilated. (**b**) In the 'bipartite' RV, the inlet and infundibulum are well developed, but the apical 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)

the RV is 'bipartite' where there is near obliteration of the apical part by muscles, while the remaining two are well developed (Fig. 19.1a, b). The atretic membranous valve usually is 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. 19.2a, 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. 19.2c, d). In a small minority, the RVOT is markedly dilated such that the usual tunnel-like configuration is lost with the small valve and small annulus facing the dome-like infundibulum (Fig. 19.2d). 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.

The morphologic subtype illustrated by Fig. 19.1a, 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.

(ii) *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. 19.1d). The coronary circulation is RV dependent because of atresia or severe stenoses of the proximal coronary arteries and has been described in anywhere from 3–34 % of these patients. 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 inserting onto the dome of the main pulmonary artery (MPA) away from the orifice of the

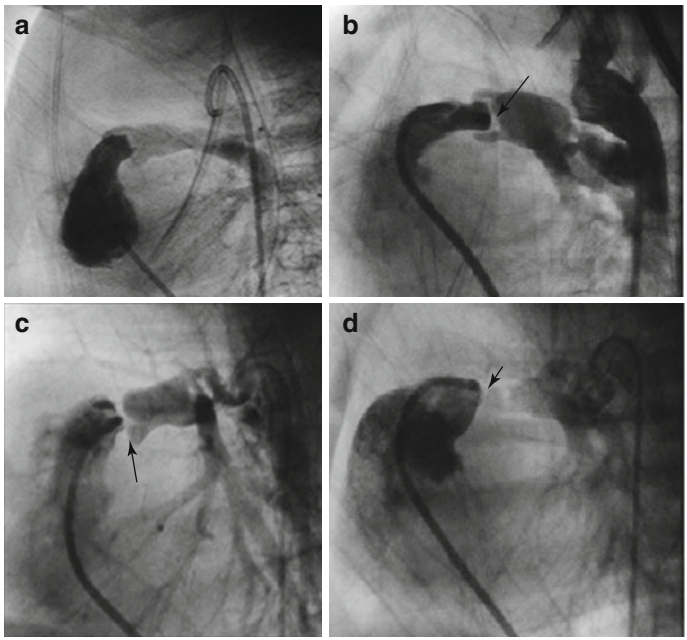


Fig. 19.2 The RVOT valve and pulmonary artery. Simultaneous angiograms in the RVOT and descending aorta opposite the PDA are performed to outline these structures. (**a**) Well-developed infundibulum, thin membranous valve and mildly hypoplastic annulus. The main pulmonary artery and sinuses are well 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)

proximal left pulmonary artery (LPA). Though they are usually elongated, they are seldom tortuous. However, as in other cyanotic heart disease, the PDA may have a more complex morphology and insert onto the proximal LPA causing stenosis of this branch.

19.3 Pathophysiology and Clinical Presentation

Because of complete RV outflow obstruction and major aorto-pulmonary 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 most common presentation. However in the current era, diagnosis is often made antenatally during fetal 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.

19.4 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.

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.

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 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. This is conventionally managed with closed surgical valvotomy or an open procedure with RVOT reconstruction. Many surgeons electively perform concomitant BT shunt as a significant number remain cyanotic after a successful valvotomy procedure.

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. PDA stenting may be performed at the same time in patients whose RV is deemed small and who likely require augmentation of the pulmonary blood flow following RFV-BD. The assignment of individual patients towards the single-ventricle vs 2-ventricle track is based on the initial echocardiographic assessment and cardiac catheterization.

19.5 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, i.e. whether the patient is a likely candidate for RFV-BD, i.e. a membranous atresia with well-developed infundibulum and at most moderate RV hypoplasia (tripartite or bipartite RV), or one destined for the Fontan track, i.e. muscular atresia of the infundibulum and a diminutive, unipartite RV.

The measurement of TV Z-score and TV/MV annulus ratio provides a semiquantitative measure of RV size. Doppler

echocardiography provides additional information with the degree of tricuspid regurgitation and an estimate of the RV systolic pressure.

Finally, branch pulmonary artery size and confluence is 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.

19.6 Technique, Materials, Tips and Tricks

(i) Ideally the patient should weigh > 2.5 kg to prevent access-related complications. The procedure is done under general anaesthesia with ICU backup for postoperative recovery. Biplane fluoroscopy is essential and echocardiography should be on stand-by in the event of suspected tamponade.

(ii) PGE1 infusion should be stopped 4–6 h 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 and 5 F sheath, respectively, and heparin 50–100 ű/kg given. A 4 F pigtail catheter is placed in the aorta opposite the PDA for angiogram of the pulmonary artery and PDA.

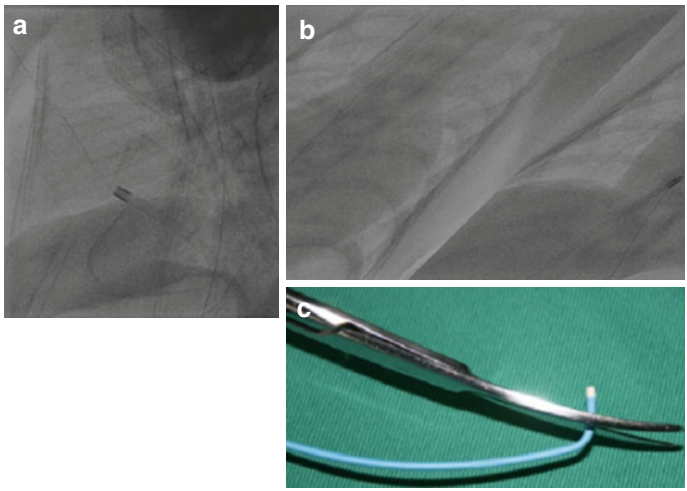


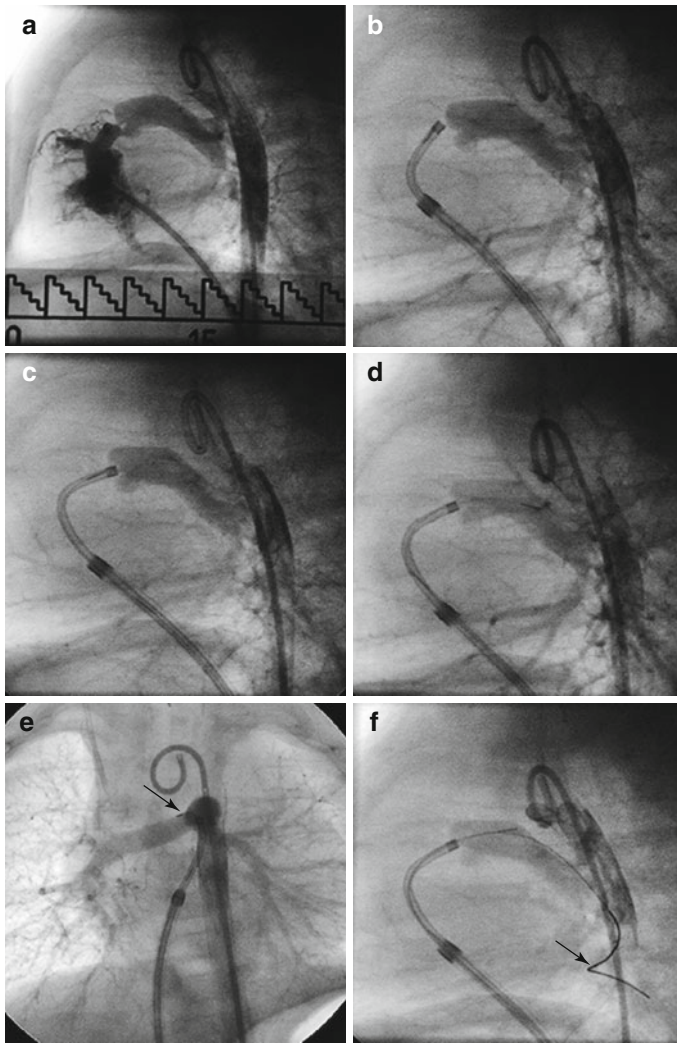
Fig. 19.3 (**a**, **b**) A 5 F Mullins' sheath placed in the RA has its tip facing the TV, facilitating quick access to the RV, and provides catheter stability during angiography and wire perforation of the valve. (**c**) Cutting the distal tip of a JR catheter facilitates manoeuvring of the catheter 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)

(iii) Initial haemodynamic 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 the tricuspid valve, enabling easy passage of a catheter into the RV (Fig. 19.3a, b).] We recommend a 4 F or 5 F Judkins right (JR) catheter for both the diagnostic

and interventional purposes. As the RV cavity is usually small hand, had injections would normally suffice. An RV angiogram is performed in the AP and lateral projections to assess the size of the RV and morphology of the RVOT (Fig. 19.4a). 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.

(iv) 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.

Fig. 19.4 Step-by-step illustration of the procedure. (**a**) Diagnostic RV angiogram in the lateral projection with a JR catheter. Small RV cavity but with well-developed infundibulum and membranous atresia. (**b**, **c**) RF wire advanced to the tip of JR catheter ready for perforation. Performing minor adjustments of catheter tip position and checking with small doses of contrast to ensure correct positioning and avoid misperforation. (**d**, **e**) Angiogram in descending aorta opposite the PDA to opacify the pulmonary arteries. RF wire tip (*arrow*) is within the lumen of the RPA. (**f**) A choice PT extra support coronary guidewire is passed alongside the RF wire and anchored in a distal RPA (*arrow*) branch. The RF wire is then withdrawn. (**g**) Initial balloon dilation with a 2.0 mm × 15.0 mm coronary balloon. A 5 F JR guiding catheter (*arrow*) provides support during passage of balloon across the valve. Without this support the balloon catheter shaft and wire have a tendency to loop in the RA instead of advancing across the valve. (**h**) Final balloon dilation with a 8.0 mm × 30 mm balloon. (**i**) RV angiogram in lateral projection post dilation showing a widely opened valve (*arrow*). (**j**) Due to the small RV cavity, the PDA was concomitantly stented (*RV* right ventricle, *RF* radiofrequency, *JR* Judkins right, *PDA* patent ductus arteriosus, *RPA* right pulmonary artery, *RA* right atrium)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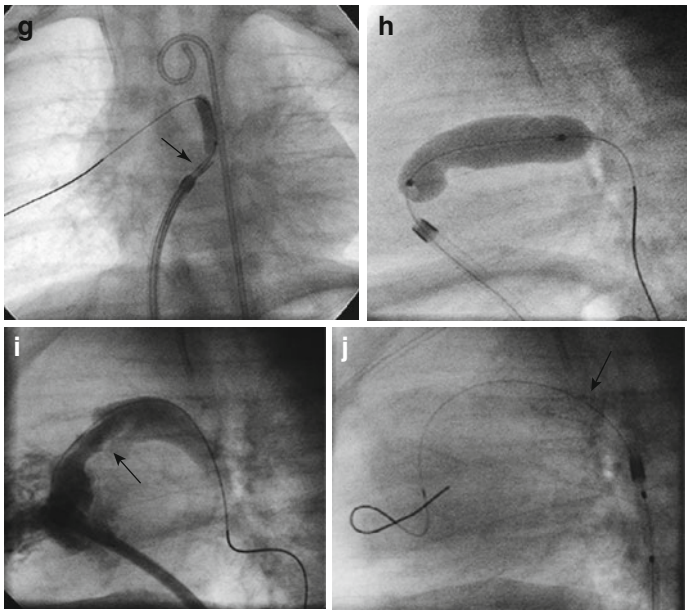

Fig. 19.4 (continued)

The size of the pulmonary annulus and thickness of the valve membrane/plate can be measured (Fig. 19.2). In favourable cases, the pulmonary root and sinuses 'cup' over the membranous atretic valve and infundibulum (Fig. 19.2a, 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. 19.3c).]

(v) *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 (Fig. 19.4b, c).

(a) *Perforation with radiofrequency (RF) wire – equipment and materials*

The Baylis radiofrequency coaxial system (Baylis Medical Company, Montreal, Canada H4T 1A1) with its generator and wire is the most widely used system for valve perforation. It consists of BMC radiofrequency perforation generator and the Nykanen RF perforation catheter (wire). The active tip diameter of the wire is 0.012″, and body diameter is 0.024″.

The Baylis system has largely replaced the older Osypka system (Dr Osypka GmbH, 79618 Rheinfelden, Germany) with its cereblate PA 120 wire (diameter 0.018″) and the HAT 300 generator. In difficult cases we prefer the Osypka wire powered by the Baylis generator as the wire is more flexible and the smaller diameter probably reduces the likelihood of tamponade in the event of misperforation.

With the Baylis system once the wire is across the pulmonary valve, the coaxial microcatheter is advanced over the wire. This wire is then exchanged with a coronary wire for graded balloon dilation. With the Osypka system, one can 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. 19.5a).

A Y-connector is attached to the JR catheter to prevent blood loss and for hand shot angiograms from 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

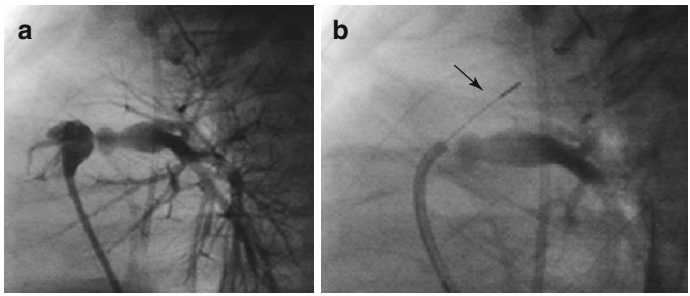

Fig. 19.5 Misperforation. (**a**) RVOT angiogram in lateral projection in a patient with associated Ebstein's anomaly and severe TR. The RVOT is markedly dilated, with a small valve annulus and pulmonary artery root. (**b**) The RF wire has perforated the RVOT wall anteriorly. Aortogram in descending 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)

at the usual setting of 5 W for 2 s. 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 RF administration is stopped once the wire is across the valve. An aortogram opposite the PDA is performed to ensure that the wire is in a pulmonary artery lumen (Fig. 19.4d, e). A misperforation outside the heart would be demonstrated. The wire following the outline of the heart in the pericardial space is also suggestive of a misperforation. The RF wire is replaced with a coronary wire of medium stiffness and parked either into one of the branch pulmonary arteries (Fig. 19.4f) or through the duct into the descending aorta. The JR catheter is then removed and replaced by a 3 mm coronary balloon for the initial dilation. [*Tip:* A 5 F JR guiding catheter with a lumen of 0.058″ is used for smooth delivery of the balloon catheter as otherwise the 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. 19.4g).] Subsequent balloon dilation is carried out with a Tyshak Mini balloon (NuMED Canada Inc., 45 s St West Cornwall, ON K6J 1G3) 150–200 % larger than the annulus size (usually 6–8 mm diameter balloon) (Fig. 19.4h).

(b) *Perforation with stiffer coronary wire*
Alternatively in the more straightforward cases, i.e. very thin, membranous valve with well-developed pulmonary artery root and sinuses cupping over the doming valve, perforation can be accomplished with stiffer coronary wires 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).
The same guidewire may then be used for balloon dilation and PDA stenting if indicated. Simplifying the procedure is attractive, but the risk of misperforation is higher than with the RF system. The stiff end of a coronary guidewire is not recommended.

(vi) Subsequent haemodynamic data is obtained, and RV angiogram is done. In patients with borderline RV size, PDA stenting can be done at the same time if the PDA is less than 2 mm and morphology is favourable.

19.7 Expected Results

The expected outcome of a successful procedure is establishment of forward flow into the pulmonary artery (Fig. 19.4i), normalization of RV pressure and marked improvement in oxygen saturation in the short term. Immediate full saturation is not 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 the possible reasons for that include insufficient dilation, infundibular spasm or the presence of fixed subvalve stenosis. In patients with RV-coronary connections, the RV angiography will show immediate disappearance of these vessels if the RV has been adequately decompressed. If the PDA is also stented at the same procedure (Fig. 19.4j), pulmonary overcirculation is common and diuretics may be required for a few weeks.

19.8 Pitfalls, Complications and Their Management

(i) The major specific complication of this procedure is misperforation of the wire outside of the heart. Fortunately most misperforations do not lead to tamponade. To avoid misperforation, it is vital that the catheter tip and wire are 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 in the pulmonary artery lumen before balloon dilation (Fig. 19.5). Misperforation tends to occur in patients with dilated RVOT and small annulus. Retrograde perforation via the PDA is an alternative.
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

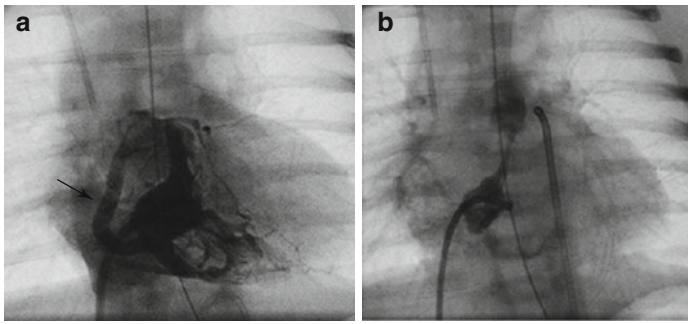

Fig. 19.6 Spasm of RV and RVOT following successful RFV-BD. (**a**) Initial RV angiogram in AP projection. The infundibulum is well developed, but the RV is essentially 'bipartite', with heavy trabeculations of the apical part. A major RV-coronary connection to the RCA (*arrow*) and minor connection to the LCA. (**b**) Post balloon dilation the patient became more hypoxic. RV angiogram in AP projection showing RV spasm with a smaller inlet and infundibulum and loss of intertrabecular spaces in the apex. The PDA was immediately stented (*RFV-BD* radiofrequency valvotomy and balloon dilation, *RVOT* right ventricular outflow tract, *RV* right ventricle, *AP* anteroposterior, *RCA* right coronary artery, *LCA* left coronary artery, *PDA* patent ductus arteriosus)

haemopericardium in a neonate may lead to more inadvertent punctures of the RV.

(ii) Following valvotomy balloon dilation, the RVOT and RV may go into spasm, causing worsening of hypoxia and haemodynamic instability after an apparently successful procedure, i.e. the well-recognized phenomenon of 'the suicidal RV' (Fig. 19.6). Prostaglandin (PG) should be restarted. If oxygenation remains poor, the PDA should be stented.

(iii) If valvotomy and balloon dilation are 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 atresia of the coronary arteries.

(iv) 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.
(v) Severe or significant pulmonary regurgitation may occur following balloon dilation. This, combined with severe tricuspid regurgitation, a large PDA and PFO, may lead to a 'circular shunt' causing severe cyanosis. This requires surgical repair of the tricuspid ± pulmonary valves and reduction of the interatrial communication, though likely to be of high risk in a sick neonate.

19.9 Post Procedure

The patient is transferred to the ICU for monitoring of arterial blood pressure and blood gases. If the patient remains haemodynamically 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 may require ductal stenting or BT shunt. Routine echocardiogram is performed the next day to assess any residual gradient across RVOT and to exclude pericardial effusion.

19.10 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

interatrial communication are the main areas of focus. Residual obstruction which could be valvar or subvalvar may increase with time requiring re-intervention, either by 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 term.

Patients with borderline RV size need close monitoring for growth of the RV and level of cyanosis. They may be candidates for bidirectional Glenn shunt to off-load the RV (1½-ventricle circulation).

In early adult life, tricuspid and pulmonary regurgitation may require surgical intervention in patients who have done well following the initial catheter intervention.

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Chapter 20
Percutaneous Transcatheter Balloon Mitral Commissurotomy

Raghavan Subramanyan and Anuradha Sridhar

20.1 Introduction

Mitral valve (MV) dilatation by balloon catheter also known as percutaneous transcatheter balloon mitral commissurotomy (PTMC) is now recognized as the treatment of choice to relieve rheumatic mitral stenosis (MS) in patients with suitable valve anatomy. PTMC was first developed by Dr. Kanji Inoue in 1982. Since then, a large worldwide experience has accumulated showing both immediate and sustained improvement. This chapter describes

R. Subramanyan (✉)
Department of Cardiology, Frontier Lifeline Hospital,
R-30-C, Ambattur Industrial Estate Road,
Chennai, Tamil Nadu 600101, India
e-mail: cardio@frontierlifeline.com

A. Sridhar
Department of Pediatric Cardiology, Frontier Lifeline Hospital,
R-30-C, Ambattur Industrial Estate Road,
Chennai, Tamil Nadu 600101, India
e-mail: cardio@frontierlifeline.com

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a practical approach to patient selection, workup, technique, materials, limitations, and complications in patients undergoing PTMC.

20.2 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 transmitral 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 valve area 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.

20.3 Indications and Patient Selection

Selection of a patient for PTMC is based upon both clinical and anatomical factors (Table 20.1). It is usually indicated in a symptomatic patient with MVA < 1.5 cm2 and favorable valve

Table 20.1 Indications for PTMC

Indication	Class
Symptomatic patients with favorable characteristics[a] for PTMC	I
Symptomatic patients with contraindication or high risk for surgery	I
As initial treatment in symptomatic patients with unfavorable anatomy but without unfavorable clinical characteristics[a]	IIa
Asymptomatic patients without unfavorable characteristics[a] and high thromboembolic risk	IIa
Previous history of embolism	
Dense spontaneous contrast in the left atrium	
Recent or paroxysmal atrial fibrillation	
Asymptomatic patients without unfavorable characteristics[a] and high risk of hemodynamic decompensation	IIa
Systolic pulmonary pressure > 50 mmHg at rest	
Need for major noncardiac surgery	
Desire of pregnancy	

Adapted from Vahanian et al. [1]. Table 14, Eur Heart J 33:2476 (with permission from Oxford University Press)

[a]Unfavorable characteristics: advanced age, previous valvotomy, class IV symptoms, atrial fibrillation, severe pulmonary hypertension, echocardiographic MV score >8, MV calcification, very severe MS, or severe tricuspid regurgitation

anatomy [1]. In general, PTMC is contraindicated if there is commissural calcification, more than mild mitral regurgitation, LA thrombus, or an associated valvular or coronary disease which will benefit from surgery. PTMC is also not advisable if the predominant obstruction is due to subvalvular thickening rather than due to commissural fusion.

20.4 Pre-procedural Evaluation

Transthoracic 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. A semiquantitative score of the MV pathology has been used to predict the efficacy of PTMC and select the appropriate patient (Table 20.2). A score of less than 9 with not more than mild MR is considered most suitable for PTMC [2]. 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 an option if the patient is a high-risk candidate for OMC. The MV morphology score however does not quantify commissural fusion or calcification. As splitting of the fused commissure is essential for relief of MS, commissural fusion and calcification must be studied in detail.

Transesophageal 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 suboptimal TTE images.

20.5 Balloon Catheters for PTMC

Inoue's method using an inflatable nylon-latex balloon catheter (NLBC) is the most widely used technique and is described in detail below. Two types of NLBC are currently 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 dumbbell shape centered in the narrowest part of the mitral valve and expand to the desired diameter (Fig. 20.1). The differences between Inoue and Accura balloon catheters are shown in Table 20.3.

Table 20.2 Mitral valve morphology score by echocardiography

Grade	Mobility	Subvalvular thickening	Leaflet thickening	Calcification
1	Highly mobile valve with only leaflet tips restricted	Minimal thickening just below the mitral leaflets	Leaflets near normal in thickness (4–5 mm)	A single area of increased echo brightness
2	Leaflet mid- and base portions have normal mobility	Thickening of chordal structures extending to one-third of the chordal length	Mid-leaflets normal, considerable thickening of margins (5–8 mm)	Scattered areas of brightness confined to leaflet margins
3	Valve continues to move forward in diastole, mainly from the base	Thickening extended to distal third of the chords	Thickening extending through entire leaflet (5–8 mm)	Brightness extending into midportions of the leaflets
4	No or minimal forward movement of the leaflets in diastole	Extensive thickening and shortening of all chordal structures extending down to the papillary muscles	Considerable thickening of all leaflet tissue (>8–10 mm)	Extensive brightness throughout much of leaflet tissue

Adapted from Wilkins et al. [5], Table 2, p 300 (with permission from BMJ Publishing Group Ltd)

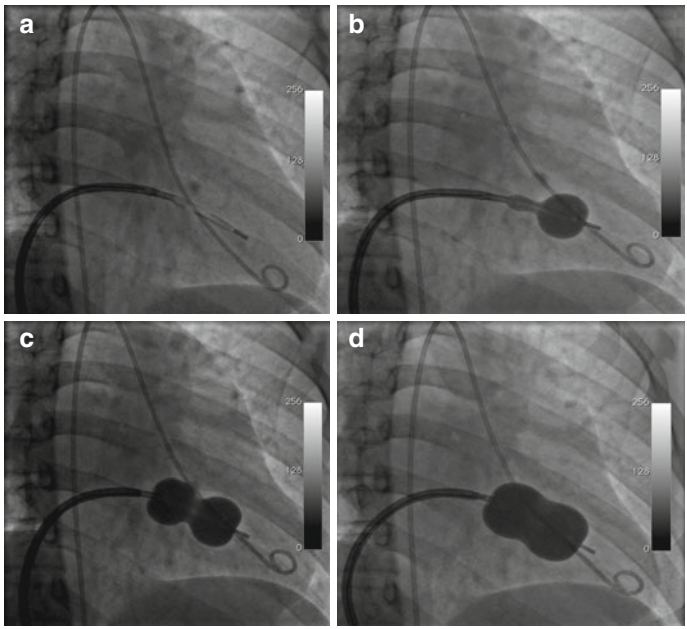

Fig. 20.1 Fluoroscopy in right anterior oblique view: (**a**) ideal position of the balloon catheter on entry pointing toward the LV apex; (**b**) and (**c**) inflation of the distal balloon followed by positioning across the mitral valve and inflation of the proximal balloon; (**d**) full inflation and dilatation of the valve orifice to the desired diameter

20.6 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, transseptal puncture is done observing usual precautions. Too low a puncture can make subsequent entry into the MV

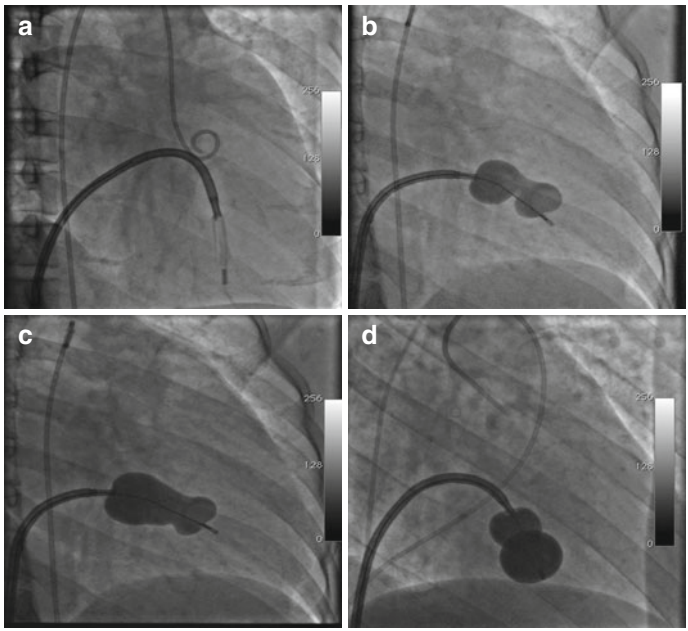

Fig. 20.2 Fluoroscopy in right anterior oblique view: (**a**) improper position of the balloon catheter pointing inferiorly and posteriorly into the chordae; (**b**) and (**c**) distortion of the distal and proximal parts of the balloon by subvalvular apparatus; (**d**) both distal and proximal parts of the balloon inflated in the LV below the valve orifice. These clues indicate entrapment of the balloon catheter

difficult and must be avoided. Heparin (5,000–10,000 units) is administered once the LA is entered. The LA and LV pressures are recorded simultaneously for the gradient (Figs. 20.3a).

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-French long stiff dilator.

Table 20.3 Differences between Accura and Inoue balloon catheters

Feature	Inoue catheter	Accura catheter
Construction	Triple lumen	Double lumen
Shaft size	12 French	11 French
Contrast dilution recommended	1:4	1.6–1.8
Blood seepage between layers	Possible	No
Prevention of deflation failure	Yes	No
Vent holes	Yes ($n=2$)	No
Vent tube	Yes	No
Balloon size achievable	+ 4 mm (22–26 mm)	+ 3 mm (23–26 mm)
Cost	More	Less

Adapted from Harikrishnan et al. [6], Table 22–4, p 225 (with permission from Jaypee Brothers Medical Publishers (P) Ltd)

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 (cms)/10), although the relationship is not linear. A slightly more objective method measures the minimum MV annulus diameter on TTE in apical 4-chamber or 2-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. An 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

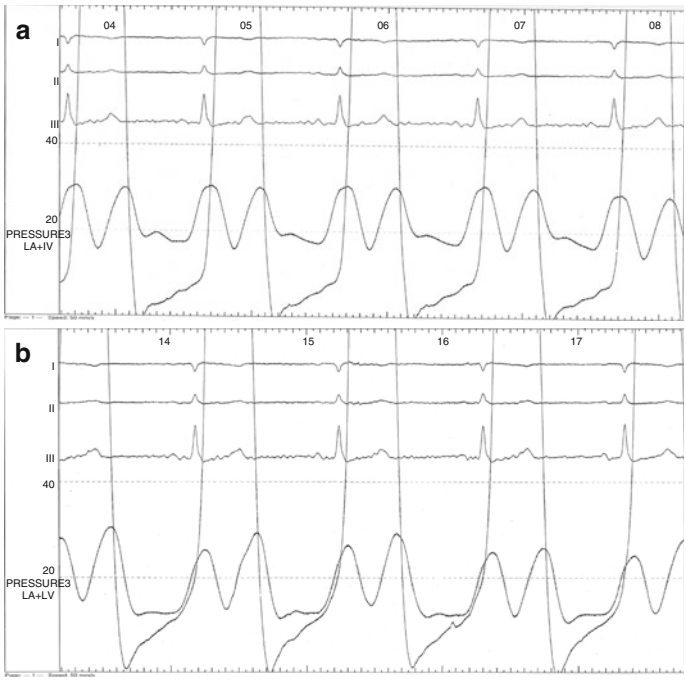

Fig. 20.3 Simultaneous LA and LV pressure tracings before (**a**) and after PTMC (**b**), showing abolition of the end-diastolic gradient and marked reduction in the mean diastolic gradient across mitral valve with successful PTMC (Pressure recording: courtesy Dr. Rajagopal S)

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 dumbbell shape of the balloon ensures a stable position in the orifice and avoids inflation in the subvalvular area (Fig. 20.1).

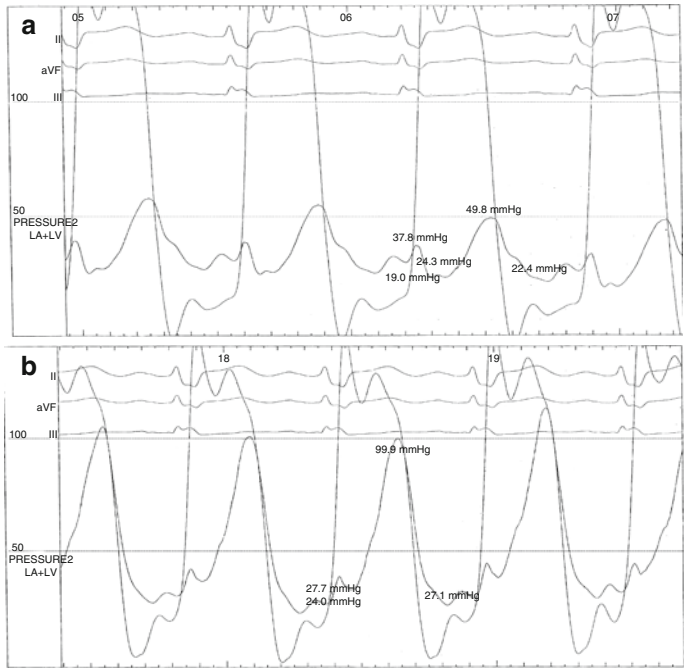

Fig. 20.4 Simultaneous LA and LV pressure tracings before (**a**) and after PTMC (**b**), showing reduction in the diastolic gradient across mitral valve. However, the high LA pressure and large V waves indicate severe acute mitral regurgitation (Pressure recording: courtesy Dr. Rajagopal S)

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. 20.3 and 20.4).
8. Subsequent dilatations are done with 1-mm increments. The desired endpoint may be one or more of the following without worsening of MR: (1) abolishment of the gradient

by pressure measurement, (2) one or both commissures are fully opened, (3) two-dimensional MVA 1.50 cms or greater. Increase in MR to more than mild grade is a definite endpoint to avoid catastrophic MR.

9. A LV angiogram can be 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 avoid trauma to the tissues.

20.7 Other Methods 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 either over two wires or in a monorail fashion over a single guidewire (MULTI-TRACK system, NuMED Co). The retrograde transarterial 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 reusable, 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.

20.8 PTMC in Special Situations

Pregnant Patients with MS: Pregnant patients with 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. It should be performed by experienced operators in the second or third trimester with minimal radiation and proper shielding.

Mitral Restenosis: 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 restenosis are similar to de novo MS.

Juvenile Rheumatic MS: PTMC can be safely performed in children with rheumatic MS with excellent immediate and follow-up results. The indications are similar to that 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 a 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 interatrial septum is advisable in this approach to achieve a more direct course toward the mitral orifice.

20.9 Tips and Tricks for Successful PTMC

The key factors for a safe and successful PTMC are awareness of pitfalls, meticulous technique, and cumulative experience. The following tips and tricks have been found useful by experienced operators:

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, although the procedure has been performed through the internal jugular vein in unusual cases with iliofemoral venous or inferior vena caval occlusion. The entry site into the femoral vein should be clearly > 2–3 cms below the inguinal ligament to allow easy passage of the stiff dilator and catheter.
3. The atrial septum is ideally punctured in the fossa ovalis, avoiding the muscular upper part of the septum. A high location of the septal entry 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. Fluoroscopy in two planes and continuous pressure monitoring are recommended for transseptal catheterization. In difficult cases either TEE or intracardiac echocardiography can be used to guide septal puncture.
4. Avoid entrapment in the pulmonary vein or in the interatrial septum. This is best achieved 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 on the spring stylet. It also avoids entry into the left atrial appendage which is the usual site for LA thrombus.

5. Entry into LV: The most common method to find the MV orifice with the tip of the NLBC is to gradually withdraw it so as to straighten its curved course in the LA and orient it along the MV to apex axis. This is best done with fluoroscopy in the right anterior oblique view (Fig. 20.1). 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 difficult cases especially with a 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. Another method is to place the guidewire in the LV and then advance the NLBC over the wire. This method is particularly useful if the LA is much dilated or the septum is markedly bulging and distorted.
6. Avoid MV trauma: 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 right anterior oblique view. The NLBC should form a smooth curve and point toward the

apex (Fig. 20.1a). 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. 20.2a). The next step is to move the NLBC back and forth on the stylet indicating 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.

7. 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. 20.2b, c and d) may indicate entrapment in the mitral chordae and is more likely to occur with severe subvalvular pathology.
8. Avoid damage to the interatrial septum by stretching the NLBC on both introduction and withdrawal and making a gentle loop 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.
9. PTMC must be monitored by echocardiography and by pressure gradient measurement to know the endpoint and for early recognition of complications such as MR or perforation (Fig. 20.4).

20.10 Expected Results

Immediate hemodynamic improvement indicates successful valvotomy and is assessed during PTMC by mean LA pressure, pressure gradient across mitral valve, and severity of MR. 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, or baseline mitral regurgitation are predictors for poor immediate outcome [1].

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 valve area or loss of the initial gain in area on late follow-up. At 10 years event-free survival has been reported to be about 79 % in younger patients [3] and 56 % in slightly older patients [4]. The MV characteristics also affect the late results of PTMC. Among younger subjects with a 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) [3].

20.11 Complications and Management

Significant complications or failed valvotomy have been reported in 1.0–15.0 % patients [1]. 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 interatrial septum puncture or during manipulation of the balloon catheter. Management consists of protamine administration, volume expansion, and, if necessary,

pericardiocentesis and surgical repair. Local vascular site complications, embolic complications, or death are uncommon (0.5–5.0 %). Major complications and sequelae are more common among patients who had inadequate immediate result of PTMC.

20.12 Summary of Critical Points for Safe and Successful PTMC

- Careful evaluation and selection of case
- Choosing the appropriate balloon diameter
- Stepwise dilatation with 1-mm increment in diameter
- Defining endpoints: abolition of gradient, valve area (>1.50 cms^2), or worsening MR
- Careful watch for complications
- No substitute for meticulous technique and experience

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Part V
Step-by-Step Procedures: Vessel Treatment

Chapter 21
Pulmonary Artery Branches

Peter Ewert and Andreas Eicken

21.1 Anatomic Description and Physiopathology

- Normal pulmonary vessels have an adequate caliber and an appropriate distensibility and they provide active support to forward flow by their windkessel function. Pathologic vessels can be either stenotic on different levels of the vessel tree, they can be hypoplastic in segments or in total, or finally vessels can be compressed by other structures like the aorta or bronchi.
- Hypoplasia can be due to flow restriction by a localized stenosis or can be due to an intrinsic disorder like Williams-Beuren syndrome (WBS).
- A specially generalized vessel abnormality is present, whenever an aortopulmonary collateral has been unifocalized to the pulmonary vessel tree. From its embryologic origin, these collaterals are more systemic and not pulmonary arteries.

P. Ewert • A. Eicken (✉)
Pediatric Cardiology and Congenital Heart Defects, Klinik für Kinderkardiologie und angeborene Herzfehler, Deutsches Herzzentrum München, Lazarettstr. 36, München, D-80636, Germany
e-mail: ewert@dhm.mhn.de; eicken@dhm.mhn.de

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- In many patients the obstructions in the pulmonary vessels are partially a consequence of surgical or – less often – transcatheter procedures and are caused by the formation of scar tissue or restrictions due to small conduits or stents.

21.2 Clinical Scenarios

- Pulmonary arteries are involved in a huge variety of CHD.
- In the clinical setting, almost always stenotic or hypoplastic vessels are a problem leading to a pressure burden of the corresponding ventricle, which in most of the cases is an anatomic right ventricle.
- Stenosis of the bifurcation – either native or after surgery – is associated with tetralogy of Fallot (TOF), PA with VSD, d-TGA after arterial switch operation, and Williams-Beuren syndrome (WBS).
- In patients after Norwood stage 1 or Damus-Kaye-Stansel operations, stenosis of the central pulmonary arteries behind the neo-aorta can occur, since the pulmonary arteries have to be reconstructed after separation from the pulmonary trunk and excision of ductal tissue.
- A special feature is the coarctation of the pulmonary artery, which is caused in analogy to aortic coarctation by ductal tissue spreading to the pulmonary artery. When the duct constricts, the contracting tissue in the pulmonary vessel produces a typical circumscript stenosis.
- On the other hand, in general, the older the patients, the higher is the likelihood that pulmonary bifurcation stenoses are residua after surgery or intervention.
- *Stenosis of pulmonary arteries distally of the bifurcation.*
  - While in some forms of WBS or TOF a generalized hypoplasia of the pulmonary vessels is present, there are clinical settings, where multiple stenoses are localized

preferably wherever the vessels branch into smaller segments.
- Despite the aspect of multiple stenosis, in most cases these visible restrictions indicate a generalized arteriopathy.
- If smallest arteries are affected, stenoses are difficult to visualize, and the transition into pulmonary artery hypertension is fluent.
- These forms are not treatable by transcatheter means.

21.3 Indications and Patient Selection

Most indications for transcatheter treatment of pulmonary arteries are stenotic vessels. The indications, however, are very variable dependent on the sort of heart defect, the specific morphology and etiology, the technical possibilities, the age, and the individual overall situation of the patient.

21.4 Pre-procedural Imaging

- Echocardiography is the most important imaging modality in congenital heart disease.
- For the direct visualization of pulmonary stenosis, however, it is often only of limited use, since peripheral stenosis cannot be seen.
- Despite these restrictions, echocardiography can detect pulmonary stenosis at the bifurcation and helps to quantify the pressure burden and the function of the subpulmonary ventricle; thus, it is a valuable screening tool.
- MRI can visualize the whole pulmonary artery tree, offers quantification of ventricular function, and has the advantage

of selective flow quantification of the right and left pulmonary artery. By the analysis of flow velocities in front and behind the stents, MRI can indirectly detect possible in-stent stenosis.
- A CT scan offers a high-resolution image of the pulmonary vessel and can be performed also in the presence of a pacemaker.
- Sometimes rotational angiography may be helpful to find the best angle for visualization of the target region.

21.5 Technique (Step by Step)

- With the help of other imaging modalities (MRI, CT), angiographies should be performed to visualize the target region in the best way, i.e., with only minimal foreshortening and, if ever possible, in a biplane approach with perpendicular angulations.
- After crossing the lesion with wire and catheter, pressure measurements across the stenosis can be performed, but are not mandatory. If the aim of the procedure is an improvement of flow distribution, like in isolated branch stenosis without markedly elevated right ventricular pressure, or the intervention yields toward optimizing the morphology of the pulmonary arteries in order to establish optimal laminar flow in patients with Fontan circulation, pressure measurements might be dispensable.
- If necessary, selective angiographies are helpful for the fine-tuning of projections and visualization of the lesion as preparation for optimal vessel measurements.
- Meticulous measurements of the vessel diameter and the length of the stenosis can then be performed.
- Now all relevant informations about the patient are complete, and a decision how to treat the lesion has to be made.

21.6 Treatment Options

Transcatheter methods to treat pulmonary arteries are *balloon dilation,* the use of *cutting balloons*, and *stent implantation*. In atretic vessels, recanalization by wires or radiofrequency perforation might be an option as well.

21.6.1 Balloon Dilation

- Historically, the balloon dilation of pulmonary stenosis was the first possible option.
- Balloon dilation is relatively easy to perform. It needs the smallest vascular access, and the insertion of a short sheath is sufficient.
- Nowadays, however, the role of balloon dilation is limited: if it is done with balloons only slightly larger in diameter as the treated vessel, the stenosis might be dilated but without permanent effect.
- On the other hand, when considerably larger balloons were used, the chance of effective stenosis reduction is higher, but the rate of complications like dissection or vessel rupture increases as well. Thus, cutting balloons and stents offer better possibilities for effective treatment with less risk.

21.6.2 Cutting Balloon

- A cutting balloon is highly effective in the dilation of even extremely firm stenosis.
- It should be considered as first choice, whenever there is doubt, whether a conventional dilation balloon will not be able to break the stenosis.
- Cutting balloons may also be used for pre-dilation before consecutive stent implantation.

- Its relative disadvantage is that it has to be used with a long sheath of considerable diameter to avoid entanglement or damage of valves, vessels, or of the blades from the balloon itself during withdrawal. In particular, to avoid this problem, it is mandatory to avoid resheating of the cutting balloon at an angle and avoiding any excessive pulling if any resistance is felt.
- Cutting balloons are available only in relatively small diameters (maximum 8 mm).

21.6.3 Stent

- Balloon-expandable stents provide the best immediate result with respect to gaining diameter, lack of recoil, safety from dissection, or vessel rupture.
- An additional option is the availability of covered stents.
- Their implantation might be technically challenging. Larger sheaths are mandatory and are not always easy to place over the stenosis.
- Premounted stents might be advantageous, if available in the desired length on the appropriate balloon.
- Open-cell design stents (Max/Mega LD, Andra) are stent constructions wherein some or all internal inflection points of the structural members are not connected by bridging elements. In contrast to that are closed-cell design stents (CP Stent) which show less longitudinal flexibility. During balloon expansion, open-cell design stents show less foreshortening than closed-cell design stents.
- Stents might exclude branch arteries by squeezing their orifices.
- Small stents might cause restenosis by tissue ingrowth.
- In general: the larger the vessel, the better the result with stents.
- In small vessels, cutting balloons is a good choice.
- The classic balloon dilation can be used as an adjunct for depicting the sort of stenosis.
- One very important issue in the use of stents in infancy and childhood is the *need of re-dilation* during somatic growth.

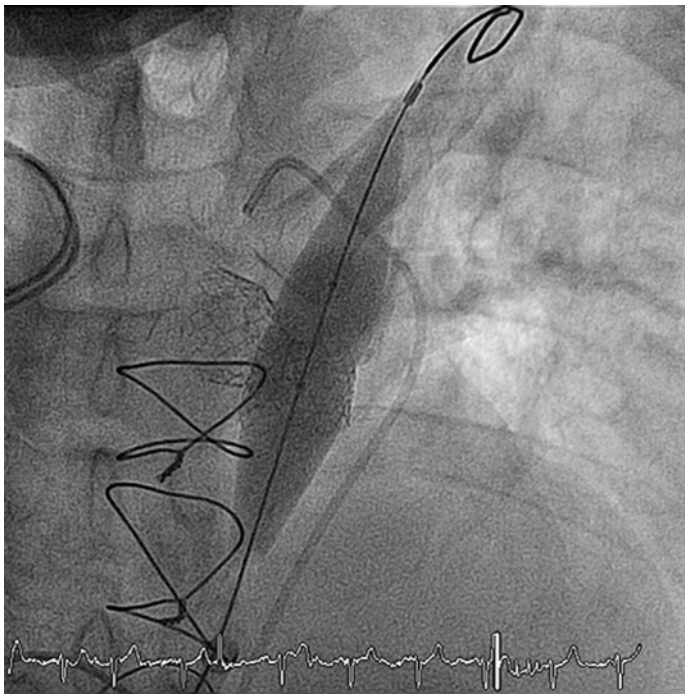

Fig. 21.1 After deliberate overstenting of the left pulmonary artery, an ultrahigh-pressure balloon was inserted across the struts and dilated to its nominal diameter, breaking the stent struts at about 8 atm dilation pressure to achieve unrestricted access to the left pulmonary artery

Thus, whenever possible, only those stents should be implanted which can be re-dilated to diameters appropriate for an adult-sized vessel. Small stents rarely have these properties, because they are not intended for this purpose.

- An interesting exception, beside experimental stent designs like the growth stent or the baby stent, is the Formula stent from Cook. With ultrahigh-pressure balloons, the stent cells can be opened and thus give way for the implantation of large stents (Fig. 21.1).

- A special issue is the *treatment of bilateral bifurcation stenosis*. To overcome the problem of obstructing the contralateral vessel by stent implantation, a simultaneous implantation of two stents in each side can be performed.
  - The disadvantage is, however, that the creation of a double lumen in the pulmonary trunk might be an obstacle for interventional treatments like pulmonary valve implantation.
  - An alternative concept is the creation of a Y-stent, in which two stents were placed through their meshes directly into the bifurcation (Fig. 21.2).
  - For this indication, preferably large open-cell design stents are used. Free access without jailing of a vessel is possible because stent struts can be cracked open by ultrahigh-pressure balloons at about 9–14 atm – a fact which has considerably lowered the threshold to overstent a pulmonary branch.

21.7 Materials

- After angiographic depiction and exact measurements of the target lesion, a distal guide wire position has to be achieved.
- This is done with an end open catheter (e.g., Arrow wedge catheter 4–7 F, Judkins right coronary catheter, multipurpose catheter).
- Balloon catheters are less prone to get caught in tricuspid valve chordae on their way up to the pulmonary arteries.
- Terumo guide wires (0.018, 0.025, 0.035) straight or curved can be very helpful to reach this distal position.
- In infants and small children, a coronary guide wire (e.g., BMW universal guide wire (Abbot) 0.014) may be helpful.
- This wire is then exchanged for a stiffer guide wire (Amplatz extra or ultra-stiff wire (Cook medical) 0.035/0.025); in

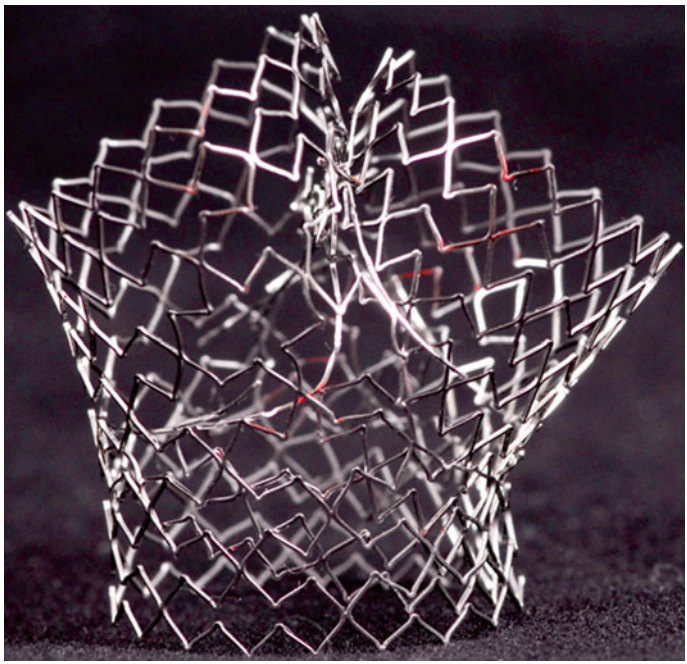

Fig. 21.2 A Y-stent, created from two balloon-expandable stents, which were placed across the struts of each other and consecutively dilated with an ultrahigh-pressure balloon (see Fig. 21.1) to achieve unrestricted flow and access into both branches

adults and adolescents even a stronger wire may be needed (Meier wire Boston Scientific 0.035, Lunderquist Cook medical 0.035). The Meier wire has a rather long soft tip which is shorter in the Lunderquist wire.

- The Multitrack catheter (PFM) may be used for pullback assessment and for angiograms without losing guide wire position.
- If only a balloon angioplasty is planned, a short sheath of adequate size is sufficient.

- For stent placement and cutting balloons, a long sheath needs to be placed across the target lesion.
- Stents augment the external balloon diameter by 1 or 2 French sizes depending on the stent type.
- Typically used *sheaths* are Flexor (Cook) 5 F–10 F (70–80 cm), Arrow-Flex (Arrow) 6 F–11 F (45–80 cm), and Mullins Sheath straight and curved (Cook) (5–22 F)(63–85 cm); coronary guiding catheters (5–8 F) may also be useful.
- Common *balloons* for angioplasty and stent deployment are Powerflex (Cordis) (6–12 mm; 2–4 cm), VACS (Osypka) (9–30 mm, 2–6 cm), and Z-med/BiB (NuMed)(10–30; 2.5–6 cm).
  - For resistant lesions, ultrahigh-pressure Kevlar balloon are used: Dorado (Bard) (8–10 mm 2 cm)/Atlas Gold (Bard)(12–24 mm, 2–4 cm) or the Mullins (NuMed) (12–25 mm; 4 cm) balloons are available.
- Various *stents* (usually balloon expandable, premounted, or hand crimped) are used.
  - For stenting of segmental pulmonary arterial stenosis, premounted coronary bare metal stents can be implanted. These small stents can only be enlarged by 1 or 2 mm and hence cannot reach large diameters.
  - Premounted stents at medium size are, for example, the Cordis Palmaz stents on Opta balloon (on 5–10 nm balloons). These stents can be dilated up to 12 mm.
  - The Cook formula stent is a rather new premounted stent crimped on a 6, 8, and 10 mm balloon. The largest stents can be dilated up to 16 mm. It offers, however, the possibility to reach unlimited diameters by overdilation with ultrahigh-pressure balloons. The stent struts break away; thus, the stent is no longer restrictive (Fig. 21.3).
  - Other frequently used open-cell design stents are EV3 Mega LD Stents (length 16 and 26 mm), the EV3 Max LD Stents (16 and 26 mm), and the AndraStents (Andramed

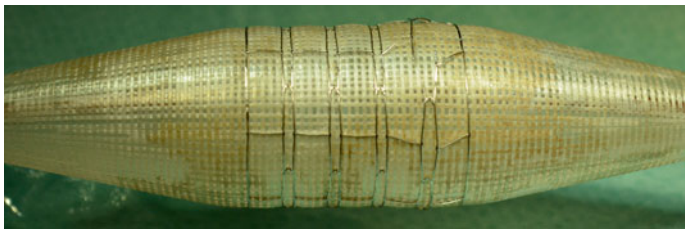

Fig. 21.3 A Cook Formula stent dilated on a 18 mm ultrahigh-pressure balloon. The stent struts begin to rupture and, thus, give way to unrestricted further dilation

xl 13 and 17 and xxl 21, 26, 39, 48, and 57 mm). The Andramed xxl open-cell design stents can be dilated to a diameter >25 mm without severe foreshortening.

– Finally, the closed-cell design 8 zig CP stent family is frequently used (bare or covered with ePTFE). They come in various lengths up to 45 mm and can be expanded to 24 mm. If expanded to larger diameters, they shorten significantly.

21.8 Expected Results

- In isolated circumscript 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. Since in these cases even very small vessels are involved, cutting balloon dilation and stent implantation techniques can be combined. The overall result, however, is variable. Assessment of a beneficial therapeutical 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 after unifocalization.
- If unobstructed flow is directed to a small pulmonary bed with normal vasculature, pulmonary hypertension may result in vascular damage due to pressure and volume overload.
- In generalized pulmonary arteriopathy with stiff and hypoplastic vessels, transcatheter interventions are of very limited effect.

21.9 Tips and Tricks

- *Challenging anatomies*
  In smaller patients, it is sometimes especially 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.

  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 of help to bring the stent into position.

- *Choosing the right balloon diameter*
  Care has to be taken to avoid undersizing of balloons when the vessels do pump during the cardiac cycle. Best is orientating on 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.

21.10 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.

21.11 Complications and Their Management

21.11.1 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.

21.11.2 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.

21.11.3 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.

21.12 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.

21.13 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.

Chapter 22
Aortic Coarctation

Raul Ivo Rossi Filho and João Luiz Langer Manica

22.1 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.

R.I.R. Filho • J.L.L. Manica (✉)
Department of Interventions for Congenital Heart Disease,
Hemodynamics Laboratory, Instituto de Cardiologia do Rio Grande do Sul,
395 Princesa Isabel Ave., Porto Alegre, CEP 90620-001, Brazil
e-mail: jocamanica@yahoo.com.br

G. Butera et al. (eds.), *Cardiac Catheterization for Congenital Heart Disease: From Fetal Life to Adulthood*,
DOI 10.1007/978-88-470-5681-7_22,

22.2 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 also can be found in routine medical screening in 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.

22.3 Indications for Treatment of Aortic Coarctation

Systemic hypertension with resting pressure gradient between upper and lower limbs greater than 20 millimeters of mercury (mmHg), which may be associated with:

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:

    - Abnormal blood pressure response to exercise
    - Left ventricular dysfunction
    - Symptoms of exercise intolerance
    - Associated lesions such as coronary artery disease and aortic insufficiency
    - Exercise gradient of more than 20 mmHg
    - Presence of left ventricular hypertrophy and/or left ventricular diastolic dysfunction

22.4 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 (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.

22.5 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.

22.6 Balloon Angioplasty

Initial reports on balloon angioplasty demonstrated good results in patients that previously underwent surgical repair, despite of 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.

22.6.1 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.

22.6.2 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 (e.g., 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 are performed using any of the previously

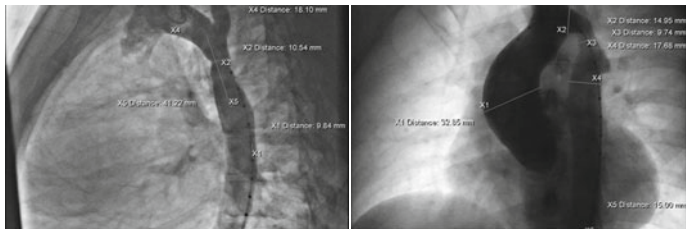

Fig. 22.1 Aortic coarctation in LAT and AP projections

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. 22.1).

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 and minimum diameter of the coarctation, aorta below the coarctation, and the descending aorta at the level of diaphragm.
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:
    - Balloon diameter should not exceed the diameter of the aorta above the coarctation or at the level of the diaphragm.
    - It should not also exceed three times the diameter of the coarctation.
    - 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.

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 10–15 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 anatomic result of dilation. Measure the diameter of coarctation.
18. Repeat the hemodynamic measurements – pressures in the ascending and the descending aorta with pullback method.

22.6.3 Expected Results

1. Systolic pressure gradient less than 10 mmHg.
2. Increase diameter of aorta at the level of the coarctation.

22.6.4 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.

22.6.5 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.

22.6.6 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.

22.6.7 Main Complications

1. Aortic wall dissection
   - 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.
   - Larger dissection – implantation of a bare or covered stent during the same procedure.
2. Small aneurysm
   - Repeat CT or magnetic resonance imaging scans to follow the progress of the aneurysm.
   - If necessary (when the diameter increases or there is a spiral aneurysm), implantation of a stent may be indicated.
3. Larger or increasing aneurysm
   - Immediate implantation of a covered stent.
4. Other complications include aortic rupture (emergency surgery or covered stent implantation) and femoral artery damage (thrombolysis or surgical repair).

22.6.8 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

22.7 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 continuing to provide beneficial effects at medium-term follow-up, mainly in patients weighing more than 20 kg [2].

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 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 bailout situation, associated dilation of the ascending aorta, and patients with an irregular aortic wall and those previously treated with the use of surgical patches [3]. Some authors recommend its use in all cases of adolescent and

adult with aortic coarctation or recoarctation, although this is not yet common practice.

There are two commercially available covered stents. The CP Stent (NuMed Inc., Hopkinton, NY, USA) is a regular bare CP Stent, which is involved with an expandable sleeve of e-PTFE. It is available in lengths of 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-, 14-, and 16-mm balloons and can be dilated to a maximal diameter of 22 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.

22.7.1 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

22.7.2 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-sized 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 one or two French larger than the sheath needed for the balloon when bare stents are implanted and three to four French larger for covered stents. Usually, long sheaths ranging from 8 F 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 (Fig. 22.2).

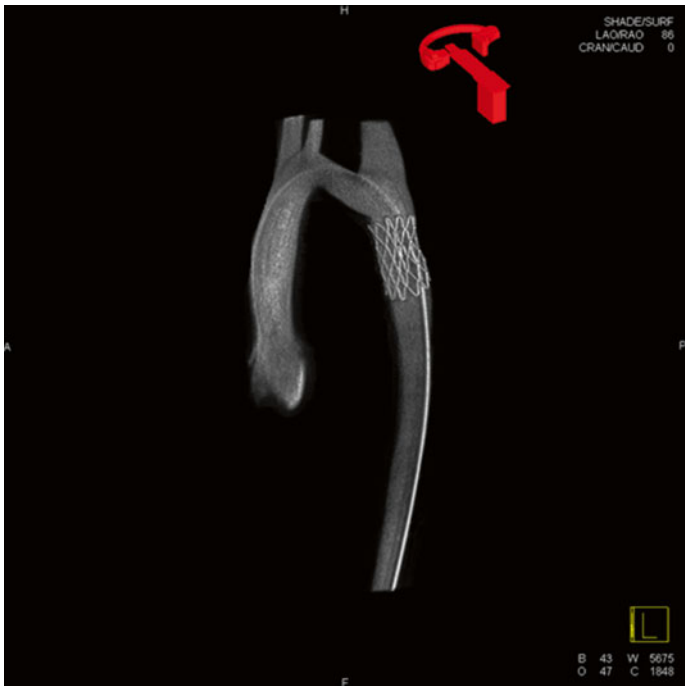

Fig. 22.2 Final result of a bare stent implantation with 3D-RA. 3D-RA: Three-dimensional rotational angiography

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 (cefazolin for 24 h).

22.7.3 Tips and Tricks

The key cornerstones of stent implantation in coarctation of the aorta are:

1. Preimplantation 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 also can 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 were 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. Don't 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 e-PTFE 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 e-PTFE 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 transseptal puncture.
6. In younger patients, surgical cutdown 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 (e.g., 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 if 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 forward.
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.

22.7.4 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).

22.7.5 Complications

22.7.5.1 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 bailout situations, an inflated balloon in the site of the injury can be a lifesaving approach while covered stent is prepared for implantation.

22.7.5.2 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.

22.7.5.3 Access-Related Complication

Acute arterial occlusion is a concern in patients in the first year of life.

Bleeding, local hematoma, and arterio-venous 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.

22.8 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 and somatic growth or, less frequently, due to neointimal hyperplasia.

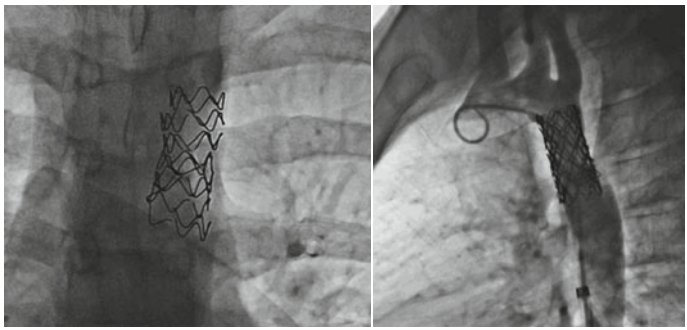

Fig. 22.3 (**a**) Type 2 Fractures in a previously implanted CP bare stent. (**b**) CP covered stent implanted within the fractured bare stent

22.9 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. 22.3).

22.10 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 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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Chapter 23
Reopening of Peripheral and Central Arteries and Veins

Henri Justino and Athar M. Qureshi

23.1 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.

H. Justino (✉) • A.M. Qureshi
Department of Pediatrics, Texas Children's Hospital,
6621 Fannin, MC 19345-C, Houston, TX 77030, USA
e-mail: hjustino@bcm.edu; axquresh@texaschildrenshospital.org

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23.2 Clinical Scenarios

The most common clinical scenarios leading to the consideration of preforming 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.

23.3 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 1,100 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.

23.4 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, consideration should be given to performing surgical repair if feasible, but catheter-based techniques may be a viable option in cases that are at high operative risk.

23.5 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, either in the radiology or echocardiography department. 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.

23.6 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

23.6.1 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:

- 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.
- 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.
- 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.
- 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.

23.6.2 Obtaining Vascular Access

Once the optimal access site is selected, vascular access may be obtained percutaneously using ultrasound guidance or "blindly" (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).

23.6.3 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 G 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, or ASAHI MiracleBros (Abbott, Abbott Park, IL). 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 6 F JR or multipurpose guide catheter). As a general rule, a given catheter requires a guide catheter of 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.

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, Bridgewater, NJ) 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).

23.6.4 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).

23.6.5 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, Bayer Healthcare, Indianola, PA or Pronto Catheter, Vascular Solutions, Minneapolis, MN) and various systems for mechanical thrombectomy and fragmentation, such as AngioJet (Bayer Healthcare) and Trellis (Covidien, Bacchus Vascular, Santa Clara, CA). In addition, acute thrombotic occlusions may necessitate infusion of thrombolytic agents such as tissue plasminogen activator (tPA).

23.6.6 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. For lesions measuring <8 mm in diameter, we often utilize cutting balloons to treat resistant lesions (Flextome, Boston Scientific). For lesions >8 mm in diameter (or for resistant lesions within previously placed stents), we resort to noncompliant 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. It is mandatory to eliminate the waist on a given balloon before proceeding to a larger diameter balloon. 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.

23.6.7 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. 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 inferior and superior venae cavae and central pulmonary arteries) and >22 mm for the 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, Covidien, Plymouth, MN) to facilitate enlargement of the cells jailing the ostium.

23.7 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 due to their length and complicated nature. 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.

23.8 Expected Results

Acute procedural success during recanalization procedures can be commonly achieved, as long as the operators are persistent and willing to change courses 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. Despite this, we commonly try to avoid stent placement in young children, knowing that a stent will commit the patient to numerous repeat procedures to redilate the stent. However, if avoidance of a stent at the initial procedure results in an early reocclusion or severe restenosis, we will generally implant a stent at the time of reintervention.

23.9 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 whereby the right common iliac artery compresses the left common iliac vein.

Lastly, perhaps the most important tip during 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. 23.1, 23.2, and 23.3).

23.10 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

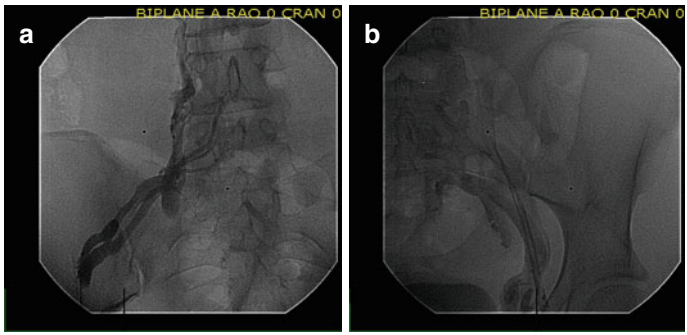

Fig. 23.1 Angiograms performed in the right femoral vein (**a**) and left femoral vein (**b**) show total occlusion of the common iliac veins bilaterally as well as of the infrarenal inferior vena cava, with numerous venous collaterals seen

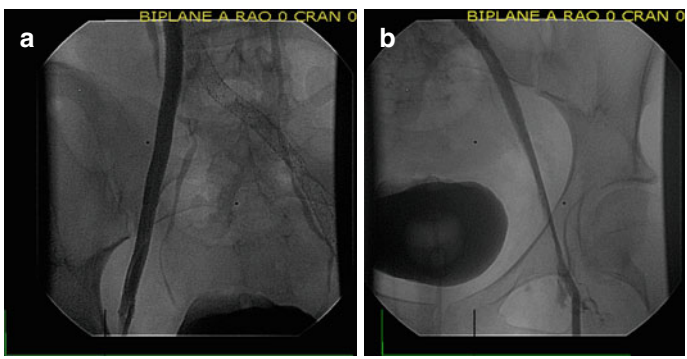

Fig. 23.2 Angiograms performed after placement of stents in the right (**a**) and left (**b**) external and common iliac veins, showing restored patency of these veins, with very little opacification of the collateral veins

procedure is extremely important in this regard. As stated above, 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.

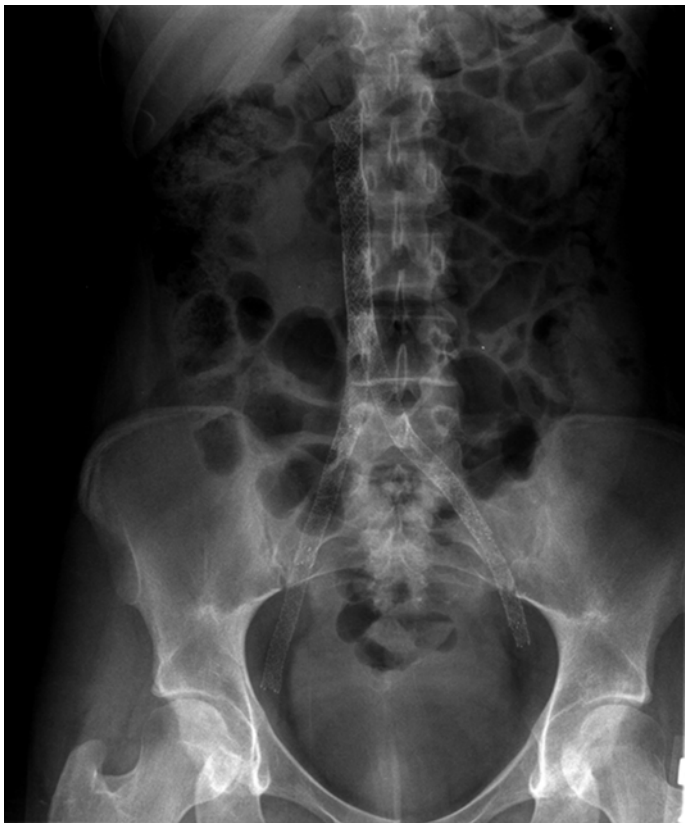

Fig. 23.3 Abdominal x-ray showing the configuration of the stents in the external and common iliac veins bilaterally entering jointly into stents placed in the infrarenal inferior vena cava

23.11 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, 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 cases such as these.

23.12 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. Placement of a covered stent may be necessary, or placement of an occlusion device at the side 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.

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.

23.13 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.

23.14 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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Chapter 24
PDA Stenting in Duct-Dependent Pulmonary Circulation

Kothandam Sivakumar

24.1 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 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. 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.

K. Sivakumar, MD, DM
Department of Pediatric Cardiology, The Madras Medical Mission, No 4A Dr J J Nagar, Mogappair, Chennai, Tamil Nadu 600037, India
e-mail: drkumarsiva@hotmail.com

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24.2 Anatomic Description and Physiopathology

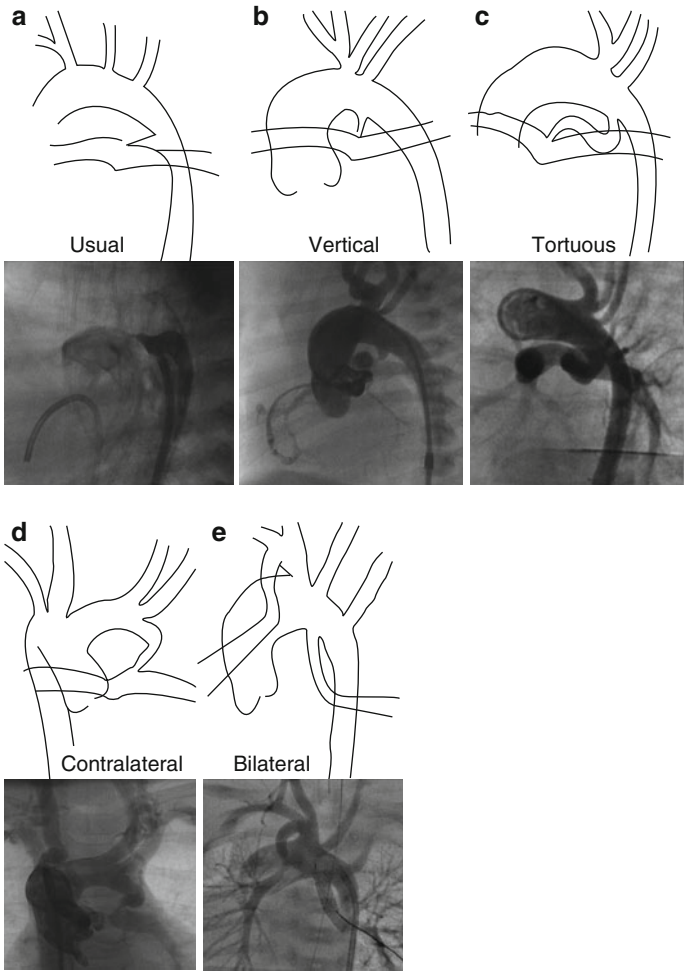
24.2.1 Anatomy of Ductus Arteriosus

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. 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. 24.1).

24.2.2 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

Fig. 24.1 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

days; anatomical closure with fibrous tissue occurs later. Duct closure in pulmonary atresia results in severe hypoxia. In a few patients, ducts remain patent for a few weeks or a few months to maintain pulmonary circulation.

24.2.3 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].

24.2.3.1 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*.

24.2.3.2 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 right ventricle-dependent coronary circulation, and complex heterotaxies. They are palliated later by bidirectional Glenn and Fontan surgeries.

24.2.3.3 Group C: Transient Inadequacy of Pulmonary Circulation

(i) 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
(ii) *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, and tricuspid valve dysplasia with regurgitation. In both of these groups, longer ductal patency is needed.

24.3 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 6th 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

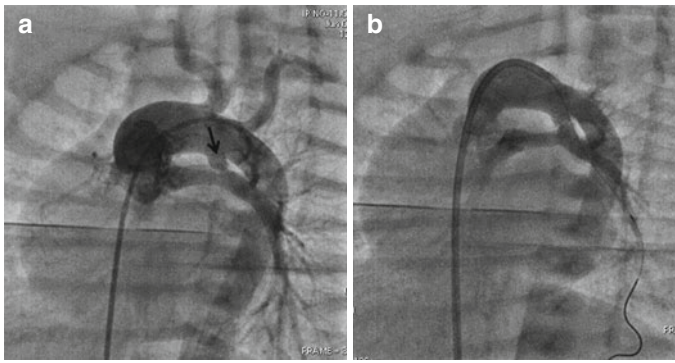


Fig. 24.2 Aortogram in shallow left anterior oblique view with a pigtail catheter advanced from femoral vein through the right ventricle into the left aortic arch (**a**) shows a vertical duct (*arrow*) arising from undersurface of the aortic arch opposite to the right innominate artery. In such cases, 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

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. 24.2). 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. 24.3). Elective conduit repair was done at 1 year of age.

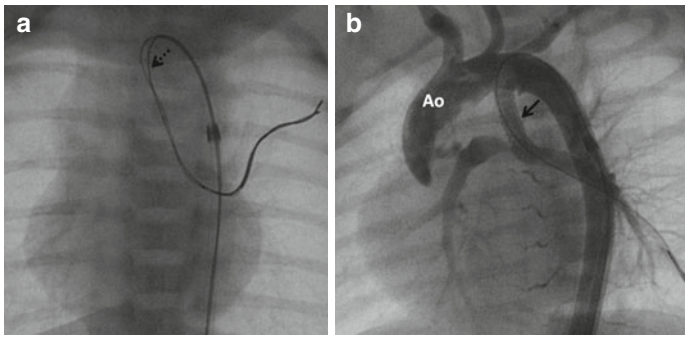


Fig. 24.3 Through a 4 F long sheath, a coronary guidewire was advanced (**a**) through a vertical duct. An additional buddy wire (*dotted arrow*) was advanced to facilitate the passage of the stent through the acute angulation of vertical duct. (**b**) After stenting, aortogram in shallow left anterior 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. 24.4). 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

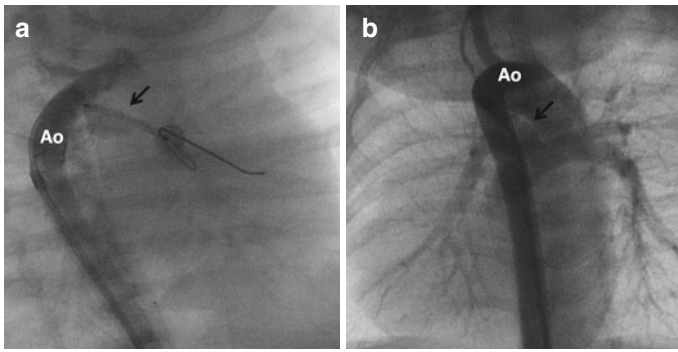


Fig. 24.4 Angiogram through a 4 F long sheath (**a**) in shallow right anterior oblique projection was done to check the position of a 4 mm stent (*arrow*) placed on a coronary guidewire through the duct advanced into the left pulmonary artery. After stenting, a repeat aortogram (**b**) showed good filling of the pulmonary arteries through the stented duct (*arrow*)

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 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. 24.5). 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. 24.6). DS with a 4 mm coronary stent was done from the femoral artery.

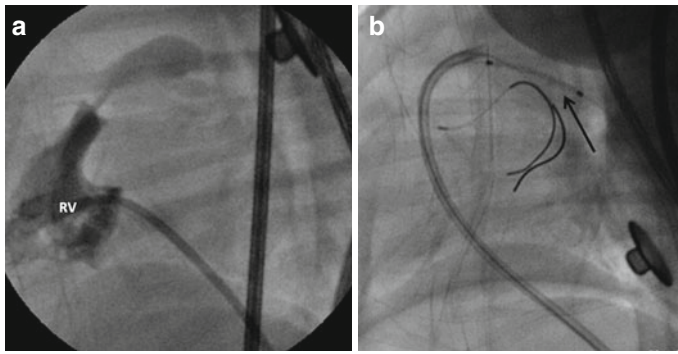


Fig. 24.5 Right ventricular (*RV*) angiogram in lateral view (**a**) demonstrates thick pulmonary valve with a narrow contrast jet into the pulmonary artery indicating severe stenosis which was dilated with a balloon. The persistent hypoxia due to inadequate antegrade pulmonary flows was an indication 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

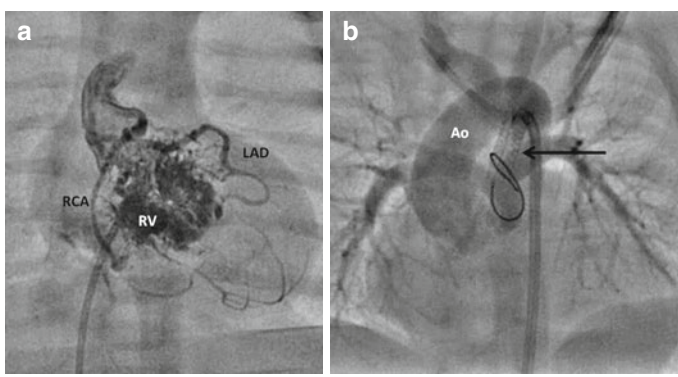


Fig. 24.6 Right ventricular angiogram (**a**) shows a hypoplastic right ventricle (*RV*) filling multiple sinusoids which fill the right coronary artery (*RCA*) and left anterior descending interventricular artery (*LAD*) indicative of right ventricle-dependent coronary circulation (RVDCC). This precludes 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*)

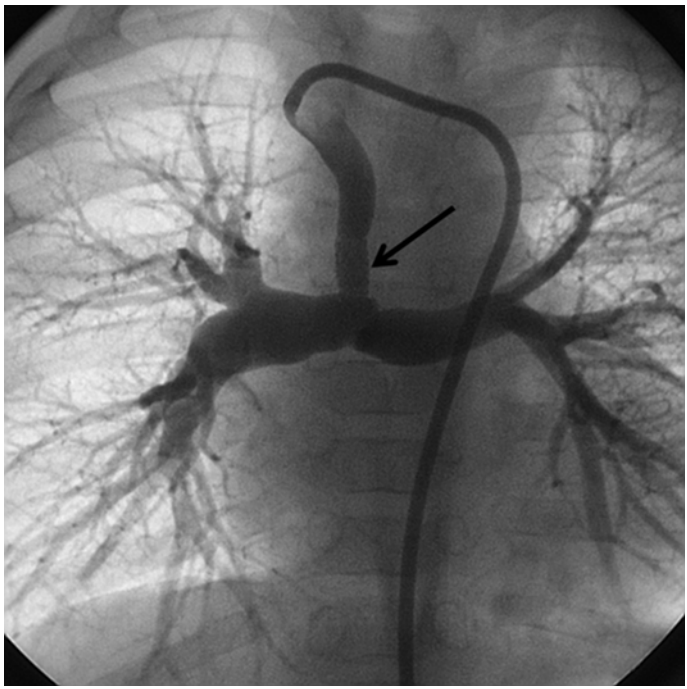


Fig. 24.7 Injection through a guide catheter passed from the left aortic arch into the right subclavian artery shows the long contralateral duct which inserts into the confluence. The narrowing in the distal insertion of the duct was stented (*arrow*) with a 3.5 mm stent

Case Study 6

A 45-day-old infant with single ventricle, pulmonary atresia, common atrioventricular valve with no regurgitation, presented 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. 24.7).

24.4 Indications and Patients Selection

24.4.1 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 mm or 4 mm grafts, DS offers a palliation lasting only for 6–12 months. In this group, DS is only indicated in:

(i) High-risk surgical candidates: low birth weight below 2.5 kg, syndromes like trisomy 21, comorbidities like bronchopneumonia, lung disease of prematurity, and other organ malformations.
(ii) 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.
(iii) 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.
(iv) If surgical experience is low in neonatal shunts, elective DS may be performed in low-risk candidates also.

24.4.2 Group B: Univentricular Hearts with Pulmonary Atresia

Both DS and surgical shunt increase the pulmonary blood flow and cause ventricular volume overload. Prolonged ventricular

dilatation and 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 shorter-term palliation, DS is more ideal than surgical shunt as it avoids surgical morbidity.

24.4.3 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.

24.5 Treatment Options

24.5.1 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 [2].

DS is preferred only 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

24.5.2 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.

24.5.3 Group C: Transient Neonatal Dependence on Duct

Prolonged PGE1 infusion for 2–6 weeks as an option is associated with (i) high costs due to prolonged intensive care stay, (ii) venous thrombosis and sepsis, (iii) drug adverse effects, namely, gastric antral hyperplasia and hyperostosis of bones, and (iv) 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.

24.6 Preprocedural Imaging

Echocardiogram is the most vital imaging tool to record the following features:

(i) Duct – morphology, origin and insertion, length, and diameter at aortic and pulmonary end

(ii) Aortic arch – side of the aortic arch and arch branches for axillary or carotid arterial access
(iii) Pulmonary arteries – mediastinal and hilar pulmonary artery sizes, stenosis of confluence at duct insertion site
(iv) Intracardiac anatomy – differentiate groups A,B, and C
(v) Ventricular systolic function, atrioventricular valve annulus size and function, aortic root diameter, and aortic valve function
(vi) Venous anomalies for transvenous approaches
(vii) Interatrial communication and need for balloon septostomy

24.7 Technique (Step-By-Step)

- PGE1 infusion should be stopped at least an hour before DS to allow ducts to be well constricted.
- A dose of *aspirin* 3–5 mg/kg is given before the procedure.
- *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 is reserved for procedural blood loss.
- Intubation and general anesthesia is preferred if axillary or carotid arterial access is anticipated.
- Intravenous fluids counter PGE1-related hypotension and facilitate quick vascular access.
- 4 F 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 (more than 180 s).
- For showing the pulmonary arteries and their confluence, aortogram should be done in shallow left anterior oblique projection (LAO 20° cranial 20°) in type I–III ducts (Fig. 24.2 and 24.3).

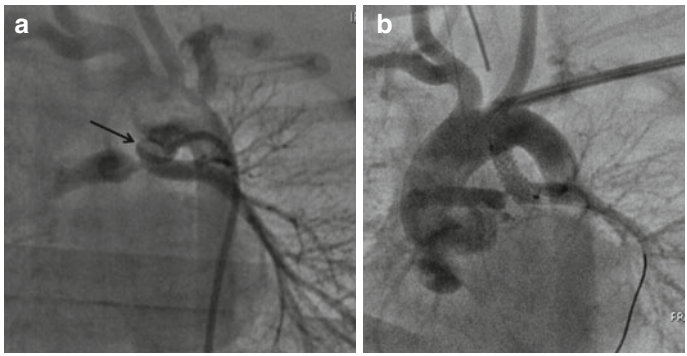


Fig. 24.8 Femoral access angiogram (**a**) with an endhole catheter shows a tortuous type III duct (*arrow*) which needed axillary artery access for successful ductal stenting with a 4 mm coronary stent (**b**). The tip of the guidewire is parked in the lower lobe of left lung

In ducts originating from right arch, RAO 20° cranial 20° projection is chosen (Fig. 24.4).

- Ductal origin and insertion, morphology, length, and diameter are delineated in lateral view (Fig. 24.1c) in type I–III ducts. In type IV and V ducts, aortogram is done in anteroposterior view (Fig. 24.6).
- Some operators prefer to access type II and III ducts through left axillary artery access which is obtained percutaneously (Fig. 24.8) or left carotid artery obtained by surgical cut down. The following techniques apply to type II and III ducts approached from femoral artery and the catheters used and wires are specifically meant for femoral route. If axillary or carotid access is obtained, a 4 French multipurpose catheter or Judkins right coronary catheter is often used.
- In type II and III ducts, the tip of the pigtail catheter is cut to a J shape and reinserted into the aortic arch to engage the duct. A Judkins 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 (Tuohy 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 *micro-catheter* can facilitate further advances of the guidewire.
- The stenting is done in most ducts through long 4 F sheaths but sometimes with 5 F Judkins right coronary guide catheter.
- Premounted nondrug-eluting *coronary stents* are chosen; length of stent is chosen based on echocardiography and angiography. Care should be taken to stent the entire length of the duct and not to leave any ductal portion unstented.
- After positioning the guidewire which 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. A repeat angiogram is done and the length of the duct is assessed in comparison to the balloon length (Fig. 24.12).
- 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.
- 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. 24.3) 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.
- 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.

24.8 Materials

Catheters: 4 F pigtail catheter, 4 F Judkins right coronary catheter, 5 F Launcher Judkins RCA guide catheter (Medtronic Co), 0.018″ lumen Cantata microcatheter (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

24.9 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. If all these attempts fail, alternative options of percutaneous ipsilateral *axillary artery access* or ipsilateral *carotid artery* entry through a cut-down exposure are attempted. As the ductal aortic origin is directly opposite to these arteries, cannulation is often easily done (Fig. 24.8). 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.

24.10 Tips and Tricks

- Even though transvenous ductal stenting helps in type II and III ducts, the stiff guide catheters through the tricuspid and aortic valves lead to their leak and cause hypotension. They may also rub the His bundle and cause conduction blocks.
- 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. 24.9).
- Systemic hypotension immediately after DS due to large aortic run off into pulmonary artery is managed by dopamine infusions.

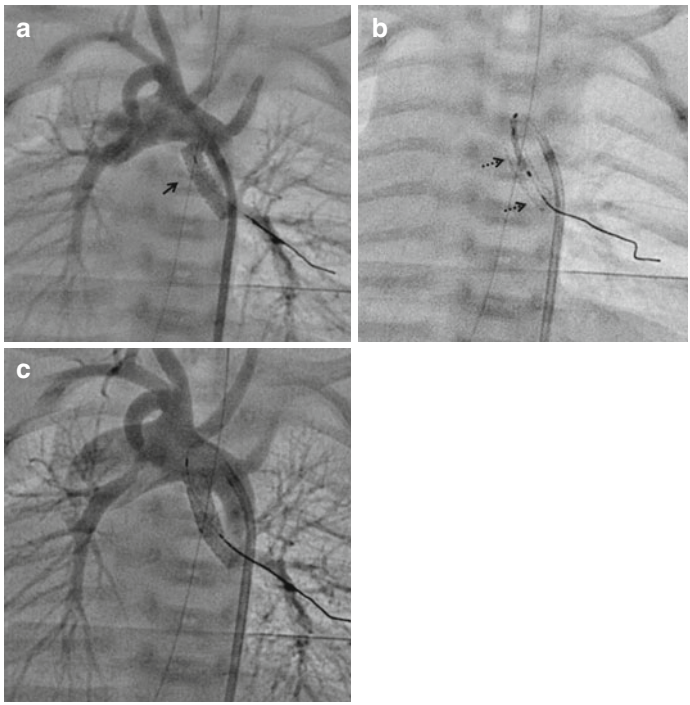


Fig. 24.9 In a type V bilateral duct in a neonate with single ventricle, pulmonary atresia, aortogram (**a**) after stenting of the left-sided duct from the undersurface of the aortic arch shows that the aortic end of the duct is uncovered by the stent (*arrow*). The guidewire is still in place. A second 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

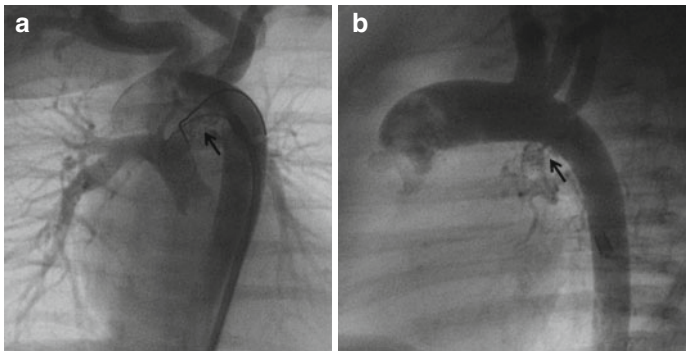


Fig. 24.10 Aortogram in shallow left anterior oblique view (**a**) shows successful stenting (*arrow*) of a type III duct from undersurface of aortic arch in a patient with d-transposition of great arteries, large ventricular septal defect, and pulmonary atresia. After removal of the guidewire, repeat aortogram shows complete thrombotic occlusion (**b**) of the stent

24.11 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 endhole catheters into the aortic end of the ductal stent may occasionally cause preferential flows into one lung (Fig. 24.10). In such instances, if echocardiogram 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.

24.12 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. 24.11) 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.

24.12.1 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. 24.10). 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 (2,000 units/kg body weight bolus followed by infusion of 1,000 units/kg/h) or tissue plasminogen activator (1 mg/kg bolus).
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 occurs 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.

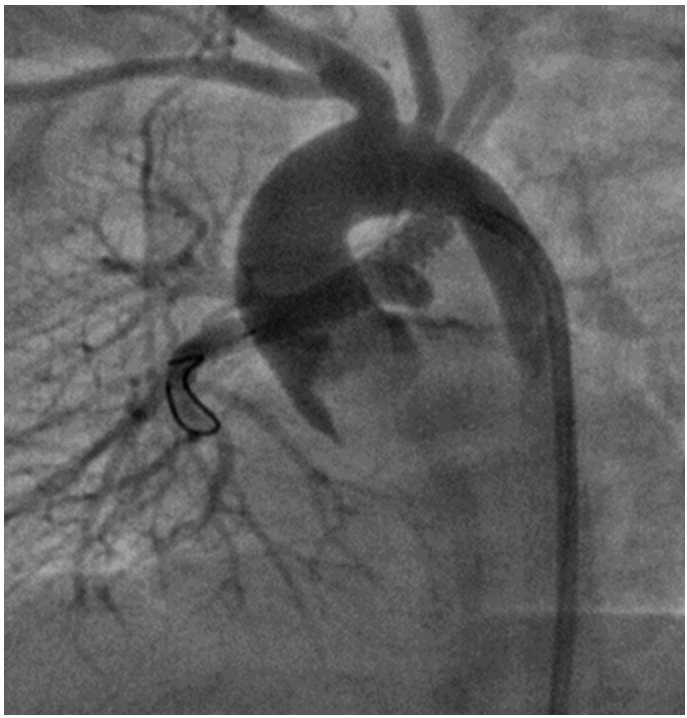


Fig. 24.11 Unilateral right lung hyperperfusion after ductal stenting in a patient with tetralogy of Fallot with pulmonary atresia

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. 24.13).
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 will prevent too distal stent embolization beyond the hilar branches.

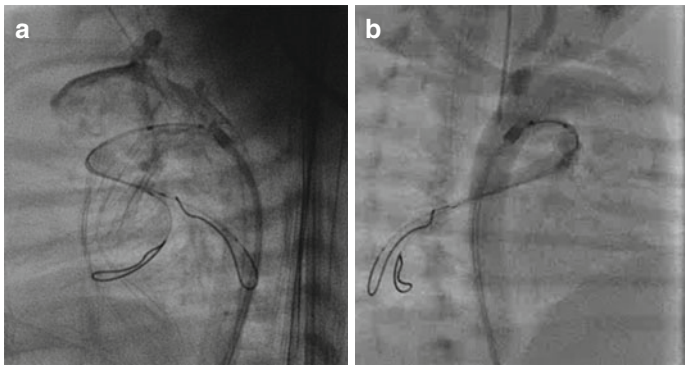


Fig. 24.12 Aortogram was repeated in lateral (**a**) and right anterior oblique (**b**) projections after an uninflated 1.5 mm × 10 mm balloon with two markers at either ends to indicate the length is placed in the duct. When the contrast fills the duct, the balloon length is compared to the straightened 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

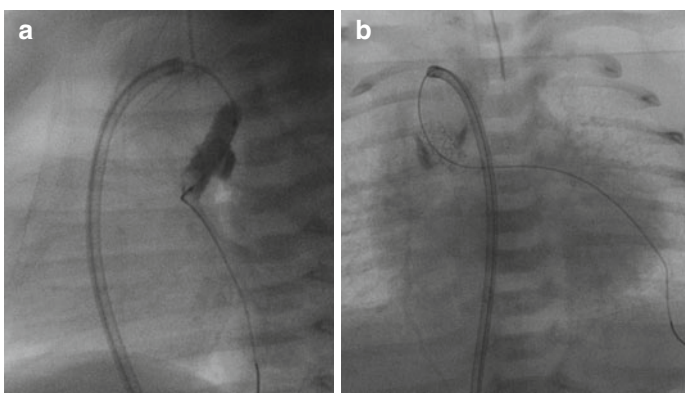


Fig. 24.13 Dissection of the duct during stent positioning (**a**), shown by contrast staining of the periductal tissues. This is immediately addressed by quick stent deployment (**b**). The staining of the ductal tissue with radiographic contrast disappears soon. Often the stent patency is not affected by this dissection and staining of the periductal tissues with contrast

24.13 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 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.

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Chapter 25
Patent Ductus Arteriosus Stenting in Duct-dependent Systemic Circulation

Dietmar Schranz

25.1 Anatomic Description and Physiopathology

Cardiac malformations causing duct-dependent systemic blood flow can be summarized as "left-sided" defects ranging from severe aortic valve stenosis, hypoplastic left heart (Shone)-complex (HLH-C) to hypoplastic left heart-syndrome (HLH-S) and aortic coarctation to interrupted aortic arch (see Chap. 39 on the hybrid approach).

25.2 Clinical Scenarios

Newborns with left-sided congenital heart malformations can have the *disadvantage* of late detection. The symptom of tachypnea is often missed or is wrongly interpreted (e.g., as sepsis). Many babies with undetected postnatal duct-dependent systemic blood flow become ill before an appropriate diagnosis is

D. Schranz
Pediatric Heart Center, University Clinic Giessen,
Giessen, Germany
e-mail: Dietmar.schranz@paediat.med.uni-giessen.de

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made. To summarize, a newborn with tachypnea is seriously ill, probably with a life-threatening illness; in these newborns, a congenital heart defect with duct-dependent systemic blood flow must always be considered. Prenatal fetal echocardiography helps to avoid postnatal heart failure and even the cardiogenic shock caused by unperceived duct obstruction. Therefore, congenital left-sided heart defects need to be immediately treated on detection, either in the fetus or after birth. Regarding to a prophylactic therapeutic strategy in which prostaglandin infusion is employed, a low-dose infusion (5–10ng/kg × min) is safe, but a high dose (25–50 or 100ng/kg × min) is dangerous. However, for the employment of a prophylactic treatment regimen, a prenatal or immediate postnatal diagnosis is needed. Importantly, the treatment of newborns before any symptoms are developed highlights the need for sophisticated medical education.

25.3 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, its Sano modification, or, rarely, a challenging biventricular repair. Indications for duct stenting in duct-dependent systemic blood flow could be prostaglandin-refractory duct obstruction or as part of an elective therapeutic strategy.

In the past, duct morphology, in particular, the junction of the duct to the descending aortic arch, seemed to be unfavorable for stenting in about 10 % of patients. This feature was even more pronounced when balloon-expandable stents were the only option for duct stenting. The use of a large introducer sheath or hemodynamic instability in the patient did not allow for the

short flow interruption that occurred during balloon inflation for stent expansion. Currently, from the technical point of view, with the use of new self-expandable stents, which can be delivered through a 4 F sheath, duct stenting in duct-dependent systemic blood flow is easily feasible in all newborns, independent of morphology or hemodynamic stability.

25.4 Treatment Options (Also See Chap. 39)

Duct stenting combined with bilateral pulmonary artery banding (bPAB), and if necessary, interatrial septum manipulation, was developed as a hybrid method [1]; this is offered as an alternative first-step approach at a number of centers worldwide, but is mostly used as a hybrid procedure in newborns with high-risk HLH-S or HLH-C [2, 3].

25.5 Pre-procedural Imaging

In newborns the diagnosis of HLH-S or HLH-C is obtained by echocardiography in a standardized manner with transverse and longitudinal planes, with special emphasis on demonstrating the 4-chamber view, outflow tracts, and a 3-vessel view using two-dimensional (2-DE) color flow and spectral Doppler interrogation. The focus of the echo examination needs to be on the interatrial communication, the tricuspid and systemic ventricular function, and the duct morphology, in terms of its length and width and its relationship to the descending aortic arch (Fig. 25.1). When considering duct stenting, the strategy should be based on the pre-procedural imaging, which could include magnetic resonance imaging (MRI) to answer any questions regarding pulmonary vein morphology or aortic arch malformations (Fig. 25.2).

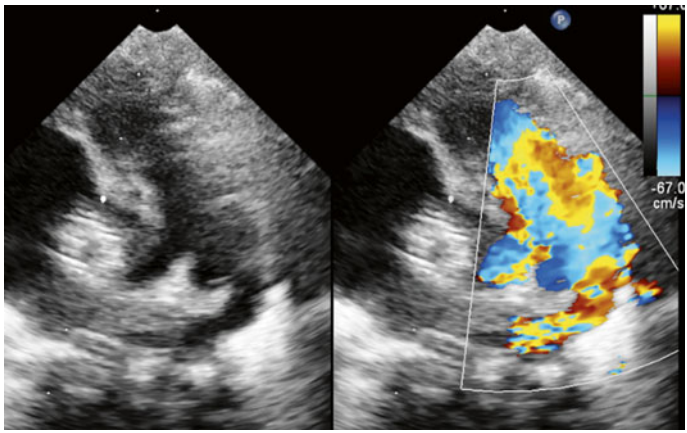

Fig. 25.1 Echocardiography evaluation of duct morphology, in terms of its length and width and its relationship to the descending aortic arch

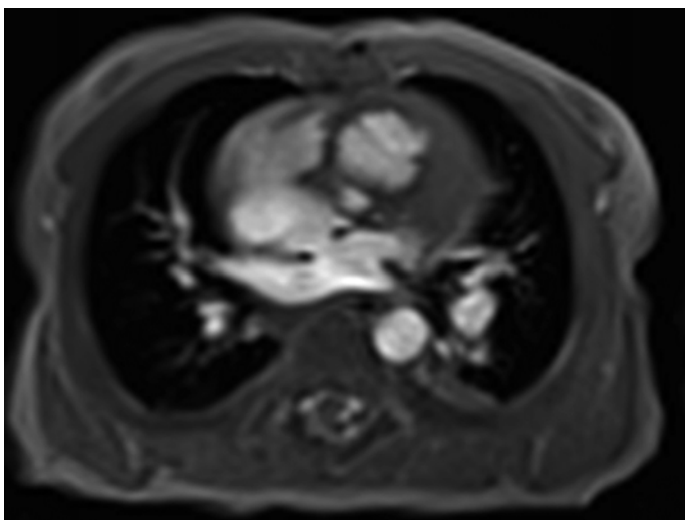

Fig. 25.2 Magnetic resonance imaging (MRI) to analyze pulmonary vein morphology or aortic arch malformations

25.6 Technique (Step-By-Step)

Duct stenting is mostly performed as part of a hybrid procedure during an open chest – beating heart scenario immediately after bilateral PAB [2] or as an elective transcatheter approach in a spontaneously breathing, sedated newborn, with or without an additional atrioseptostomy procedure [1, 3]

The percutaneous transcatheter approach is usually performed by femoral vein or arterial access. In the past, the placement of a 4 F multipurpose catheter was done by passing it through a 4 F sheath (Terumo, Frankfurt, Germany), which was placed in the femoral artery and served for blood pressure monitoring and delineation of the duct-aortic junction. Currently, percutaneous duct stenting is easily performed by *femoral artery access* utilizing a 4 F delivery system (see Chap. 39).

The step-by-step approach by *femoral vein access* can be summarized as follows (Fig. 25.3a, b).

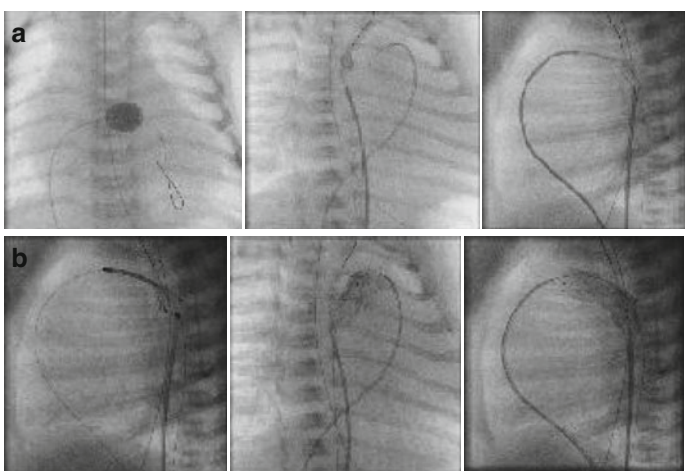

Fig. 25.3 (**a**) Duct Stenting in Ductus arteriosus dependent Systemic Blood Flow (DA-SBF). How to do it. (**b**) Duct stenting in DA-SF. How to do it

Step 1 – Evaluation of Interatrial septum/patent foramen ovale (*IAS/PFO*) (*Rashkind* +): the interatrial communication must be evaluated. In HLH-S the communication should be unrestrictive or should have no, or only a minimal pressure gradient; in HLH-C patients the interatrial communication should guarantee a sufficient preload for the growing left ventricle.

Step 2, for free crossing of the tricuspid valve a 4 F open-end balloon catheter is used in ensemble with a soft coronary guidewire (BMW, Abbott, Wetzlar, Germany) which is advanced over the duct in the descending aorta (DAO) without the need to follow with a wedge catheter.

Step 3, the balloon catheter is exchanged for a 4 F right Judkins catheter (rJC), which is advanced in the pulmonary artery (PA) and may be advanced over the duct in the DAO.

Step 4, the soft coronary guidewire (BMW) is exchanged for a stiff 0.014 coronary guidewire (S port, Abbott). After pullback pressure measurement from the DAO to the PA is obtained by using a hemostat valve at the Judkins catheter, the rJC is positioned closed to the pulmonary end of the duct. The hemostat valve avoids bleeding and allows angiography by the manual injection of contrast medium. Additionally, a 4 F multipurpose catheter is advanced to the aortic duct junction guided by the BMW coronary guidewire, which can easily be advanced in the aortic arch or through the duct in the pulmonary system; the wire and catheter serve as a marker and the hemostat valve can be used for angiographies from the arterial side performed prior to and during duct stenting.

Step 5, delineation of the duct morphology is performed with biplane angiography in the 30° right anterior

oblique (RAO) and 90° lateral planes. This imaging strategy dramatically reduces the interventional time required for percutaneous duct stenting, in addition to improving safety. If the decision for duct stenting is made, the diameter of the stent to be utilized must be at least 1–2 mm greater than the minimal measured duct diameter, but in any case it must exceed the diameter of the DAO. This recommendation is of particular importance in newborns with an interrupted aortic arch.

Step 6, after the preparation of a pre-packed self-expandable Sinus-Superflex-Duct Stent (SSF-DS, OptiMed, Karlsruhe, Germany) and flushing of the 4 F delivery system, the system in total is carefully advanced over the stiff S port wire through the tricuspid valve, the right ventricle, and the PA with the tip advanced to the DAO.

Step 7, the multipurpose catheter together with the BMW guidewire placed in the DAO serves as a marker during expanding of the stent. Slow pulling back of the covering of the delivery system enables full stent expansion to be performed with fluoroscopy control; in some patients short cine scenes are necessary to sufficiently visualize the very thin struts of the open-cell stent.

Step 8, the lateral 90° plane is used to control the stent position at the PA end of the duct (Fig. 25.4).

Step 9, the right anterior oblique (RAO) 30° is the plane of choice to observe the exact stent position in relation to the junction of the duct to the DAO.

Step 10, before the delivery system is carefully removed the stent should have had a short time period during which it has fully expanded with the aid of nitinol conditioning at a temperature of 37° C; then the

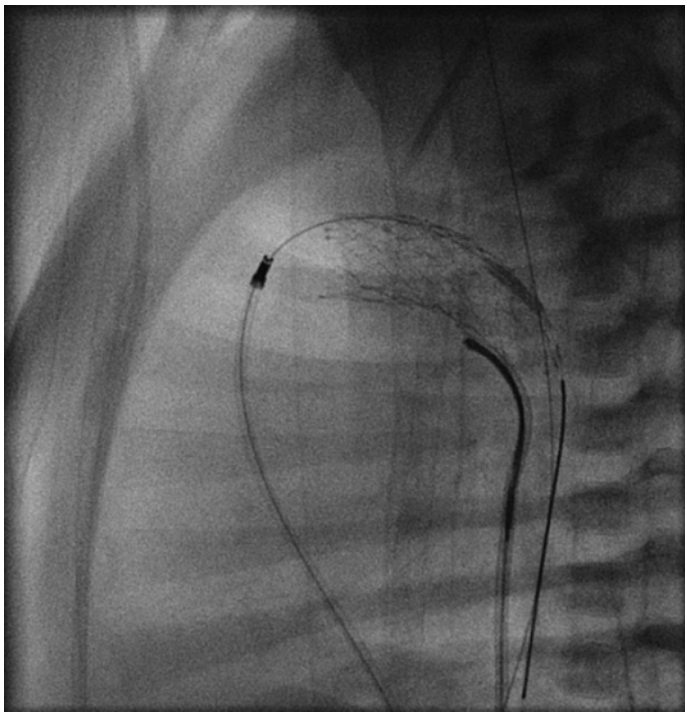

Fig. 25.4 Lateral view showing stent implantation

open-cell device has the chance of expanding within the arterial wall of the duct, and the risk of destabilizing the stent position or inducing inadvertent stent slipping is minimized.

Steps 11 and 12, before the S port wire is carefully removed by re-advancing the rJC to the pulmonary end of the stented duct, the appropriate stent position has to be evaluated by angiographies from the pulmonary and arterial sides, through the Judkins and multipurpose

catheters, respectively. The junction of the descending aortic arch 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 important that the duct is fully covered by the stent, and the retrograde access to the aortic arch is carefully analyzed.

In most patients there is no need to over-stent the retrograde access to the descending aortic arch; the multipurpose catheter in ensemble with the BMW guidewire easily guides duct stenting and its relation to the descending aortic arch. The indication for the placement of an additional stent within the isthmus is based on pull back pressure measurements and the results of the angiography performed through the multipurpose catheter (see Chap. 39).

Step 13, an additional hemodynamic analysis is recommended after removal of the guiding catheter at the end of the procedure. Noninvasive blood pressure measurement at the right arm, to estimate the coronary and cerebral perfusion pressure, has to be performed simultaneously with invasive pressure measurements of the PA (by the rJC), and DAO pressure measurement by the multipurpose catheter in order to exclude significant pressure gradients across the stented duct and within the aortic isthmus, respectively. The decision for stopping low-dose prostaglandin therapy is based on the hemodynamic and angiographic data obtained; usually the prostaglandin infusion is stopped after the placement of a self-expandable stent, in contrast to the procedure with a balloon-expandable stent, in which the prostaglandin infusion is stopped before heart catheterization to Provoke slight obstruction.

Step 14, the procedure of duct stenting is finished when the hemodynamic and clinical condition of the patient is stable; this may occur after some hours or perhaps days; in other words, the interventionalist knows best if he/she has achieved an optimal or only a sufficient result Fig. 25.3a, b.

25.7 Materials

In the past, duct stenting was almost exclusively performed with balloon-expandable stents. However, now balloon-expandable stents have been almost completely replaced by self-expandable stents, which can be used even in large vessels without any obstruction. Here, the materials for duct stenting in newborns with duct-dependent systemic blood flow are summarized.

The Sinus-Superflex-DS™ (SSF-DS™; Opti-Med, Karlsruhe, Germany) stent is used in our institution; it is a self-expandable, open-cell nitinol stent available with diameters of 7, 8, and 9 mm and stent lengths of 12, 15, 18, and 20 mm, and in future a stent length of 24 mm may be available. This new generation of self-expandable stents is deliverable through a 4 F sheath, which means the stent can be safely placed by femoral vein access, as well as through a 4 F sheath placed in the femoral artery.

The SSF-DS™ has received a CE (Conformité Européene) mark for its specific indication of duct stenting in newborns.

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
0.036 in. wire for introducing the catheter (Cordis, Hamburg, Germany)
0.014 in. coronary floppy wire (middle weight balanced, Abbott, Wetzlar, Germany)
0.014 in. super stiff (support, S sport wire, Abbott, Wetzlar, Germany)
SinusSuperFlex-DS™ (Optimed, Karlsruhe, Germany)

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.

25.8 Expected Results

Duct stenting in duct-dependent systemic blood flow can be performed with a mortality rate of less than 1 % (data presented at the World Congress of Pediatric Cardiology and Cardiac Surgery [WCPCCS] 2013, Cape Town, South Africa). In uncomplicated cases the procedure performed by venous access can be done with a fluoroscopy time of less of 10 min, and when performed by arterial access the fluoroscopy time is less than 5 min. However, the results are dependent on the institutional experience, the general therapeutic strategy, and the materials utilized. In this context it should be mentioned that experience is defined by how complications are managed.

The goal of any procedure should be an optimum outcome; possible complications should not be a surprise, but a calculated and well-prepared-for event.

25.9 Tips

25.9.1 Tip 1: Preparation

The interventionalist not only has to obtain the parents'/carers' written consent, but also to plan his/her catheter strategy based on the available or self-performed echocardiography. If there are any open questions, MRI, in relation to pulmonary vein connection, single or multiple vein stenosis, or unusual aortic arch morphology, should be performed and included in the decision-making. Such information is better to have *before* the catheter approach than arising as a surprise during the approach.

25.9.2 Tip 2: Avoidance of Hemodynamic Instability

Patients with duct-dependent systemic blood flow need high pulmonary vascular resistance or immediate and sufficient BPB. Therefore, anesthesia and controlled ventilation are sometimes more dangerous than the transcatheter procedure itself. In our institution catheterization is usually performed in spontaneously breathing patients, in most of them immediately after extubation following surgical pulmonary banding; sedation is performed with diazepam and ketamine at very low, but repeated single doses of 0.5–1 mg diazepam and 1 mg ketamine.

As mentioned above, a 4 F open-end inflated balloon catheter is used to cross the tricuspid valve to avoid later hemodynamic instability; the 4 F delivery system is advanced across the tricuspid and pulmonary valves over the stiff coronary guidewire. Therefore, before the stent is delivered the hemodynamic stability must be checked during the previous advancing of the right 4 F Judkins catheter. Although the SSF-DS™

delivery sheath is only 4 F, the relatively stiff ensemble might compromise a borderline hemodynamic situation by causing artificial tricuspid and/or pulmonary valve regurgitation. This is one more reason for carefully observing the patient's pre-interventional hemodynamic status and avoiding any artificial blood pressure decrease. Currently, any significant pulmonary and tricuspid valve regurgitation or associated systemic ventricular failure would indicate duct stenting by femoral artery access (see Chap. 39).

25.9.3 Tip 3: Stent Placement

Stent delivery through the tricuspid valve, right ventricle, and pulmonary valve within the duct must be performed without any feeling of obstruction. A stent should never be advanced with force; the interventionalist should observe the blood pressure measured invasively by the multipurpose catheter positioned in the descending aorta, as well as the blood pressure measured noninvasively at the right arm.

25.10 Pitfalls (See Sect. 25.11)

1. Morphology-dependent pitfalls:
   Duct obstruction
   Duct aneurysm
   Aortic coarctation
2. Material-related pitfalls
   Stent expansion problems
   Stent-dependent obstruction of the duct-aorta junction
   Stent slipping caused by removal of the relatively stiff delivery system
   Open strut-related obstruction in advancing a second stent

25.11 Complications

1. Stent slipping
2. In-stent obstruction or insufficient stent expansion
3. Retrograde aortic arch obstruction
4. Hemodynamic instability

25.12 How to Manage Complications

To manage (*1*), if stent slipping is observed, the stent usually migrates to the pulmonary side; to avoid slipping to the aorta, a stent diameter that is 1–2 mm greater than the diameter of the descending aorta has to be chosen. To resolve such a complication, a 5-mm snare has to be available in stock, to have the chance of holding the stent from the arterial side while a second stent is advanced from the venous side to enable fixing the first one by placing a stent-in-stent, so-called telescope stenting (Fig. 25.4).

To manage (*2*), immediate in-stent obstructions may be caused by the relatively weak radial force of the very thin struts and the open-cell design of the SSF-DS™. When there is insufficient stent expansion with a residual obstruction, re-dilation should be performed with an 8 × 30 mm Sterling balloon catheter, which can be advanced from the venous as well as the arterial side even through a 4 F sheath. Considering the high flow through the duct, re-dilation of an already stented duct is more effectively performed with a balloon of 30 mm length than with one with a length of 20 mm.

To manage (*3*), retrograde aortic obstruction induced by a previously placed duct stent 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 this serious complication. The reasons for the 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, or if over-stenting of the descending aortic arch is needed because of unfavorable anatomy, then stenting with a SinusRepo-DS™ closed cell device with a 5 or 6 × 9 mm design should be performed to treat or to avoid a severe coarctation (see Chap. 39).

To manage (*4*), any severe hemodynamic instability must be analyzed to determine its reasons; duct stenting is, in most cases with the rare exception of prostaglandin-resistant duct obstruction, not mandatory, but an elective approach to replace prostaglandin infusion.

Currently, for cases presenting with severe hemodynamic instability despite BPB having already been performed, duct stenting should be performed by femoral artery access (Chap. 39).

25.13 Post-Procedural Care

When the stent placement is successful, prostaglandin is stopped immediately after the procedure. However, when the result is suboptimal, prostaglandin should be continued for 24 or 48 h until the stent and the immediate foreign body-dependent

vessel proliferation that may be associated with a delayed duct obstruction is less probable. If duct obstruction seems probable, close observation needs to be performed during the few days following the delayed stopping of prostaglandin therapy. Covering the duct fully with the stent seems to be the best option to avoid duct obstruction.

In any case, heparin 300 U/kg per day is administered for 24 or 48 h following the transcatheter procedure. The cyclooxygenase inhibitor acetylsalicylic acid (aspirin, Bayer, Leverkusen, Germany) is not routinely administered; 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 has been placed within an aortic coarctation (see Chap. 39).

One of the most important points in post-procedural care is blood pressure monitoring. It must be emphasized that systolic and diastolic, and not – MEAN – blood pressure, 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 must be measured.

The patients' parents/carers are taught to observe the respiratory rate in their sleeping baby, because the clinical condition of the patient is correlated with the respiratory rate during sleep. As long as the respiratory rate is within the age-dependent range, the patient is not seriously ill.

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 outpatient.

25.14 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 case 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

Chapter 26
Step-by-Step Closure of Atrial Septal Defects (ASDs)

John D.R. Thomson

26.1 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. 26.1a) 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-centering (with a core) and the non-self-centering devices (with a thin central stalk). Self-centering devices are most commonly deployed due to their ability to deal with defects of most sizes.

J.D.R. Thomson
Department of Cardiology, Leeds General Infirmary,
Great George Street, E Floor, Jubilee Wing, Leeds LS1 3EX, UK
e-mail: thomsonjdr@mac.com

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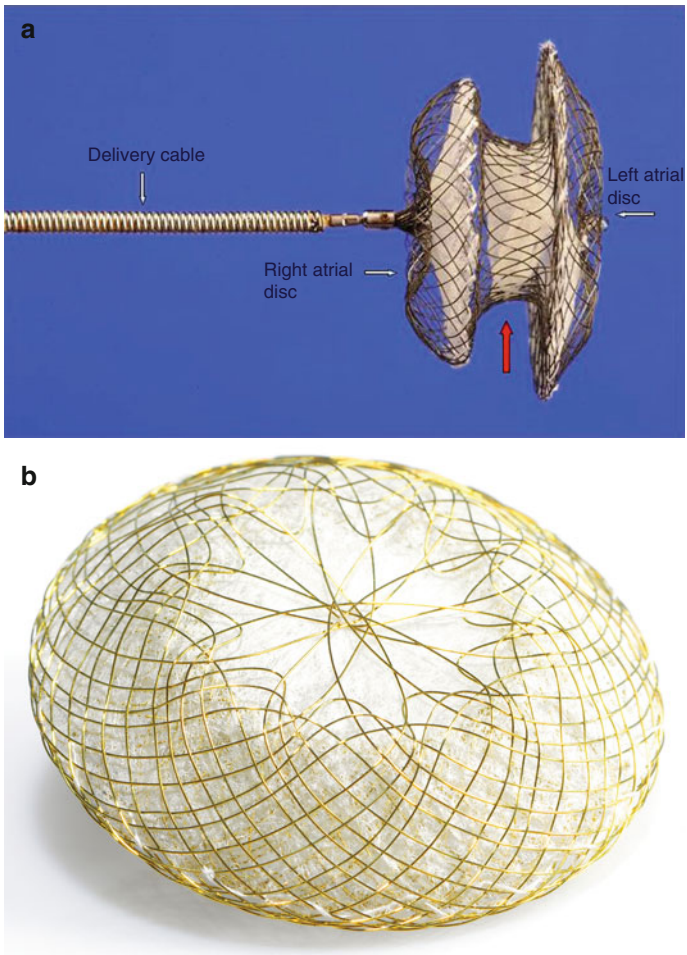

Fig. 26.1 (**a**) Amplatzer septal occluder (ASO). The *red arrow* points to the self centring core of the device. (**b**) Occlutech Figulla occluder, left atrial aspect. (**c**) Occlutech Figulla occluder, articulation with the delivery cable. (**d**) Cardia Ultrasept occluder. Showing the left atrial disc and the central portion N.B. the right atrial disc is undeployed within the delivery sheath. (**e**) Gore septal occluder: left atrial aspect. Note the five interlocking wires forming the frame covered in ePTFE

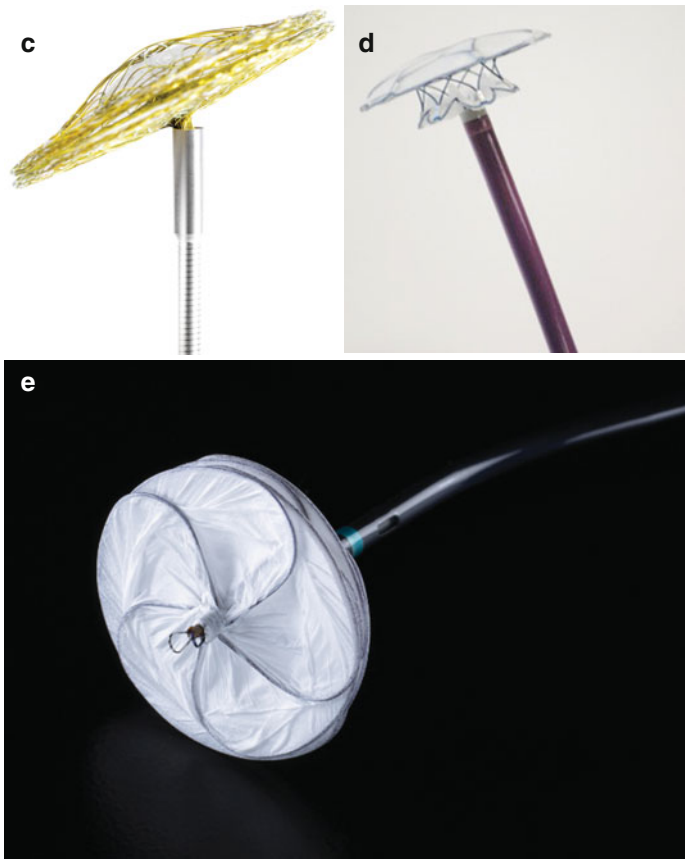

Fig. 26.1 (continued)

26.2 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

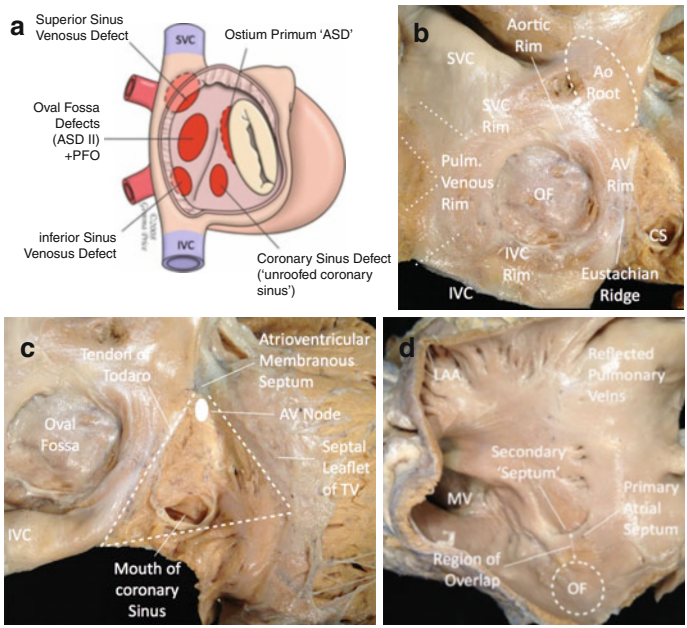

Fig. 26.2 (**a**) Schematic diagram of defects of the atrial septum, viewed through the right atrium (Image courtesy of Dr Andrew Cook/Gemma Price). (**b**) Cadaveric specimen. View from the right atrium showing structures adjacent to the oval fossa (*OF*). *SVC* superior vena cava, *IVC* inferior vena 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

heart (Fig. 26.2a). The atrial septum is not symmetrical. When 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. 26.2b). 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. 26.2c).

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. 26.2d).

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 are 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.

26.3 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.

26.4 Clinical Scenarios: Natural History

The vast majority of infants and children with ASDs are asymptomatic, although many have tendency to recurrent chest infections and respiratory symptoms. Very occasionally an infant will be encountered 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.

26.5 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 3rd 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 resolved, the right heart is usually remodelled 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, patients are encountered 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.

26.6 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 procedures so that an informed decision can be made.

26.7 Device Options

1. Currently available self-centering 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. 26.1a).

   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. 26.1b) and the delivery system is innovative, allowing articulation of the device on the delivery wire to facilitate closure of larger or asymmetric defects (Fig. 26.1c).

   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. 26.1c).

   Device delivery systems: All 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 all loaded into a short tube before being introduced into the delivery sheath. Modern self-centering devices are generally retrievable and repositionable prior to final release from the delivery cable.

2. Non-self-centering devices:

   Occlutech, Cera and St Jude (Amplatzer) all produce a variant of their self-centering 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-centering 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/): A newer form of occluder based on five interlocking nitinol/platinum wires covered with an ePTFE shell. The GSO has a soft "scalloped" construction that allows the device to adapt to the contours of the heart with an extremely low profile (Fig. 26.1d). The device comes pre-attached to a delivery handle, and the delivery sheath is part of the pre-assembled system. The device is advanced to the atrial septum using a monorail port at the end of the wire. Device deployment is by a simple movement of the delivery button and the device can be retracted and deployed as many times as required.

26.8 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 flows 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 function can be masked by an atrial shunt and closure of the defect in these patients can (rarely) precipitate pulmonary oedema. In older adults (>50 years), it is important to rule out and if necessary treat coexistent coronary abnormalities/disease.

26.9 Techniques: Step by Step

1. Pre-procedure

   Ensure appropriate patient selection.
   Is there adequate pre-procedural imaging?
   Has all co-morbidity 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. 26.3a, 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 guide wire
   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 into central, small- to medium (<20 mm)-sized defects. The chosen device is inserted into the delivery sheath using the proprietary loading tube (Fig. 26.4a) and thoroughly flushed with heparinised saline. Via the delivery sheath, the device is then advanced to the left atrium (Fig. 26.4b). 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,

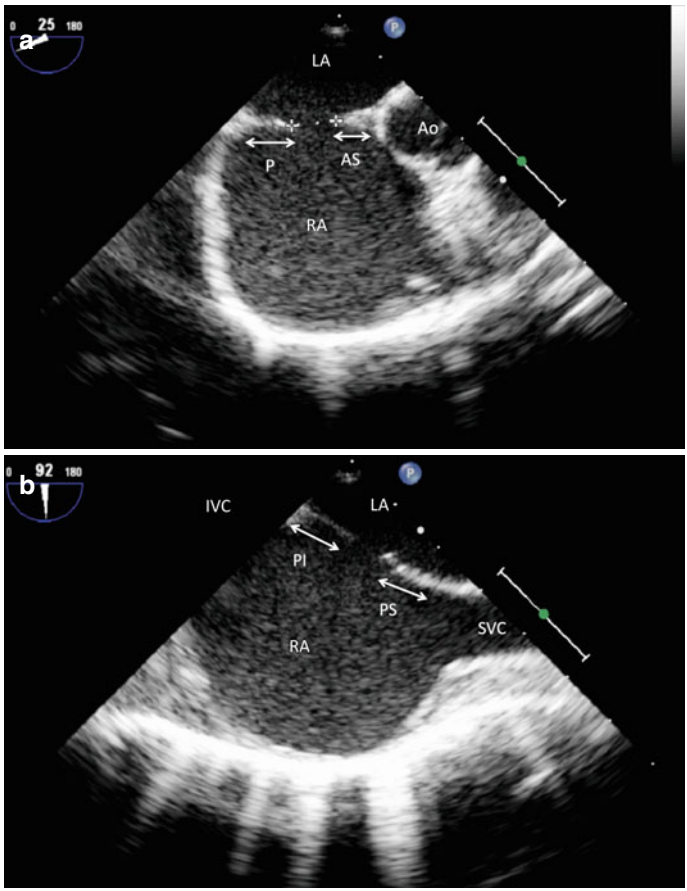

Fig. 26.3 (**a**) Trans-oesophageal echocardiography, "30°" view. *RA* right atrium, *LA* left atrium, *Ao* aorta, *P* posterior atrial rim, *AS* anterior-superior atrial rim. (**b**) Trans-oesophageal echocardiography, bi-caval view. *PI* postero-inferior atrial rim, *PS* postero-superior rim. The *arrows* delinate the extent of the tissue margin around the defect

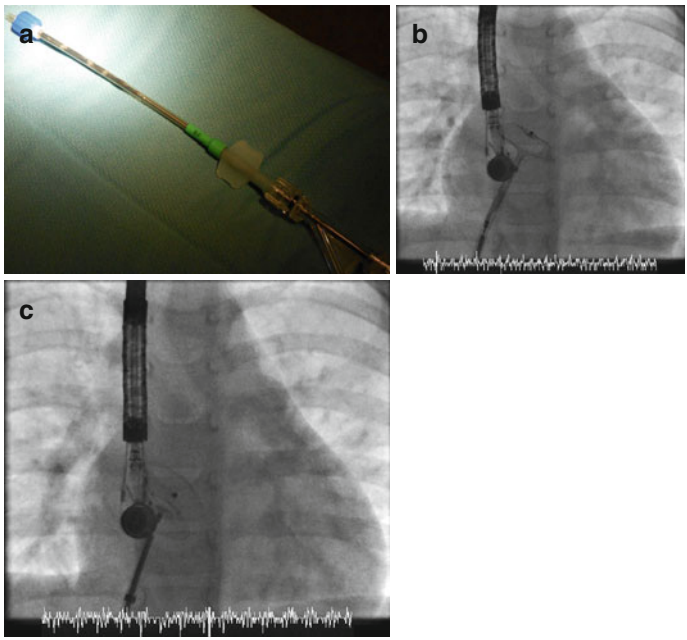

Fig. 26.4 (**a**) ASO loaded in delivery sheath. (**b**) Fluoroscopy. ASO, left atrial disc deployed. (**c**) Fluoroscopy. ASO, device fully deployed

the core is developed, the right disc developed (Fig. 26.4c) (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.

26.10 Materials

26.10.1 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 2 wires to create a double-length cable) in the event of an emergency.
   (g) Cook Flexor 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)

26.11 Tips and Tricks

1. Sizing: For single ASDs many operators prefer to simply measure the defect using either TEE or ICE in 2 perpendicular planes, taking the largest diameter on colour Doppler flow and selecting the next device size up to avoid significant oversizing (Fig. 26.5). 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

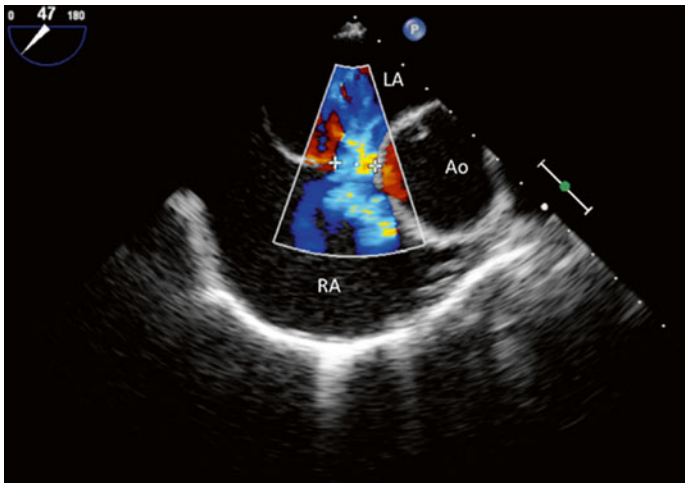

Fig. 26.5 Trans-oesophageal echocardiography, colour Doppler: markers showing sizing using colour Doppler. *RA* right atrium, *LA* left atrium, *AO* aorta

used for gentle inflation. Care must 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 manufacturers 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.

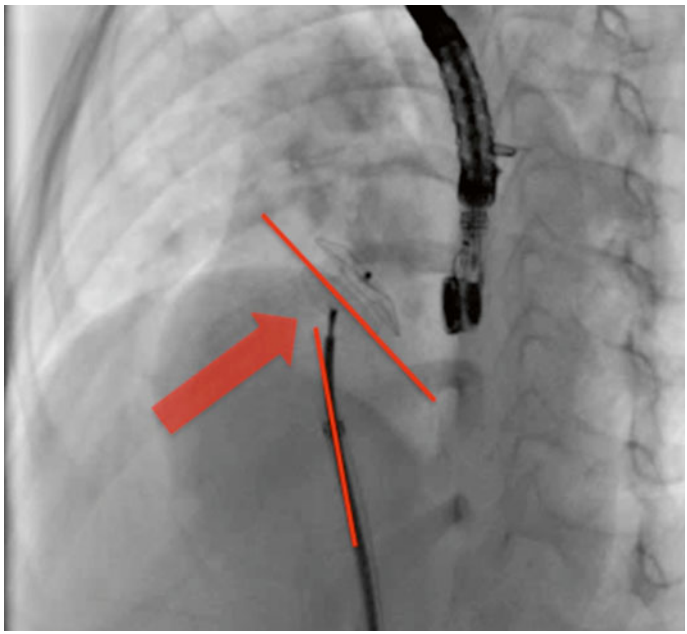

Fig. 26.6 Fluoroscopy: the angle between the septum and the delivery cable (*lines*)

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 usual approach from the inferior vena cava means that the Amplatzer septal occluder approaches the atrial septum at an angle (Fig. 26.6). 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. 26.7), which have a delivery cable which

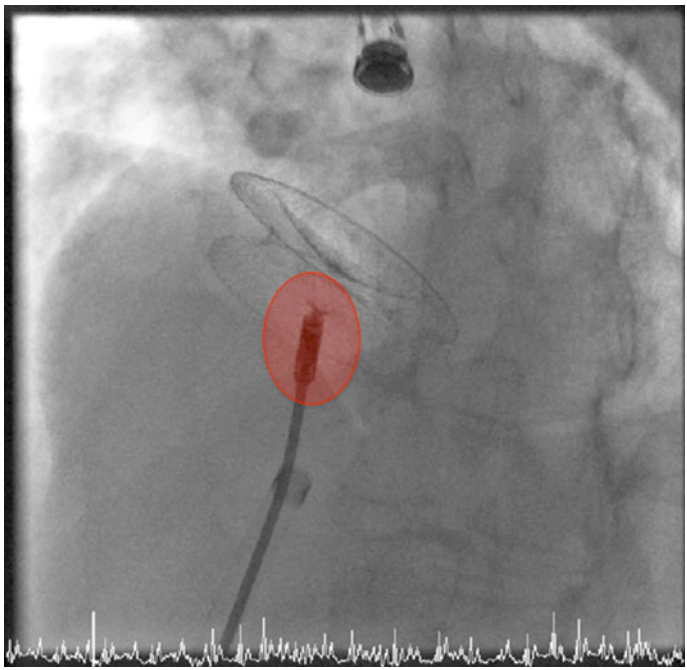

Fig. 26.7 Occlutech occluder. The flexible connection between the delivery cable and the device is *circled*

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:

   (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 to

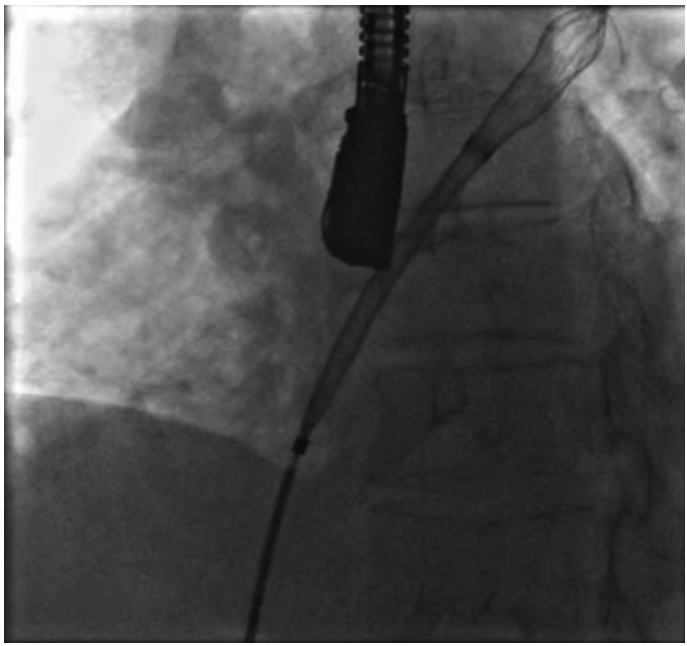

Fig. 26.8 Delivery of the left disc within the left upper pulmonary vein

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 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. 26.8). 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.

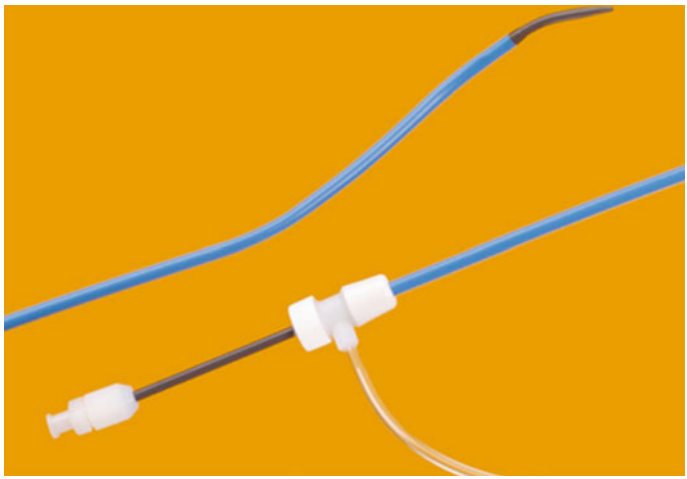

Fig. 26.9 Hausdorf delivery sheath. Note the "three-dimensional" curve which alters the approach of the device to the septum

(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. 26.9), 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" 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. 26.10). 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.

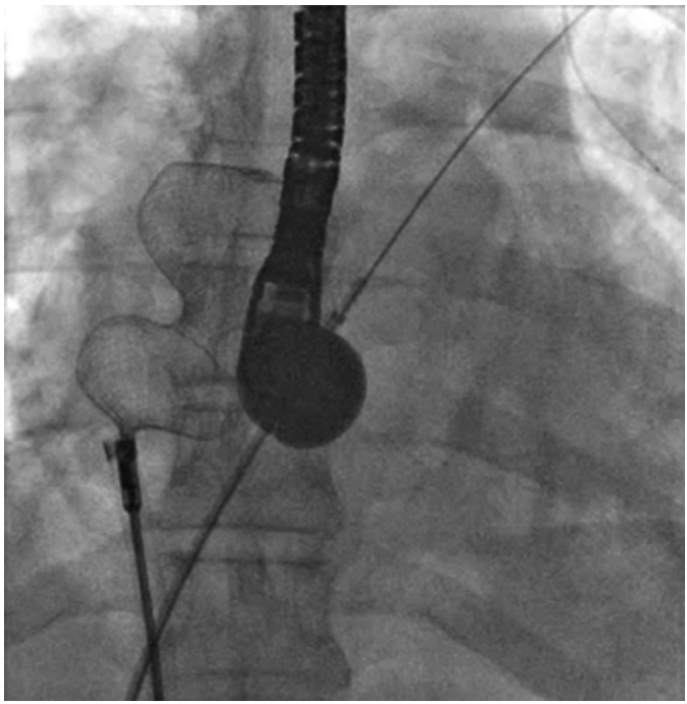

Fig. 26.10 Balloon-assisted technique

(d) Other techniques: The use of a small snare threaded over the delivery cable to hold onto the device screw such that the main guide wire 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. 26.11a, b).

5. Multiple defects: Multiple ASDs vary from distinct defects within a relatively firm atrial septum to multi-fenestrated

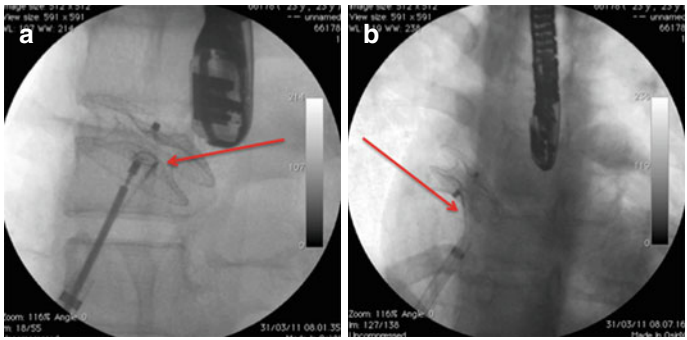

Fig. 26.11 (**a**, **b**) Snare (*arrowed*) around the screw of an ASO before (**a**) and after (**b**) release from the cable (Images courtesy of Dr Gianfranco Butera)

holes within a mobile structure. Sometimes multiple defects can be tackled with a single occluder, assuming that the edge of the defects are less than a few mm 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 2 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.

26.11.1 Using Non-Self-Centering 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 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 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 take 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.

26.12 Pitfalls

26.12.1 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 what data there is suggests 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.

26.12.2 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 in 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 challenge; 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.

26.13 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

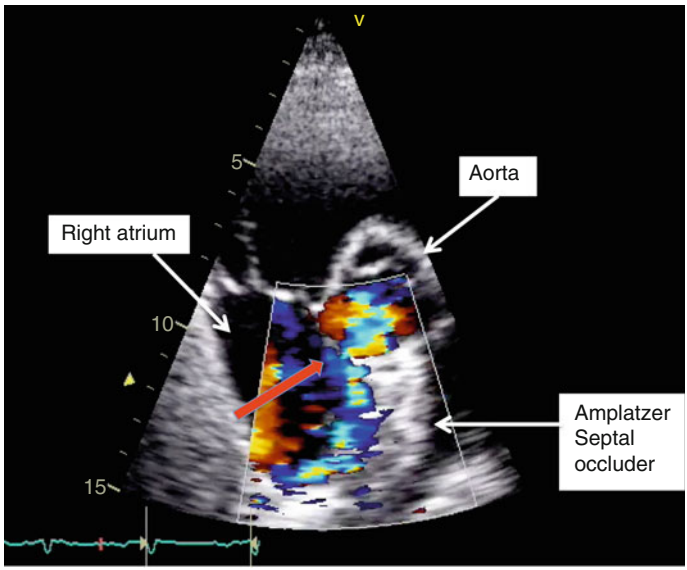

Fig. 26.12 Device-related perforation into the aorta (*arrowed*)

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. Migrainous 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. 26.12). 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 being more common with relatively oversized devices and in patients with absent retro-aortic rims.

26.13.1 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.

26.14 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.

26.15 Follow-Up

All patients: Review at 3 months/12 months.

26.15.1 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 indefinitely, although there is debate about review interval based on the occurrence of late erosion. What is clear is that there patients should remain under indefinite cardiac follow-up.

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Chapter 27
Fontan Fenestration Closure

Derize E. Boshoff and Marc H. Gewillig

27.1 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

D.E. Boshoff (✉) • M.H. Gewillig
Department of Pediatric Cardiology, University Hospital Leuven,
49 Herestreet, Leuven 3000, Belgium
e-mail: derize.boshoff@uz.kuleuven.be; marc.gewillig@uz.kuleuven.ac.be

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the atriopulmonary connections 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].

27.2 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.

27.3 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

27.4 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 and, 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.

27.5 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. 27.1), additional interatrial leaks and possible venous collaterals.
- 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.

27.6 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,

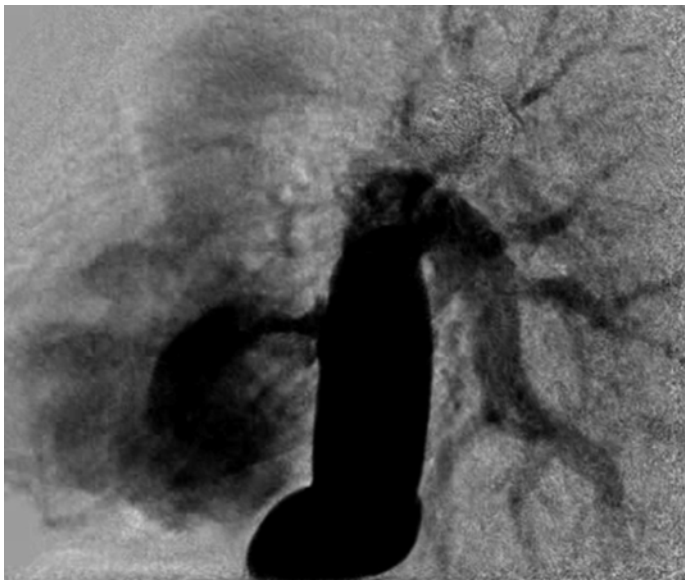

Fig. 27.1 Lateral view of contrast injection in inferior caval vein: a 20 mm conduit is mounted between the inferior caval vein and pulmonary artery; a 4.5 mm fenestration allos righ-to-left shunt into the left atrium

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. 27.2).
- 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 during 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.

27.7 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

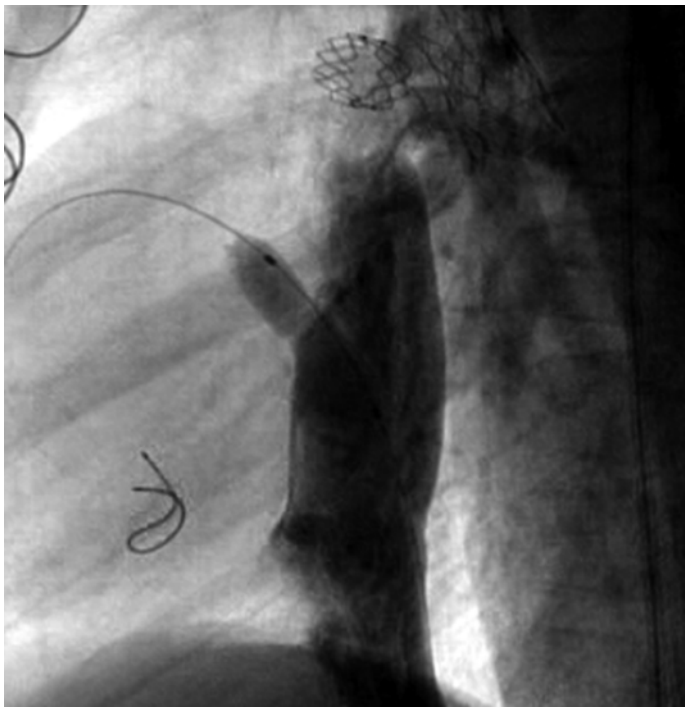

Fig. 27.2 Low-pressure balloon occlusion of 4.5 mm fenestration with a 6 mm Tyshak balloon; contrast injection through venous sheath confirms total occlusion

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.

27.8 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.
- 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. 27.3a, b) and on TEE if necessary. Haemodynamic measurements and saturations should be repeated.

27.9 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

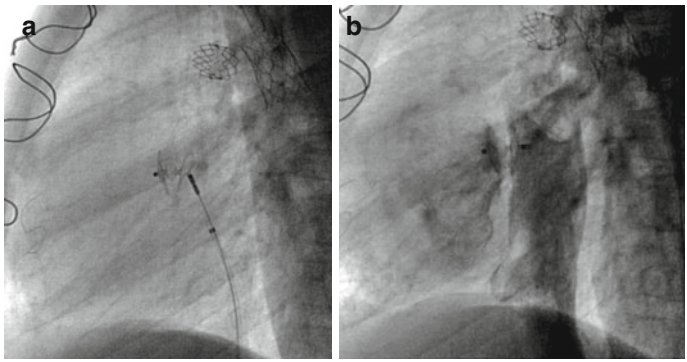

Fig. 27.3 (**a**) deployment of Amplatzer duct occluder type II in fenestration, device still attached to delivery cable. (**b**) cavogram after release of device: both disks are clearly at appropriate end of the fenestration; there is still some contrast like "smoke" through the device which will disappear within minutes after release

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.

27.10 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.

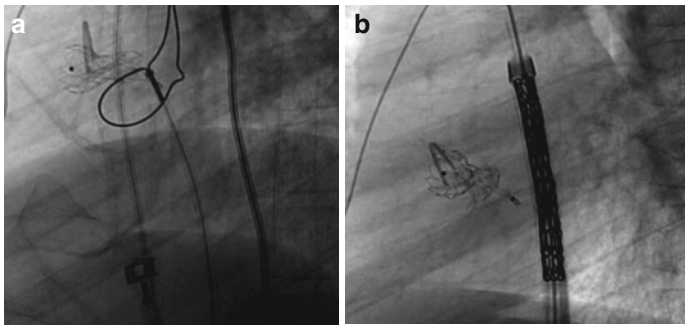

Fig. 27.4 Stented fenestration with previous attempt of closure with an Amplatzer duct occluder type II; now complete closure with a covered CP stent (see text)

- Future reclosing of these stented fenestrations after patients have improved haemodynamically might pose some challenges. Figure 27.4a 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. 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. 27.4a). 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. 27.4b), obtaining complete closure of the fenestration and a non-obstructive conduit.

27.11 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.

27.12 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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Chapter 28
Ventricular Septal Defects

Massimo Chessa and Gianfranco Butera

28.1 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, PVSD) with or without aneurism, and they can be native of 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.

M. Chessa (✉) • G. Butera
Pediatric and Adult Congenital Heart Center,
IRCCS Policlinico San Donato-University Hospital,
Via Morandi 30, San Donato, MI 20097, Italy
e-mail: massichessa@yahoo.it;
gianfranco.butera@grupposandonato.it

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Large defects give signs and symptoms of cardiac failure in early infancy, and they have to be treated surgically during 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 a VSD

A shunt is considered significant when the following are found:

(i) Left atrial enlargement, defined as a left atrial-to-aortic ratio >1.5
(ii) 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

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.

28.2 Patient Selection

- Absence of active infection: if a source of potential infection is found, 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 of 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.

28.3 Technical and Equipment Issues

28.3.1 Device for MVSD

The Amplatzer muscular ventricular septal defect occluder (Amplatzer Muscular VSD Occluder, AGA Medical Corporation, St. Jude, MN) 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) (Fig. 28.1). 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.

28.3.2 Device for PMVSD

The Amplatzer membranous ventricular septal defect occluder has two discs of unequal size (Amplatzer Membranous VSD Occluder, AGA Medical Corporation, St. Jude, MN). The aortic rim of the asymmetric left ventricular disc exceeds the dimensions of the connecting waist by only 0.5 mm, so as to avoid impingement on the aortic valve, whereas the apical end

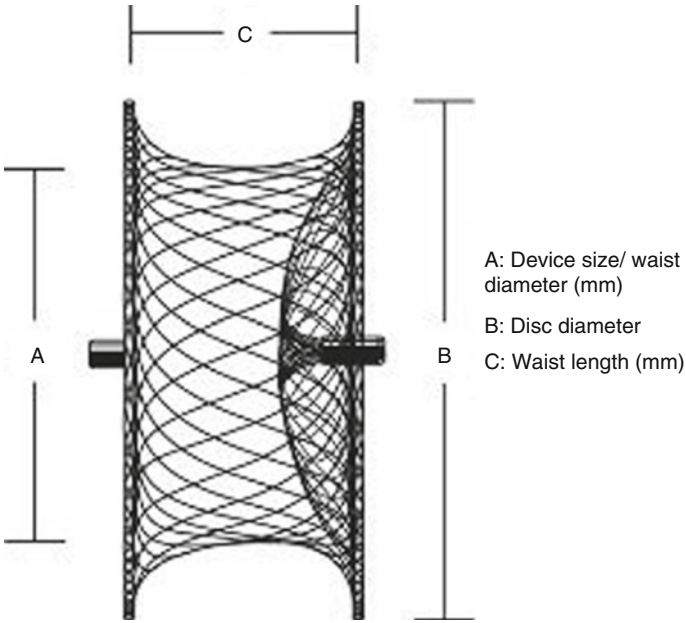

Fig. 28.1 The Amplatzer Muscular VSD Occluder, AGA Medical Corporation, St. Jude, MN. *A* device size/waist diameter (mm), *B* disc diameter (mm), *C* waist length (mm)

is 5.5 mm larger than the waist. This apical end of the left ventricular disc contains a platinum marker to facilitate correct orientation during implantation. The right ventricular disc is symmetrical, and it exceeds the diameter of the connecting waist by 2 mm throughout its circumference (Fig. 28.2). The device is available in sizes from 4 to 18 mm and requires delivery sheaths from 7 to 9 French.

A MVSD Amplatzer device II is also available. It was redesigned to prevent conduction abnormalities with a 75 % reduction in radial force, 45 % reduction in clamping force and 10 % increase in stability. The left disc of this new device has an

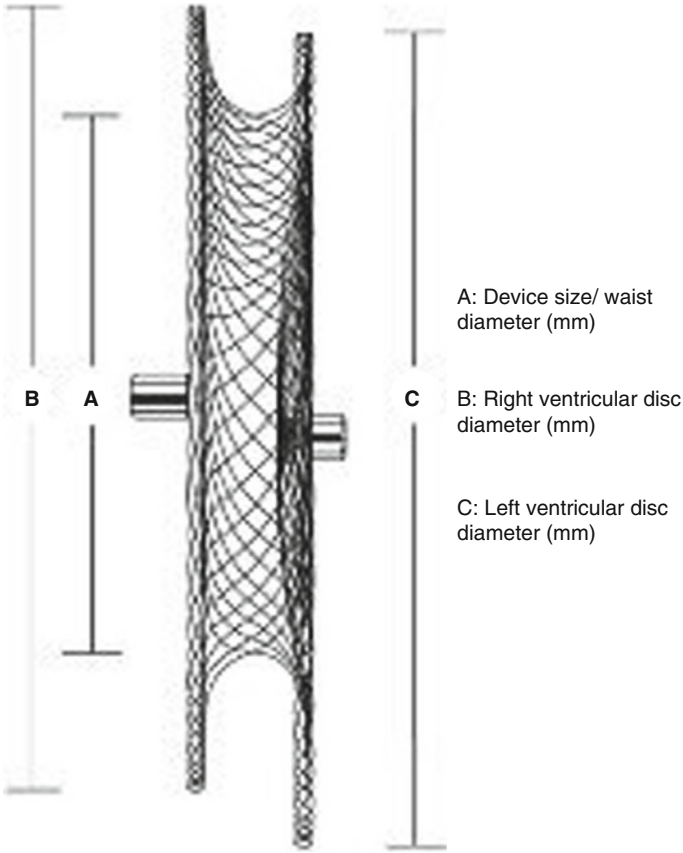

Fig. 28.2 The Amplatzer Membranous VSD Occluder, AGA Medical Corporation, St. Jude, MN. *A* device size/waist diameter (mm), *B* right ventricular disc diameter (mm), *C* left ventricular disc diameter (mm)

elliptical and concave shape that adapts to the LV outflow tract. It is available in two configurations:

1. Eccentric with a 1-mm superior rim and a 3-mm inferior rim
2. Concentric with a 3-mm superior and inferior rims

The waist length is increased from 1.5 to 3 mm, and the waist diameter ranges from 4 to 10 mm in 1-mm increments plus 12 and 14 mm. Polyester patches are sewn into the discs.

The delivery system consists of a delivery cable and a pusher catheter having a sharp curvature of 180° inferiorly. This allows correct orientation of the left ventricular disc during implantation. It has a flattened part of the socket that matches the flat portion of the microscrew, in order to force larger part of the left ventricular disc to be oriented downwards so that it points to the left ventricular apex.

28.4 Procedure

28.4.1 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 and vesical catheter for diuresis evaluation.
- A transoesophageal echocardiography 2D (2D-TEE) or 3D (3D-TEE) must be used to monitor the procedure.
- Heparinization with IV administration of 100 UI/kg heparin. Check hourly the activated clotting time >250 s. In case

administer heparin intravenously 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 has to wear masks and hats.

28.5 Access Site

- A femoral vein (FV) access is used to approach the closure of a PMVSD, and an internal jugular vein (IJV) can be used for MVSD closure.
- An arteriovenous circuit must be created (internal jugular vein-femoral artery for MVSD and femoral vein-femoral artery for PMVSD closure).
- Both sides for vascular femoral access are prepared.

28.6 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. 28.3).

The VSD is crossed from the left side by using a right Judkins catheter and a soft Glidewire (0.035″, J Tip, Terumo); the wire is advanced to the pulmonary artery or to the SVC/IVC,

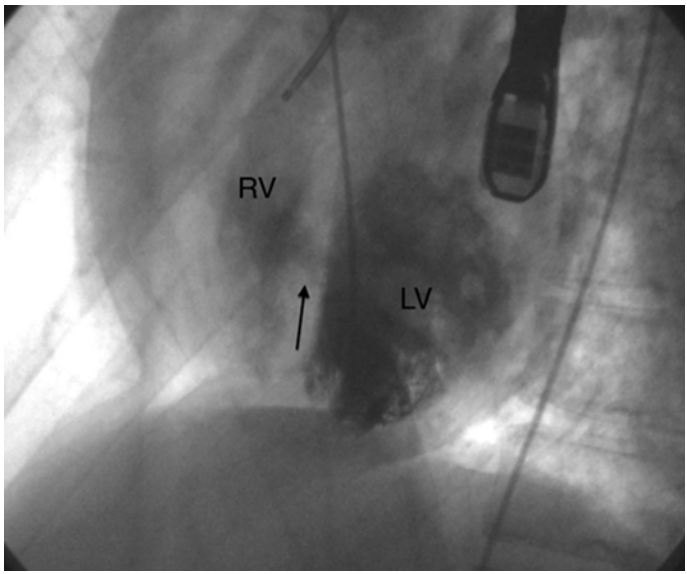

Fig. 28.3 Left ventricular angiogram: *LV* left ventricle, *RV* right ventricle, *Arrow* mid muscular VSD

where it is snared with a GooseNeck Snare (Microvena Corporation, 20–25 mm in adults, 10–15 mm in children) and exteriorized out of the right internal jugular vein or femoral vein establishing an arteriovenous circuit (Fig. 28.4).

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 to obtain additional information on the position and the size of the VSD.

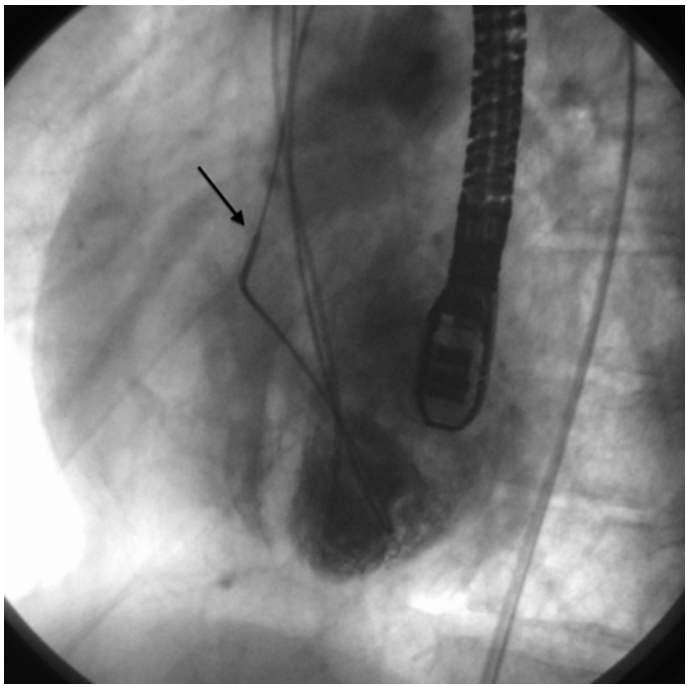

Fig. 28.4 Left ventricular angiogram: the *arrow* shows the arteriovenous circuit (femoral artery-internal jugular vein)

According to both angiographic and echocardiographic information, a muscular 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. 28.5a), and the central waist and the proximal disc are deployed; a test angiogram is done to verify the correct position of the device (Fig. 28.5b).

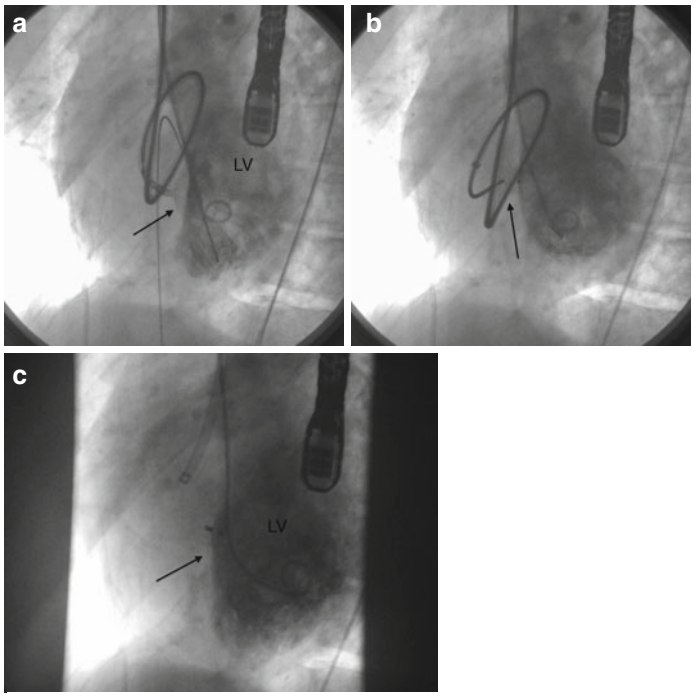

Fig. 28.5 Left ventricular angiogram: (**a**) The system is withdrawn towards the septum (*arrow*); (**b**) the central waist and the proximal disc are deployed (*arrow*); (**c**) a test angiogram is done to verify the correct position of the device (*arrow*). *LV* left ventricle

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. 28.5c)

Patients receive acetyl salicylic 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.

28.6.1 Alternative Techniques

28.6.1.1 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″ J Tip Terumo 260-cm exchange wire introduced through a 5-Fr Judkins right coronary artery catheter.

The wire is then advanced in the pulmonary artery. The catheter is exchanged with a 80-cm delivery sheath (AGA medical) over the wire to the right ventricle apex. The wire and dilator are removed slowly in order to avoid air suctioning.

The chosen device is prepared and advanced into the long sheath. The distal disc is opened in the RV apex paying attention to the ventricular wall and tricuspid wall.

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.

Left ventricular angiograms and echocardiographic evaluations are performed to confirm the position of the device and the absence of complications.

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.

28.6.1.2 Hybrid Approach

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).

The chest and the pericardium are opened, under TEE control; an 18-gauge 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 0.025 short guide wire 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-sized Amplatzer muscular VSD device is delivered using TEE monitoring

28.7 Catheterization and Haemodynamic Evaluation for PMVSD Closure

Angiographies are performed using 60° left atrial oblique plus 20° cranial view (Fig. 28.6).

An angiogram of the ascending aorta is also performed in 50° left atrial oblique view to check for aortic insufficiency.

The size of the defect, and its relationship to the aorta, is confirmed by 2–3D TEE (Fig. 28.7a, b).

The defect is crossed from the left ventricle by using a right Judkins or a right Amplatzer catheter and a Terumo wire. The catheter is advanced to the pulmonary arteries or to the SVC/IVC;

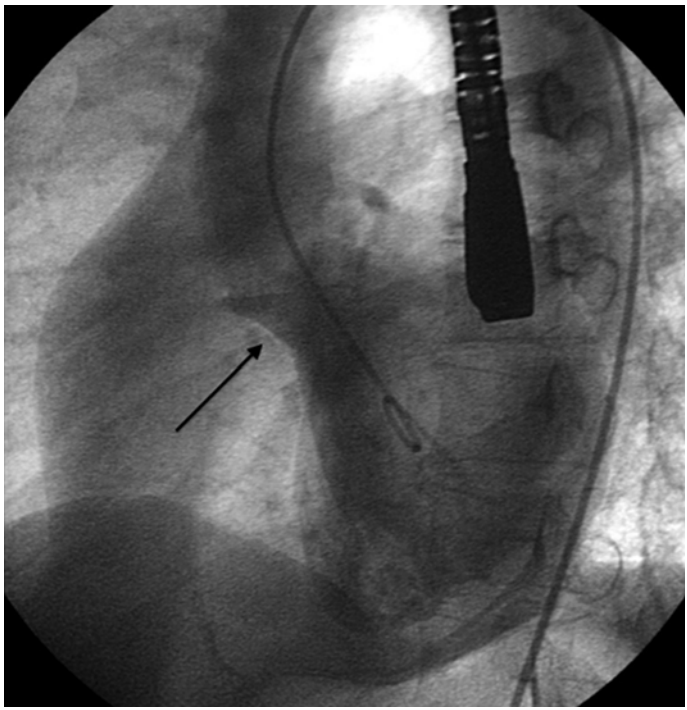

Fig. 28.6 Left ventricular angiogram: the *black arrow* shows the PMVSD

the Terumo wire is then replaced by the soft exchange noodle wire (a dedicated 300-cm exchange guide wire, AGA Medical Corporation, Golden Valley, MN), snared with a GooseNeck Snare, and exteriorized from the femoral vein (arteriovenous circuit) (Fig. 28.8a, b).

The AGA braided sheath is advanced over the wire up to the 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.

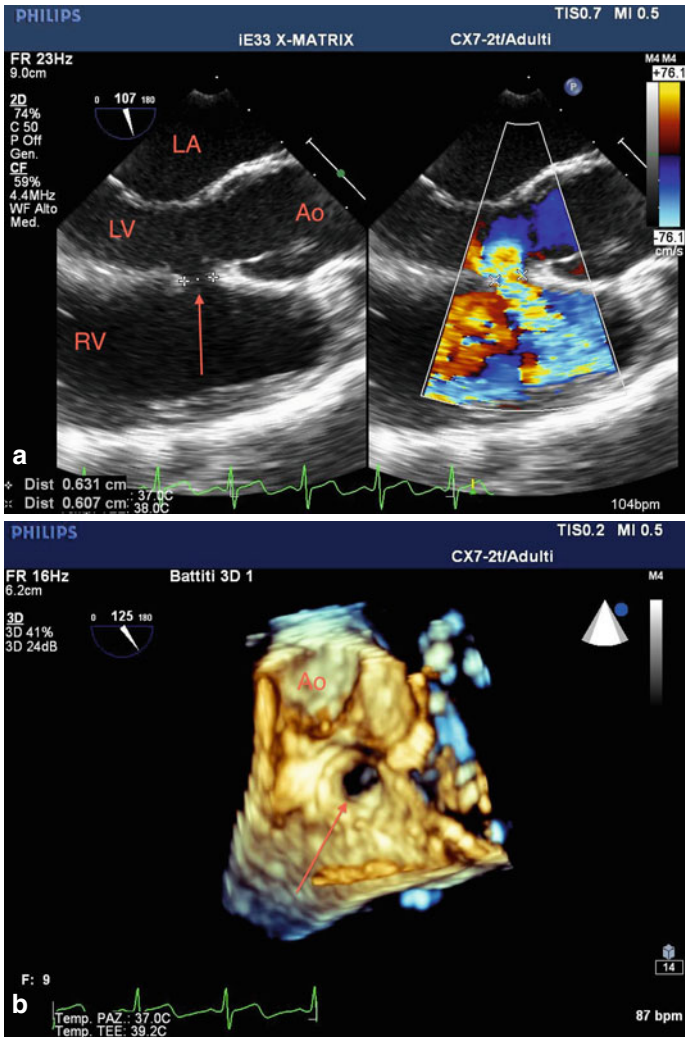

Fig. 28.7 (**a**) 2D-TEE long-axis view: *arrow*, PMVSD; *RV* right ventricle, *LV* left ventricle, *LA* left atrium, *Ao* aorta. (**b**) 3D-TEE: *arrow*, PMVSD; *Ao* aorta

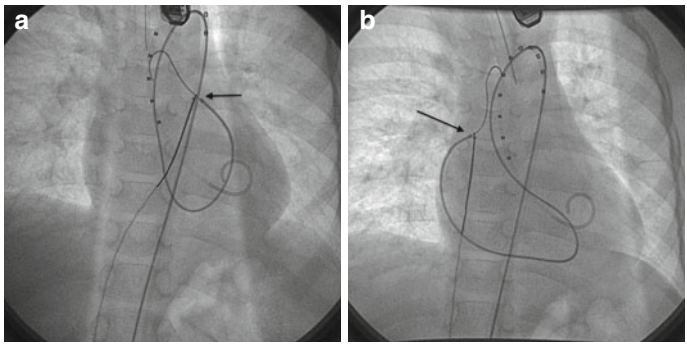

Fig. 28.8 The wire is snared with a GooseNeck Snare in the left pulmonary artery (**a**) or in the superior caval vein (*arrow*) (**b**) and exteriorized from the femoral vein (*arrow*) (arteriovenous circuit)

When the long sheath is in ascending aorta, hold the guide wire 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. 28.9).

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 microscrew 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. 28.10).

When the left disc is deployed, 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.

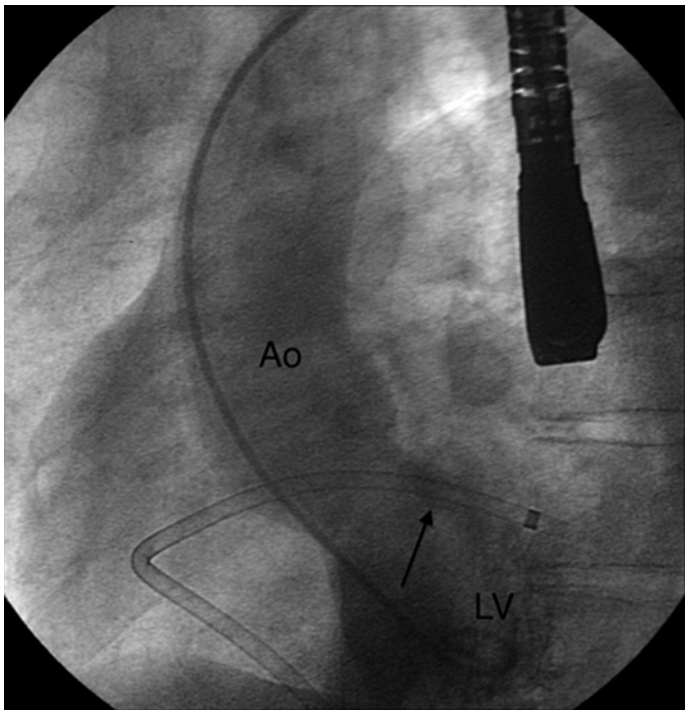

Fig. 28.9 The long sheath (*arrow*) into the left ventricle (*LV*). *Ao* aorta

The proximal disc is then deployed on the right side of the septum, and angiographic testing is done before releasing the device (Fig. 28.11a–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 and 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. 28.12a, b).

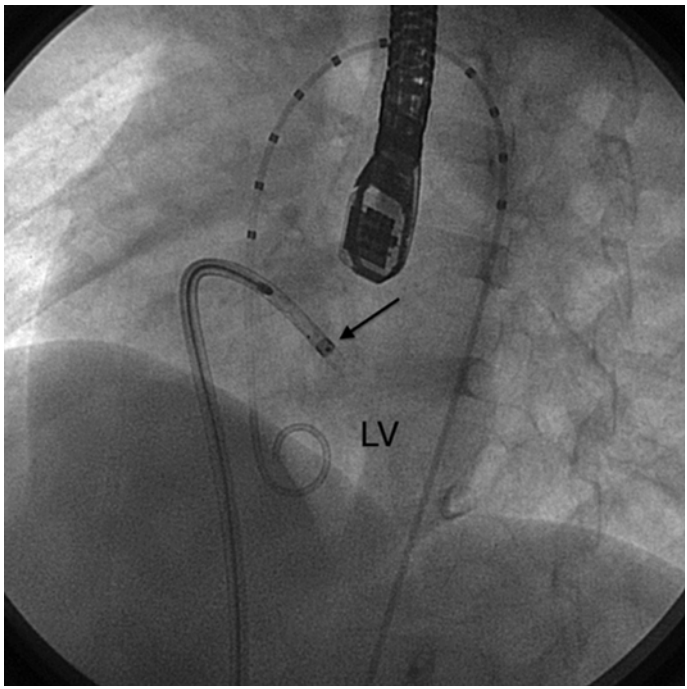

Fig. 28.10 The device is up to the tip (*arrow*) of the long sheath, and the entire system is withdrawn to the left ventricular outflow tract. *LV* left ventricle

After 10–15 min, the left ventricular angiogram and aortogram are repeated to assess possible residual shunting or aortic regurgitation (Fig. 28.13a, b). 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. 28.14).

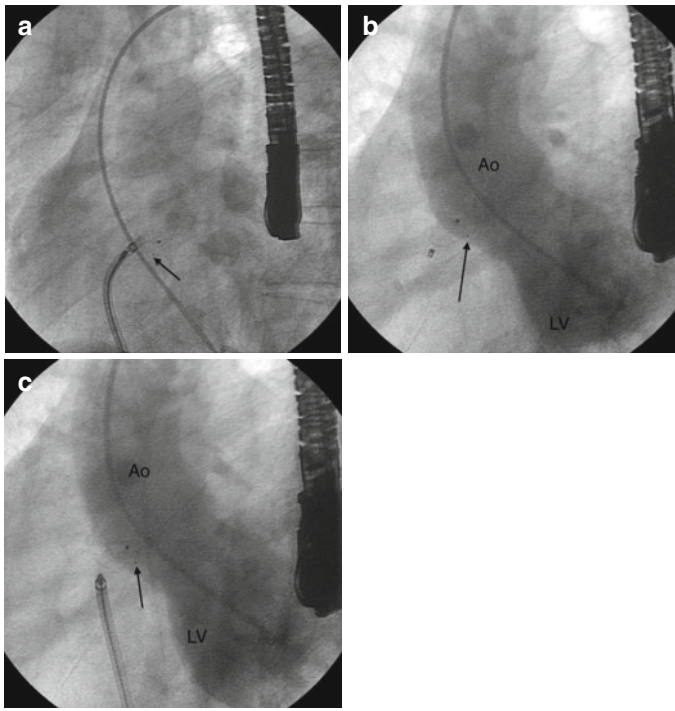

Fig. 28.11 Left ventricular angiogram: (**a**) the left disc is open and withdrawn to the interventricular septum. (**b**) The proximal disc is then deployed on the right side of the septum, and angiographic testing is done before releasing the device. The *arrow* shows the platinum marker of the distal disc pointing downwards. (**c**) The final angiography shows the complete closure of the defect. *Ao* aorta, *LV* left ventricle

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. 28.15).

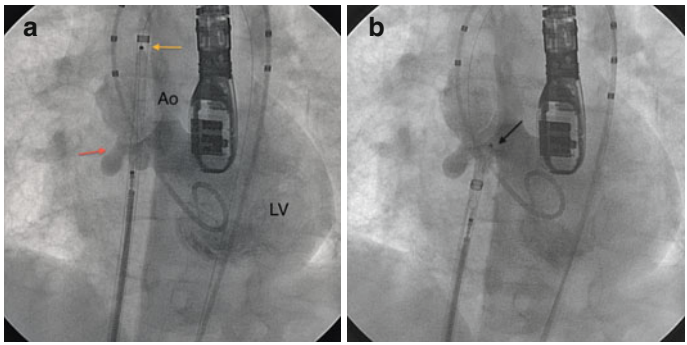

Fig. 28.12 Left ventricular angiogram: (**a**) the sheath can be left in the ascending aorta (*yellow arrow*). The *red arrow* shows the aneurysm of the PMVSD. (**b**) The left ventricular disc opened under the aortic valve while coming with the sheath from the aorta (*arrow*). *Ao* aorta, *LV* left ventricle

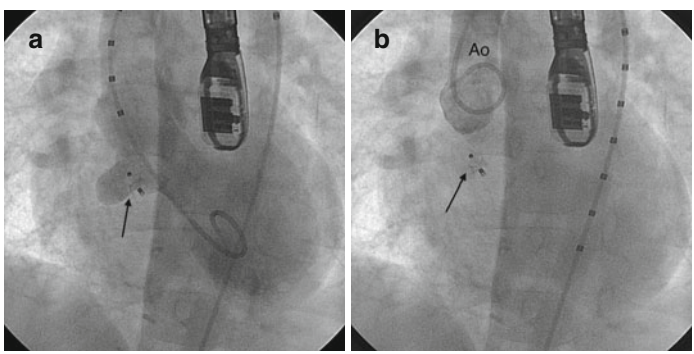

Fig. 28.13 The left ventricular angiogram (**a**) and aortogram (**b**) are repeated at the end of the procedure to assess possible residual shunting or aortic regurgitation. The *arrows* show the device setting into the aneurism. *Ao* aorta

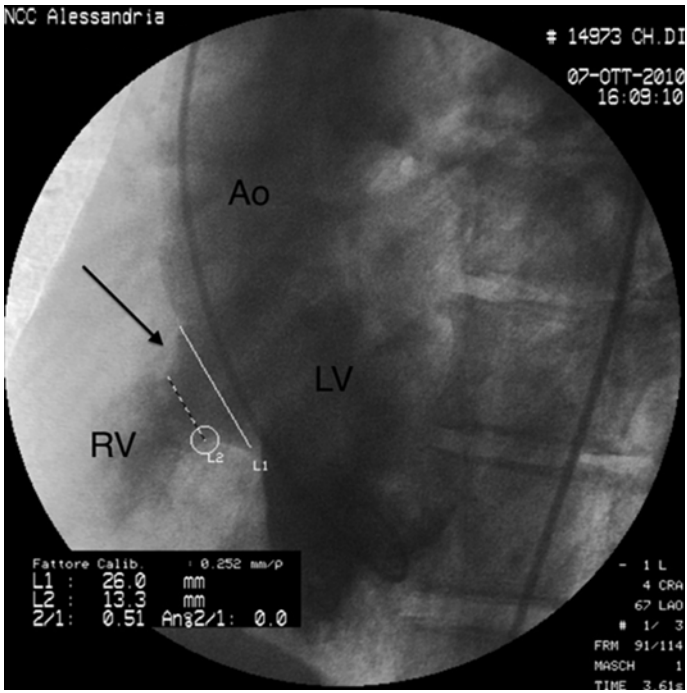

Fig. 28.14 Left ventricular angiogram showing a PMVSD with an aneurysm (*arrow*). *Ao* aorta, *LV* left ventricle, *RV* right ventricle

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 aneurism, different devices (PDA I AGA Medical Corporation, St. Jude, MN, or a Nit-Occlud® Lê VSD pfm medical ag.Köln, Germany) may be taken into consideration.

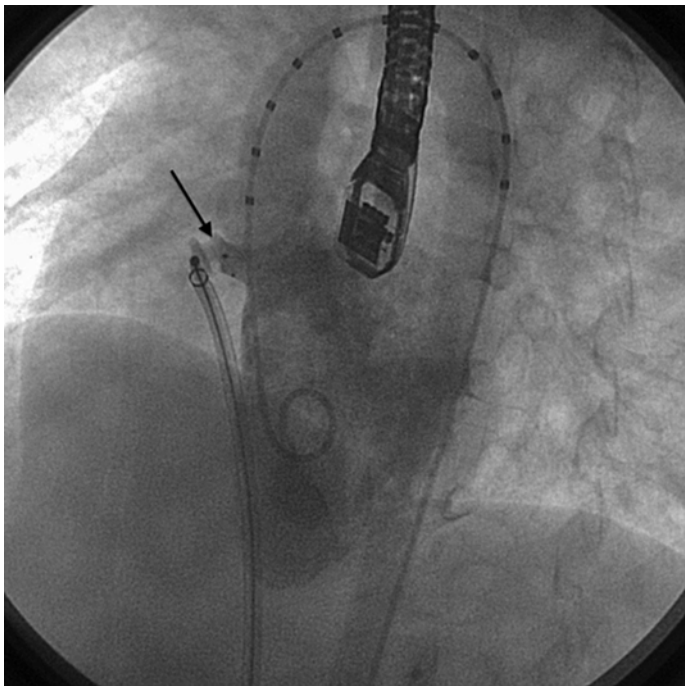

Fig. 28.15 Left ventricular angiogram. The *black arrow* shows the device deployed into the aneurysm

28.8 Specific Technical Aspects for Postsurgical Residual VSD

Balloon sizing of the defect: due to the varied anatomy of the substrate (presence of patches and patch leaks), sizing at the TEE and angiography can be more difficult. Balloon occlusion of the shunt and assessment with TEE and angiography provide 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.

The standard anterograde approach may be more difficult (presence of the surgical patch, less space in the subaortic region to deploy the LV disc and increased risk of complications).

A retrograde approach may overcome these issues.

28.9 Complications

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 usually was 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 a serious 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 oversizing the device size by more than 1 mm.

Suggested Reading

1. Chessa M, Carminati M, Cao QL, Butera G, Giusti S, Bini RM, Hijazi ZM (2002) Transcatheter closure of congenital and acquired muscular ventricular septal defects using the Amplatzer device. J Invasive Cardiol 14:322–327
2. Carminati M, Butera G, Chessa M, De Giovanni J, Fisher G, Gewillig M, Peuster M, Piechaud JF, Santoro G, Sievert H, Spadoni I, Walsh K (2007) Transcatheter closure of congenital ventricular septal defects: results of the European Registry. Investigators of the European VSD Registry. Eur Heart J 28(19):2361–2368
3. Butera G, Carminati M, Chessa M, Piazza L, Micheletti A, Negura DG, Abella R, Giamberti A, Frigiola A (2007) Transcatheter closure of perimembranous ventricular septal defects: early and long-term results. J Am Coll Cardiol 50(12):1189–1195

Chapter 29
Patent Ductus Arteriosus Closure

Ahmed Mohammed Alkamali

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.

29.1 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. 29.1). The most common is type A.
- Different approaches and devices may be needed in different morphologies.

A.M. Alkamali
Paediatric Department, Dubai Hospital, Dubai,
United Arab Emirates
e-mail: aalkamali@hotmail.com

G. Butera et al. (eds.), *Cardiac Catheterization for Congenital Heart Disease: From Fetal Life to Adulthood*,
DOI 10.1007/978-88-470-5681-7_29, © Springer-Verlag Italia 2015

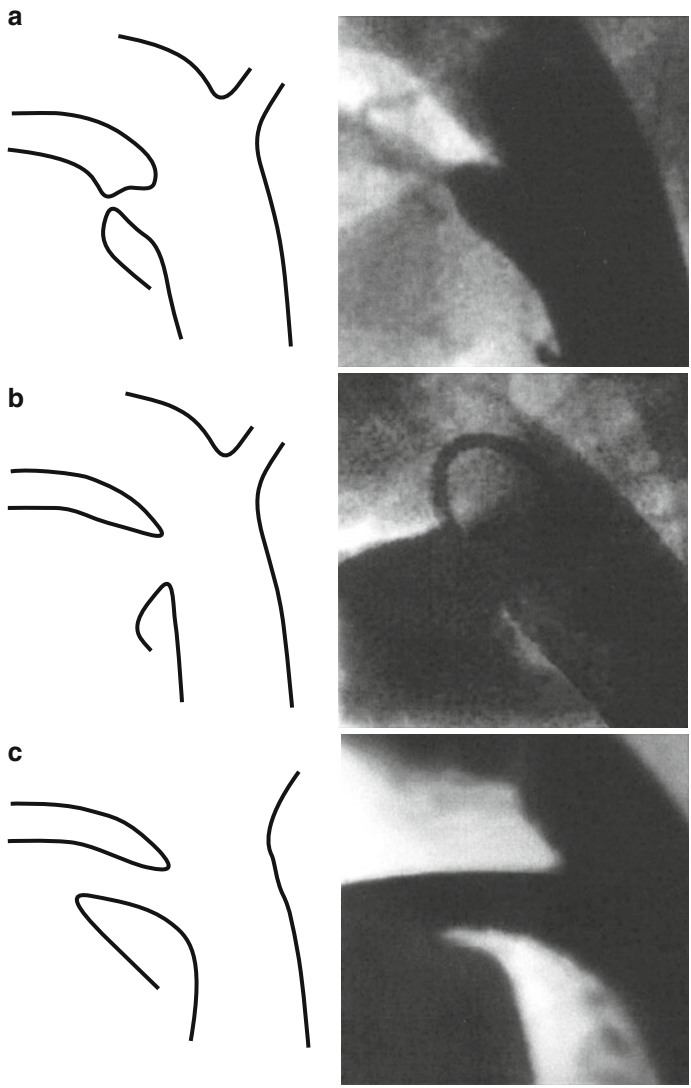

Fig. 29.1 Krichenko classification of PDA morphologies, (**a**) conical shape, (**b**) short and wide ductus "Window", (**c**) long tubular ductus without constriction, (**d**) with multiple construction " complex", (**e**) elongated ductus

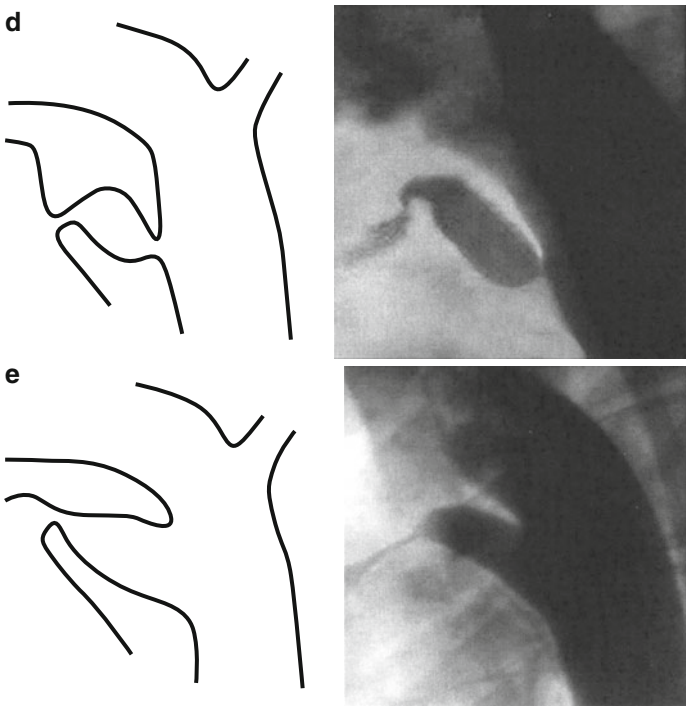

Fig. 29.1 (continued)

29.2 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.

- In symptomatic infants weighing less than 5 kg, surgical closure of the PDA is recommended. In fact, the use of 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.
- 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 contraindicated. 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.

29.3 Devices

29.3.1 Coil

- In this review, 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 3 loops up to 8 mm diameter by 5 loops. The label in coil package will have 2 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. 29.2 and 29.3).
- The Flipper Cook coil is made from 0.035″ wire and can go easily through 4-F end whole only multipurpose catheter that accommodates 0.038″ wire.

29.3.2 Devices

1. 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 pulmonary 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

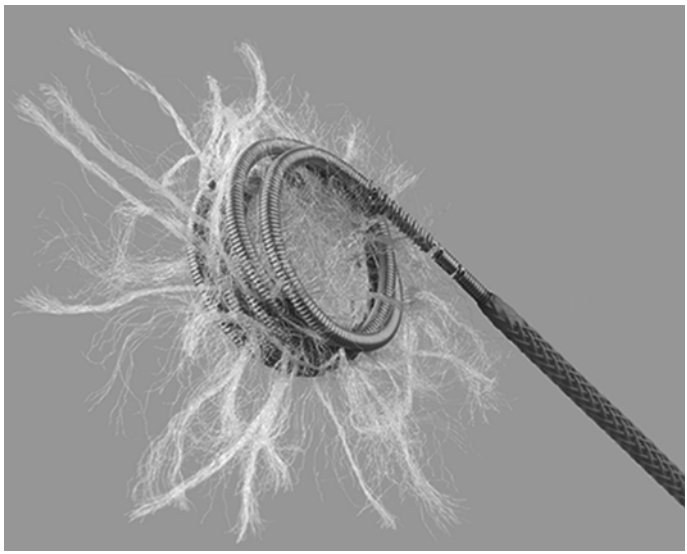

Fig. 29.2 Cook coil

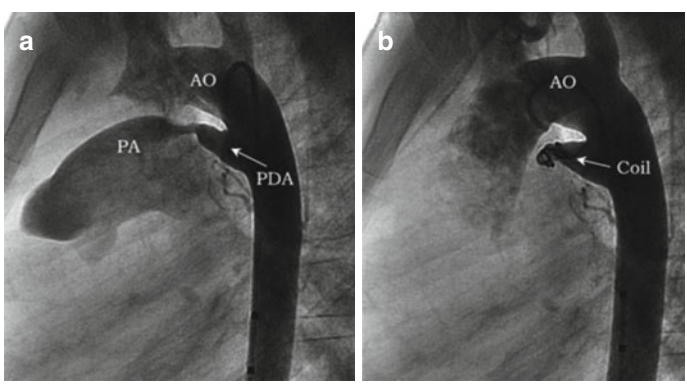

Fig. 29.3 Angiography in lateral view showing a small PDA (**a**) before and (**b**) after closure with detachable Cook coil

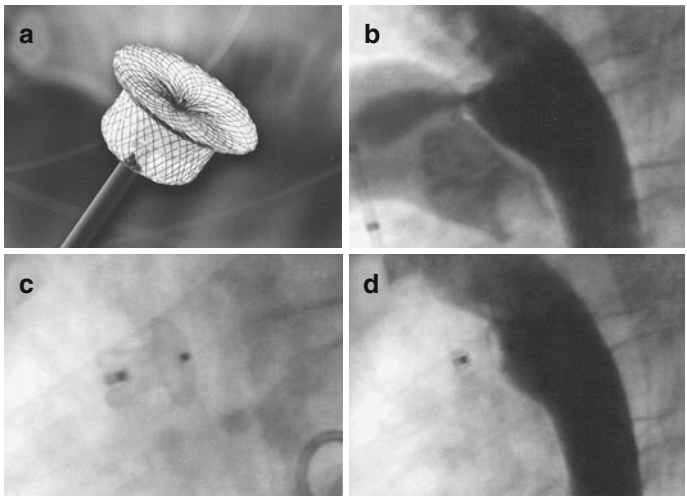

Fig. 29.4 (**a**) Amplatzer ADO I, (**b**) angiographic lateral view showing PDA, (**c**) ADO I in position, (**d**) aortography in lateral view showing complete PDA closure

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. 29.4 and 29.5).

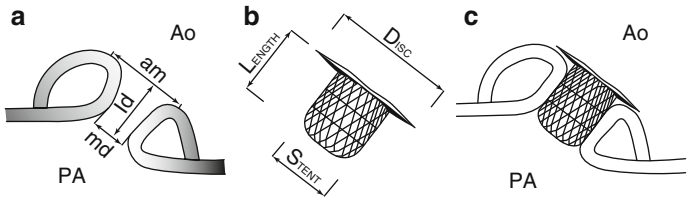

Fig. 29.5 Schematic view of duct measurements and ADO I occluding the PDA

2. The use of new 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, 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.
   - This design allows deployment from both the arterial and venous sides (Figs. 29.6 and 29.7).
   - 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. 29.6) 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.

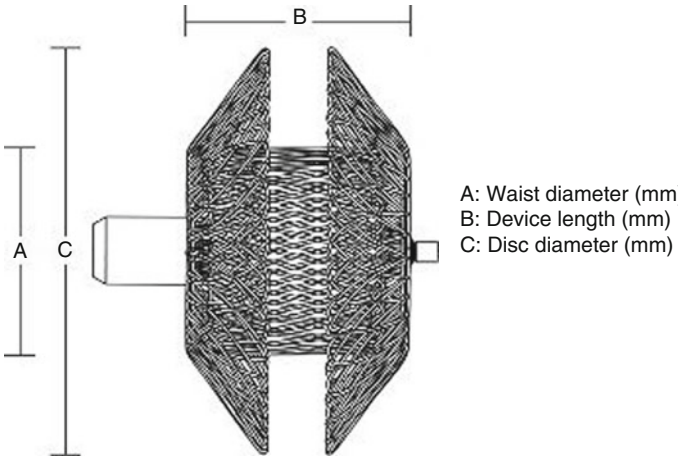

Fig. 29.6 Amplatzer Duct Occluder II. *A* Waist diameter (mm), *B* device length (mm), *C* disk diameter (mm)

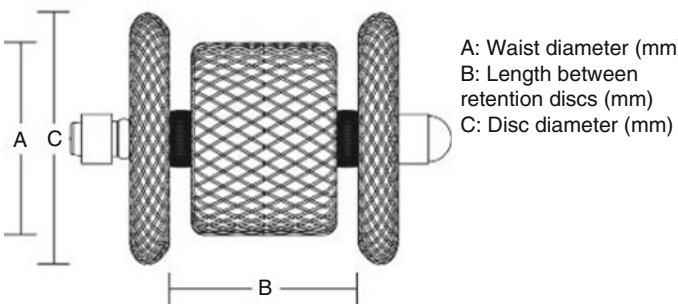

Fig. 29.7 Amplatzer Duct Occluder II AS. *A* Waist diameter (mm), *B* length between retention disks (mm), *C* disk diameter (mm)

- 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 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.

3. The ADO II additional size (ADO II AS) (Fig. 29.7) 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 allows 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 go through 4-Fr TorqVue LP delivery system.
   - 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 the more suitable device. For the short window-type PDA with no ampulla, the atrial septal occluder is more suitable.

29.4 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.
- 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.
- 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.
- 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).

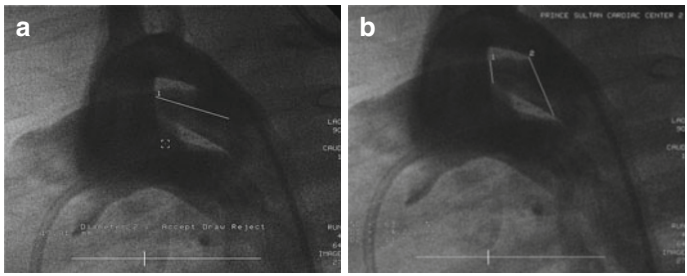

Fig. 29.8 PDA measurements: (**a**) pulmonary end, (**b**) length

- The device is chosen according to PDA morphology and narrowest diameter (Fig. 29.8).
- 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.

29.5 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. 29.2 and 29.3).

- 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 3 diameters of the PDA (total length, ampulla diameter, and narrowest diameter) (Fig. 29.8), 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 cathe-

ter. Under lateral projection fluoroscopy guide push the coil to the tip of delivery catheter in main pulmonary artery. When tip of coil out, withdraw the mandril till the coil completely free from mandril. You will noticed 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 to 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.

29.6 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 introducing and 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 cases 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 a 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 ADO II AS, the procedure of device delivery is very similar to ADO I.

- 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.

29.7 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.

29.8 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.

29.9 Device Embolization

Retrieval of an ADO device can be performed by using a 10–15-mm gooseneck snare and a Mullins Cook long sheath appropriate for the device.

Other Possible Complications

- Hemolysis
- Left pulmonary branch obstruction
- Aortic isthmus obstruction
- Be careful when closing large ducts in small babies!

Further Reading

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Chapter 30
Catheter Closure of Coronary Artery Fistula

Kothandam Sivakumar, Ajit Mullasari, and Bharat Dalvi

30.1 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,

K. Sivakumar, MD • A. Mullasari, MD
Department of Cardiology, Madras Medical Mission,
No 4A Dr J J Nagar, Mogappair, Chennai, Tamil Nadu 600037, India
e-mail: drkumarsiva@hotmail.com; sulu_ajit57@yahoo.co.in

B. Dalvi, MD (✉)
Department of Cardiology, Glenmark Cardiac Centre,
10 Nandadeep 209 D, Dr Ambedkar Road Matunga (E), Mumbai,
Maharashtra 400019, India
e-mail: bharatdalvi@hotmail.com

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fistulae may arise from more than one coronary artery. Over 90 % of the CAFs drain in the right heart chambers.

30.2 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.

30.3 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. 30.1).

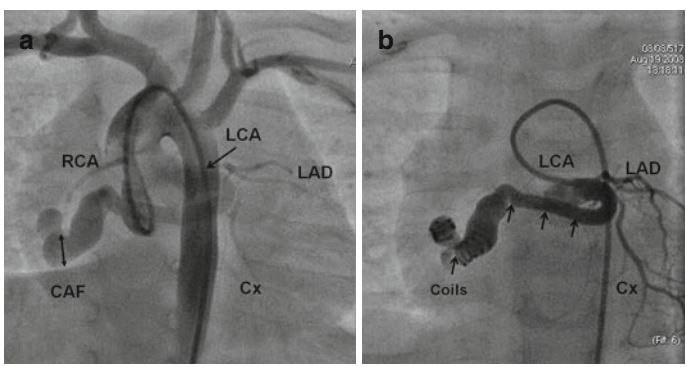

Fig. 30.1 (**a**) Aortic root angiogram in anteroposterior view shows tortuous coronary artery fistula (*CAF*) (*arrow*) arising from the left coronary artery (*LCA*) entering right atrium (*RA*). Normal branching of the LCA into the left anterior descending (*LAD*) and left circumflex (*Cx*) is well seen. Proximal portion of the right coronary artery (*RCA*) is also opacified. (**b**) It was closed with two Gianturco coils (*arrow*)

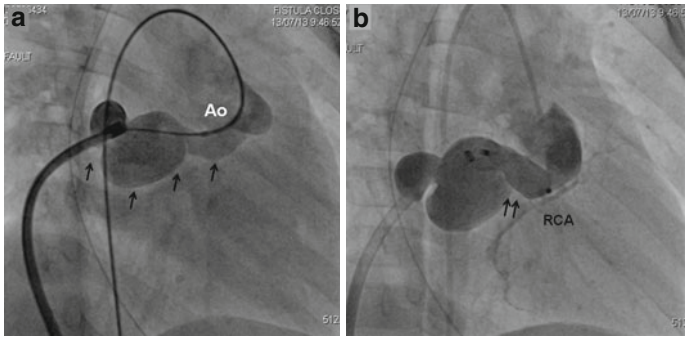

Fig. 30.2 (**a**) Angiogram through venous sheath after AV loop formation in right anterior oblique projection shows a large fistula (*arrows*) from the proximal part of right coronary artery (*RCA*) coursing posteriorly to enter the right atrium. Faint opacification of the aortic root (*Ao*) is also seen. The RCA branches are not well seen due to high flows through the fistula. (**b**) After closure with a 14–12 duct occluder I device (*arrows*), RCA fills well

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. 30.2).

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

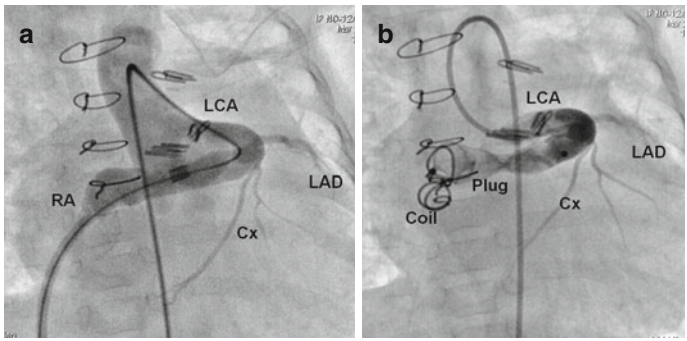

Fig. 30.3 (**a**) Angiogram through venous sheath after arteriovenous (A-V) loop formation in a patient with a significant residual flow from left coronary artery (*LCA*) to right atrial (*RA*) fistula after surgical ligation shows the long fistulous tract and normally branching LCA into the left anterior 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

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 fistula was prompted by persistent heart failure, growth failure, and recurrent pneumonia warranting ventilatory support (Fig. 30.3).

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. 30.4).

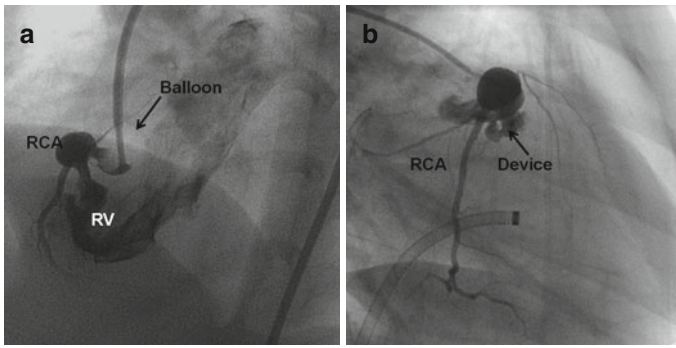

Fig. 30.4 (**a**) Aortogram in left anterior oblique view with cranial angulation using a balloon tipped catheter shows large fistula from proximal right coronary artery (*RCA*) to the right ventricle (*RV*). The branches of the RCA distal to the fistula are seen due to proximal balloon occlusion. (**b**) After closure with a duct occluder I, angiogram in right anterior oblique view shows 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. 30.5).

Case Study 6

An 8-year-old asymptomatic child with RCA to the RV fistula had entire RCA which was aneurysmally dilated to 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. 30.6).

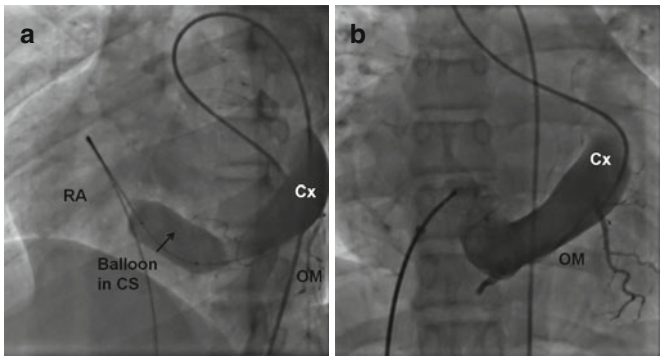

Fig. 30.5 (**a**) Selective angiogram in the left circumflex artery (*Cx*) after occluding the fistula opening into the right atrium (*RA*) by a 25 mm Tyshak II valvuloplasty balloon shows the obtuse marginal (*OM*) branches of the Cx. (**b**) After the distal end of fistula was closed with a large duct occluder I, the contrast is seen to opacify the Cx and OM branches more intensely

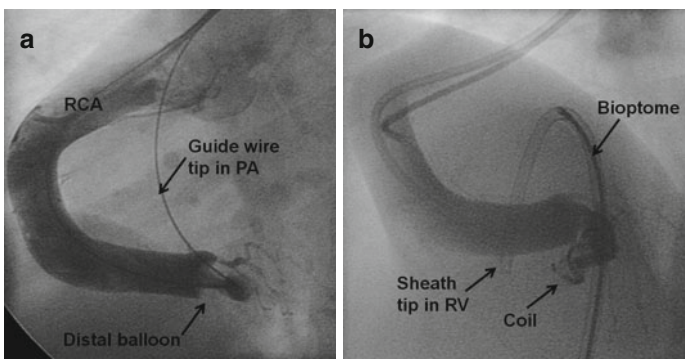

Fig. 30.6 (**a**) Selective right coronary artery (*RCA*) angiogram in left anterior oblique view shows a large RCA to right ventricle (*RV*) fistula. Through a second arterial access a guidewire was advanced through the fistula into the pulmonary artery and a distal balloon occlusion was done. The posterior 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

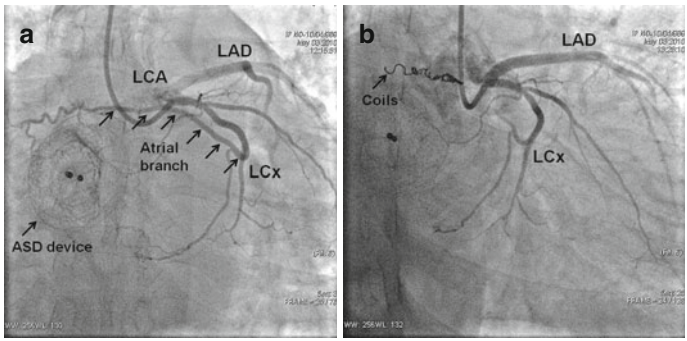

Fig. 30.7 (**a**) Left coronary artery (*LCA*) injection through right radial arterial access in right anterior oblique view shows a small fistula (*multiple arrows*) from the left circumflex artery (*LCx*) coursing posteriorly and terminating in the right atrium in a patient who had closure of atrial 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

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. 30.7).

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.

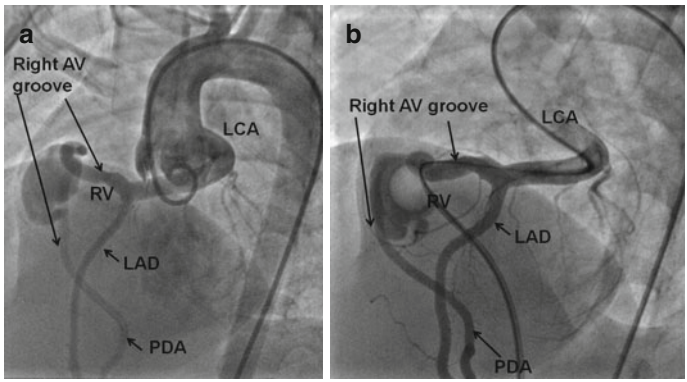

Fig. 30.8 (**a**) Aortic root angiogram in left anterior oblique projection shows absence of right coronary artery with faint opacification of the left coronary artery (*LCA*). A dilated left anterior descending (*LAD*) artery continues beyond the apex in the posterior interventricular groove as posterior 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*)

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 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. 30.8).

30.4 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

30.5 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.

30.6 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.

30.7 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. 30.9). This crucial information avoids surprises 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.

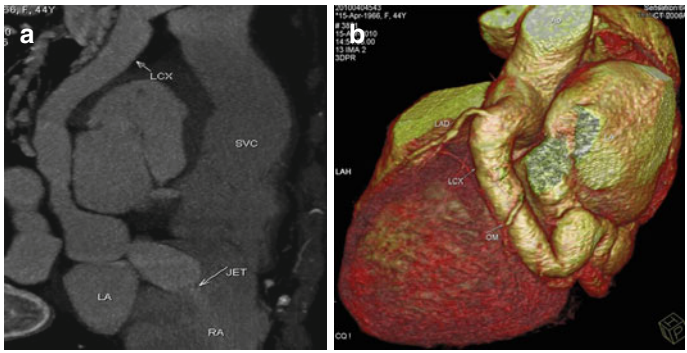

Fig. 30.9 (**a**) CT coronary angiogram showing a fistula arising from the left circumflex artery (*LCx*) coursing in front of the left atrium (*LA*) and draining into the RA just below the entry of superior vena cava (*SVC*). Severe restriction at the point of exit into the RA is producing a jet effect. 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

30.8 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. 30.7). 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 runoff 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. 30.8).
8. In high-flow fistula, the coronary branches are delineated better by proximal or distal balloon occlusion angiogram (Figs. 30.4, 30.5, and 30.8).
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. 30.4).
10. Distal occlusion with balloon floatation wedge catheters (Figs. 30.5 and 30.8) or compliant occlusion balloons (Fig. 30.6) is carried out after forming an arteriovenous

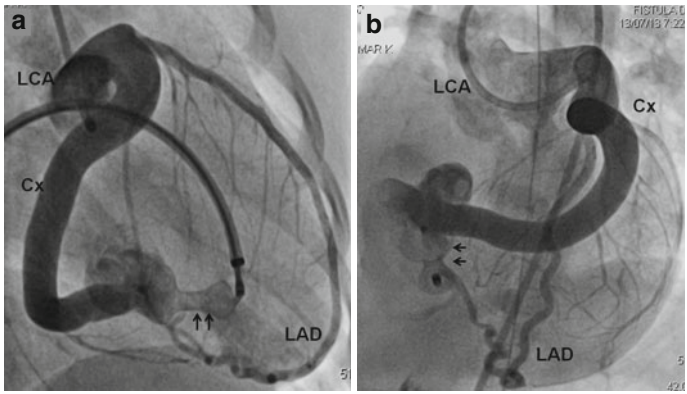

Fig. 30.10 Left coronary artery (*LCA*) injections in right anterior oblique projection (**a**) and left anterior oblique view (**b**) show a large feeder from the dilated left circumflex (*Cx*) artery and smaller additional feeder from the terminal part of the left anterior descending (*LAD*) artery 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*)

(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. 30.2). 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. 30.5 and 30.10).
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 Noodlewire (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. 30.5 and 30.6).

30.9 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)

30.10 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

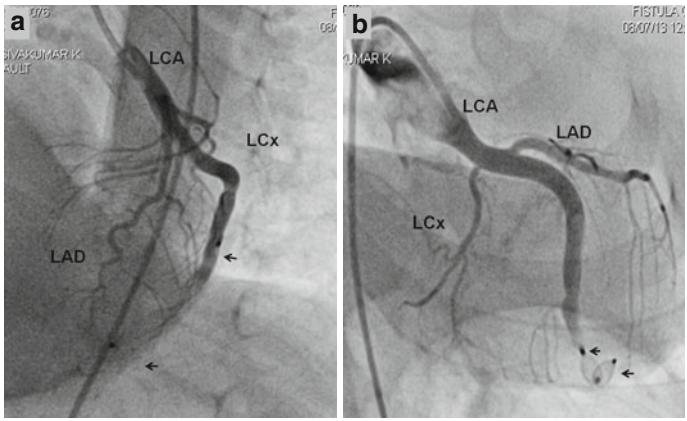

Fig. 30.11 (**a**) Left coronary artery (*LCA*) injection in left anterior oblique and (b) right anterior oblique views show a fistula from the ramus intermedius branch of the LCA arising between the left anterior descending (*LAD*) and the left circumflex (*LCx*) branches. This fistula was closed through 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.

catheters (Fig. 30.11). 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.
6. When multiple coils are chosen, they are intertwined together and delivered with the aid of bioptome in the desired location (Fig. 30.6). Bioptome helps in controlling the coil delivery.

30.11 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.

30.12 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.

30.13 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.

30.14 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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Chapter 31
Vessel Embolization: Transcatheter Embolization of Pulmonary Arteriovenous Malformations and Aortopulmonary Collateral Arteries

Liang Tang, Zhen-fei Fang, and Sheng-hua Zhou

31.1 Transcatheter Embolization of Pulmonary Arteriovenous Malformations

31.1.1 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.

L. Tang, MD • Z.-f. Fang, MD • S.-h. Zhou (✉)
Department of Cardiology, The Second Xiangya Hospital of Central South University, No.139, middle Ren-min Road, Changsha, Behind Hunan 410011, China
e-mail: tangliang1226@126.com; zhenfeifang@sohu.com; zhoushenghua_guo@163.com

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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.

PAVMs can be further characterized according to 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].

31.1.2 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.

31.1.3 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.

31.1.4 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 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.

31.1.5 Pre-procedural Imaging

31.1.5.1 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.

31.1.5.2 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.

31.1.5.3 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.

31.1.6 Technique (Step by Step)

31.1.6.1 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.

31.1.6.2 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 interventionalist. In general, PAVM with feeding artery diameters of 3–8 mm is treated with coils, whereas those with diameters ≥9 mm may be treated with Amplatzer duct occluder or vascular plugs.

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 coils are reported in the chapter on ductus arteriosus closure.

ADO

Before the advent of AVP, the 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. 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. 31.1a, b). The recent developed AVP IV (Fig. 31.1c) 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.

31.1.6.3 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.

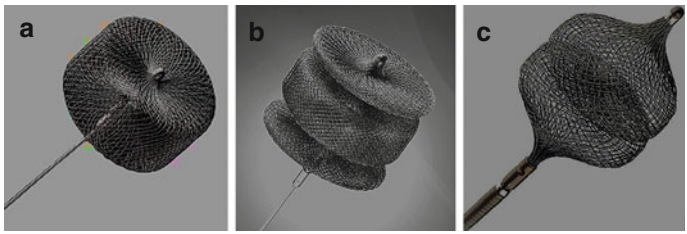

Fig. 31.1 Amplatzer plug I (**a**). Amplatzer plug II (**b**). Amplatzer plug IV (**c**)

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.

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.

Hand-injected angiography(usually 4 frames/s) 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.

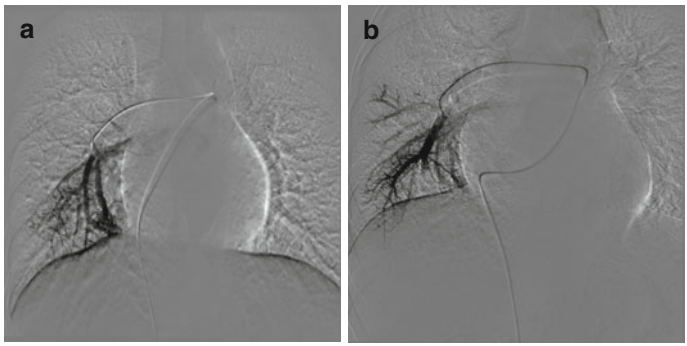

Fig. 31.2 (**a**) Pulmonary angiogram showing a PAVM of the right lower lobe. (**b**) Dense packing of two 6 mm coils producing complete occlusion

It is important to achieve as distal an embolization as possible in the feeding artery to avoid occluding branches to normal adjacent lung. 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 interventionalist 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. 31.2).

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].

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 microcatheter (Excelsior; Boston Scientific)) is advanced coaxially through the catheter into the aneurysmal sac. Several microcoils (0.018 in.) are densely filled within the venous sac until a large matrix is established.

31.1.6.4 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 time consuming.

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.

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 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.

The Amplatzer duct occluder (ADO) can also be an alternative for closing large PAVM (Fig. 31.3). 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.

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 might also require embolization.

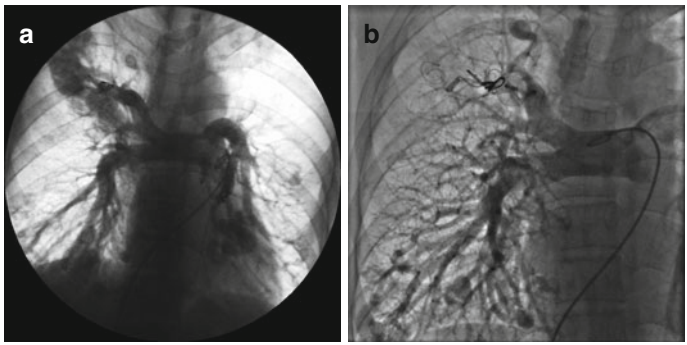

Fig. 31.3 A 29-year-old male with multiple PAVMs was referred for repeated PAVM occlusion. (**a**) Pulmonary angiogram revealed recanalization of previously embolized PAVMs. (**b**) PAVMs in the right upper and lower lobe were completely occluded with ADO

31.1.7 Expected Results

1. Pulmonary angiogram post-embolization 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.

31.1.8 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:

31.1.8.1 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.

31.1.8.2 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.

31.1.8.3 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 l5 min. A continuous flushing of catheters, observation for back-bleeding, and removal of catheters or wires "underwater" can largely prevent this complication.

31.1.8.4 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.

31.1.8.5 PAVM Recanalization

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, collateralization from adjacent arteries, or missed accessory pathways. Recanalization of PAVM can be attributed to coil elongation (use of oversized coils), poor coil packing, or use of an insufficient number of coils. This complication can be reduced by good closure technique and by selecting appropriate occluder for large PAVM.

31.1.9 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.

31.2 Transcatheter Embolization of Aortopulmonary Collateral Arteries

31.2.1 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.

They may be masked by another predominant cardiac lesion and are not discovered until after surgical repair of the major lesions.

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 (Fig. 31.4a–c).

They frequently run a retroesophageal course. Occasionally, the collateral arteries may arise from the coronary artery.

In patients with cyanotic congenital heart disease 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

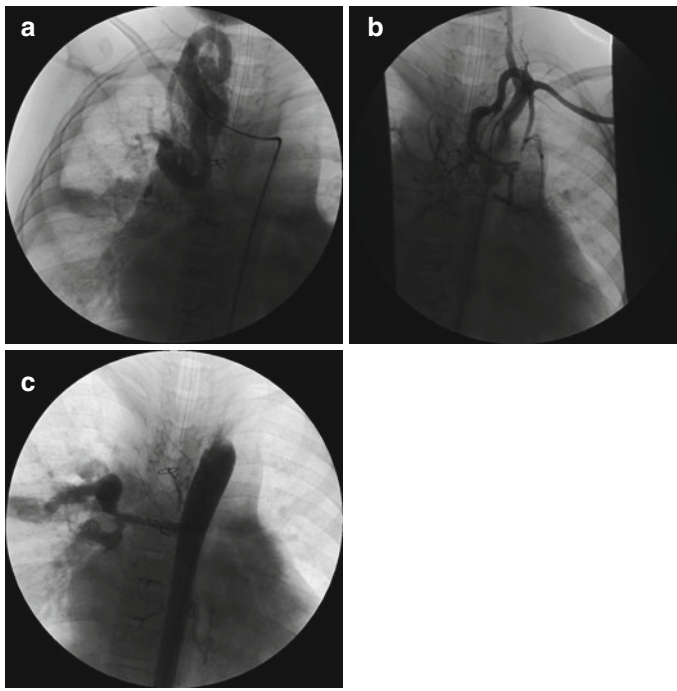

Fig. 31.4 A 10-year-old boy who had previously undergone surgical correction for TOF was referred for transcatheter occlusion of APCs. Selective angiogram in the innominate artery (**a**), the left subclavian (**b**), and the descending aorta (**c**) demonstrating multiple APCs. Repeated angiogram (**d**–**f**) after AVP and coil deployed, confirming complete occlusion of the major APCs

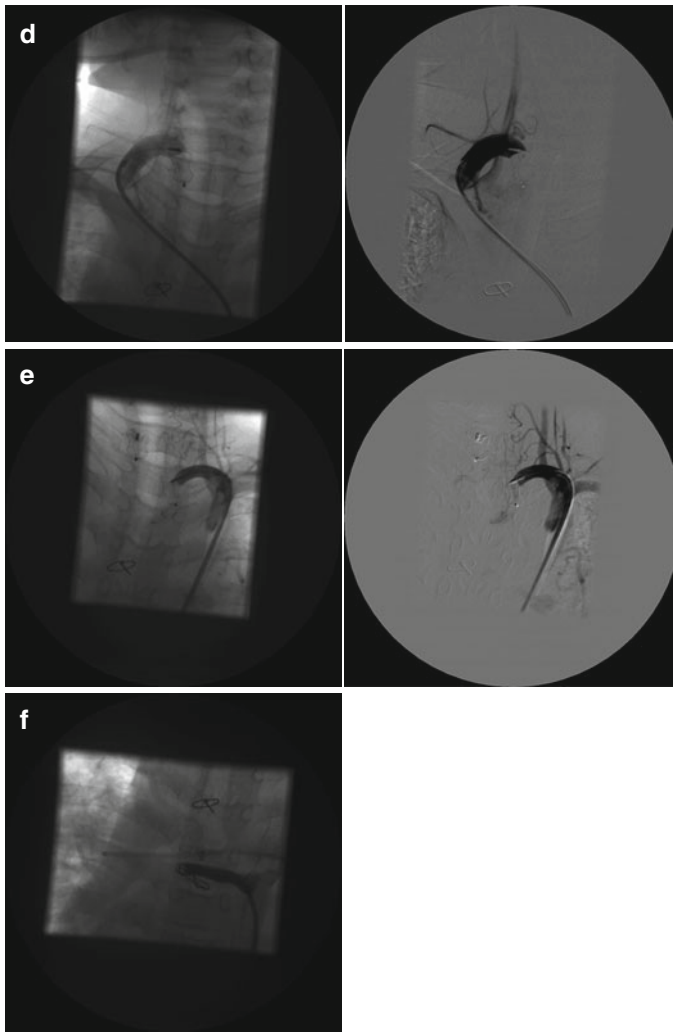

Fig. 31.4 (continued)

limit blood flow via the pulmonary arteries, and increase pulmonary arterial pressure and vascular resistance during the postoperative period of corrective surgery.

31.2.2 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.

31.2.3 Indications and Patient Selection [3]

31.2.3.1 Indications

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.

31.2.3.2 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.

31.2.3.3 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.

31.2.4 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.

31.2.5 Pre-procedural Imaging

Conventional angiography remains the gold standard for morphological assessment of the APCs. Noninvasive imaging modalities such as contrast-enhanced magnetic resonance angiography (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.

31.2.6 Technique (Step by Step)

31.2.6.1 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 circumstance, 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.

31.2.6.2 Occlusion Techniques and Devices

Currently, transcatheter occlusion of APCs is performed most commonly with detachable or undetachable coils and Amplatzer Vascular Plug (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 (Fig. 31.4d–f).

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.

31.2.7 Expected Results

1. Cardiac catheterization post-embolization 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.

31.2.8 Complications and Its Management

31.2.8.1 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.

31.2.8.2 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.

31.2.8.3 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 is not effective, the residual shunt should be eliminated by further embolization, and surgical removal is an alternative option.

31.2.9 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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Chapter 32
Closure of Residual Postsurgical Defects

Gerrit Kaleschke and Helmut Baumgartner

32.1 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

G. Kaleschke • H. Baumgartner (✉)
Division of Adult Congenital and Valvular Heart Disease,
Department of Cardiovascular Medicine, University Hospital Muenster,
Albert-Schweitzer-Campus 1, A1, Muenster 48149, Germany
e-mail: gerrit.kaleschke@ukmuenster.de;
helmut.baumgartner@ukmuenster.de

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overload. Inferior residual defects may cause right-to-left shunt in the 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. 32.1).

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

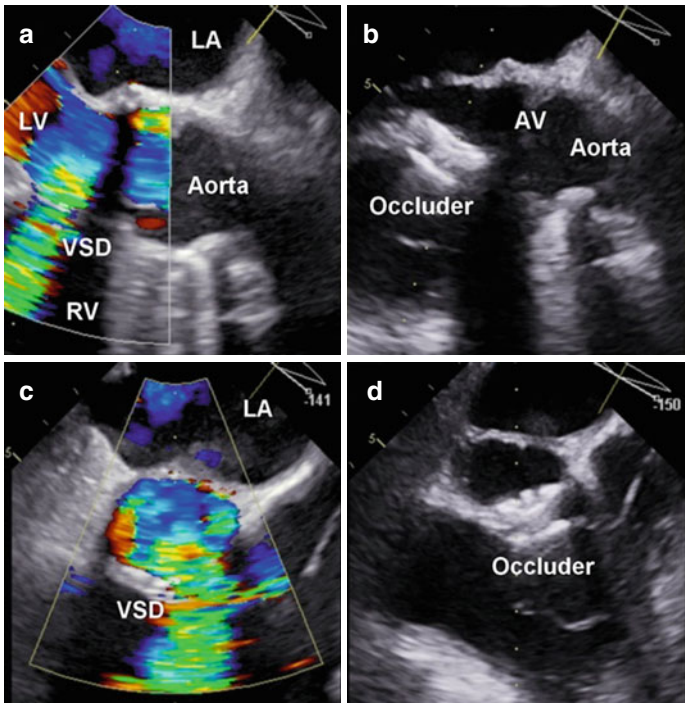

Fig. 32.1 Example of postsurgical muscular VSD closure. Transesophageal echocardiography of a postsurgical muscular VSD in a 73-year-old male after recurrent aortic valve surgery and myectomy (Qp/Qs ratio 2, evidence of pulmonary hypertension, (**a**) color Doppler long axis and (**b**) short axis view 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 (**c**: 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

are rare. Given the increased risk of thromboembolism after permanent pacemaker implantation, every patient after atrial switch operation should meticulously 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 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.

32.2 Clinical Scenario

Residual defects may be diagnosed in asymptomatic patients during routine follow-up visits. Patients may present 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.

32.3 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*

32.4 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 covered stents such as covered CP Stents™ (NuMed Inc. NY, USA).

32.5 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.

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.

32.6 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 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).

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 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, accessibility of the defect).

Although superior baffle leaks may be easier 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 guide wire 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 guide wire (e.g., Amplatz Extra Stiff, Cook Medical, IN, USA) is recommended.

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 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. 32.2). When covered stents are used for the treatment of leaks (especially in combination with baffle

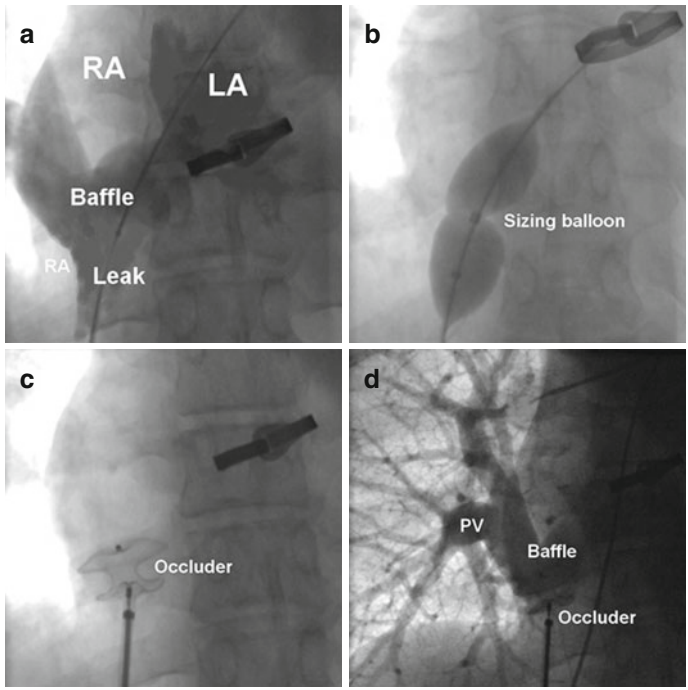

Fig. 32.2 Example of baffle leak closure. A 45-year-old female with correction of scimitar syndrome at the age of 10 years, recurrent embolic events (TIA, myocardial infarction without evidence of coronary artery disease, meningitis). Diagnosis of baffle leak based on contrast echo was confirmed during 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

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.

Multiple devices. If there are two or more adjacent leaks that necessitate the implantation of 2 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 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 circumscriptive 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. This issue has been addressed by the development of the membranous VSD Occluder 2 (not yet available) that may have a better profile that prevents damage to the conduction system (lower clamp force, softer outer waist, but also larger wing span to enhance stabilization of the occluder). 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. Beside the Amplatzer™ VSD devices, the Duct Occluder 2 may be an option and fit within the aneurysm with its left disk.

32.7 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.

32.8 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. 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.

32.9 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.

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Chapter 33
ASD Closure in Special Situations: Elderly, PA-IVS

Giuseppe Santoro, Luca Giugno, Cristina Capogrosso, Gianpiero Gaio, and Maria Giovanna Russo

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) submitted to right ventricular decompression.

33.1 ASD Closure in Elderly

Secundum type of atrial septal defect (ASD) is the most commonly found, previously undetected, congenital heart disease (CHD) in adulthood and elderly, accounting for nearly 80 % of ASDs [1]. In this subset of patients, atrial shunt tends to progressively increase over time due to physiologic decrease of left ventricular (LV) compliance and/or associated chronic diseases,

G. Santoro, MD (✉) • L. Giugno • C. Capogrosso • G. Gaio • M.G. Russo
Pediatric Cardiology and G.U.C.H Unit,
A.O.R.N. "Ospedali dei Colli", II University of Naples,
Via Camillo Sorgente, 31, Salerno 84125, Italy
e-mail: santoropino@tin.it

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such as aortic valve sclerosis, essential arterial hypertension, and coronary artery disease. These pathophysiologic changes may cause progressive increase of pulmonary over-circulation and relative LV preload decrease and deconditioning, so resulting in high risk of atrial arrhythmias and progressive drop of systemic cardiac output.

Thus, elderly patients with significant atrial shunt tend to be often highly symptomatic due to pulmonary hypertension, atrial arrhythmias, and low LV output, thereby indicating ASD closure at any age. In this subset of patients, percutaneous approach might be highly advisable to avoid sternotomy and postsurgical respiratory rehabilitation. Furthermore, percutaneous ASD closure results in significant positive remodeling both of right and left cardiac chambers, as well as clinical improvement of short- and long-term *cardiopulmonary function*.

Another significant advantage of transcatheter approach is the potential to "test" pathophysiologic consequences of defect closure by temporary *balloon test occlusion* before final device deployment. In fact, abolishing atrial shunt results in significant sudden changes of both pulmonary and systemic circulations. Indeed, abrupt decrease of pulmonary blood flow in a high-resistance pulmonary circulation might be poorly tolerated by the right ventricle (RV) and may result in decrease of pulmonary venous drainage and, hence, systemic cardiac output. At the same time, sudden decrease of left chamber preload may be poorly tolerated by an under-trained LV, so potentially resulting in pulmonary edema.

33.1.1 Indication

Elderly patients potentially candidate to ASD closure should be always submitted to a thorough baseline *hemodynamic evaluation* as well as pharmacologic/mechanical tests, in order to evaluate amount and site of *pulmonary vascular resistance* and to check their potential changes during vasodilator challenge or

temporary shunt closure as well as to highlight LV behavior during temporary balloon-driven ASD closure.

33.1.1.1 Elderly Subjects with Increased Pulmonary Artery Pressures

According to current guidelines, pulmonary hypertension is defined as a systolic pulmonary artery pressure >40 mmHg and a mean pressure >25 mmHg. Patients with significant shunt (signs of RV volume overload) and PVR <5 WU should undergo ASD closure regardless of symptoms.

Patients with PVR >5 WU but <2/3 SVR or PAP <2/3 systemic pressure (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy) and evidence of net L–R shunt (Qp:Qs > 1.5) may be considered for intervention

33.1.1.2 Elderly Subjects with Reduced LV Compliance

Before cardiac catheterization, there are several criteria that may anticipate abnormal compliance of LV:

Age >60 years
Left ventricular hypertrophy
Restrictive physiology at echocardiographic Doppler evaluation

Balloon occlusion test is a critical step in patients with borderline LV compliance, in whom a sudden increase of LV preload may precipitate pulmonary edema and systemic low output.

The following data obtained during balloon testing are considered as relative contraindications to shunt closure:

- Persistent 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 post-expiratory peak pressure during mechanical ventilation or breath fatigue in awake patients)

This subset of patients may benefit from various approaches:

- Partial ASD closure with a fenestrated device
- 3–5 days trial with intravenous anti-congestive drugs (diuretics and ACE inhibitors) or 3 months trial with oral drugs (diuretics and ACE inhibitors) 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.

33.1.2 Technique

Technical steps of transcatheter ASD closure in elderly are not significantly different from what widely described in younger patients [2]. However, based on the previous pathophysiologic considerations, the procedure is quite more time-consuming and challenging as well as demands some additional tips.

- In this view, a critical step to reliably record the hemodynamic data is adequacy of respiratory pattern, obtained either with a comfortable position in awake patients or optimizing mechanical ventilation in anesthetized ones.
- Systemic artery and left ventricle pressures should be recorded by small-size catheters both at baseline and during provocative tests.
- Right atrial, pulmonary artery, and pulmonary capillary wedge pressures as well as systemic pressure must be recorded at the end of a quiet respiratory cycle. SpO_2 in

superior vena cava, inferior vena cava, pulmonary artery, and femoral artery must be measured in triplicate. Pulmonary venous saturation must be measured in patients with ASD or assumed as 96 % in the remaining cases, according to medical literature on CHD–PAH. Pulmonary and systemic blood flows can be obtained with the Fick principle using table-derived oxygen consumption values and calculated oxygen content at the corresponding sites in order to rule out residual shunts.

- Finally, pre-procedural diagnostic work-up of the elderly patient potential candidate to ASD closure should always include coronary angiography and/or intracoronary pressure and/or flow recordings. Indeed, the significant increase of LV volume load resulting from atrial shunt closure might potentially unmask subclinical, borderline coronary artery stenoses.
- Temporary balloon occlusion test for 15 min may mimic final device deployment.
- In our opinion, ASD dynamic balloon occlusion from right atrial aspect (Equalizer Balloon Occlusion Catheter, Boston Scientific, Natick, Massachusetts, USA) is preferable to static balloon occlusion (AGA sizing balloon, AGA Medical Corporation, Golden Valley, MN, or NuMED sizing balloon, NuMED, Hopkinton, NY) in that less interfering with volume and compliance of the left heart chambers.
- 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.
- Left ventricular diastolic pressures and systemic arterial pressures are carefully evaluated before, during, and after balloon testing (see before) (Fig. 33.1).
- Pulmonary artery pressures are evaluated during and after balloon testing in subjects with high baseline values.

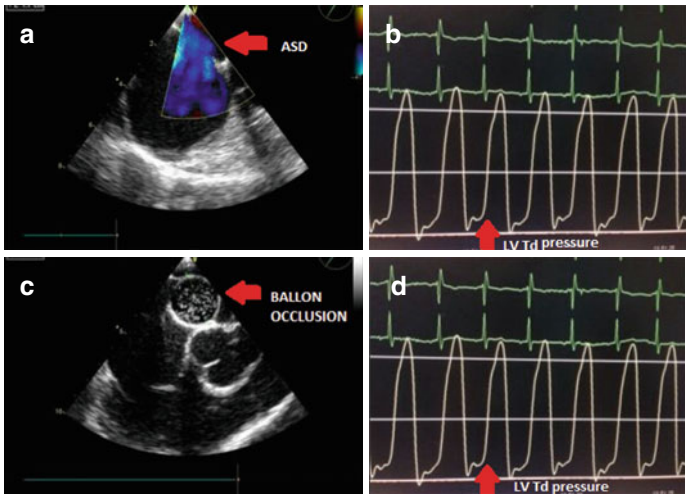

Fig. 33.1 Percutaneous closure of large ASD in adult. (**a**) Large ASD with significant left-to-right shunt at color Doppler analysis. (**b**) LV end-diastolic pressure before ASD temporary occlusion. (**c**) Dynamic ASD balloon occlusion test performed from the right atrium. (**d**) No change in LV end-diastolic pressure during balloon inflation is recorded

- In patients with chronic atrial fibrillation, percutaneous ASD closure may be followed by DC shock conversion and prophylactic anti-arrhythmic therapy, hoping that a favorable long-term atrial remodeling might maintain sinus rhythm over long-term follow-up.
- If needed, fenestration is created in the device.
- Fenestration is 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.
- Inhaled 100 % oxygen, via a rebreathing mask, was the most commonly used agent to test pulmonary artery vasoreactivity.

It is now more common that acute vasodilator testing be performed using short-acting vasodilators such as inhaled nitric oxide, intravenous administration of epoprostenol, or adenosine. An acute reduction of mean pulmonary arterial pressure as high as >10 mmHg, with resultant mean pulmonary arterial pressure drop to less than 40 mmHg without fall in cardiac output, is considered as positive vasoreactivity response.

33.1.3 Follow-Up

ASD closure is expected to abolish systemic-to-pulmonary shunt, reduce pulmonary artery pressure, and significantly increase systemic output, thereby resulting in steadily and progressively clinical and functional improvement also in elderly. However, arrhythmic risk of these patients does not seem to change over time, although a favorable atrial remodeling might halt the trend toward atrial instability. Mortality and morbidity of percutaneous ASD closure in elderly are not significantly different from younger age but pharmacologic long-term therapy with pulmonary vasodilators or anti-congestive drugs might be advisable in patients with borderline pulmonary hypertension or LV compliance, respectively. In subjects with a fenestrated device, the fate of fenestration is usually spontaneous closure within a few months.

33.2 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 unloading by

percutaneous or surgical valvotomy. RV hypoplasia and abnormal compliance almost always burden on long-term pathophysiology of this malformation [2, 3].

In this pathophysiologic setting, right-to-left atrial shunt occurs whenever right atrial pressure is higher than left atrial pressure and atrial shunt might act as a safety valve either to unload the right chambers or to increase the systemic ventricle output.

Thus, ASD closure may be hemodynamically dangerous and clinically poorly tolerated, resulting in systemic pressure venous increase (particularly harmful at hepatic and renal level) and systemic output decrease due to drop of LV filling volume.

33.2.1 Indication and Procedure

Ideally, atrial shunt closure should be always performed in patients with PA-IVS submitted to RV decompression. This approach avoids the inevitable right-to-left shunt caused by a borderline right chamber compliance resulting in systemic hypoxia during effort or potential paradoxical embolization.

Indication to ASD closure derives from baseline clinical and instrumental findings, as well as uneventful balloon occlusion test of the septal defect.

Anatomic and/or functional findings of RV unsuitability to biventricular physiology, such as *tricuspid valve hypoplasia* (z-score >–3), RV hypoplasia (bipartite morphology or severe apical hypertrophy), and 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.

Procedure

- Two venous lines and one arterial access are obtained.
- The first step is complete right and left heart catheterization.
- Transesophageal echocardiographic and fluoroscopic monitoring are needed.
- Balloon occlusion testing is performed and maintained for 10–15 min.
- Ideally, balloon occlusion test should be performed from the left atrial aspect in order to avoid any interference with the right chambers volume and compliance. Therefore, dynamic balloon testing is preferable to static ASD occlusion.
- The following parameters are monitored: *central venous pressure*, systemic arterial pressure, and systemic oxygen saturation.
- In ideal conditions, oxygen saturation should increase to >94 %, systemic arterial pressure should not decrease to >20 %, and central venous pressure should not increase to >20 % as compared to baseline values.
- However, in case of borderline changes, a short-term course of diuretics may be given to proceed to a second attempt of closure some few days after or a short-term (3–6 months) trial of diuretic therapy may be set up after device deployment. In alternative, device fenestration could be considered.
- ASD/PFO closure in PA-IVS is performed following a routinely, well-described technique (Fig. 33.2).

33.2.2 Follow-Up

Scanty data are so far published about long-term follow-up following ASD/PFO closure in PA-IVS [3], but the theoretical advantages deriving from right-to-left shunt closure in terms of restoration of normal oxygen saturation at rest, avoidance of desaturation during exercise, and improvement of work capacity seem to be maintained over long-term follow-up.

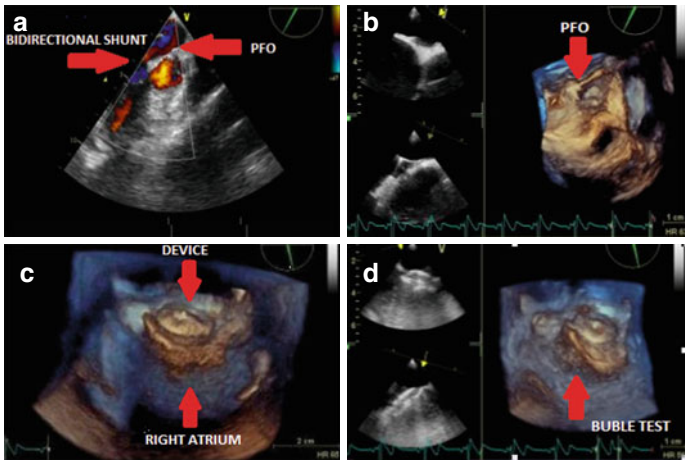

Fig. 33.2 PFO closure in a patient with PA-IVS submitted to RV decompression. (**a**) Bidirectional shunt at level of a tiny PFO as imaged at color Doppler analysis. (**b**) 3D transesophageal echocardiography showing the patent foramen ovale probed by a guide wire during the occluding test and (**c**) after the device implantation. (**d**) The bubble test after the final device deployment fails to show any right-to-left shunt

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Chapter 34
Creating an Interatrial Communication

Derize E. Boshoff 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 perfusion, 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

D.E. Boshoff (✉) • M.H. Gewillig
Department of Pediatric Cardiology, University Hospital Leuven,
49 Herestraat, Leuven 3000, Belgium
e-mail: derize.boshoff@uzleuven.be;
marc.gewillig@uzleuven.be

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to create or enlarge an interatrial communication. These include balloon atrial septostomy, blade atrial septostomy, static balloon dilation of the septum, and radiofrequency 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 may provide a more durable solution.

34.1 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.

34.2 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 radiofrequency (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.

34.3 Imaging Techniques

34.3.1 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 positional abnormalities. Several different angiographic projections have been described to best visualize the interatrial septum during transseptal procedure (discussed in detail in Chap. 15).

34.3.2 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.

34.4 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.

34.4.1 Balloon Catheters

Balloon atrial septostomy (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).

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].

34.4.2 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 balloon atrial septostomy 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.

34.4.3 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 pre-shaped 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.

34.4.4 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 radiofrequency (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.

34.5 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.

34.6 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 preexist-

ing 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.

34.7 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. 34.1). Balloon diameter will depend on patient/atrial size and underlying cardiac anomaly.

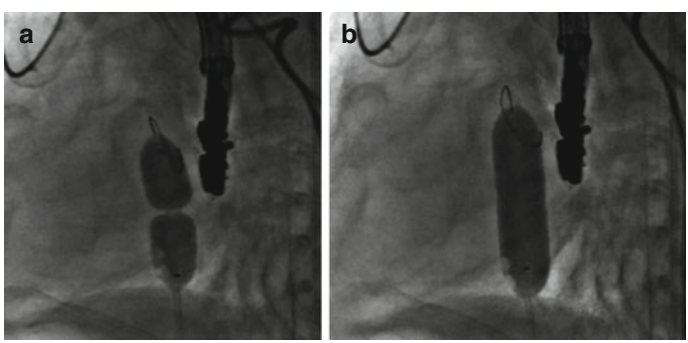

Fig. 34.1 Static balloon dilation of restrictive atrial communication (**a**) 12 mm balloon inflated at mild pressure delineating the small atrial communication (**b**) full inflation stretches-tears the septum.

34.8 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.

34.8.1 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. 34.2). In contrast to conventional balloon atrial septostomy, stent implantation requires anti-aggregation treatment (acetylsalicylic acid 2–5 mg/kg/day) to prevent thrombus formation.

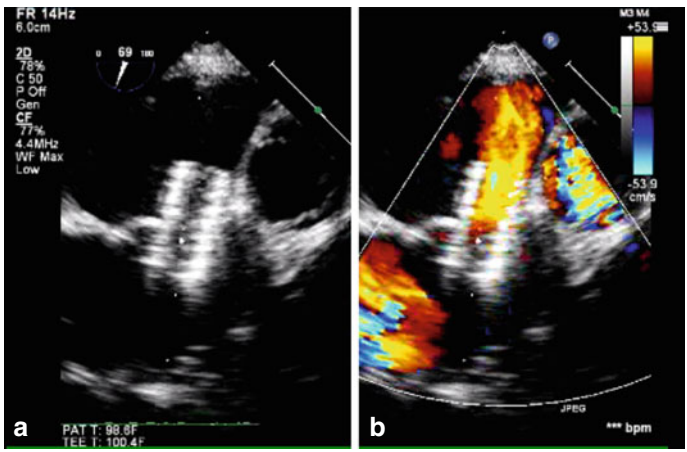

Fig. 34.2 TEE of stent across atrial septum (**a**) a Genesis 1910 stent is nicely positioned across the atrial septum (**b**) color flow mapping shows right-to-left shunt across the stent

34.9 Stenting of the Interatrial Septum: Restrictive Technique

34.9.1 Pulmonary Arterial Hypertension

Atrial septostomy for severe pulmonary arterial hypertension (PAH) improves cardiac index and functional class by the creation 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.

34.9.1.1 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. 34.3). The stent is then manually crimped, and its stability tested.

34.9.1.2 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. 34.4). 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%.

34.9.2 Fontan Circulation

Secondary fenestration of a failing Fontan circulation is a valuable technique to improve the hemodynamic condition of the

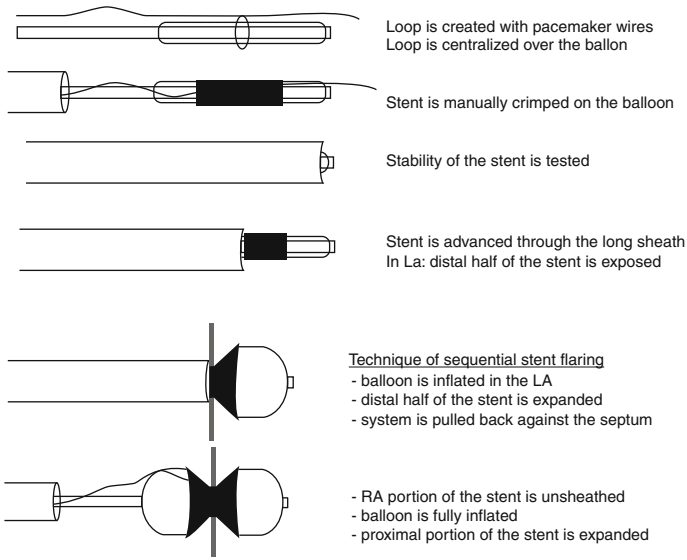

Fig. 34.3 Cartoon with various steps for a diabolo stent

patient. The fenestration is created to allow a restrictive 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

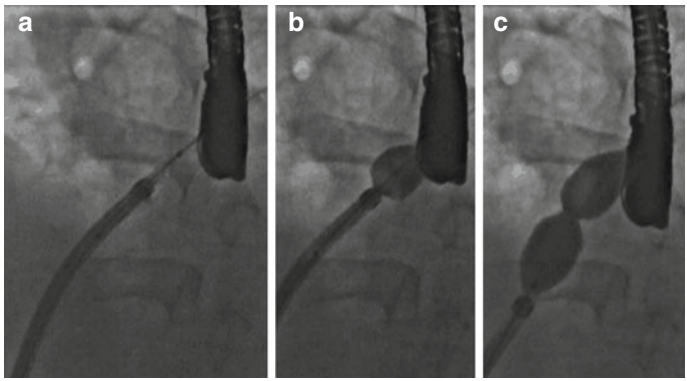

Fig. 34.4 Deployment of a 1910 Genesis stent (**a**) 10F sheath into left atrium, stent is partially uncovered (**b**) inflation of 15 mm balloon results in flaring of distal end of the stent (**c**) after pulling whole system against the septum, the sheath is pulled back to uncover the RA end, and further inflation of the balloon results in diabolo shape of the stent across the atrial septum

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.

34.10 Complications

Complications of balloon atrial septostomy 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.

34.11 Conclusions

Creation or enlargement of interatrial communications can be achieved using a variety of transcatheter techniques including transseptal needle puncture or RF perforation, balloon septos-

tomy, 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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Part VII
Step-by-Step Procedures: Valve Implantation

Chapter 35
Melody Valve Implantation in Pulmonary Position

Gianfranco Butera, Massimo Chessa, and Philipp Bonhoeffer

35.1 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.

Despite of this, the consensus indications are:

1. RV systolic pressure >2/3 systemic with clinical symptoms
2. RV systolic pressure >3/4 systemic without clinical symptoms

G. Butera (✉)
Pediatric and Congenital Heart Disease, Policlinico San Donato IRCCS, Via Morandi 30, San Donato Milanese, Milan 20097, Italy
e-mail: gianfranco.butera@grupposandonato.it

M. Chessa
Pediatric and Adult Congenital Heart Center, IRCCS Policlinico San Donato, Via Morandi 30, San Donato Milanese, Milan 20097, Italy
e-mail: massimo.chessa@grupposandonato.it

P. Bonhoeffer
Fondaxione Toscana Gabriele Monasterio, CNR Regione Toscana, Pisa, Italy

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3. Severe pulmonary valve regurgitation associated with one or more of the following:
    - RV dysfunction and/or dilatation (assessed on echocardiography and/or cardiovascular magnetic resonance imaging (cMRI: RVEDvolume index >140–150 ml/m^2)
    - Decreased exercise capacity (peak VO_2 <65–70 % predicted for normal)
    - Arrhythmias: atrial or ventricular sustained arrhythmias

35.2 Patient Selection

- Absence of active infection.
- Check and treat all sources of potential infection: submit the patient to dental checkup.
- Complete and deep analysis of previous medical history, cardiac catheterization, and surgeries: in particular check surgical notes.
- Check renal and hepatic function, and blood cell count.
- Candidates for Melody valve have a conduit or an anatomical stenosis in the RVOT or main pulmonary artery.
- Check MR and/or CT.
- Check if associated anomalies are present: pulmonary branch(es) stenosis, bifurcation stenosis, residual intracardiac defects, coronary position being a potential risk factor for compression during the procedure.
- The ideal candidate for Melody valve should have a dysfunction of a previously implanted conduit ranging between 16 and 22 mm in diameter.

35.3 The Valve and the Delivery System

35.3.1 Device

The Melody transcatheter pulmonary valve (Medtronic, USA) 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 34 mm CP stent and a diameter of 18 mm that can be crimped to a size of 6 mm and re-expanded from 18 mm up to 22 mm (Fig. 35.1). 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

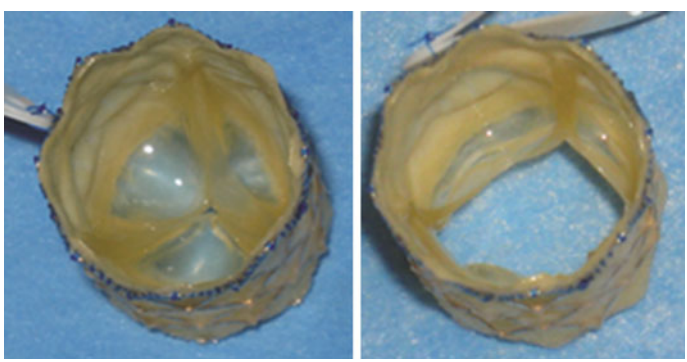

Fig. 35.1 Melody percutaneous pulmonary valve, in the close (*left*) and open (*right*) position, mounted on a platinum-iridium stent

insertion. The suture is clear colored 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 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.

35.3.2 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. 35.2). The system is available with three outer balloon diameters: 18, 20, and 22 mm. The tip of the system is blue to

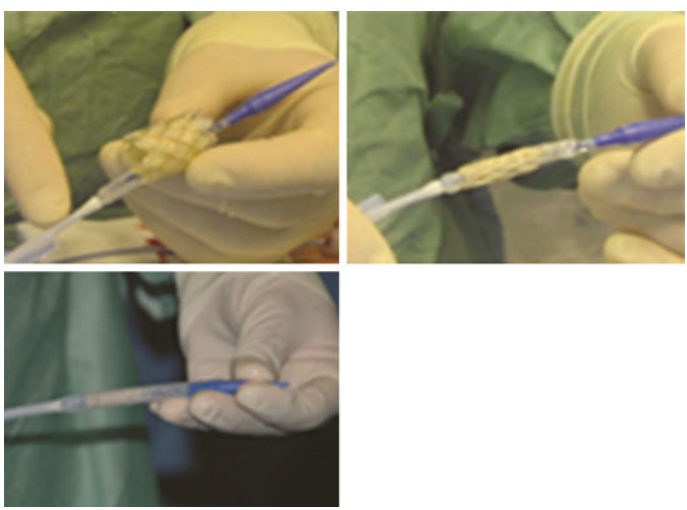

Fig. 35.2 Loading of the PPVI stent onto the delivery catheter

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 while optimizing flexibility and retaining the pushability required for the procedure. There is a retractable sheath which covers the stented valve during delivery 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).

35.4 Procedure

Preparation

- General anesthesia and orotracheal intubation.
- Biplane catheterization laboratory preferred.
- Patient position with arms lifted up, behind the 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, and vesical catheter for diuresis evaluation.
- Full anticoagulation is obrained by administering intravenously 100 UI/kg of heparin. 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 for strict asepsis. Special attention has to be given to operators' scrubbing and patient's preparation (including careful depilation). The personnel involved has to wear masks and hats.

35.5 Access Site

- Usually a femoral access is used. Sometimes the internal jugular access can be used.
- Both sides for vascular femoral access are prepared.
- A 12 Fr femoral sheath is placed: this helps because the need for changing femoral sheath is avoided during diagnostic catheterization, balloon sizing, balloon testing, angiographies 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 5 Fr sheath.

35.6 Catheterization and Hemodynamic Evaluation

- Right heart catheterization is performed using standard techniques to assess pressures and saturations with a right coronary catheter, JR 3.5, 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 curves as much as possible, to place the tip of the wire as distal as possible (ideally in the pulmonary artery bed at the level of the diaphragm) and to be sure that 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).
- The preferred guidewire is usually a 0.035″ exchange wire (Amplatz Ultra Stiff wire 260 cm long (Boston Scientific

Corp., Natick, MA), Lunderquist 260 cm long (Cook Europe, Denmark), Back-up Meier 300 cm long (Boston Scientific Corp., Natick, MA).

- Special care has to be paid to wire stability and position. This should not move during the procedure. If, in any phase of the procedure, the wire moves and the position is not satisfactory, replace in the ideal position with the use of a catheter. Never push forward the stiff wire because of the risk for pulmonary vascular bed injuries.
- Angiography is performed using a 6 Fr multitrack 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. 35.3).
- Angiographies are performed in the pulmonary artery trunk, right ventricle, and ascending aorta (Fig. 35.3).
- Usually the following imaging planes are used:

Lateral View

  - To visualize the anterior chest, landing zone, and proximal end of pre-stent
  - Useful to check coronary arteries involvement during balloon testing

Anteroposterior View with Cranial Angulation Usually Associated to Left Anterior Oblique Angulation

  - To visualize relation to bifurcation and distal end of stent.
  - Useful to check coronary artery involvement during balloon testing.

- If needed, selective coronary artery angiography is performed.
- Delineate landing zone and decide about final target diameter to be achieved with Melody valve.

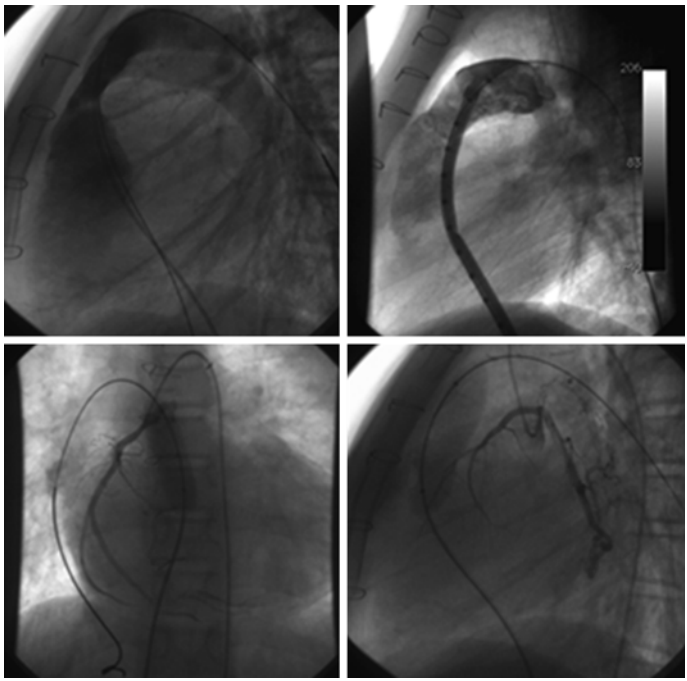

Fig. 35.3 *Left upper*: Angiography in lateral view showing severe stenosis and regurgitation at the level of the RVOT; *right upper*: Angiography in lateral view of the RVOT through the lateral port of the Mullins sheath after pre-dilatation of the RVOT; *left and right bottom*: balloon RVOT testing during simultaneous coronary angiography

In case of doubt on the characteristics of the landing zone, it can be useful to do as follows:

- Use a sizing balloon (34 mm Amplatzer ASD sizing balloon(NuMED Inc., Hopkinton,NY). This gives a precise negative image of the RVOT and landing zone.
- Then use a low-pressure angioplasty balloon (e.g., a Z-Med II balloon, NuMED Inc., Hopkinton, NY) or Crystal balloons (Balt, Montmorency, France) in order to check for tissue distensibility

- As a further step, if the tissue looks poorly distensible with low-pressure balloons, use a high-pressure balloon (Mullins X Ultra high-pressure balloon catheter, NuMED Inc., Hopkinton, NY), Atlas PTA balloon dilatation catheter (Bard, Tempe, AZ, USA)
- In this latter case, two situations should be considered:
  - Presence of a lesion with high risk of tear. This can be anticipated in case of 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. 35.3).
    - 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.
      - In this case, there could be a potential for conduit tear.
- Control RVOT angiography is performed to rule out possible extravasation:
  - If a tear is confirmed, then covered stents (Covered CP Stent™, 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.

35.7 Pre-stenting

Exchange the 12 Fr sheath for an 18 Fr venous short sheath in order to make it easier to exchange the balloons and long sheaths for pre-stenting (Fig. 35.4).

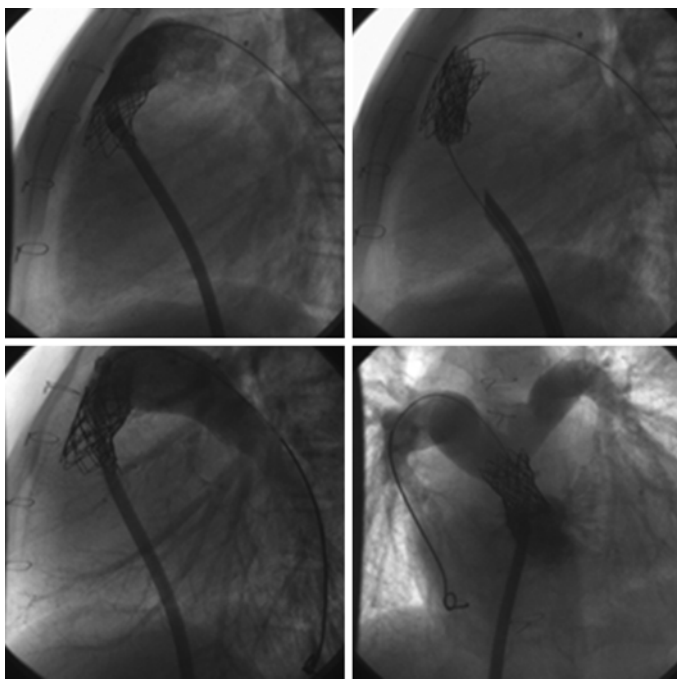

Fig. 35.4 *Left upper*: angiography in lateral view after pre-stenting; *right upper*: Ensemble is in place and inner balloon is inflated; *left bottom*: Melody is correctly placed and angiography in the pulmonary trunk shows no pulmonary regurgitation; *right bottom*: angiography in anteroposterior view with cranial and left anterior oblique angulation: normal flow in both pulmonary arteries

35.7.1 Stent Choice

- Bare stent (CP stent, NuMED Inc., Hopkinton, NY), IntraStent (ev3 Inc, Plymouth, MN), and Andra Stent (Andramed, Reutlingen, DE)
  - When no extravasation is seen
  - Full balloon expansion achieved by pre-dilatation
- Covered stent (Covered CP Stent™, NuMED Inc., Hopkinton, NY)
  - When extravasation is seen
  - Anticipated risk of substrate tear or fracture

Length of the stent should be enough to cover the stenotic area and the entire length of the Melody valve.

35.7.2 Stent Implantation

- Use a 14 Fr Mullins long sheath.
- If a bare stent is used and there is jailing of a PA, it is possible to dilate cells with Atlas ultrahigh pressure balloons >20 atm to reopen the stent to the PA (it is usually preferred to use open-cell or partially open-cell stent as the Andra Stent).
- If a covered stent is used, special care should be done to avoid jailing. Furthermore, because the use of covered stent 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 expected tear.
- After stent implantation, record on fluoroscopy both balloon inflation and deflation during stent implantation. In case of significant recoil or presence of compressing forces on the

stent, implant a second or a third (or even more) stent until the area looks stable (this is to avoid the risk of Melody valve stent fracture during follow-up).
- In case of any doubt, repeat coronary angiogram before further dilatation.
- Dilate stent until desired internal diameter is achieved.
- Recheck hemodynamics (RV and PA pressure and gradient).
- Repeat RVOT and PA angiographies for extravasation.
  - Place additional covered stent(s) in case of extravasation.

35.8 Melody Valve Implantation

35.8.1 Prepare Melody Valve and Ensemble

- The valve is taken out of the packaging by using a sterile forceps in order to avoid contamination.
- The valved stent is prepared in three sequential saline baths (one to three 5 min in each) to wash off the glutaraldehyde, in which it is stored.
- The label of the valve is taken out.
- 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, while a saline flush is administered via the side port to remove air bubbles from the system.

- A special care is paid during all these phases, in particular concerning strict asepsis.
- Predilate the groin with 22 and 24 Fr dilators.
- Insert the Ensemble in the groin and push hemostatic sleeve valve in the access site
- Advance Melody to the preferred landing zone:
  - It is important to have a simultaneous view of the 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, in particular when a 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 because of the characteristics of the Ensemble itself. In these cases, pushing on the Ensemble will pull back the wire.
      - Push on Ensemble while applying a gentle pulling on wire (beware of significant wire coming back). With this maneuver, usually, the dilator comes a bit distant from the RVOT and allows advancement.
      - Push on the guidewire/delivery system in order to create a loop in the right atrium. This maneuver 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 the tricuspid valve.

  - With the right atrial loop in place, pull back the Ensemble/guidewire system. This may help a jump of the system into the landing zone.
  - Consider to change the wire position to the other pulmonary artery.
  - Consider using the internal jugular vein access.

- Uncover Melody by pulling back the sheath to the double marker on the Ensemble system. Usually, there can be a forward movement of the balloon/Melody valve. Sometimes, operators cannot uncover the valve because of too much friction. In this case, the Ensemble can be advanced into a PA, the system is straightened, and the valve partially uncovered. Then, the Melody/balloon system is pulled back in the landing zone.
- Position and complete uncovering of the valve can also be 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. Ensemble may move forward when the inner balloon is inflated (Fig. 35.4).
- After final confirmation of the position, the outer balloon is also hand inflated to complete the deployment.
- The balloons are deflated (inner balloon first) and the delivery system withdrawn carefully and slowly.
- The Ensemble is exchanged for the 18 Fr sheath.
- Pressure measurements are obtained to confirm the result.
- Angiographies are performed (Fig. 35.4).
  - 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 preexisting conduit that could not be enough for patient size/body surface area) or another sub- or supravalvular structure.
  - Use appropriately sized ultrahigh-pressure balloon with a maximum balloon size of 24 mm.
- Pressure measurements are obtained to confirm the result.
- Angiographies are performed.

35.8.2 Complications

A rate of 5–7 % has been reported.

- Device instability and migration/embolization: This is a very rare event. If it occurs, surgery is usually needed. It is possible to attempt to recapture and reposition the valve if the guidewire is still correctly in place.
- Homograft rupture: It has been reported previously how to avoid this complication. If bleeding occurs (hemothorax), a chest tube is placed and autotransfusion should be initiated as soon as it is possible to reestablish a sufficient circulation for further intervention. Acute thoracotomy cannot usually be advised, since decompression of the chest may exacerbate bleeding and lead to later difficulty in locating the source of bleeding. If it is possible to identify the rupture point, covered stents can be implanted.

  Strategies to avoid RVOT conduit rupture include sequential balloon dilatation, starting 2–3 mm $\geq$ the stenotic area, and increasing by around 2–3 mm with each balloon.

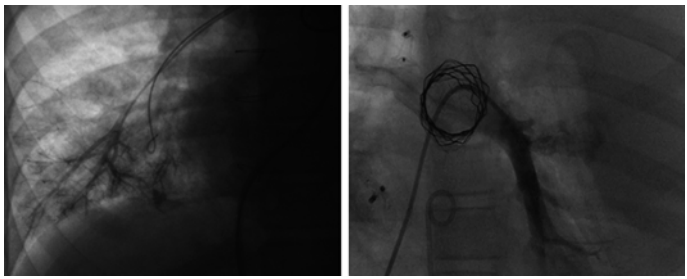

Fig. 35.5 Perforation of distal (*right*) or main (*left*) pulmonary artery

Repeat conduit angiography between angioplasties. If possible, avoid overdilatation of the conduit to beyond the value of the diameter of the conduit itself.

Once CA 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 is 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 cannot be enough to avoid complications.
- 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 maneuvering of the catheter through the right heart. In case of damage in a pulmonary branch, the approach is similar to that of the conduit rupture (Fig. 35.5).
- Melody stent fractures (Fig. 35.6): They are a potential complication of all cardiovascular stent applications. In a report by Nordmeyer et al., the prevalence of stent fractures

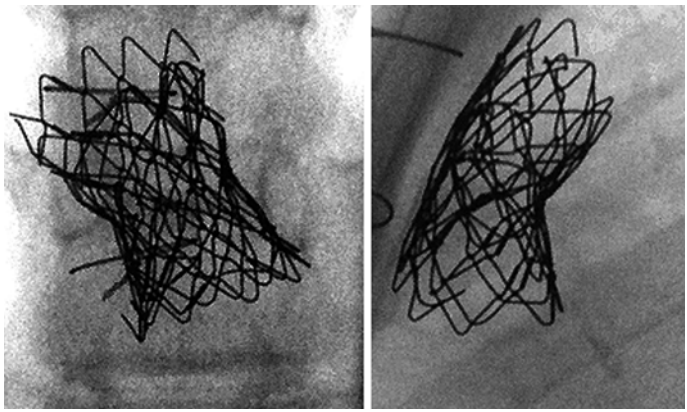

Fig. 35.6 Type II stent fractures in anteroposterior (*left*) and lateral view (*right*)

was 21 %. Implantation into a native RVOT, absence of RVOT calcification, and qualitative recoil of the valved stent just after implantation may be predictors of stent fracture. A classification that can guide management has been formulated by the same author, 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 repeated PPVI (valve-in-valve procedure) or surgery, and type III fractures (separation of fragments/embolization) necessitate surgery.

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.

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 hemodynamic results.

- Endocarditis has been documented on both the venous wall and the valve itself. Long-life antibiotic prophylaxis is mandatory. It may or not result in device dysfunction, and surgical or medical management strategies should be employed accordingly.
- Vascular access complication: as in other procedures. Getting groin ultrasounds in patients having had previous interventions could be useful.

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Chapter 36
SAPIEN Valve Implantation in the Pulmonary Position

Noa Holoshitz and Ziyad M. Hijazi

36.1 Introduction

The SAPIEN valve is made up of three bovine pericardial leaflets, hand sewn into a stainless steel, balloon expandable stent (Fig. 36.1). A fabric cuff 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 valve is currently available in 23 and 26 mm diameter with heights of 14.5 and 16 mm, respectively. It can therefore be used in conduits measuring up to 24 mm at the time of transcatheter valve replacement.

The delivery system used with the SAPIEN valve is the Retroflex III. It is a tapered nose cone-shaped balloon catheter with a deflectable tip (Fig. 36.2). It requires either a 22 or 24 Fr

N. Holoshitz, MD • Z.M. Hijazi, MD, MPH (✉)
Rush Center for Congenital and Structural Heart Disease, Rush University Medical Center,
1653 West Congress Parkway Jones 770, Chicago, IL 60612, USA
e-mail: zhijazi@rush.edu

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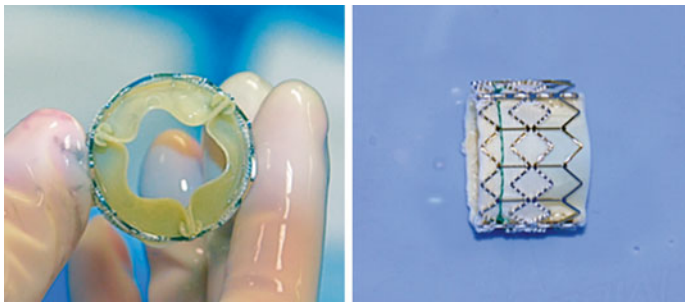

Fig. 36.1 The Edwards SAPIEN transcatheter heart valve. The valve (shown en face on the left and from the side on the right) is available in 23 or 26 mm diameter. It consists of three bovine pericardial cusps mounted into a stainless steel balloon expandable stent

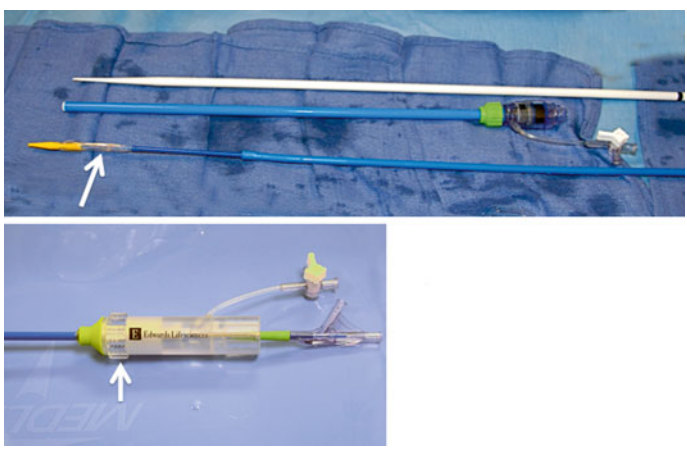

Fig. 36.2 The delivery system that includes the 35 cm-long delivery sheath and dilator and the RetroFlex delivery catheter that has a tapered steerable tip (shown in the *top panel*) which facilitates valve crossing; *white arrow* indicates the location of the valve on the catheter. The handle of the Retroflex catheter (shown in the *bottom panel*) has a knob (*arrow*) to steer the tip of the catheter

hydrophilic sheath for the 23 and 26 mm valves, respectively. The hub of the guiding catheter has a control knob, which can deflect the catheter through passage into the right ventricular outflow tract (RVOT) (Fig. 36.2).

Currently, the next generation of the SAPIEN valves, the SAPIEN XT, is being evaluated for use in the aortic position in the USA with the PARTNER II trial. In this newer valve, the stent material has been changed from stainless steel to a cobalt chromium alloy, which allows for a smaller delivery profile and sheath. In addition, the SAPIEN XT valve is available in a 3rd size, a 29 mm diameter. The valve is available for use outside the USA, and there are case reports of this valve being used in the pulmonic position [1]; however, there is no available long-term data for this newer generation valve.

36.2 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. Quite honestly, it is this group of patients we believe that constitutes the largest population who may benefit from such technology.

36.3 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.

36.4 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 [1]. 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 36.1. These criteria were based on surgical 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/m^2 as determined by cardiac MRI. However, if the patient is

Table 36.1 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

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.

36.5 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 available valve systems for tPVR in the USA. Other than the SAPIEN 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 36.2. The SAPIEN valve is available in larger sizes than the current Melody system and therefore may be appropriate for placement in larger conduits,

Table 36.2 Comparison of Melody and the SAPIEN valves

Characteristic	Melody valve	SAPIEN valve
Stent material	Iridium 10 %, platinum 90 %	Stainless steel
Valve material	Bovine jugular vein	Bovine pericardium treated with ThermaFix
Available size (diameter)	18–22 mm	23, 26 mm (SAPIEN XT available in 29 mm outside the USA)
Stent height	34 mm	14.5, 16 mm
Delivery sheath size	22 French	22 French, 24 French

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 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 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 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.

36.6 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.

36.7 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 and the shape of the conduit and presence of calcifications (Fig. 36.3a).
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. 36.3b). 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 is the distance, and then deploy a bare metal stent as a landing zone [2].
5. Given the short height of the Edwards SAPIEN valve, bare-metal stent implantation (pre-stenting) as a landing zone is performed routinely. The stent is deployed on a BiB

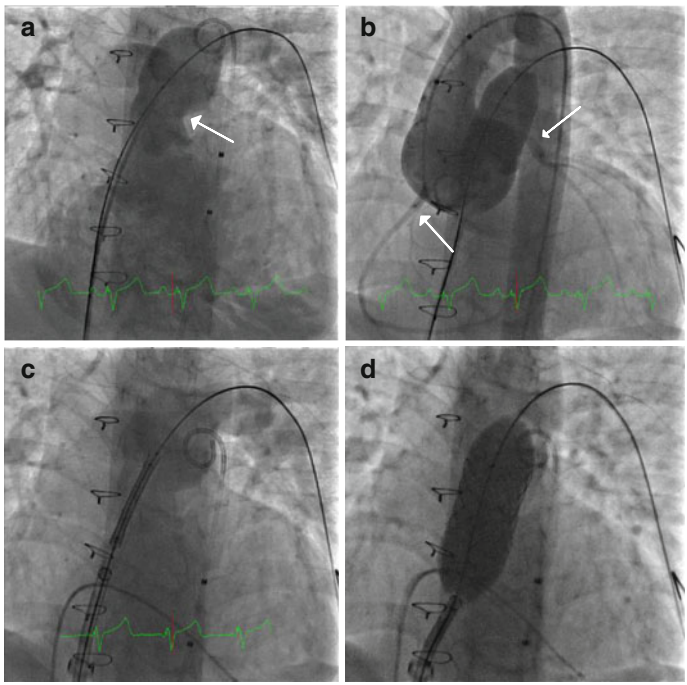

Fig. 36.3 Angiographic stepwise approach to TPVR. (**a**) Angiography of degenerated conduit showing pulmonic regurgitation and narrowing (*arrow*). (**b**) Simultaneous balloon inflation and aortic root injection to assess distance from the conduit to the coronary arteries (*arrows*). (**c**) Positioning of the bare metal stent in the conduit. (**d**) Deployment of the bare metal stent in the conduit. (**e**) angiography in the RVOT demonstrating free pulmonic regurgitation (*arrow*) following pre-stenting. (**f**) Positioning of the Edwards SAPIEN valve (*arrow*) in the RVOT. (**g**) angiography in the RVOT following valve deployment demonstrating no narrowing and no pulmonic insufficiency. (**h**) En-face view of the stent and valve showing uniform expansion

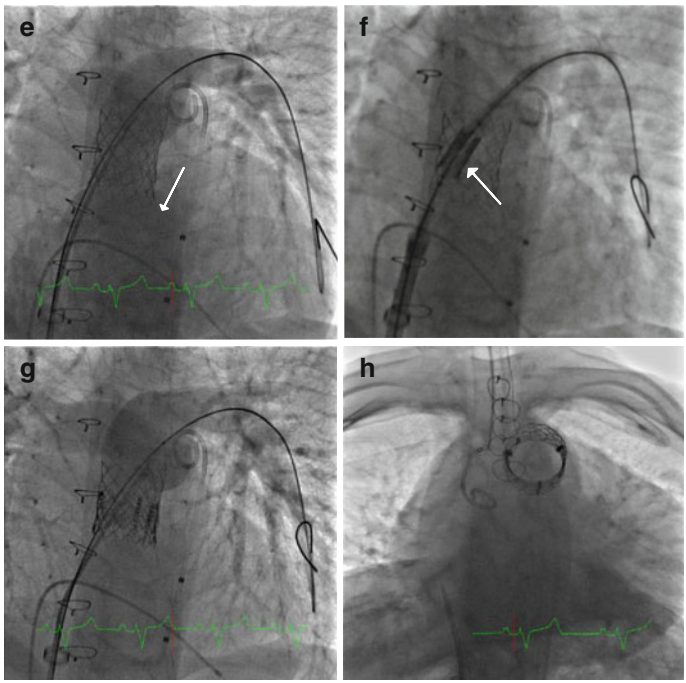

Fig. 36.3 (continued)

(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 (Fig. 36.3c–e). 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. Covered stents of this size are unfortunately not commercially available in the USA, but may be created in the catheterization laboratory by sewing Gore-Tex onto 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. Typically, we expand the 23 mm valve to be no less than 21 mm in diameter and the 26 mm valve to be no less than 23 mm in diameter. The valve has been tested for functionality and durability at these diameters.
7. The valve stent is crimped symmetrically using a specialized crimping tool onto a 30 mm long pre-sized balloon catheter (Fig. 36.4).
8. The valve is then delivered across the pre-stented RVOT over a stiff guidewire (Meier wire or Lunderquist). Angiograms are performed prior to balloon inflation to assess for proper positioning of the valve (Fig. 36.3f, g).
9. Valve performance is evaluated either angiographically (Fig. 36.3g) 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. 36.5).
10. 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.

Fig. 36.4 *Left*: the valve is positioned on the delivery balloon, green suture line lined up away from the yellow tip. *Right*: valve being crimped using the crimping tool

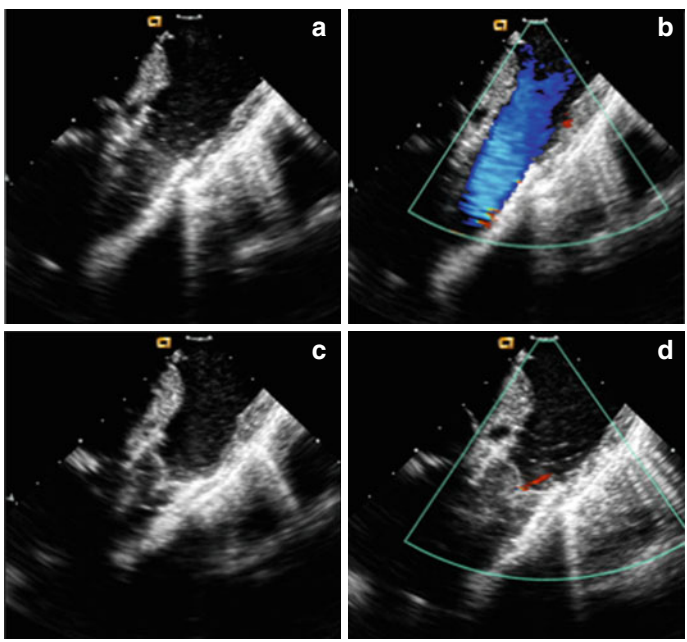

Fig. 36.5 Intracardiac echocardiography. (**a**) At baseline color Doppler demonstrates free pulmonic regurgitation and narrowed valve. (**b**) Continuous wave Doppler across the RVOT at baseline showing significant gradient. (**c**) After presenting the RVOT, (**d**) color Doppler demonstrates wide open pulmonic insufficiency. (**e**) After Melody valve implantation, (**f**) color Doppler demonstrates no residual insufficiency

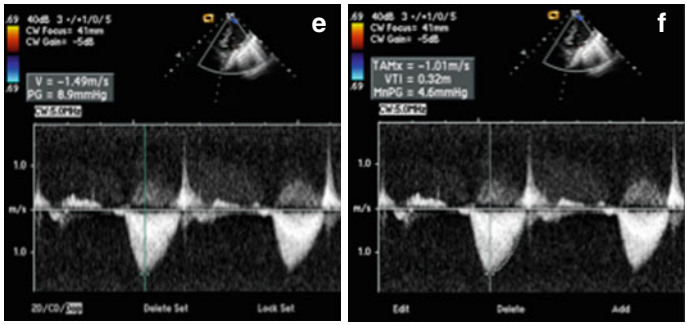

Fig. 36.5 (continued)

36.8 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 [3]. 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.

The most recent data on tPVR with the SAPIEN valve is from a European series of 22 patients [4]. 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.

36.9 Tips and Tricks

The RVOT can be a tricky area to navigate, especially with the bulky Retroflex III delivery system. 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.

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.

36.10 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 [3]. The European multicenter registry reported a major complication rate of 13.6 % in the first 22 procedures performed [4]. We expect that the complication rates will decrease significantly as operators become more experienced with the use of the SAPIEN valve, as was shown to hold true for the Melody valve. Expected complications can be broken down into several categories which are discussed below.

36.10.1 Vascular Complications

Given the large caliber of the delivery sheath of the SAPIEN 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. There were no reported vascular complications in the COMPASSION trial or the European cohort [3].

36.10.2 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 [1]. This catastrophic complication can be avoided by thorough evaluation of the coronary anatomy prior to the procedure to 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 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.

36.10.3 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.

36.10.4 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 [3].

36.10.5 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.

36.10.6 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.

36.10.7 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 % [1]. 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, most likely because of the shorter stent height and routine use of pre-stenting.

36.10.8 Endocarditis

In Melody follow-up studies, five patients (3.2 %) were diagnosed with endocarditis over a mean follow-up of 5 months. Although there have not been any reports of endocarditis with the SAPIEN valve following implantation in the pulmonic position, there have been reports when the valve was used in the aortic position. The updated AHA guidelines recommend continuing lifelong endocarditis prophylaxis for patients who have a conduit.

36.11 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.

36.12 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.

36.13 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 for 6 months in patients with a native RVOT and lifelong for patients with a conduit.

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Chapter 37
Percutaneous Tricuspid Valve Implantation

Andreas Eicken and Peter Ewert

37.1 Anatomic Description and Physiology

Since the advent of percutaneous valve implantation in the year 2000, many patients were treated successfully by a percutaneously implanted valve in the aortic and less frequently in the pulmonic position. So far, only a few patients were treated by percutaneous tricuspid valve implantation (PTVI) and even less by percutaneous mitral valve implantation. This current chapter describes PTVI. The initial treatment of severe tricuspid valve dysfunction (regurgitation or stenosis or both) is surgical plasty at our center. Only if a surgical valvuloplasty does not seem feasible or the result of this operation is unsatisfactory, tricuspid valve replacement is performed. Our local surgical team in Munich prefers to implant a biological valve prosthesis in the tricuspid position. However, with elapsing

A. Eicken, MD (✉) • Peter Ewert
Clinic for Pediatric Cardiology and Congenital Heart Defects,
Deutsches Herzzentrum München, Technische Universität,
Lazarettstr. 36, 80636 Munich, Germany
e-mail: eicken@dhm.mhn.de; ewert@dhm.mhn.de

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time biological valves undergo degeneration and need to be replaced once valve dysfunction reoccurs. Valve degeneration leads to tricuspid regurgitation and/or stenosis, right atrial enlargement, onset of clinical deterioration, atrial dysrhythmias, and signs of right heart failure. To avoid repeated open heart surgeries and to reduce the number of surgical procedures during a patient life, PTVI is an alternative treatment option. The failing bioprosthesis usually is an ideal "landing zone" for a percutaneous valve, if the dimensions are well defined.

37.2 Indication for Treatment (PTVI)

PTVI is indicated in patients with severe dysfunction of the bioprosthesis in the tricuspid position. In general, the same guidelines for patient selection are applied for PTVI as for surgery (Table 37.1) [1]. Patients should be treated prior to severe clinical deterioration. After a detailed cardiological examination (patient and medical history including operation reports, clinical examination, echocardiography (transthoracic and transesophageal) with Doppler interrogation, chest x-ray, and an exercise test with assessment of VO_2 max, the decision for treatment is made. Cardiac MRI may be used to assess the right ventricular end-diastolic volume index which can be used to monitor the effect of PTVI during follow-up. Severe tricuspid regurgitation causes volume loading of the right ventricle. Serial cMRI examinations may be used to determine the timing for PTVI. It is essential to know the bioprosthesis (size and manufacturer) which is in place. For selection of the optimal percutaneous valve, the internal diameter of the current tricuspid valve prosthesis is important.

Table 37.1 Indication for tricuspid valve surgery [1]

	Class[a]	Level[b]
Surgery is indicated in symptomatic pts with severe TS[c]	I	c
Surgery is indicated in pts with severe TS undergoing left-sided valve surgery[d]	I	c
Surgery is indicated in pts with severe primary or secondary TR undergoing left-sided surgery	I	c
Surgery should be considered in pts with moderate primary TR undergoing left-sided valve surgery	IIa	c
Surgery should be considered in pts with mild or moderate secondary TR with dilated annulus (≥40 mm or >21 mm/m^2) undergoing left-sided valve surgery	IIa	c
Surgery should be considered in asymptomatic or mildly symptomatic pts with severe isolated primary TR and progressive RV dilatation or deterioration of RV function	IIa	c
After left-sided valve surgery, surgery should be considered in pts with severe TR who are symptomatic or have progressive RV dilatation dysfunction, in the absence of left-sided valve dysfunction and severe pulmonary vascular disease	IIa	c

TR tricuspid regurgitation, *pts* patients, *TS* tricuspid stenosis
[a]Class recommendation
[b]Level of evidence
[c]Percutaneous balloon valvuloplasty can be attempted as a first approach if TS is isolated.
[d]Percutaneous balloon valvuloplasty can be attempted if percutaneous mitral commissurotomy can be performed on the mitral valve

37.3 Technique of PTVI

Patients are treated using conscious sedation or general anesthesia. If not performed before, a transesophageal echocardiogram is done for assessment of valve regurgitation and the inflow

gradient across the tricuspid valve. Then vascular access (vein and artery) is achieved usually in the groin. Valve delivery via a venous neck vessel has been described as well. Pressures at right atrial and right ventricular level are recorded. The difference between the A wave in the right atrium and the right ventricular end-diastolic pressure in mmHg is assessed (TrV-inflow gradient). Most bioprostheses in the TrV position are mounted in a non-distensible ring offering a safe landing zone for a percutaneous valve. Using fluoroscopy the metallic cage of the bioprosthesis is usually visible and is an excellent landmark during valve implantation. An aortogram depicts the course of the right coronary artery in relation to the bioprosthesis. A "balloon interrogation," however, is rarely necessary since the metallic frame of the bioprosthesis prevents coronary compression. An angiogram within the right ventricle is performed (RV function and degree of tricuspid regurgitation) (Fig. 37.1). Accurate valve placement requires coaxial positioning within the bioprosthesis. The x-ray system is rotated to a complete perpendicular position in the anterior posterior plane which is used for valve delivery. A 0.0035 in. superstiff guidewire is placed distally into one pulmonary artery. This position adds additional safety during valve delivery since a guidewire position in the right ventricle is less stable. Even if the type of bioprosthesis in tricuspid position is known, a balloon test is performed to determine the actual inner diameter of the prosthesis (Fig. 37.2). The balloon chosen for this test should be at least 2 mm larger than the suspected internal bioprosthesis valve diameter. Valuable information on current biological valves and their dimensions are available in the literature [2]. In our practice, a Melody valve is chosen if the internal diameter is <24 mm (Fig. 37.3). In larger internal valve diameters, the Edwards Sapien 26 mm or Sapien 29 mm may be used. If the internal diameter of the bioprosthesis is at least 2 mm less than the external diameter of the inflated implanted valve, pre-stenting is not necessary. In large valves, the internal diameter may be reduced by pre-stenting

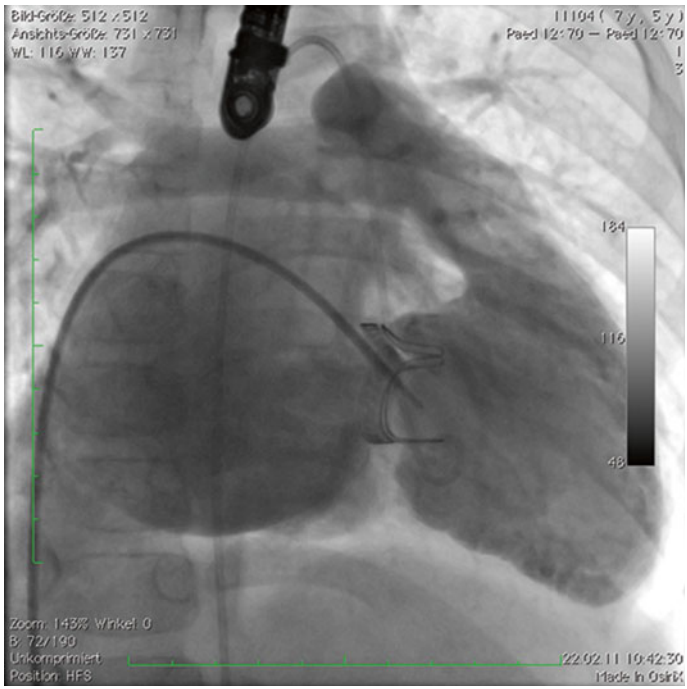

Fig. 37.1 Injection of contrast into the right ventricle of a patient with Ebstein's anomaly after implantation of a Carpentier-Edwards Perimount 27 mm bioprosthesis (Edwards Lifesciences, Irvine, CA). Severe tricuspid regurgitation stains the enlarged right atrium

with CP stents. Dilation of a covered 8 zig CP stent to a large diameter leads to significant shortening of the stent. For this reason in our practice, at least a 39 mm or preferably a 45 mm long covered CP stent should be used. If this stent is mounted on a large balloon, a large long sheath for stent delivery is necessary. The groin needs to be dilated prior to valve implantation. For the Melody valve a short 22 F dilator is used since the delivery catheter is 22 F. The valve is mounted in the same way

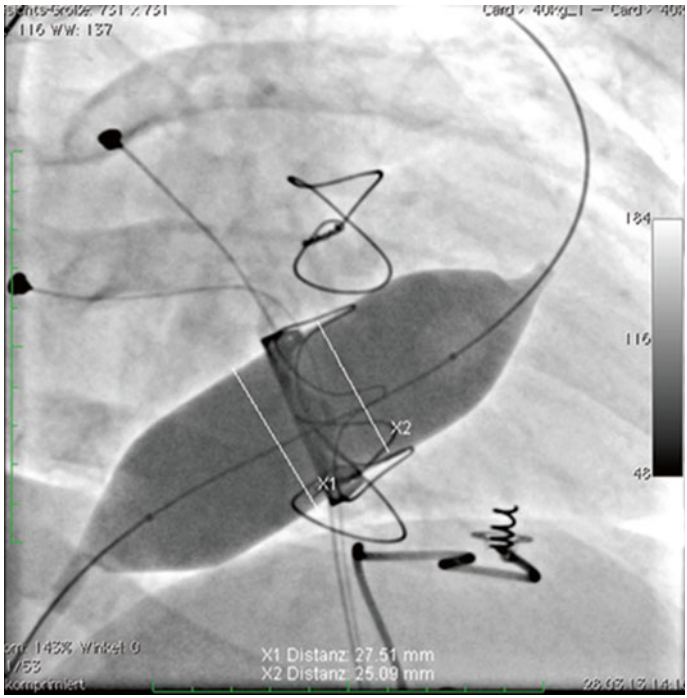

Fig. 37.2 A balloon interrogation test is performed with a 28 mm diameter Z-MED II-X™ balloon (NuMed Inc. Best, The Netherlands) in a patient with Ebstein's anomaly after three operations. Finally a Carpentier-Edwards Perimount 31 mm bioprosthesis had been implanted. The minimal diameter was 25 mm

as it is described for percutaneous pulmonary valve implantation. Edwards supplies excellent hydrophilic dilators for predilation of the groin. In contrast to the retrograde positioning in the aortic valve, the Sapien 26 and 29 valves need to be premounted the other way around (180° opposite than in aortic valve delivery) to account for the antegrade positioning. The Sapien 26 valve is mounted on the Retroflex 3 catheter system

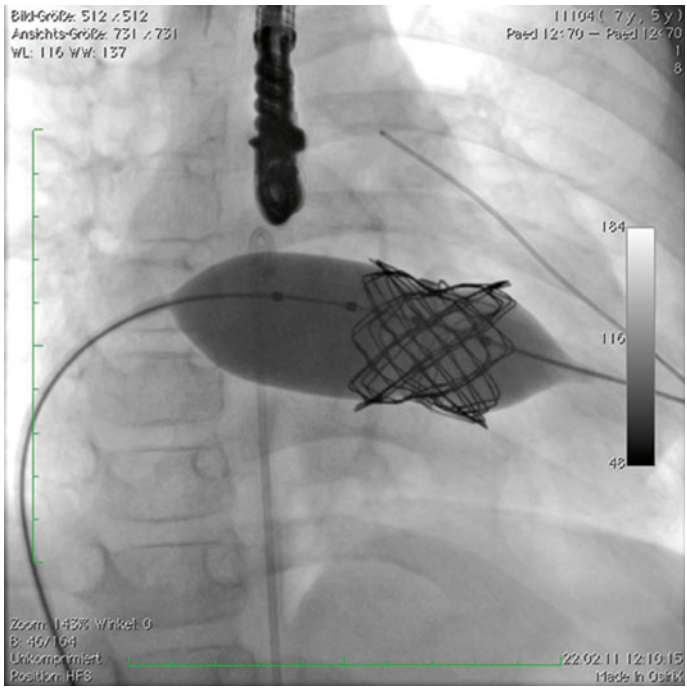

Fig. 37.3 After pre-stenting with a bare 34 mm CP 8 zig stent (NuMed Inc. Best, The Netherlands), the Melody valve (Medtronic Inc. Minneapolis, MN) was implanted on a 22.0 mm delivery system. It was post-dilated with a 22 mm Mullins XTM balloon (NuMed Inc. Best, The Netherlands). The patient was a 5-year-old girl (17.7 kg) with tricuspid dysplasia after valve plasty and two subsequent surgically implanted biological valves. At the last operation a Perimount 21 mm bioprosthesis had been implanted which now was severely regurgitant. In retrospect, pre-stenting was not necessary in this patient

which is introduced through a 24 F hydrophilic sheath into the inferior vena cava. The Sapien XT 29 valve is mounted on the shaft of the NovaFlex + transfemoral system which is introduced via the eSheath (20 F) into the patient. Within the patient this

valve is then pulled onto the delivery balloon against the direction of the valve leaflets. Although this described procedure is "off label" and there is some concern relating to damage of the valve leaflets when pulling them against the direction of blood flow, in our experience all those implanted Sapien XT valves show excellent function during short- and medium-term follow-up.

Valve implantation is similar for all three available valves. They are advanced slowly on the stiff guidewire until they reach their desired position within the tricuspid valve prosthesis. Slow balloon inflation enables readjustment of the implanted valve to avoid delivery in an angulated position. The valve should be as horizontal to the plane of the bioprosthesis as possible. A stable wire position in the pulmonary artery allows repositioning of the valve during delivery. There is, however, a self-adjusting effect when the delivery balloon is fully inflated, since it will slip by itself into a perpendicular position to the bioprosthesis. Strict perpendicular positioning of the x-ray system (best in the PA plane) enables a valve position where the proximal struts just peak out of the bioprosthesis (Fig. 37.4). Positioning too far into the bioprosthesis may lead to valve embolization into the right ventricle. Pressure measurements (RV and RA), a final RV angiogram, and the TEE examination confirm the result (Fig. 37.5). A list of the material used is depicted in Table 37.2, and Fig. 37.6 shows the Melody valve and the Sapien 26 and 29 valves.

37.4 Post Interventional Treatment and Follow-Up

Periprocedural treatment includes antibiotic prophylaxis (usually a second-generation cephalosporin (cefuroxime 100 mg/kg/day or 3×1.5 g/day in adults). Acetylsalicylic acid

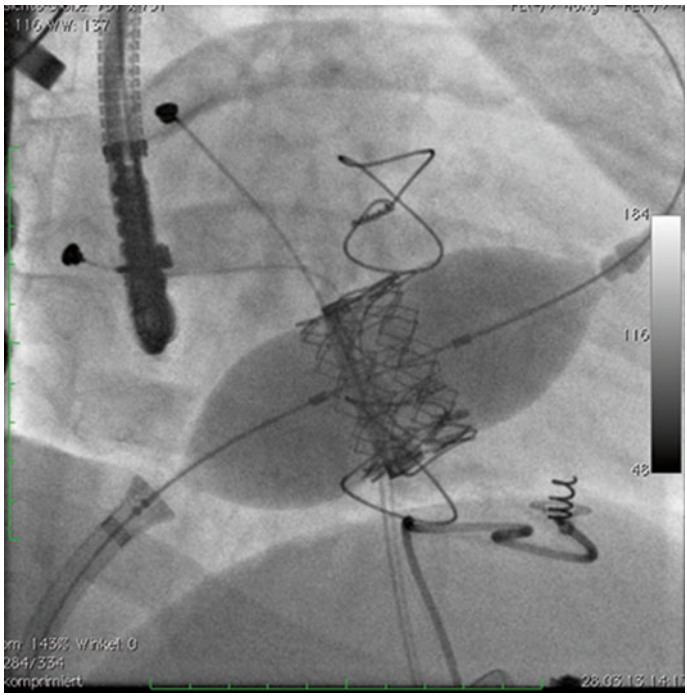

Fig. 37.4 An Edwards Sapien XT 29 mm valve is positioned within a 31 mm Perimount prosthesis under fluoroscopic and TEE guidance. The anterior posterior x-ray plane was rotated to completely perpendicular level, and only the last proximal struts of the Sapien valve "peak" out of the bioprosthesis. This position enables optimal valve function and prevents valve embolization into the ventricle. The patient was an 18-year-old girl after three open heart surgeries and two additional pacemaker implantations who presented with severe regurgitation of the Perimount valve

(100 mg/day or 3–5 mg/kg) is administered for 6 months. After 6 months a TEE, a cMRI, an exercise test, and a clinical examination are performed to document valve function and the patient's clinical status. Then annual cardiac examinations are

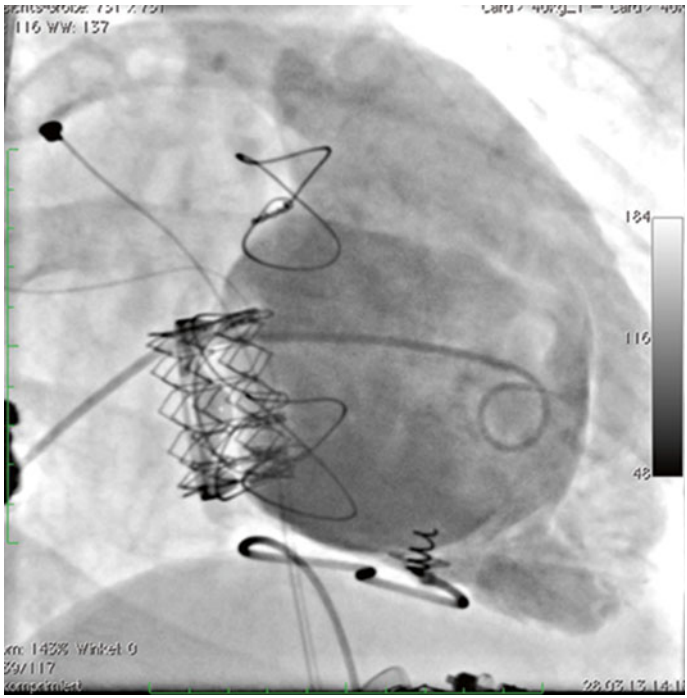

Fig. 37.5 After implantation of a 29 mm Edwards Sapien XT valve, a final angiogram into the right ventricle assesses a good result

recommended. So far there is only one case series with 15 patients published in the literature including some of our patients [3]. The longest follow-up in our experience is now >5 years, and this patient is in NYHA I and hence doing very well clinically. Table 37.3 shows all eight patients treated in Munich since December 2006 since the percutaneous program was initiated. In all but one case, the original implanted valve does not show any sign of valve dysfunction. The only patient with early failure (severe Melody valve regurgitation 18 months after PTVI) was treated surgically with a Perimount 31 mm valve. However, 6 months later valve dysfunction reoccurred,

Table 37.2 Material used during percutaneous tricuspid valve implantation PTVI

0.0035 in. superstiff guidewire	Lunderquist Wire and Amplatz ultrastiff, Cook Medical, Bloomington, IN; Backup Meier wire, Boston Scientific, Natick, MA
Balloon for interrogation of the valve	2 mm larger than the expected diameter of the bioprosthesis – Z-MED II-X™ balloon, NuMed Inc., Best, The Netherlands; Cristal balloon Balt Extrusion, Montmorency, France
Stents for reduction of diameter	CP 8z 45 mm covered stents, NuMed Inc. Best, The Netherlands
Large long sheath	Flexor 22 F, Cook Medical, Bloomington, IN
Short dilator	22 F, Cook Medical, Bloomington, IN
Hydrophilic dilators	Edwards Lifesciences, Irvine, CA
Melody valve	Medtronic, Minneapolis, MN
Sapien 26	Retroflex 3 catheter system, Edwards Lifesciences, Irvine, CA
Sapien XT 29	NovaFlex + transfemoral system with eSheath (20 F), Edwards Lifesciences, Irvine, CA

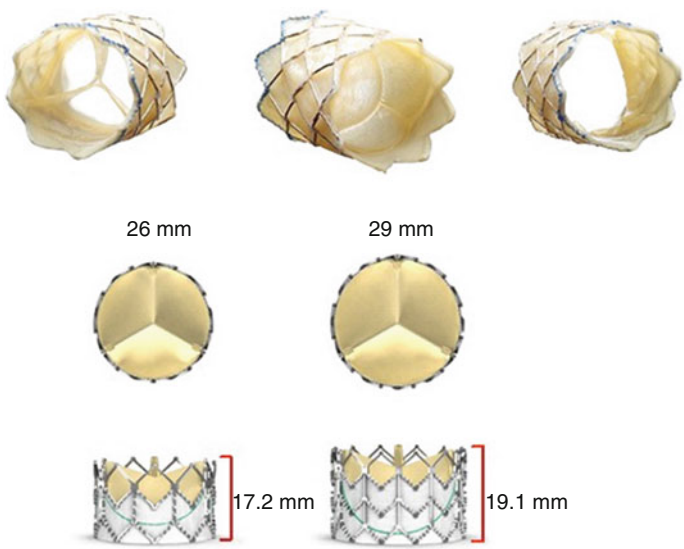

Fig. 37.6 Considerable bleeding may occur and blood in the bronchi may impede an effective gas exchange

Table 37.3 Patients with percutaneous tricuspid valve implantation at the German Heart Center Munich (8 patients – 9 procedures)

Patient	Diagnosis	Bioprosthesis	Dysfunction	Treatment	Follow-up results
18 years, 76 kg	Ebstein	Perimount 27	TR grade 4	2x stent Sapien 26	8 months: no TR, absent TS
31 years, 53 kg	DCRV, TVR	Mosaic 25	TR grade 4	2010 stent + Melody	
			2012 TR 4	Perimount 27	After 6 months severe TR of Perimount
			re TR 4	Sapien 29	23 months:TR 2, absent TS
5 years, 18 kg	TrV dysplasia	Perimount 21	TR + TS	Stent + Melody	2.2 years good valve function
31 years, 75 kg	Fontan RA-RV	Homograft 26	"TR" 3–4	Stent Melody	5.3 years, 3 struts, good valve function, NYHA 1
43 years, 100 kg	Fontan RA-RV	Hancock 30	"TR" 3–4	Stent Melody	4.5 years, good valve function, NYHA 2–3
54 years, 68 kg	critical PS	Perimount 27	TR 4	Sapien 29	3 months good function
17 years, 68 kg	Ebstein	BDG + Perimount 31	TR 4	Sapien 29	1 month good function
55 years 64 kg	Ebstein	Heterograft 33	TR 4	Sapien 29	3 months good function

TR tricuspid regurgitation, *TS* tricuspid stenosis, *DCRV* double-chambered right ventricle, *TVR* tricuspid valve replacement, *TrV* tricuspid valve, *RA-RV* right atrium to right ventricle anastomosis (Fontan Björk), *NYHA* New York Heart Association functional classification, *PS* valvar pulmonary stenosis, *BDG* bidirectional Glenn shunt

and the patient was treated successfully with a Sapien 29 mm. The reasons for early valve dysfunction in tricuspid position remain speculative so far. Biological valves may fail early after surgery and after PTVI.

In conclusion, in selected patients with bioprosthesis failure in tricuspid position, PTVI is a safe, effective, and elegant alternative to repeated surgery, and this may reduce the number of open heart surgeries during a patient life.

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Part VIII
Step-by-Step Procedures: Principles of Hybrid Approach

Chapter 38
Hybrid Approach in Hypoplastic Left Heart Syndrome (HLHS)

Dietmar Schranz

38.1 Anatomic Description and Physiopathology

Hypoplastic left heart syndrome is a rare congenital heart defect in which the left side of the heart is underdeveloped and the right ventricle supporting both systemic and pulmonary circulation. HLHS is accounting for 2–3 % of all congenital heart defects [1]. Untreated, HLHS is a fatal congenital heart defect. HLHS is morphologically subdivided in mitral and/or aortic atresia or stenosis, respectively. About 6 % of HLHS patients have a completely intact atrial septum at birth and up to 22 % a severely restrictive atrial septum. Intact or highly restrictive atrial septum (IAS) is associated with a high rate of mortality. Another form of pulmonary venous flow obstruction might be associated with total anomalous pulmonary venous return (TAPVR). Neonates that present with hypoplastic left heart syndrome (HLHS) are clinically stable as long as the parallel circulation is balanced.

Newborns with HLHS and intact atrial septum (IAS) are critically ill born. Therefore, in newborns with profound hypoxemia

D. Schranz
Pediatric Heart Center, Justus-Liebig University, Giessen, Germany
e-mail: dietmar.schranz@paediat.med.uni-giessen.de

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and acidosis that require immediate attention, HLHS has always to be considered as a differential diagnosis. On the other side, HLHS neonates with an unrestricted interatrial communication have the risk for systemic low cardiac output. Postnatal adaptation usually leads to decrease in pulmonary vascular resistance, which disposes in a patient with HLHS to pulmonary runoff. In prenatally undetected HLHS, patients are often admitted in cardiogenic shock because of severe obstruction of the ductus arteriosus.

38.2 Clinical Scenarios

Prenatal fetal echocardiography improves the postnatal outcome of newborns with HLHS. Heart failure or even cardiogenic shock can be avoided if with the parents the postnatal strategy is early discussed. In this context, prophylactic prostaglandin infusion in a low dosage of about 5 ng/kg×min can be used safely with nearly no side effects (Chap. 25). Surgical options are based on three-stage procedures. However, in terms of the physiopathology, hybrid approach consisting of bilateral pulmonary banding and duct stenting and atrioseptostomy, if necessary, might be an innovative alternative to the classical Norwood or Sano operation as an elective procedure or as a highly effective treatment in high-urgency patients even for bridging to heart transplantation.

38.3 Indications and Patient Selection

Independent of the improvements of surgical and intensive care for newborns with HLHS, the parents have to decide among options that include surgical treatment, surgical-interventional treatment, or compassionate therapy. This 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, immediate performed surgical pulmonary 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.

38.4 Treatment Options

With focus on the hybrid procedure, bilateral pulmonary artery banding (PAB) combined with duct stenting and if necessary interatrial septum manipulation was developed as a hybrid method [2] and meanwhile offered as an alternative first-step approach in a number of centers worldwide, but mostly the hybrid procedure is used in newborns with high-risk HLH. However, starting a hybrid program with high-urgency cases might be problematic because the hybrid approach needs experience to make this strategy to a less invasive alternative of Norwood or Sano palliation.

38.5 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

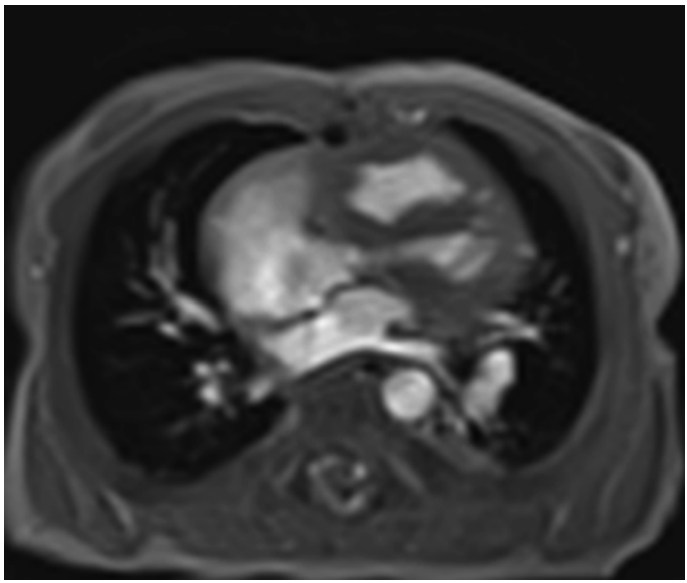

Fig. 38.1 Depicted is MRI 4ch view with a missing LV, restrictive atrial septum, and anterior positioned single right ventricle

data. In HLHS, atrioventricular, ventriculoarterial arrangement is mostly normal, but ccTGA with hypoplastic systemic right ventricle has postnatally the same pathophysiological and therapeutic consequences. Important is the knowledge concerning the interatrial communication, pulmonary venous flow characteristics, the atrioventricular valve (tricuspid valve), systemic ventricular function, the duct morphology and its relationship to the aortic arch, and the coronary perfusion in particular in patients with atretic aortic valve (Chap. 25). In complex anatomy, MRI is additionally helpful to avoid invasive imaging or to plan a surgical or interventional approach (Fig. 38.1).

38.6 Technique (Step-by-Step)

Hybrid procedure consists of bilateral pulmonary arterial banding (bPAB) and duct stenting performed as part of the approach during an open chest – beating heart scenario or as an elective transcatheter approach in a spontaneous breathing, sedated newborn with or without an additional atrioseptostomy [2, 3].

Currently, PAB is performed surgically by an open-chest approach without cardiopulmonary bypass. In our center, the percutaneous transcatheter approach is usually performed by femoral vein or arterial access (Chap. 25).

38.6.1 Step-by-Step Approach

Step 1 depends on the clinical condition of the newborn; surgical bPAB is performed as a high-urgency approach in a newborn with HLHS and pulmonary runoff, but even as the first step of the hybrid approach in a well-conditioned newborn with a still balanced pulmonary to systemic circulation (Fig. 38.2a–d). Balanced circulation in HLHS is present by a Qp-Qs of 1, as in a patient with an arterial (SaO_2) venous (SvO_2) oxygen saturation difference of about 20 % meaning SaO_2 of 80 % and SvO_{22} of 60 %, respectively. It has to be mentioned that bPAB should only be performed as the first step in newborns with free or only slightly obstructed interatrial communication with a minimal pressure gradient and in newborns with wide-open ductus arteriosus achieved by low dosages (2–10 ng/kg/min) of prostaglandin.

In newborns with a body weight of more than 3 kg, a 3.5 mm PTFE shunt is cut receiving a strip of only 2 mm, which is used for bilateral PAB; in lower body weight the same shunt is sutured that the lumen is less than 3.5 mm, and in a premature or newborn with less than 2.5 kg, a 3 mm GORE-TEX shunt is

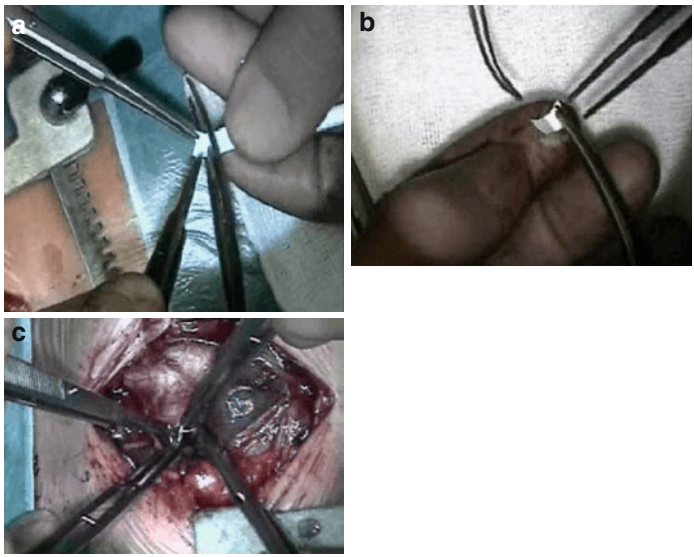

Fig. 38.2 Open-chest approach for bilateral pulmonary banding; (**a**) shows cutting of a 3.5 mm PTFE tube. (**b**) The opened "tube" strip. (**c**) Suturing the strip around the right pulmonary artery

cut and used for bilateral pulmonary banding. The surgical procedure does not consume more than 20 min; the time in the operation theater takes 2–3 h.

In *Step 2*, following the surgical bPAB, the patient is transferred to the cardiac intensive care unit, and in elective cases immediate extubation is the most important therapeutic goal. Cardiac-supporting drugs are mostly not necessary, if the SaO_2 is less than 85 %. In still high SaO_2 values, re-banding has to be considered, but mostly not necessary because after extubation in spontaneous breathing patient, the SaO_2 is mostly further decreasing. However, lowering systemic vascular resistance by

milrinone 0.2–1 μg/kg × min might be of additional benefit as well as long-acting oral vasodilative drugs. In our institution the ACE inhibitor lisinopril is used together with the highly specific ß-blocker bisoprolol both in low dosages of 0.05 mg/kg once per day. The dosage of the ß-blocker is adapted to the heart rate.

Considering the "Giessen hybrid" approach, elective duct stenting is performed following surgical bilateral PAB in an immediate extubated patient.

In *Step 3*, if the decision is made for the percutaneous completion of hybrid stage I, manipulation of the interatrial septum (IAS) is performed if necessary. Atrioseptostomy should regularly be performed before duct stenting. The techniques for atrioseptostomy range between a Rashkind procedure and static balloon dilatation with or without cutting-balloon technique or an immediate IAS stenting. In the past, we used premounted balloon expandable stents (Genesis or Valeo stents) with a high risk of stent slipping during the pullback procedure of bad deflated balloons. The change to self-expandable open-cell stents (OptiMed) of 8 × 12 or 8 × 15 mm sinus-SuperFlex-DS made this approach safe and very easy. In addition, the need for only a 4 F sheath to deliver these stents allows even using an unusual access as a transhepatic approach with low risk and good success (Fig. 38.3).

In *Step 4*, for duct stenting a multipurpose catheter is advanced through a 4 F sheath placed in the femoral artery. The catheter is guided by BMW-0.014″ floppy coronary wire from the descending aorta through the duct in the pulmonary artery. After hemodynamic determinations by pullback pressure measurements from the pulmonary and from the aortic arch to the descending aorta (DAO), biplane (RAO 30° and 90° lateral) angiography is performed to analyze the ductus morphology including the smallest width and length and to delineate the junction of the descending aortic arch and the ductus. Based on these data, stent width and length are chosen. As mentioned in Chap. 25, the

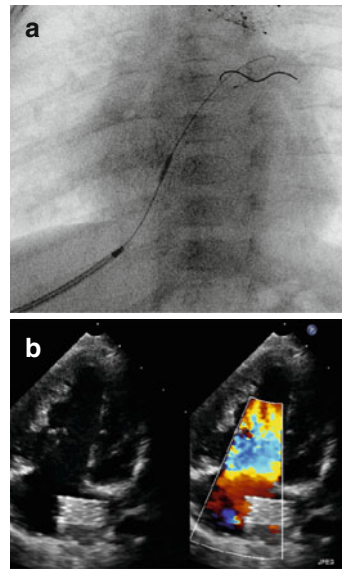

Fig. 38.3 (**a**, **b**) Stent placement within the intact atrial septum (IAS) by transhepatic approach (**a**); the thin struts of the sinus-SuperFlex-DS (8 × 15 mm) deliverable through a 4 F sheath are almost not visible; the stent positioned in the IAS is better shown by echocardiography

diameter of the utilized stent is chosen at least 1–2 mm above the minimal measured duct diameter, but in any case exceeding the diameter of the descending aorta. In most patients with HLHS, a stent with a width of 8 mm is recommended.

In *Step 5*, the venous approach for duct stenting is described in Chap. 25. Here, the approach through the femoral artery is shortly summarized. For stent placement, the soft coronary wire (BMW, Abbott) is exchanged for a stiff 0.014 coronary wire (S'port, Abbott). Utilizing even the novel developed self-expandable stents with a CE mark for duct stenting in newborns with HLHS (OptiMed, Karlsruhe, Germany), duct stenting is currently performed mostly by the arterial access (Fig. 38.4a–d).

In *Step 6*, after preparing the pre-packed self-expandable sinus-SuperFlex-Duct Stent (SSF-DS) and flushing of the 4 F delivery system, the system in total is carefully advanced over the stiff S'sport from the DAO to the PA.

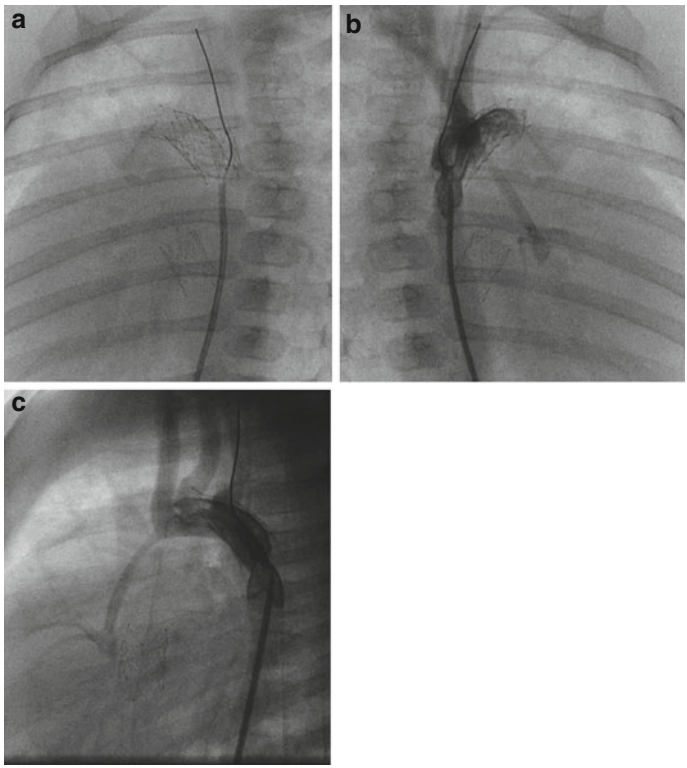

Fig. 38.4 (**a–c**) Depicted are self-expandable sinus-SuperFlex-DS of 8 × 18 mm positioned in the ductus arteriosus and one 8 × 15 mm within the IAS as well as a 5 × 9 mm sinus-Repo-DS at the side of an aortic coarctation in a newborn with HLHS (MA, AA). Lateral projections are shown in (a, c) and RAO 30° view in (b)

In *Step 7*, based on the biplane angiography, a marker for stent placement is necessary. The stent is expanded under fluoroscopy by controlled pullback of the covering part of the delivery system until the stent is fully expanded; in some

patients, short cine scenes are necessary to visualize sufficiently the very thin struts of the open-cell designed stent.

In *Step 8*, the lateral 90° plane is used to control the stent position at the end of the pulmonary artery. Right anterior oblique (RAO 30°) is the plane of choice to observe the exact stent position in relation to the junction of the duct to the descending aorta.

In *Step 9*, as mentioned in Chap. 25, before the delivery system is carefully removed, the nitinol stent should have had a short time period to expand fully at 37 °C temperature; then the open-cell device has the chance for expanding within the arterial wall of the duct, and the risk is minimized to destabilize the stent position or to induce inadvertent stent slipping.

In *Step 10*, before the S'sport wire is carefully removed, the multipurpose or right Judkins catheter is readvanced to the pulmonary end of the stented duct. The sufficient stent position has to be evaluated by angiographies from the pulmonary artery and from the aortic side.

The junction of the descending aortic arch to stented duct is carefully observed as well as the positioning of the stent in relation to the pulmonary trunk and left pulmonary artery. The duct should be fully covered by the stent, but in some morphology the decision for stenting across the retrograde perfused descending aortic arch is necessary. However, in any case the retrograde access to the aortic arch needs to be carefully analyzed.

In *Step 11,* in most patients it is not needed to cross stent the descending aortic arch.

In *Step 12*, the indication for placement of an additional closed-cell designed stent in a diameter of 5 or 6 mm with a length of 9 mm (sinus-Repo-DS, OptiMed, Karlsruhe) within the isthmus or through the struts of the open-cell designed duct stent is based on pullback pressure measurements and results of the angiography, which is made through the lumen of the multipurpose catheter.

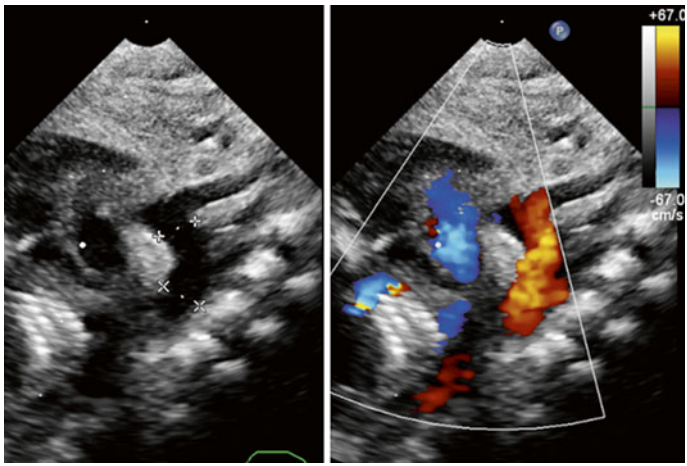

Fig. 38.5 Echocardiographic suprasternal short axis shows an unobstructed aortic arch in 2-D and by color Doppler in a newborn with HLHS (MA, AA) after hybrid stage I consisting of surgical pulmonary artery banding and percutaneous duct and atrial septum stenting, respectively

In *Step 13*, therefore, an additional hemodynamic analysis is recommended after removal of the guiding catheter at the end of the duct stenting procedure. Noninvasive blood pressure measurement at the right arm is performed to estimate the coronary and cerebral perfusion pressure. It has to be performed simultaneously with invasive pressure measurements of the pulmonary artery pressures and DAO pressure. A significant pressure gradient across the stented duct and the aortic isthmus is needed. The decision for stopping the low dosage of prostaglandin therapy is based on the obtained hemodynamic and angiographic data. In most of our patients, the prostaglandin infusion is stopped after self-expandable stent placement. Figure 38.5 demonstrates an unobstructed aortic arch and retrograde color flow by echocardiography after stent placement within the duct and IAS.

In *Step 14*, the procedure of duct stenting is finished, if after a couple of hours or maybe days the hemodynamic and clinical condition of the patient remains stable; in other words, the interventionist knows best if he/she has achieved an optimal or only a maybe sufficient result (Chap. 25).

38.7 Materials

The materials for the interventional part of the hybrid approach are summarized:

Sinus-SuperFlex-DS™ (OptiMed, Karlsruhe, Germany) is used in our institution (see Chap. 25); as mentioned above, the self-expandable, open-cell nitinol stent available with diameters of 7, 8, and 9 mm and stent lengths between 12, 15, 18, and 20, respectively, is used for duct stenting. The 12 or 15 mm × 8 mm sinus-SuperFlex-DS™ is also used for IAS stenting, if necessary.

For additional stenting of an aortic coarctation, sinus-Repo-DS™ (OptiMed, Karlsruhe, Germany) in a width of 5 and 6 mm and a length of 9 mm is available.

The sinus-DS™ stents are deliverable through a 4 F sheath, and both have received a CE mark for its specific indication of duct stenting and stenting of the coarctation in newborns with HLHS or HLHC.

Currently used material:

- Puncture needle (Vygon, 2 F arterial set)
- 4 F sheath (Terumo)
- 4 F wedge, balloon end-open catheter (Cordis)
- 4 F right Judkins catheter (Cordis)
- 4 F multipurpose catheter (Cordis)
- Hemostat valve
- 0.036 in. guidewire for catheter introducing (Cordis)

- 0.014 in. coronary floppy wire (middle weight balanced, Abbott)
- 0.014 in. super stiff (support, S'sport wire, Abbott)
- Sinus-SuperFlex-DS™ (OptiMed, Karlsruhe, Germany)
- Sinus-Repo-DS™ (OptiMed, Karlsruhe, Germany)

38.8 Expected Results

Hybrid approach consisting of bilateral PAB, percutaneous duct stenting, and atrioseptostomy, if necessary, can be technically performed with a mortality rate less than 1 %.

In uncomplicated patients, surgical pulmonary artery banding is an approach in-between of the planned operation program. In general PAB is performed without mortality and very less morbidity despite necessity as an open-chest approach, but without cardiopulmonary bypass. Elective patients can be extubated at the day of surgery. If PAB is performed in the afternoon, the patient is mostly extubated at the other day. Duct stenting without the need for IAS manipulation is performed as an elective approach in an extubated patient under the continuous infusion of low-dose prostaglandin; stenting by the procedure by femoral artery access can be performed in less than 5 min, in particular as a high-urgency treatment.

Results are dependent on the institutional experience, the general therapeutic strategy, and the utilized material.

38.9 Tips and Tricks

Tip 1: *hybrid mentality* between surgeons and pediatric cardiologists is the basis for successful hybrid approach. Hybrid stage I as an alternative procedure to a Norwood operation should be

started with surgical bilateral PAB if there is no need to manipulate an obstructive IAS before or to stent the duct because of a prostaglandin refractory obstruction. However, if the duct is already stented, the surgeons have to perform carefully the bPAB without touching the duct stent, in particular if a balloon expandable stent is used. In fact, stent compression may occur. This is why in the "Giessen hybrid approach," bPAB is the first step.

Tip 2: *safety* of patients with duct-dependent systemic blood flow depends on wide-open duct, high pulmonary vascular resistance despite unrestrictive interatrial communication or immediate and sufficient bilateral PAB. From this point of view, anesthesia and controlled ventilation are sometimes more dangerous than the surgical and transcatheter procedure itself. Effectively performed bPAB is the most important component of the hybrid approach. PAB avoids hemodynamic instability by pulmonary runoff. Duct stenting is not needed because usually the duct remains open with low dosages of prostaglandin ("Japanese hybrid strategy" without duct stenting). In our institution, no experience exists for duct stenting by transpulmonary approach. The combination of bPAB together with stenting the duct is also called the "Columbus hybrid approach" [3].

We prefer percutaneous duct stenting by the following reasons:

First, the anatomy of the descending aortic arch duct is important to know for the strategy of stent placement; cross stenting of the descending aortic arch is usually the consequence in the transpulmonary approach. Such a stenting across the descending aortic arch is in our institution only performed in selected patients, with an atypical junction of DAO and DA. In most patients, percutaneous DA stenting can be performed with a remaining access to the descending aortic arch without to cross the struts of the duct stent.

Second, if there is a need for stenting the coarctation in advance, as it was needed in about 5–10 % of our newborns with

HLHS/HLHC, it can be performed immediately after stent placement within the duct. In addition, any manipulation at the interatrial septum even in advance to avoid later obstruction (>25 %) can be done prior to duct stenting even in the same transcatheter procedure.

Third, we are convinced that our low-risk approach of hybrid stage I is based on focusing only on bPAB without any other surgical manipulation as duct stenting or performing a reverse shunt (Toronto hybrid approach). In addition, elective catheterization in a spontaneous breathing stable patient is an additional reason to reduce stress of the patient and to achieve safe and successful results.

Tip 3: *the use of the novel self-expandable stent systems (sinus-SuperFlex-DS, sinus-Repo-DS)* improves the percutaneous technique of duct and IAS stenting, as well as stent placement within a retrograde aortic obstruction. Therefore, "Giessen hybrid" might further expand the options for newborns with HLHS or HLH-.

Tip 4: *after surgical bPAB,* the patient needs to be carefully monitored and extubated without the risk of secondary need for reintubation; oxygen demand and consumption should be guided by the heart rate effect of the alpha-2 receptor agonist clonidine and its effect on systemic vascular resistance. In addition, the analgesic effect of clonidine allows the patient to be awake without pain. Inodilators as milrinone are usually not necessary in elective treated patients, but useful if the arterial oxygen saturation is still high after bPAB as long as the patients are not fully awake.

38.10 Pitfalls (See Chap. 25)

Morphology-dependent and material-related pitfalls and complications of duct stenting are described in Chap. 25.

Anesthesiological 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, before the surgical banding is performed.

Surgical-dependent pitfalls are cardiac ischemia by too much manipulation of the sometimes very tiny ascending aorta during band placement around the right pulmonary artery. Bleeding by a left atrial appendage injury might be a surgical complication by placing the band around the left pulmonary artery.

Intensive care pitfalls are even related to neglecting the sensitive parallel circulation together with the right ventricular single ventricle function, which is preserved despite bPAB.

38.11 How to Manage Complications

Anesthesiological-, surgical-, and intensive care-related complications are meanwhile rare after surgical bPAB as the first part of hybrid stage I.

Duct stent-related complications are described in Chap. 25.

Any retrograde aortic obstruction should be treated by stenting even in advance by placing a sinus-Repo-DS™ closed-cell design.

Stenting of the interatrial septum might have the complication of stent embolization, but since we use the self-expandable sinus-SuperFlex-DS with a width of 8 mm and mostly a length of 15 mm, it does not happen. However, the device can theoretically easily be caught by snare and removed through a 6 F sheath.

38.12 Postprocedural Care

Following hybrid stage I completion, the patient is mostly observed for a further 1 week before discharge home. The clinical condition should allow the parents to care their baby at home. Respiratory rate should be less than 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 %; in addition, echocardiography before discharge should show an unrestricted or only slightly restricted interatrial communication and a stented duct flow of less than 2.5 m/s, and a Doppler flow across the bPAB 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.

38.13 Follow-Up

In 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 stage II is successfully performed. A hybrid stage I is a palliative approach. Patients with fully duct-dependent systemic blood flow have a high mortality risk in any case of duct obstruction. The same is true for any severe obstruction within the interatrial septum or aortic arch. The art is to assume any complication and to wait for a complication. Therefore, close follow-up control is mandatory in any patient until the next therapeutic step is performed.

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Chapter 39
Hybrid Approach: Defect Closure

Gareth Morgan and Eric Rosenthal

39.1 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].

39.2 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.

G. Morgan (✉) • E. Rosenthal
Department of Paediatric Cardiology, Evelina London Children's Hospital, St Thomas' Hospital, Lambeth Palace Road, London SE1 7EH, UK
e-mail: gareth.morgan@gstt.nhs.uk; eric.rosenthal@gstt.nhs.uk

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39.3 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.

39.4 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.

39.5 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.

39.6 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.

39.7 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. 39.1 and 39.2).

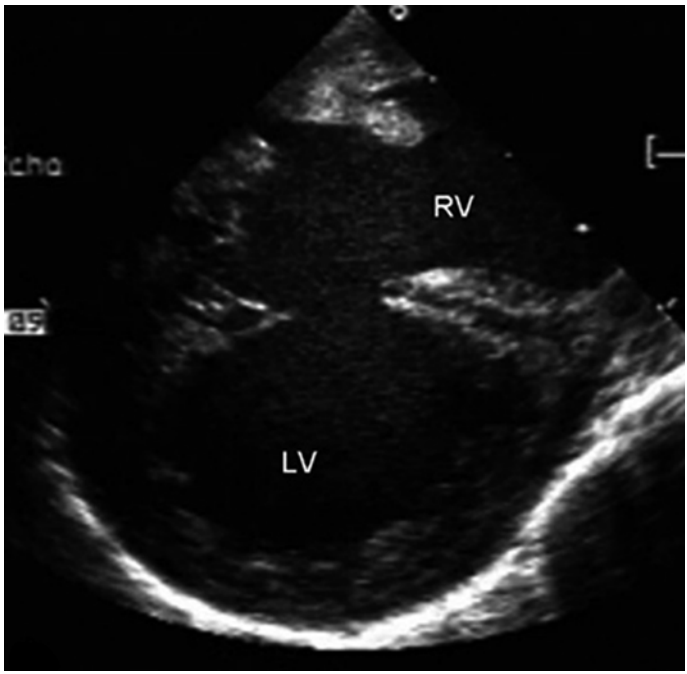

Fig. 39.1 A large mid-muscular VSD delineated with epicardial echocardiography in a 4.8 kg patient. Epicardial echo can be useful as the probe can be used to mimic the desired angle and direction for the wire and sheath passage. *RV* right ventricle, *LV* left ventricle

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.

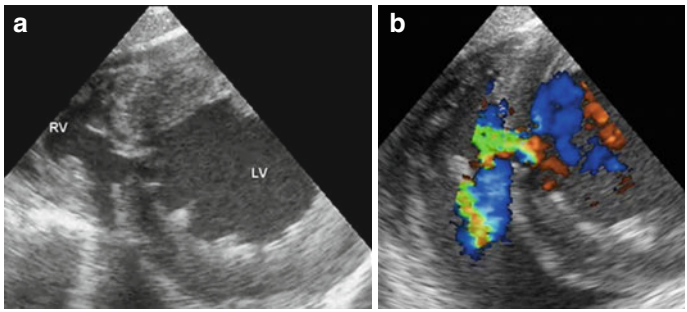

Fig. 39.2 TOE view of a moderate mid-muscular VSD with 2D imaging (**a**) and colour flow Doppler (**b**). Note that the TOE view gives a less 'surgical orientation' of the defect and requires more spatial awareness from the operators compared with the epicardial scan. *RV* right ventricle, *LV* left ventricle

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 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

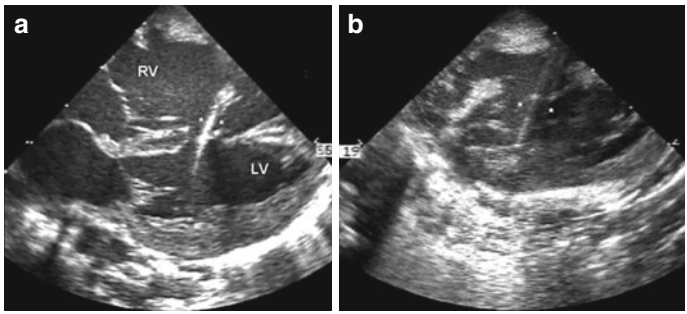

Fig. 39.3 With epicardial imaging the defect shown in Fig. 39.1 has been crossed with the sheath and wire and this has been followed with the dilator and sheath, delineated by the *asterisks* (**a**). After the dilator and wire have been removed, the 'train-track' appearance of the empty sheath is seen (**b**). *RV* right ventricle, *LV* left ventricle

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. 39.3). 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 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. 39.4). 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

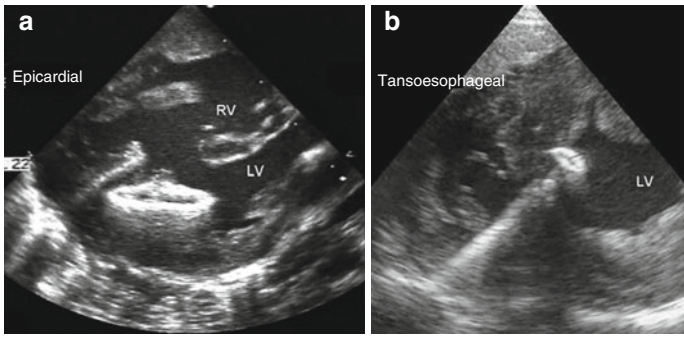

Fig. 39.4 The difference in orientation between epicardial (**a**) and TOE (**b**) guidance is demonstrated here, with deployment of the left ventricular disc. Note that careful planning and good imaging have allowed the disc to be opened free in the LV cavity in both cases. *RV* right ventricle, *LV* left ventricle

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. 39.5).

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.

39.8 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. 39.6).

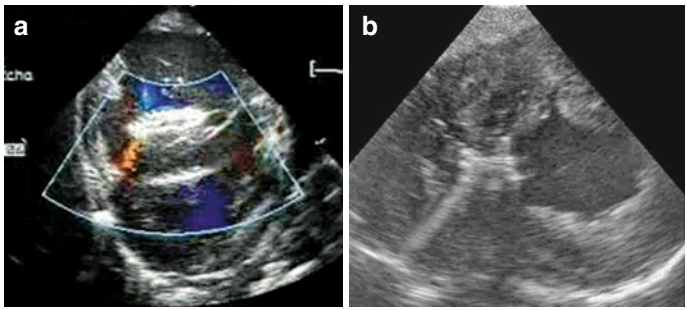

Fig. 39.5 Assessment of the conformation of the waist and RV disc is one of the most important steps. Time should be taken to assess the device and possibly multiple modalities including fluoroscopy can be used. Here we see satisfactory conformation of both discs on the epicardial echo (**a**) and TOE (**b**) – the waist of the device can also be seen to have conformed well (**b**)

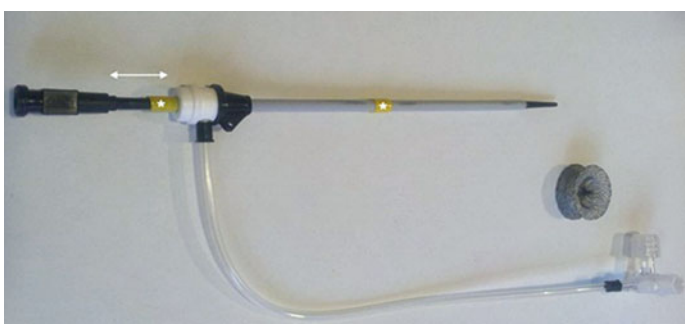

Fig. 39.6 Modification of the sheath to decrease excursion of the stiff dilator towards the LV posterior wall and managing carefully the depth of the sheath into the ventricle can be done by placing two rubber shods (*) onto the ensemble. The first goes onto the dilator and limits its protrusion from 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

2. Some operators have advocated soaking the device in the patient's blood prior to insertion to decrease the risk of post-operative 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.

39.9 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.

39.10 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.

39.11 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.

39.12 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.

39.13 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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Chapter 40
Hybrid Approach: Stent Implantation

Ralf J. Holzer

40.1 Introduction

Hybrid therapies that involve the cooperation between cardiothoracic surgeon and interventional cardiologist have increased significantly over the last 10 years [1]. A CCISC survey conducted by Dan Gruenstein 2 years ago 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 [1–3]. 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.

R.J. Holzer, MD, MSc, FSCAI
Cardiology Division, Department of Pediatrics,
The Heart Center, Nationwide Children's Hospital,
The Ohio State University, 700 Children's Drive,
Columbus, OH 43205, USA
e-mail: ralf.holzer@nationwidechildrens.org

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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. 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 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 that a stent placed preoperatively in the catheterization laboratory potentially being an obstacle at the time of surgical therapy.

On the downside, 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. 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 either direct visualization with 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. This section will provide examples and technical descriptions for each of the more common approaches.

40.2 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 [1]. The most common setup is a patient who requires pulmonary valve or conduit replacement and who has a concomitant kink of the proximal left pulmonary artery (Fig. 40.1). 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

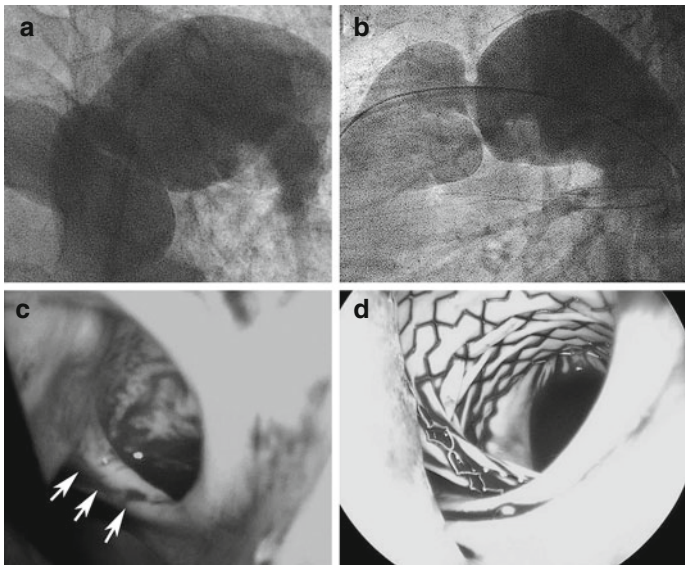

Fig. 40.1 Adult patient with a proximal LPA stenosis (**a**, **b**) undergoing hybrid stent therapy using an endoscopic approach with direct visualization, as well as surgical pulmonary valve replacement. Image (**c**) documents the ridge/kink (*arrow*) at the proximal LPA while image (**d**) documents the same lesion after placement of an intraoperative stent

the correct size of the stent and balloon. Most commonly, these patients have recently undergone cardiac catheterization and if a patient is identified of needing surgery for associated lesions (such as valve replacement), while also having a vascular stenosis that is amenable to intraoperative stent therapy, the findings should be discussed at the time of cardiac catheterization with the cardiothoracic surgeons to make a decision "there and then" whether a percutaneous stent delivery is to be performed or whether an intraoperative hybrid therapy is the preferred therapeutic option.

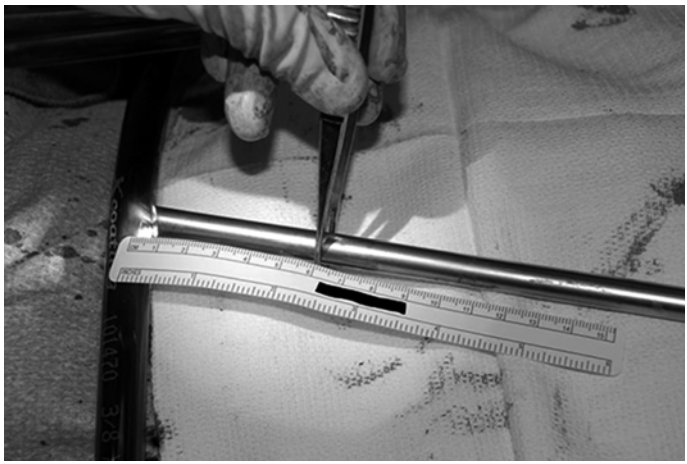

Fig. 40.2 Endoscope used during intraoperative stent placement via direct visualization. The endoscope facilitates imaging of the side branches distal to the stenotic lesion and allows estimate of the distance to those branches

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 a variety of Hagar dilators. Endoscopic guidance is extremely helpful in aiding stent delivery (Fig. 40.2). 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. Soft wires are preferable and stiff wires as well as long sheaths are not required. Prepping the stent on the balloon should be performed in standard technique, even though elimination of all air or the use of contrast in the balloon is not quite as important as during percutaneous stent delivery.

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. 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. Once the stent is fully expanded, the endoscope is utilized to evaluate the entire stent position and stent lumen (Fig. 40.1). Any struts that expand beyond the proximal end of the vascular lesion can be folded over by the surgeon using some stronger pickups, thereby creating a smooth adherence of the struts to the vessel wall (Fig. 40.3). In addition to visual and endoscopic inspection, it is desirable to perform an exit angiography at the end of the procedure to evaluate the results of stent placement angiographically.

40.3 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 where there is very little opportunity for direct visualization, such as residual arch obstructions identified during exit angiography after a

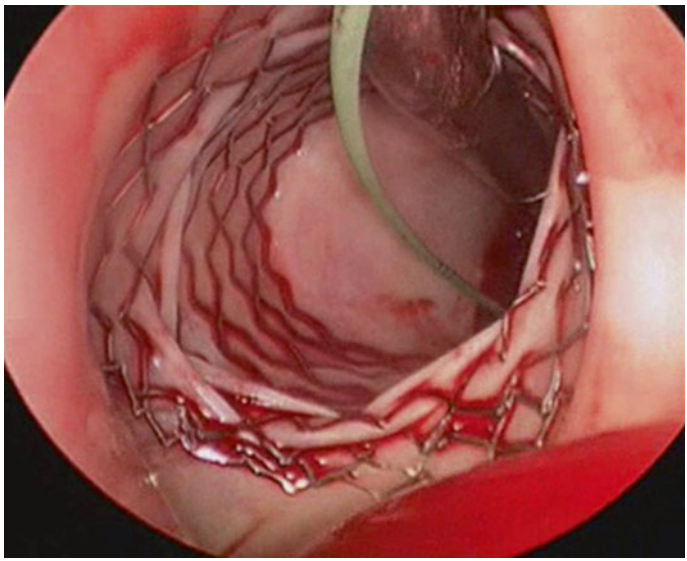

Fig. 40.3 Stent positioned in proximal LPA using direct visualization with endoscopy. Note the folded meshwork of the stent that is bent over the proximal LPA ridge to create a smooth entry site into the LPA

comprehensive arch repair (Fig. 40.4). In addition, angiographic guidance is advantageous in patients where there is very little preexisting imaging data or in smaller vascular structures, where the 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

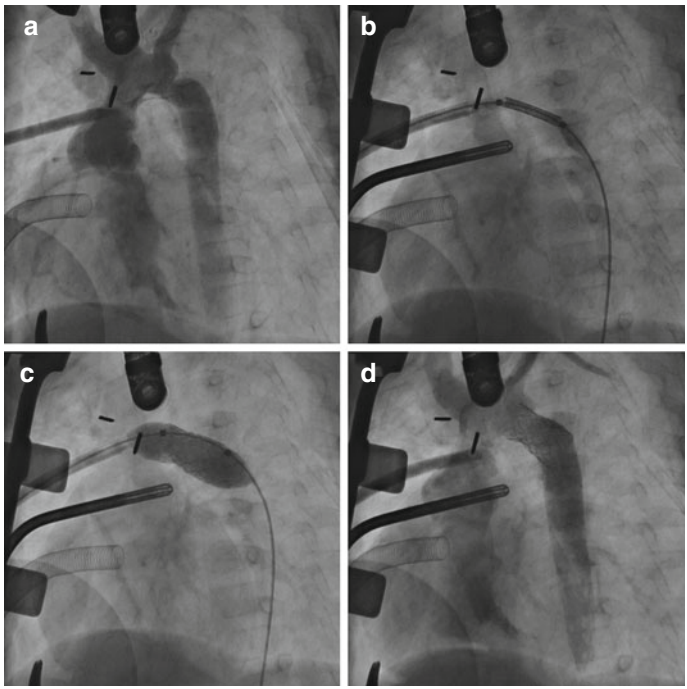

Fig. 40.4 Six-months-old infant with hypoplastic left heart syndrome undergoing a bidirectional Glenn procedure. There was a preexisting concern about an arch obstruction. Intraoperative angiography (**a**) documented a narrowing distal to the left subclavian artery. An intraoperative stent was placed through a sheath that was advanced over a wire through a purse string in the ascending aorta. (**b**) Stent positioning, (**c**) stent expansion, and (**d**) final angiogram

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.

Technically, the procedure is fairly straightforward and requires the use of a portable C-arm or a hybrid operating room or catheterization laboratory. The vascular access point

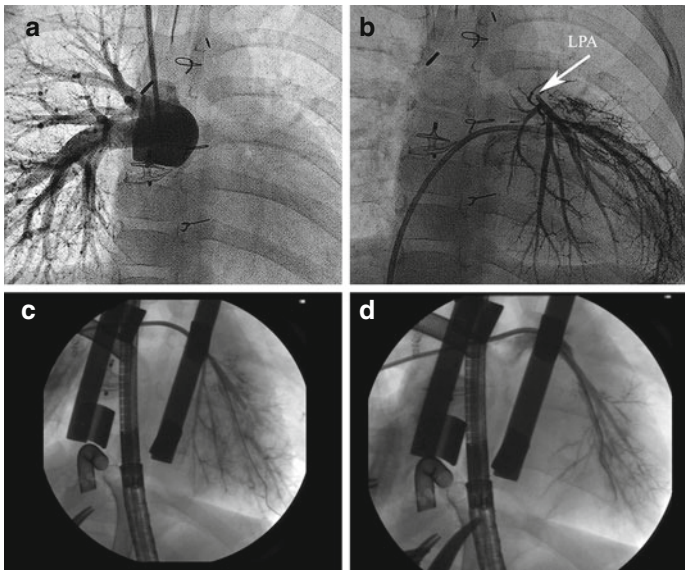

Fig. 40.5 Pre-Fontan catheter evaluation in a 3-year-old patient documented a disconnected LPA. (**a**) SVC angiography documenting no filling of an LPA. (**b**) Reverse left pulmonary venous angiography showing a small and hypoplastic LPA. (**c**) Intraoperative angiography documenting extreme hypoplasia of the LPA. (**d**) Angiography after deployment of a premounted stent documenting improved flow to the distally LPA

is chosen ideally directly opposite the lesion that requires treatment (Fig. 40.5). 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 usually utilized in a small patient, most angiographic acquisitions are performed 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 to a larger size. Stiff wires however are usually not required. 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. In most circumstances, 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 (Fig. 40.5).

40.4 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. 40.6). 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 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.

40.5 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 40.7 provides an example of a late-diagnosed 6-months-old infant with DORV, hypoplastic left ventricle, and a 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

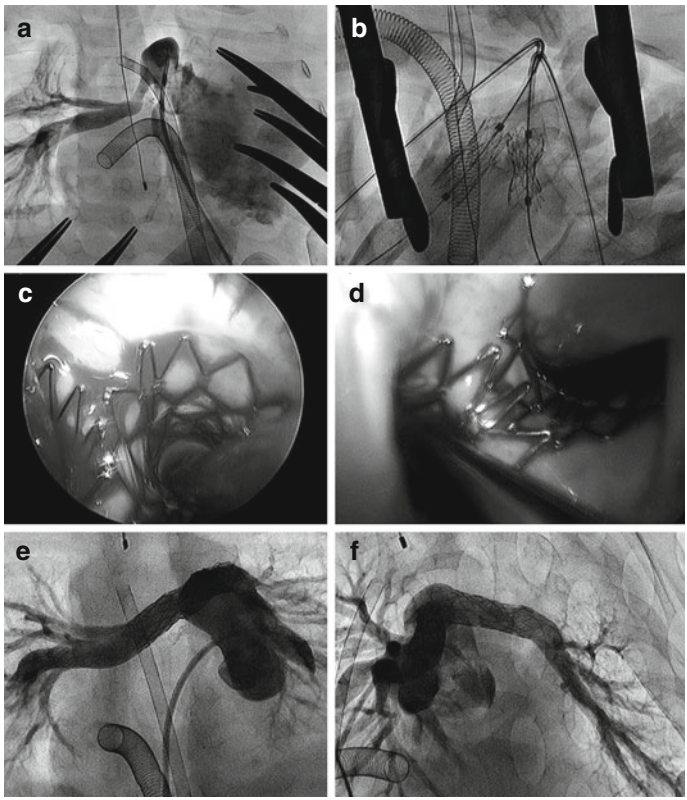

Fig. 40.6 Patient with pulmonary atresia and VSD after surgical correction who developed a large false aneurysm as well as bilateral branch pulmonary artery stenosis. (**a**) Bilateral branch PA stenosis delineated with intraoperative angiography. (**b**) Intraoperative stent expansion of RPA and LPA stent simultaneously. (**c**, **d**) Stent meshwork visualized after resection of the false aneurysm before (**c**) and after (**d**) manual folding of the mesh. Image (**e**) and (**f**): Exit angiography documenting excellent relief of the RPA and LPA stenosis

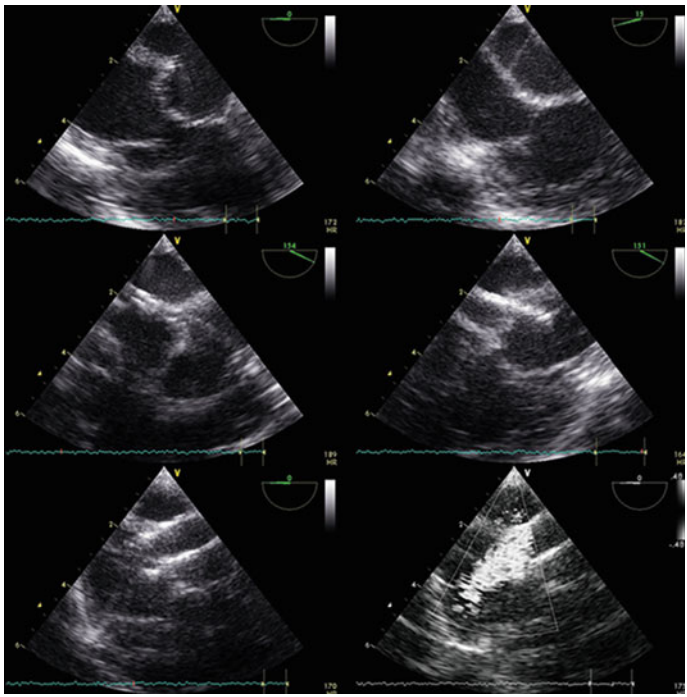

Fig. 40.7 Six-months-old infant with DORV, hypoplastic LV, mitral stenosis, and an intact atrial septum. Intraoperative stent placement across the atrial septum is performed at the time of pulmonary artery banding using guidance via transesophageal echocardiography. Needle tenting the atrial septum (*top left*). Wire crossing the atrial septum (*top right*). Sheath across the atrial septum (*middle left*). Stent positioning across the septum (*middle right*). Stent fully expanded across the septum (*bottom left*). Laminar color flow across the atrial septal stent (*bottom right*)

as a structure with increased echo-brightness. It is important to position the stent as central as possible. Once the stent has been expanded, the balloon and wire will need to be removed carefully to avoid dislocating the atrial septal stent in the process.

40.6 Exit Angiography

Any time an intraoperative vascular stent has been placed, it is important to evaluate the result using an exit angiography at the end of the procedure. Not only is exit angiography 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 transesophageal echocardiography. We published our 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 (Fig. 40.8) [3]. Exit angiography is fairly straightforward and can be performed just with the C-arm, and angiographic catheter, and a power injector.

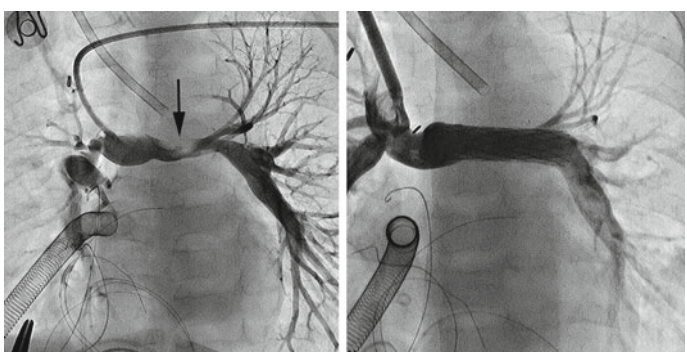

Fig. 40.8 Exit angiography after comprehensive stage II palliation in a 5-month-old infant with hypoplastic left heart syndrome documenting a stenosis of the left pulmonary artery (*arrow*) (*left*). Angiography after intraoperative placement of a stent to the left pulmonary artery (*right*)

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Part IX
Step-by-Step Procedures: Miscellanea

Chapter 41
Retrieval Techniques

Rui Anjos, Inês C. Mendes, and Duarte S. Martins

41.1 Introduction and Clinical Scenarios

Removal of foreign bodies from vessels or cardiac structures has become more frequent over recent years, with the widespread use of indwelling catheters, leads, guidewires, and devices.

Foreign bodies requiring intravascular or intracardiac retrieval usually result from 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

R. Anjos (✉) • I.C. Mendes • D.S. Martins
Department of Pediatric Cardiology, Hospital de Santa Cruz,
Centro Hospitalar de Lisboa Ocidental,
Rua Prof. Reinaldo dos Santos, Carnaxide, Lisboa 2795, Portugal
e-mail: ranjos@netcabo.pt; agnescmendes@hotmail.com;
duartesaraivamartins@gmail.com

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frequent with large devices, usually atrial septal or patent arterial duct devices and in most of these cases, a sternotomy, frequently with cardiopulmonary bypass, will be required.

41.2 Foreign Bodies and Embolized Materials Requiring Retrieval

The most frequently embolized material requiring retrieval is by far a fragment of catheter [1], usually as the result of a fractured central venous catheter. 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, and venous filters), guidewires, and pacing wires.

In most cases, embolization occurs in the systemic venous system, as the result of fractured long-term venous catheters. Embolized, fractured, or malpositioned 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. 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.

41.3 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 at that time, diagnosis and retrieval were made predominantly in symptomatic 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 some embolized foreign bodies in place may be acceptable in selected patients, especially in the presence of very small fragments, positioned in difficult access locations or in patients with a very low life expectancy [1].

41.4 X-Ray Equipment

Removal of a foreign body in the cardiac catheterization laboratory requires adequate equipment. The use of biplane X-ray equipment is extremely 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 in fact it is moving away from the target.

41.5 Planning of the Procedure

Careful planning of the procedure is of paramount importance. It is essential 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 retrieval 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. 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 concertina effect. This leads to loss of torque control and maneuverability 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, there are dedicated sheaths, 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.

41.6 Retrieving Systems and Devices

41.6.1 Snares

Snares are the most frequently used devices for retrieval procedures, employed in over 80–90 % of the cases [1–3]. Therefore,

all catheterization laboratories dealing with implantation of devices, therapeutic embolization, and retrieval procedures should have an available selection of multiple size snares.

41.6.1.1 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 snare 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 significantly 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) and the Amplatz GooseNeck® (EV3) with loop diameters ranging from 5 to 35 mm, inserted through 4–6 Fr catheters. The ONE Snare® (Merit Medical), with similar diameters and

catheters, has a small fold 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 EV3 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® (Radius) is a snare with a smooth helical loop 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 MICRO Elite Snare® (Radius) 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.

41.6.1.2 Multiple-Loop Snares

Multiple-loop snares have several loops opening simultaneously. The 2-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®, with 2–6 mm diameter, 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® (Angiotech), and the Atrieve Vascular Snare® (Angiotech). 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. It has a 40 mm diameter and requires an 8 Fr sheath. The CloverSnare 4 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.

41.6.1.3 Snares for 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 other 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.

41.7 Graspers and Forceps

Graspers for intravascular retrieval include the Vascular Retrieval Forceps® (Cook), used especially for intravascular retrieval of coils but also for catheters, guidewires, or other foreign objects, requiring a 4 F sheath or guiding catheter. The Alligator Retrieval Device® (Covidien) is designed for coil retrieval from small vessels, typically from the cerebral circulation and requires a 3 Fr microcatheter. Although these graspers are appealing, in reality, their use is generally limited to small vessels and their preferential use is for coil 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.

41.8 Baskets

Helical baskets are the only devices capable of engaging and retrieving a spherical or ovoid object (shaped 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 4-wire helical-loop basket, with 7 cm length and 3 cm diameter. The catheter shaft is 8 Fr, requiring a larger 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 the foreign body, which is frequently drawn into the basket when this is withdrawn, or with rotation of the retrieving system. The major disadvantage is that basket systems are bulky and rigid, with the potential to cause damage in their own right.

41.9 Step-by-Step Approach

41.9.1 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.

The approach should follow these steps [1, 3]:

- Obtain a thorough history and ascertain the type of catheter (length, diameter, flexibility) 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 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.
- 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 and 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 of the system 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, to avoid trauma to the heart or vessels.

41.9.2 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]:

- 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. 41.1). 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

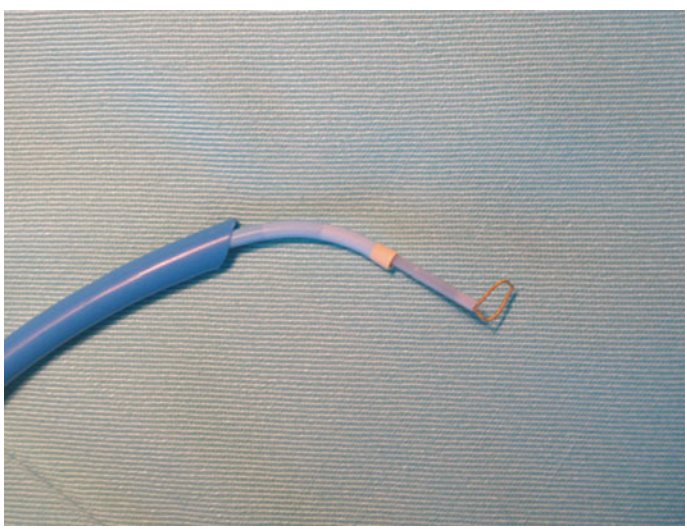

Fig. 41.1 A multiple coaxial system, formed by a snare, a guiding catheter, and a large valved sheath. The maneuverability and orientation of a guiding catheter to direct the snare are coupled with the resistance and dimension of the sheath. A coaxial system also provides extra support to retrieve large devices

the guiding catheter. With the multiple coaxial system, the maneuverability and orientation of a guiding catheter to direct the snare to an optimal position are associated with the size and support of a large sheath.

- The most commonly selected retrieval device for an occluder is a snare catheter. Single-loop catheters are often used, but multiple-loop snares are excellent options, increasing the probability of capture (Fig. 41.2). 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 into a sheath.

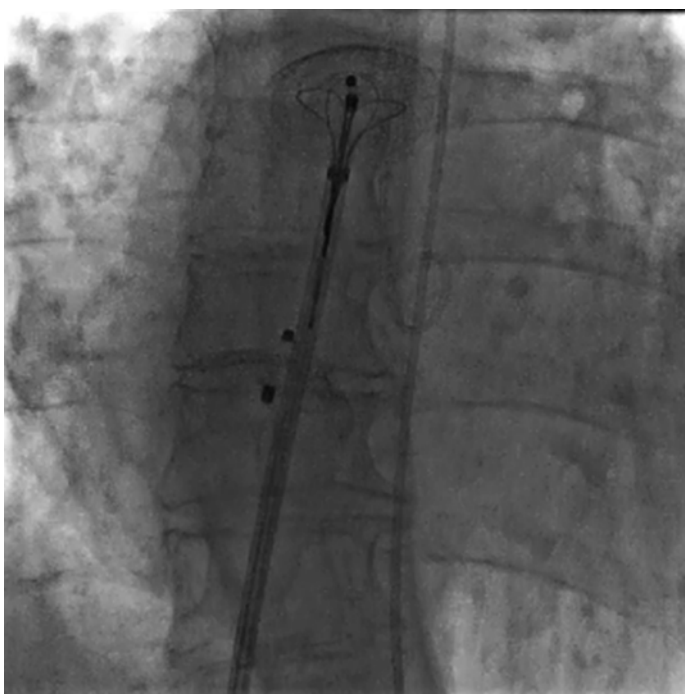

Fig. 41.2 A multiple-loop snare retrieving a large Amplatzer device embolized into the left atrium. The multiple loops increase the probability of capturing and when used on a frontal approach, as shown in this figure, has a self-centering mechanism

- 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 generally only 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. 41.3). A beveled tip is sharp and so can potentially cause damage to the vascular and cardiac structures, so care must be taken when advancing the sheath without the protection of a dilator or a coaxial catheter.
- 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.

41.9.3 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.

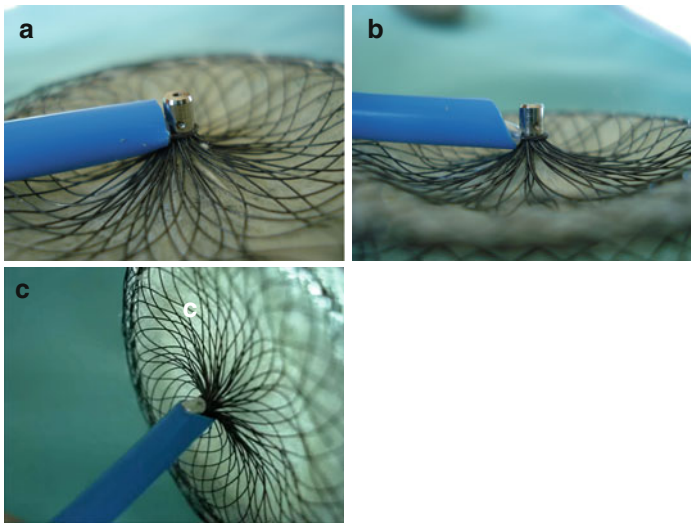

Fig. 41.3 Capture of the hub of a large Amplatzer device. Retrieval into the sheath is not possible as the hub is long and perpendicular to the sheath (**a**). Beveling the tip of the sheath (**b**) allows for hub retrieval into the sheath (**c**), as this increases the sheath extremity profile

- 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 never totally 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.

41.9.4 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 the position of the guidewire 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.

41.10 Complications of Retrieval

The most important complications related to retrieval techniques are vascular or cardiac tears or perforation/injury to cardiac valves and adjacent 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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Chapter 42
Pericardiocentesis

Maarten Witsenburg

42.1 Introduction

The pericardial space normally contains several ml of serous fluid. Due to diseases and 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 accumulation occurs over a longer period of time.

M. Witsenburg
Departments of Congenital and Pediatric Cardiology,
Thoraxcentre, Erasmus MC, PO Box 2040, 3000 CA Rotterdam,
The Netherlands
e-mail: m.witsenburg@erasmusmc.nl

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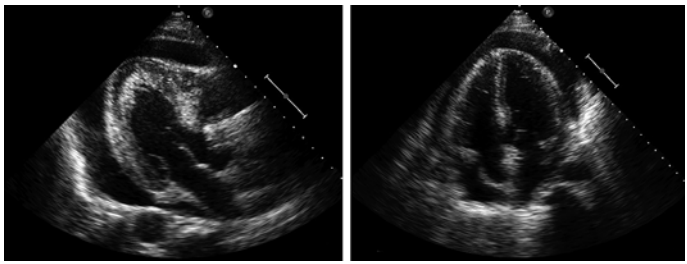

Fig. 42.1 Echocardiographic four-chamber and long-axis view showing moderate pericardial effusion

42.2 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 of a low-voltage ECG. The other extreme of the spectrum is the patient with acute low output due to cardiac tamponade where pericardial drainage is potentially lifesaving. Clinically low blood pressure, tachycardia and muffled heart sounds may raise suspicion. Echocardiography will reveal pericardial effusion (Fig. 42.1). 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.

42.3 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

42.4 Complications

Potential complications of the pericardial puncture 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.

42.5 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 is difficult to reach, and if multiple septa are present, 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.

42.6 Preparation

In a nonurgent 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.

42.7 Access and Drainage

A pericardial puncture set is prepared (Fig. 42.2).

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. 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 aspirate fluid. When necessary, the access can be echocardiographically confirmed by injecting some agitated saline.

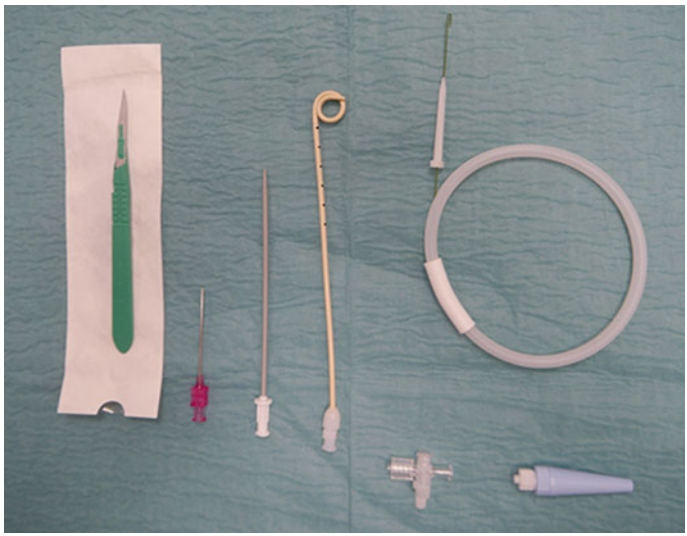

Fig. 42.2 Paediatric pericardial drainage set with scalpel (William Cook Europe, Bjaeverskov, Denmark)

(Alternative 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 3-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.

42.8 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.

Suggested Reading

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Chapter 43
Endomyocardial Biopsies

Davide Marini

43.1 Clinical Scenarios

Despite the fact that noninvasive diagnostic techniques are experiencing a period of tremendous development, endomyocardial biopsy (EMB) is the gold standard for in vivo diagnosis of rejection in cardiac allograft patients, cardiac tumours, myocarditis or other infiltrative cardiovascular diseases such as amyloidosis, sarcoidosis, Fabry disease and arrhythmogenic right ventricular dysplasia (ARVD).

Because of the invasiveness and complication risks of the procedure and the limited specificity of the noninvasive tests, it has often been difficult to identify the correct clinical setting in which EMB is necessary or it might be avoided. No randomized, controlled data exist on the use of EMB, and medical evidence is lacking especially in children. Moreover, relevant

D. Marini, MD, PhD
Department of Paediatric Cardiology, Ospedale Infantile Regina Margherita, Piazza Polonia, 94, Turin 10126, Italy
e-mail: dmarini@cittadellasalute.to.it

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published literature usually refers to specific cardiac diseases: instead, the decisions to recur to EMB are generally taken on the basis of clinical presentations, not of histopathological diagnoses, which are known only after the procedure. And this is a major obstacle to obtain clear indications to EMB in the clinical practice [1].

In order to define the current role of EMB in the management of cardiovascular disease, the American Heart Association (AHA), the American College of Cardiology (ACC) and the European Society of Cardiology (ESC) charged a multidisciplinary group of experts to provide a scientific statement by reviewing the published literature and organizing the information by clinical scenarios rather than histopathological diagnosis. The goal was to provide useful recommendations for clinical practice with classes of recommendations and levels of evidence (Table 43.1) [1]. This scientific statement was also endorsed by the Heart Failure Society of America and the Heart Failure Association of the European Society of Cardiology and published in 2007 [1].

43.2 Indications

According to this scientific statement:

A. EMB is recommended (*Class 1*) with *Level B* evidence in *Clinical Scenarios 1 and 2* (Table 43.1). In these clinical scenarios, the aim of the EMB is to detect the more aggressive forms of myocarditis, such as *giant cell myocarditis (GCM)* and *necrotizing eosinophilic myocarditis (NEM)*. Immunosuppressive treatment may improve the outcome in these forms; however, in the absence of clinical improvement, the diagnosis of GCM and NEM may require early implantation of mechanical circulation device support.

Table 43.1 Class of recommendation and level of evidence in 14 Clinical Scenarios [1]

Scenario number	Clinical scenario	Class of recommendation (I, IIa, IIb, III)	Level of evidence (A, B, C)
1	New-onset heart failure of <2 weeks' duration associated with a normal-sized or dilated left ventricle and hemodynamic compromise	I	B
2	New-onset heart failure of 2 weeks to 3 months duration associated with a dilated left ventricle and new ventricular arrhythmias, second- or third-degree heart block or failure to respond to usual care within 1–2 weeks	I	B
3	Heart failure of >3 months duration associated with a dilated left ventricle and new ventricular arrhythmias, second- or third-degree heart block or failure to respond to usual care within 1–2 weeks	IIa	C
4	Heart failure associated with a DCM of any duration associated with suspected allergic reaction and/or eosinophilia	IIa	C
5	Heart failure associated with suspected anthracycline cardiomyopathy	IIa	C
6	Heart failure associated with unexplained restrictive cardiomyopathy	IIa	C
7	Suspected cardiac tumours	IIa	C
8	Unexplained cardiomyopathy in children	IIa	C
9	New-onset heart failure of 2 weeks' to 3 months' duration associated with a dilated left ventricle, without new ventricular arrhythmias or second- or third-degree heart block, that responds to usual care within 1–2 weeks	IIb	B

(continued)

Table 43.1 (continued)

Scenario number	Clinical scenario	Class of recommendation (I, IIa, IIb, III)	Level of evidence (A, B, C)
10	Heart failure of >3 months' duration associated with a dilated left ventricle, without new ventricular arrhythmias or second- or third-degree heart block, that responds to usual care within 1–2 weeks	IIb	C
11	Heart failure associated with unexplained HCM	IIb	C
12	Suspected ARVD/C	IIb	C
13	Unexplained ventricular arrhythmias	IIb	C
14	Unexplained atrial fibrillation	III	C

Conversely, the diagnosis of *lymphocytic myocarditis* is a positive prognostic factor for spontaneous recovery, even in patients on inotropic or mechanical circulatory support.

B. EMB is considered reasonable (*Class IIa*) with *Level C* evidence in *Clinical Scenario 3* because patients may be at risk of cardiac *sarcoidosis or idiopathic granulomatosis myocarditis* and in *Clinical Scenario 4* because they might be at risk of *hypersensitivity myocarditis (HSM)* (Table 43.1). Both sarcoidosis and HSE may respond to therapy with corticosteroids.

C. EMB is also considered a reasonable procedure (*Class IIa*) with *Level C* evidence in cases of unexplained heart failure associated with *suspected anthracycline cardiomyopathy* (*Clinical Scenario 5*) or *restrictive cardiomyopathy* (*Clinical Scenario 6*) in which it may reveal either a specific infiltrative disorder, for example, *amyloidosis* or *hemochromatosis*, or myocardial fibrosis and myocyte hypertrophy consistent with *idiopathic restrictive cardiomyopathy* and in the setting of any *unexplained cardiomyopathy in children* (*Clinical Scenario 8*).

D. In cases of suspected *cardiac tumour* (*Clinical Scenario 7*), with the exception of typical myxomas, EMB is a reasonable investigation (*Class IIa*) with *Level C* evidence, if (1) the diagnosis cannot be established by noninvasive or less invasive techniques, (2) tissue diagnosis can be expected to influence the course of therapy, (3) the chances of successful biopsy are believed to be reasonably high and (4) the procedure is performed by an experienced operator [1].

E. EMB may be considered to detect a lymphocytic myocarditis in *Clinical Scenario 9* (*Class IIb*), with *Level B* evidence and in *Clinical Scenario 10,* in *unexplained hypertrophic cardiomyopathy*, in suspected *ARVD* and *unexplained ventricular arrhythmias* (respectively, *Clinical Scenarios 11,12,13) (Class IIb)* with *Level C* evidence. EMB should not be performed in the setting of unexplained atrial fibrillation (*Class III*), with *Level C* evidence.

Additionally, EMB is indicated in *clinical suspicion of allograft rejection* in posttransplant recipients [2].

43.3 Preparation of the Exam and Vascular Access

In adults with normal ejection fraction, EMB can be performed using local anaesthesia or light sedation. Infants and young children require general anaesthesia, endotracheal intubation and mechanical ventilation. Monitoring including electrocardiographic rhythm, blood pressure and pulse oximetry should be part of the routine practice. The use of heparin prophylaxis is generally avoided if the approach is exclusively venous.

The preferred venous access points are the right internal jugular vein and the right femoral vein. The choice is generally based on the operator's experience. Subclavian access is rarely employed.

Usually, the femoral artery is chosen for retrograde access to the left ventricle. In this case, aspirin or other antiplatelet agents may be added to heparin to decrease the risk of systemic embolization. It has been demonstrated that biventricular samples increase the diagnostic sensitivity of EBM for diagnoses of myocarditis; however, this may be due to a higher number of biopsy specimens taken in biventricular EMBs [3].

43.4 Materials

The bioptomes commonly employed are single-use, with sharpened cusps designed to pinch instead than to cut the myocardial muscle (Fig. 43.1). There are two different types of bioptomes: (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 approach. Flexible bioptomes requiring the use of a long sheath or a guiding catheter are used for both arterial and venous femoral approach. Some common bioptomes available on the market are listed in Table 43.2 (Fig. 43.2).

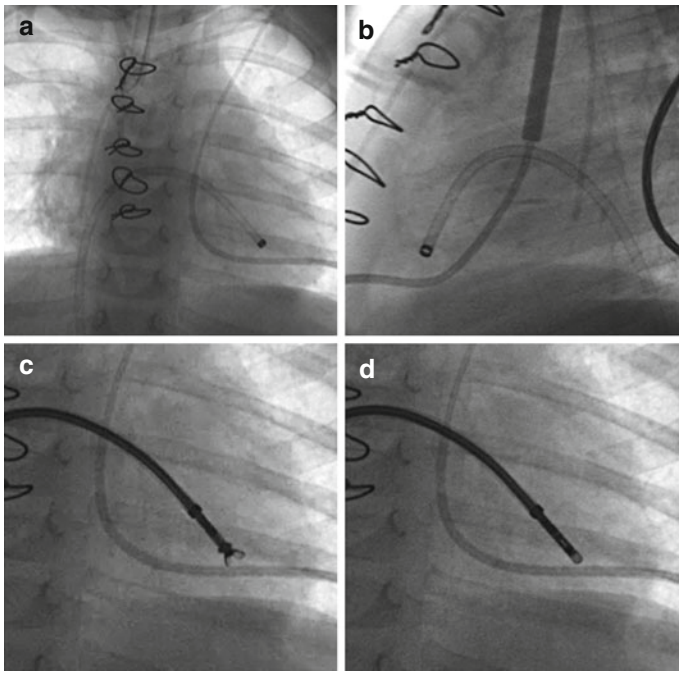

Fig. 43.1 Anteroposterior view (**A**) and lateral view (**B**) showing the position of the long sheath from femoral venous approach. The jaws of the bioptome should be open within the ventricular cavity before reaching the muscular wall (**C**). The jaws should be closed to pinch the interventricular septum keeping a gentle pressure (**D**)

43.5 Pre-procedural Imaging

Echocardiography should be routinely performed to evaluate the global and the segmental ventricular function and to exclude the presence of any other abnormalities such as pericardial effusion or valvular dysfunction before the procedure. Also, computed tomography (CT) and cardiac MRI may be of interest in patients scheduled for EMB. In order to decrease the risk of inadvertent

Table 43.2 Common bioptomes of different size and shaft length

Model	Size	Shaft length (cm)
Cook®	3.0, 5.2	60 and 120
Cordis®	5.4, 7.0, 8.0, 9.0	50 and 104
SparrowHawk®	5.0, 6.0, 7.0	50 and 105
Novatome™	5.0, 6.5, 8.0, 9.0	50 and 100
Argon®	5.0, 5.5, 6.0, 7.0,7.5	50 and 105
Meiners™	6.0	110

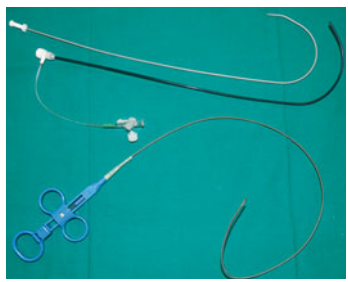

Fig. 43.2 A 50 cm long flexible bioptome (Cordis®) may be used in a 6F 40 cm long pre-shaped Flexor® Check-Flo® Introducer® (Cook®)

biopsy of the right ventricular free wall, CT may be used to assess the angle of the interventricular septum with the superior or the inferior vena cava. An interesting clinical approach to increase the diagnostic sensitivity of EMB is to use prior cardiac MRI to guide the biopsy to areas of the abnormal myocardium suggesting inflammatory disease or cellular necrosis [3].

43.6 Technique

From the femoral venous access site, a long sheath with angulated tip is advanced over a standard guide wire placed in the distal pulmonary artery and positioned within the right ventricle.

From the internal jugular approach, the preshaped and stiff bioptome is usually manoeuvred independently and advanced into the right ventricle without the protection of a long sheath. By using this approach, the bioptome has to cross the tricuspid valve each time to obtain a muscular specimen.

For EMB from the left ventricle, usually the selected long sheath has a straight tip. Alternatively, a coronary guiding catheter may be used.

The bioptome is advanced under fluoroscopic surveillance generally 0° anteroposterior and 90° latero-lateral views for the right ventricle and 30° right anterior oblique and 60° lateral anterior oblique for the left ventricle. Additionally, 2D/3D transthoracic or transoesophageal echocardiography may be used in combination with fluoroscopy to control the bioptome direction and its final position.

After each biopsy, drawing some blood from the long sheath or from the catheter and flushing with isotonic saline are wise to prevent clotting.

43.7 Expected Results

Depending on the studies to be performed on the specimens, the procedure should obtain 5–10 samples of 1–2 mm^2 in size, which should be collected from 2 to 3 different sites in order to reduce the sampling error [1].

43.8 Tips and Tricks

Obtaining biopsy specimens from the free right or free left ventricular wall risks perforation and cardiac tamponade; hence, the samples should be taken from the interventricular septum. This

is mandatory in cases of EMBs in patients with dilated cardiomyopathy or clinical suspicion of acute myocarditis, in which the right free wall is often thin and friable.

Before the insertion of the flexible bioptome through the long sheath, it may be useful to slightly bend the distal end of the bioptome to provide an adequate curve and to enhance its flexibility. In addition, it is recommended to open the jaws of the bioptome within the ventricular cavity before reaching the muscular wall. Then, the bioptome must be advanced slowly until feeling mild resistance. When the bioptome engages the wall, it usually provokes ventricular premature beats. It is important to keep gentle pressure while the jaws close to pinch (Fig. 43.3). Then the bioptome may be retrieved, always keeping the jaws closed.

Before the central venous catheter is removed or the patient leaves the catheterization laboratory, echocardiography may be used to exclude any postprocedural pericardial effusion.

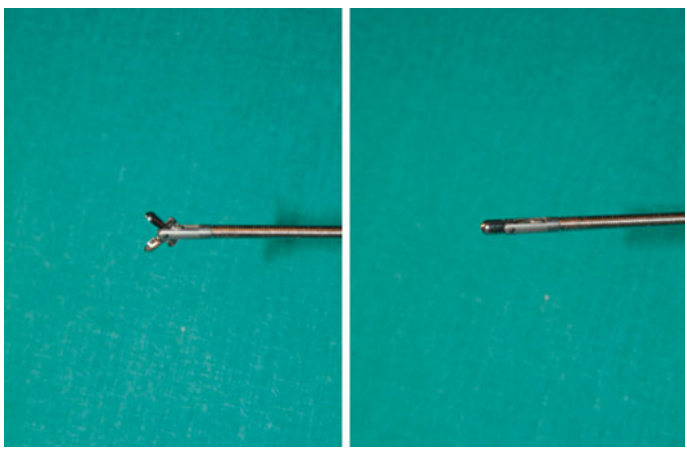

Fig. 43.3 The profile of the biopsy forceps (Cordis®) when the jaws are open (*left panel*) and closed (*right panel*)

43.9 Pitfalls and Complications

The main drawbacks of this technique are the risk of procedural complications and long-term sequelae. Furthermore, the EMB consists of taking a limited number of specimens from the myocardial muscle of the right or left ventricle. This means that the sensitivity of EMB is limited [1–3].

Many different complications may occur during EMB, related to sheath insertion, to the biopsy itself or to the clinical status of the patients. Major and well-known complications include pericardial tamponade with the need for pericardiocentesis, hemopericardium, permanent AV block and severe valvular damage. The precise frequency of these events is not known. An overall complication rate as less than 6 % is reported in most published series and registries [1–3]. Life-threatening events occur less frequently: most series report major acute complication rates of less than 1 % [1–3]. Of note, the risk of EMB is highest in sick children with suspected myocarditis on inotropic support, especially in infants. Although very rare, death may occur, in most cases as a result of perforation with cardiac tamponade in patients with cardiogenic shock or unstable ventricular arrhythmias.

43.10 How to Manage Complications

In cases of pericardial tamponade, immediate pericardiocentesis should be performed. The blood sucked from the pericardium should be promptly returned to the patient through the vascular introducer. While performing the standard manoeuvres of resuscitation, the surgical team should be contacted in order to repair the lesion and possibly implant a ventricular assist device.

Table 43.3 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

43.11 Postprocedural Care

The samples should be removed off the bioptome by using a sterile needle and placed on sterile gauze. The type of storage depends on the clinical question [1, 2]. Some standard fixatives and techniques of analysis are listed in Table 43.3.

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Chapter 44
Evaluations Before Partial and Total Cavopulmonary Connections

Gabriella Agnoletti

44.1 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 left ventricle whilst 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 precedes conversion to TCPC. PCPC forces 50 % of cardiac output to bypass the heart and directly enter the lungs, increasing oxygen saturation.

G. Agnoletti
Department of Pediatric Cardiology,
Hospital "Città della Salute e della Scienza", Torino 10100, Italy
e-mail: gagnoletti@cittadellasalute.to.it

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44.2 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 different scenarios possible:

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 catheterisation laboratory.

44.3 Indications and Patient Selection

PCPC and TCPC are often preceded by cardiac catheterisation, aimed at measuring PAP, assessing pulmonary artery size and treating possible associated anomalies. Catheterisation 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 catheterisation is commonly performed prior to PCPC include:

- Subject 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 catheterisation 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.

44.4 Imaging

Noninvasive imaging can provide definitive information on pulmonary artery anatomy (Fig. 44.1).

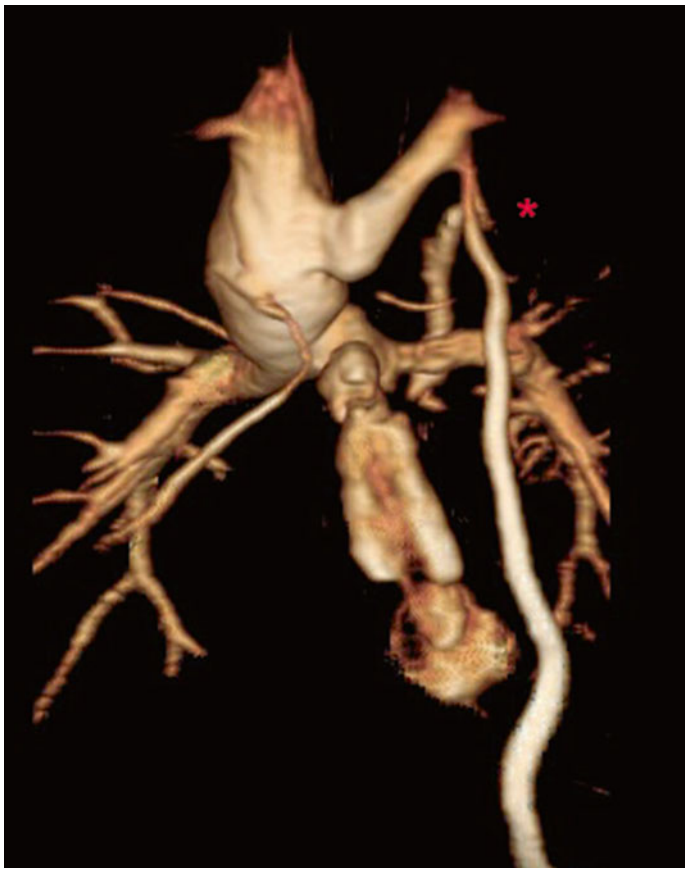

Fig. 44.1 3D MRI in AP view shows a dilated superior vena cava, the presence of antegrade flow and a venovenous collateral (*)

In addition, 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.

44.5 Pre-PCPC Catheterisation

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.

Arterial access is generally needed to visualise 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 visualise 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 visualisation of the banded pulmonary arteries, the stented arterial duct and the native aortic arch.

I 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.

44.6 Pre-TCPC Catheterisation

Catheterisation is performed, according to the presence of forward flow to the lungs, via the femoral vein or the internal jugular vein [1].

Arterial catheterisation 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.

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 performed. 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 transoesophageal 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. 44.2).

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.

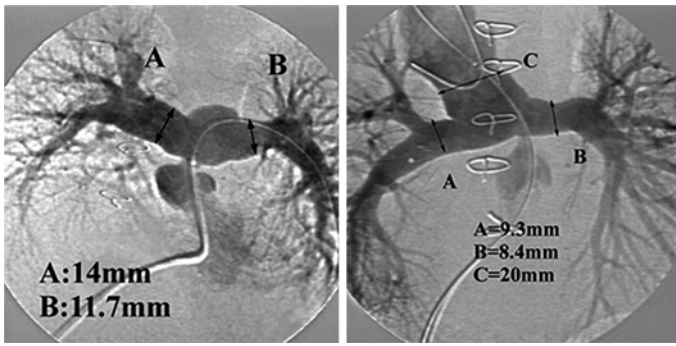

Fig. 44.2 Measurement of pulmonary artery (*double-headed arrows*) size in two different patients before (*left*) and after (*right*) PCPC. In both cases, the catheter enters the pulmonary artery in an antegrade way. In the patient with PCPC, a multi-track catheter (notice the guidewire parallel to the 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 are used to calculate cross-sectional areas (Fig. 44.2).

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 [3].

44.7 Materials

Diagnostic catheterisation 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 catheterisation 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 the anatomy and size of the vessels.

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.

44.8 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 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.

44.9 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. 44.3).

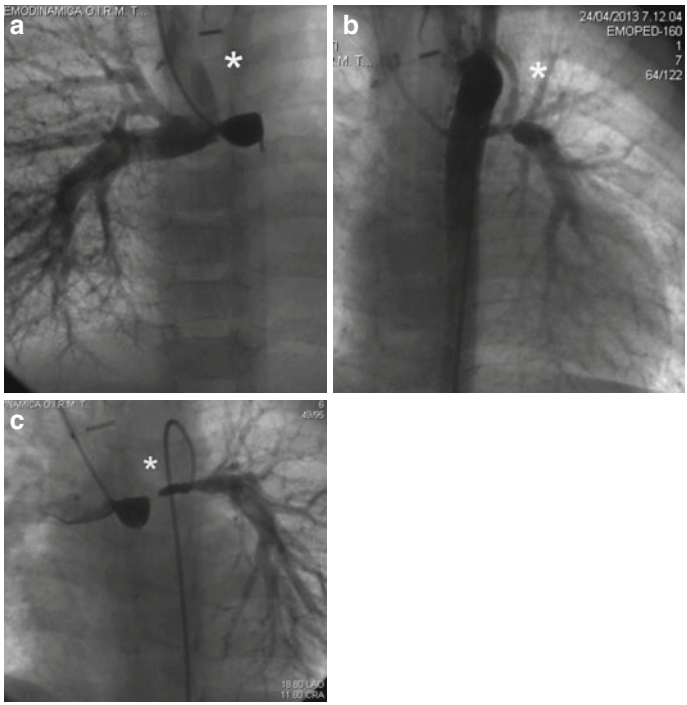

Fig. 44.3 Disconnected pulmonary arteries in a patient with competitive flow. The right pulmonary artery is fed via the superior vena cava (**a***) and the left pulmonary artery via a BT shunt (**b***). Simultaneous injection allows to appreciate the distance between the pulmonary arteries (**c***)

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 catheterisation can be performed from the venous approach.

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.

44.10 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.

44.11 Complications

Patients in whom a diagnostic catheterisation was performed have the general risks of any cardiac catheterisation.

Jugular catheterisation 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 catheterisation was performed are subject to the risks of the respective interventions. There are generic rules and specific rules to manage complica-

tions. Generic rules are be quiet, be logical and avoid useless manoeuvres. 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.

44.12 Post-procedural Care and Follow-Up

Patients having had a diagnostic catheterisation should undergo standard follow-up. In anti-aggregated patients having had a jugular catheterisation, particular attention should be paid to the risk of bleeding. After interventional catheterisation post-procedural care varies in accordance with the intervention performed.

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Chapter 45
Hemodynamics in Pericardial and Myocardial Diseases

Giacomo Pongiglione and Maria Giulia Gagliardi

45.1 Pericardial Disease

45.1.1 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

G. Pongiglione (✉) • M.G. Gagliardi
Medical and Surgical Department of Pediatric Cardiology
and Cardiosurgery, Bambino Gesù Children's Hospital, IRCCS,
Piazza San't Onofrio, 4, 00165 Rome, Italy
e-mail: giacomo.pongiglione@opbg.net; mgiulia.gagliardi@opbg.net

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3. Distribute hydrostatic forces over the heart
4. Exclude extracardiac and/or intrathoracic disease from extension into the heart [1]

45.1.2 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:

Transmural filling pressure = intracavitary pressure (EDP) – 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. 45.1).

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].

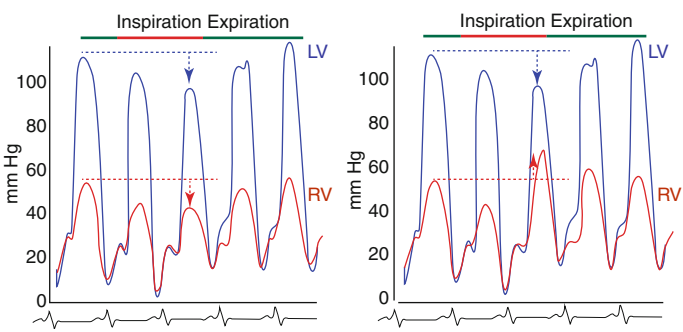

Fig. 45.1 Left tracing, normal; right tracing typical of constriction. Normally there is a concordant fall in LV and RV pressures in inspiration. In constriction, there is discordance of LV and RV pressure changes in inspiration: the LV pressure falls, and the RV pressure rises

45.2 Clinical Scenarios

45.2.1 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 descendant; 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. 45.2). 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].

[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.

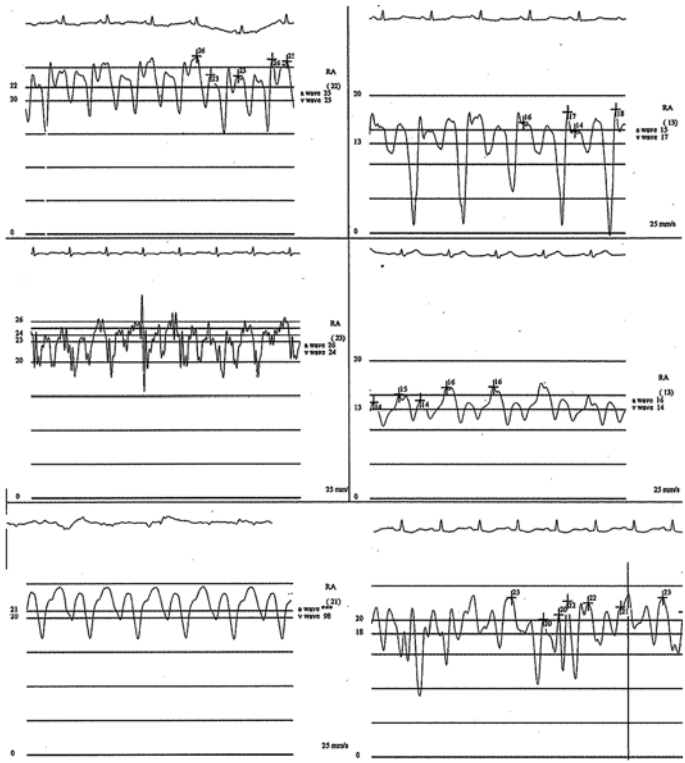

Fig. 45.2 Right atrial pressure in proven constriction cases. The mean pressures are all elevated. Although the *x* and *y* descent are all exaggerated, it can be seen how variable the depth of the *x* and *y* descent may be. In most cases, the magnitude of the *x* and *y* descents are similar, producing 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 and the pressure increases – Kussmaul's sign. It is better appreciated by a mean pressure tracing

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].

45.2.2 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 fares far less well with surgical *pericardiectomy* than do other causes of constriction. In constrictive disease, the pericardium presents thickened, sometimes with calcification, but it can be apparently normal.

45.2.2.1 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, 3].

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 Table 45.1, there are echocardiographic and hemodynamic data helping differentiate between constrictive pericarditis and restrictive cardiomyopathy.

45.3 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.

Table 45.1 Clues to differentiate constrictive pericarditis and restrictive cardiomyopathy

Restrictive cardiomyopathy	ECHO	Constrictive pericarditis
LVH, RVH	Ventricle morphology	Normal ventricles
Typical, biatrial	Atrial enlargement	Possible left atrial enlargement
Normal systolic function	Ventricle function	Inspiratory septal bounce
Low tissue Doppler velocity	Tissue Doppler	Normal tissue Doppler velocity (TDI mitral lateral wall >8 cm/s)
Normal LV inflow velocity	Doppler	LV inflow velocity fall by 25 % with insp.
	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	No
Normal	Inspiratory variation	Exaggerated

LVH left ventricle hypertrophy, *RVH* right ventricle hypertrophy, *TDI* tissue Doppler imaging, *LVED* left ventricle end-diastolic pressure, *RVEDP* right ventricle end-diastolic pressure

Myocarditis (MYO) is an inflammation of the myocardial tissue and may need a catheterization for endomyocardial biopsy (see Chap. 44).

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.

45.3.1 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

45.3.2 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 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, Terumo guidewire, and thermodilution (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 has not been previously excluded.

Routine measurement of oxygen saturation in blood sample taken from superior vena cava and pulmonary artery to detect unsuspected shunts [2].

45.3.3 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 cardiac index (CI = CO/BSA) as a functional parameter.

- In neonates, normal C.I. ranges = 4–5 l/min/m^2.
- In children = 3–4.5 l/min/m^2.
- In adolescent and young adult = 2.5–4 l/min/m^2 [3].

Left ventricular pressure waveform: Both the rate of rise and the rate of fall of left ventricular pressures are slow. The ventricular pressure 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 (dF/dt) [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 transpulmonary gradient is >12 mmHg [2] (for vasoreactive tests, see Chap. 9).

45.3.4 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 mid-ventricular 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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Chapter 46
Imaging and Treating Coronary Arteries in Children

Teiji Akagi

46.1 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.

T. Akagi, MD, PhD, FACC, FSCAI
Cardiac Intensive Care Unit, Okayama University Hospital,
2-5-1, Shikata-cho, Kita-ku, Okayama 700-8558, Japan
e-mail: t-akagi@cc.okayama-u.ac.jp

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In about half of the cases, the conus branch is the first branch of the RCA. In the 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, has a common origin with the RCA, or has 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 toward 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 toward 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 toward 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 marginal branches supply the lateral wall of the LV. They are numbered sequentially from proximal to distal.

46.2 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 toward the patient's left, and negative angles are toward 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 toward the patient's head; negative angles are toward the patient's legs.

- Right coronary artery is best seen in 45° right anterior oblique, 45° left anterior oblique, and lateral projections.
- Left main coronary artery is usually well visualized in the "spider view" (20° caudal + 45–60° left anterior oblique view).
  - Left anterior descending artery: it is usually seen using multiple angiographic views including (a) 20–25° caudal + 15° left anterior oblique, (b) 40° cranial + 10° right anterior oblique, and (c) 30° cranial + 45° left anterior oblique.
  - Left circumflex artery is best seen with caudal angulations associated to either right anterior oblique or left anterior oblique inclinations.

46.3 Kawasaki Disease

46.3.1 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 [1]. 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 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 coronary risk factors such as hyperlipidemia, smoking, or hypertension may accelerate these conditions.

46.3.2 Procedure

- During the 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 the pediatric population.
- It is obviously true that the fundamental therapeutic regimen for ischemic heart disease after Kawasaki disease is coronary bypass surgery. However, the long-term coronary graft patency is not satisfactory. Even it the use of intrathoracic arterial bypass graft, the unsatisfactory surgical results exist, especially operative age under the 12 years old.
- 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 (PTCRA) may be the only effective treatment (Fig. 46.1). Excellent acute results for

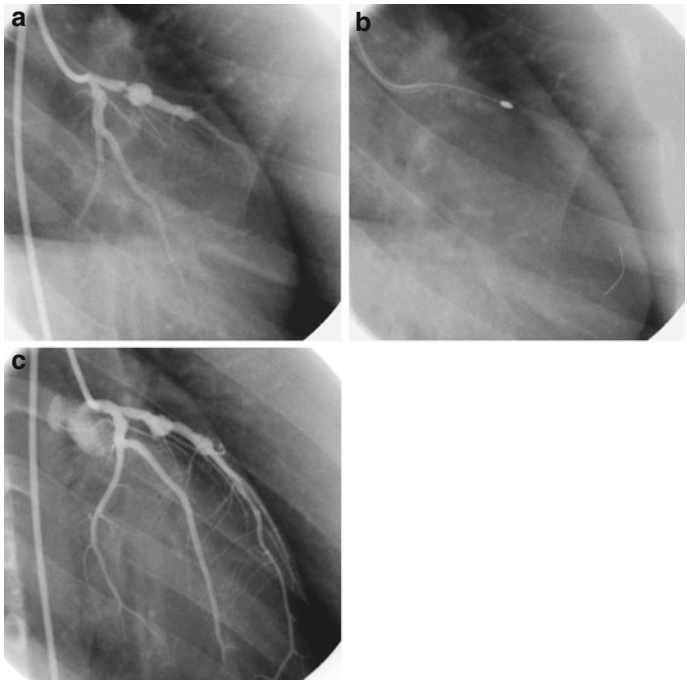

Fig. 46.1 Coronary rotational ablation in Kawasaki disease. (**a**) Before the intervention, (**b**) rotational ablation, and (**c**) post-rotational ablation. Coronary stenosis is completely resolved

PTCRA were observed in previous studies (Fig. 46.2). Although the 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. For this reason, this procedure can only be performed in older patients.

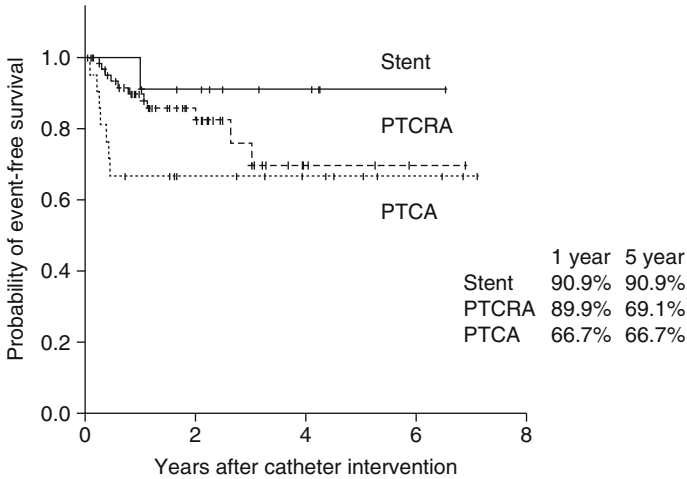

Fig. 46.2 Long-term outcome after catheter intervention in Kawasaki disease (*PTCA* percutaneous coronary angioplasty, *PTCRA* percutaneous coronary rotational ablation)

- Anticoagulation and antiplatelet medication should be continued through their life. Intravascular ultrasound imaging provides valuable information for the selection of the appropriate interventional procedure and early detection of vascular complications.

46.3.3 Other Special Conditions

46.3.3.1 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.

46.3.3.2 Post-arterial Switch Evaluation

- First of all, it is mandatory to know the spectrum of coronary artery variations seen in d-TGA (Fig. 46.3) and the surgical report.
- Secondly, usually the coronary arteries are reimplanted 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 toward the aorta in order to clearly delineate the surgically created ostium.

46.3.3.3 Pulmonary Atresia and Intact Ventricular Septum

See Chap. 19.

46.3.3.4 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 at the facing PA sinus but can occur to the main pulmonary artery or at the level of one of the two proximal branches.

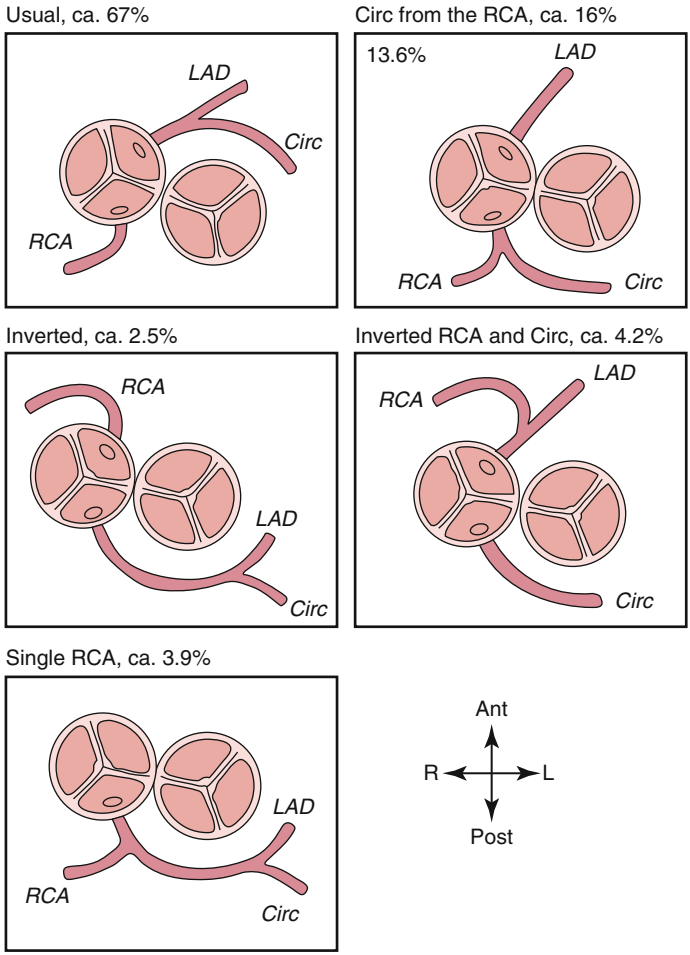

Fig. 46.3 Coronary artery anatomy in d-TGA (From Wernovsky and Sanders [2]. With permission)

46.3.3.5 Coronary Artery Disease Posttransplantation

It consists in a concentrical myointimal proliferation involving the entire vessel.

46.4 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 related to the forced injection of contrast inside a small branch of against the wall of the coronary artery.

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Part X
Tables and Charts: Appendix

Chapter 47
Hemodynamic Formulae, Calculations, and Charts

Lee N. Benson and Juan Pablo Sandoval Jones

47.1 Body Surface Area Formulae (BSA)

Mosteller [1]*:*

$$\mathrm{BSA}\left(\mathrm{m}^2\right) = \sqrt{\frac{\mathrm{height}\left(\mathrm{cm}\right) \times \mathrm{weight}\left(\mathrm{kg}\right)}{3{,}600}}$$

Dubois and Dubois [2]*:*

$$\mathrm{BSA}\left(\mathrm{m}^2\right) = 0.007184 \times \mathrm{height}\left(\mathrm{cm}\right)^{0.725} \times \mathrm{weight}\left(\mathrm{kg}\right)^{0.425}$$

L.N. Benson MD (✉) • J.P.S. Jones
Division of Cardiology, Department of Pediatrics,
The Hospital for Sick Children, 555 University Ave,
Toronto, ON M5G 1X8, Canada
e-mail: lee.benson@sickkids.ca; juanpablo.sandovaljones@sickkids.ca

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47.2 Derived Hemodynamic Data

47.2.1 Cardiac Output

$$\mathrm{SV} = \mathrm{EDV} - \mathrm{ESV}$$

$$\mathrm{CO} = \mathrm{SV} \times \mathrm{HR}$$

$$\mathrm{EF} = \mathrm{SV} / \mathrm{EDV}$$

where SV = stroke volume, EDV and ESV = end-diastolic and end-systolic volumes, CO = cardiac output, HR = heart rate, and EF = ejection fraction.

47.2.2 The Fick Equation

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

where Q is cardiac output expressed in liters per minute (l/min) and $\mathrm{VO_2}$ is the oxygen consumption in ml $\mathrm{O_2}$/min.

The denominator of the Fick equation is the arteriovenous oxygen content difference (a – v $\mathrm{O_2}$ diff) and is expressed as ml $\mathrm{O_2}$/l of blood.

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

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:

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

$$\mathrm{C_vO_2}\left(\mathrm{ml\,O_2/l}\right) = \mathrm{Oxygen\ capacity}\left(\mathrm{ml\,O_2/l}\right) \times \mathrm{venous\ oxygen\ saturation}\left(\%\right)$$

If the patient is breathing enriched oxygen ($F_IO_2 > 30\ \%$), 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.

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

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

47.2.3 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:

$$Q_p = \frac{VO_2}{\text{pulmonary venous } O_2 \text{ content} - \text{pulmonary arterial } O_2 \text{ content}} \text{ or}$$

$$Q_p = \frac{VO_2\,(\text{ml } O_2/\text{min})}{(\text{PV sat} - \text{PA sat}) \times 1.39 \times \text{Hgb}\,(\text{g/l})}$$

where PV is pulmonary vein and PA is pulmonary artery saturation.

Similarly, Q_s is estimated as

$$Q_s = \frac{VO_2}{\text{systemic arterial } O_2 \text{ content} - \text{mixed venous } O_2 \text{ content}} \text{ or}$$

$$Q_s = \frac{VO_2\,(\text{ml } O_2/\text{min})}{(\text{Ao sat} - \text{MV sat}) \times 1.39 \times \text{Hgb}\,(\text{g/l})}$$

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.

$$Q_{\mathrm{p}} = \frac{\mathrm{VO}_2}{\left(\text{pulmonary venous } \mathrm{O}_2 \text{ content} - \text{mixed venous } \mathrm{O}_2 \text{ content}\right)} \text{ or}$$

$$Q_{\mathrm{ep}} = \frac{\mathrm{VO}_2\left(\mathrm{ml/min}\right)}{\left(\mathrm{PV\,sat} - \mathrm{MV\,sat}\right) \times 1.39 \times \mathrm{Hgb}\left(\mathrm{g/l}\right)}$$

$$\text{Mixed venous saturation} = \frac{\left(3 \times \mathrm{SVC\,sat} + \mathrm{IVC\,sat}\right)}{4} \text{ or}$$

$$= \frac{\mathrm{SVC\,sat} - \left(\mathrm{SVC\,sat} - \mathrm{IVC\,sat}\right)}{4}$$

$$Q_{\mathrm{p}} : Q_{\mathrm{s}} = \frac{\left(\mathrm{Ao\,sat} - \mathrm{MV\,sat}\right)}{\left(\mathrm{PV\,sat} - \mathrm{PA\,sat}\right)}$$

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.

47.2.4 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):

$$\mathrm{O_2ER} = \frac{\mathrm{VO_2}}{\mathrm{DO_2}}.$$

47.2.5 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 $\times 80 = \mathrm{dyne} - \mathrm{sec} - \mathrm{cm}^{-5}$

Normal values:

PVRI: 1–3 Wood units $\times\, \mathrm{m}^2$ or 80–240 $\mathrm{dyn} \times \mathrm{s} \times \mathrm{cm}^{-5} \times \mathrm{m}^2$

SVRI: 15–30 Wood units $\times\, \mathrm{m}^2$ or 800–1,600 $\mathrm{dyn} \times \mathrm{s} \times \mathrm{cm}^{-5} \times \mathrm{m}^2$

47.2.6 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.

47.2.6.1 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			

47.2.6.2 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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