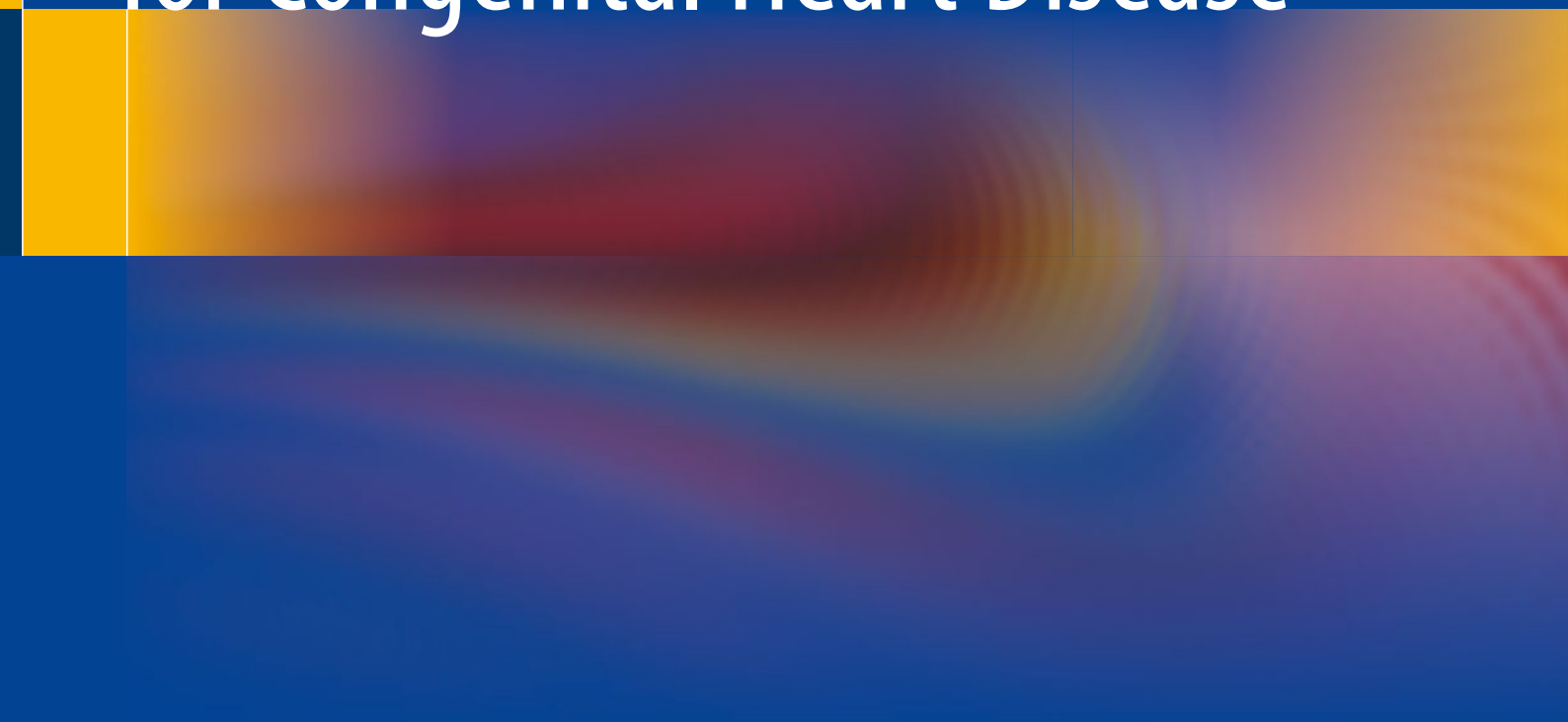
Gianfranco Butera
Massimo Chessa
Andreas Eicken
John D. Thomson
*Editors*

# Atlas of Cardiac Catheterization for Congenital Heart Disease



Springer

EXTRAS ONLINE

# Atlas of Cardiac Catheterization for Congenital Heart Disease

Gianfranco Butera • Massimo Chessa
Andreas Eicken • John D. Thomson
Editors

# Atlas of Cardiac Catheterization for Congenital Heart Disease

Springer

*Editors*
Gianfranco Butera
Department of Pediatric Cardiology and Adults with Congenital Heart Disease
Evelina London Children's Hospital
St Thomas' Hospital
London
United Kingdom

Andreas Eicken
Department of Pediatric Cardiology and Congenital Heart Disease
German Heart Center Munich
Technische Universität München
Munich
Germany

Massimo Chessa
Children & Adult Congenital Heart Disease
Policlinico San Donato
San Donato Milanese
Italy

John D. Thomson
Yorkshire Heart Center
Pediatric Cardiology
Leeds General Infirmary
Leeds
United Kingdom

Additional material to this book can be downloaded from http://extras.springer.com.

ISBN 978-3-319-72442-3 ISBN 978-3-319-72443-0 (eBook)
https://doi.org/10.1007/978-3-319-72443-0

Library of Congress Control Number: 2018968277



This Springer imprint is published by the registered company Springer Nature Switzerland AG
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

# Preface

A picture is worth a thousand words!

And think about the impact of videos!

Moreover, we are in the era of the image and of "liquid" life and society.

The ATLAS version of the handbook of interventional congenital cardiology goes exactly in that direction. The authors made the meaningful effort to keep a step further and develop an agile tool.

More than 450 images and 120 videos detail procedural steps and technical issues and are provided for reader's joy and learning.

A useful learning tool is now available worldwide!

London, UK — Gianfranco Butera
San Donato Milanese, Italy — Massimo Chessa
München, Germany — Andreas Eicken
Leeds, UK — John D. Thomson

# Contents

**Part XI Future Directions**

# About the Editors

**Gianfranco Butera, MD, PhD, FSCAI** Consultant Pediatric Cardiology of the Evelina Hospital in London. Lead of Congenital Interventional Cardiology of St Thomas'-Evelina Hospital in London (UK). He has been Chef de Clinique at Hôpital Necker Enfants Malades - Paris and worked as Consultant for Pediatric Cardiology and GUCH cardiology at the Policlinico San Donato IRCCS in Milan (Italy). He studied at the Catholic University of Rome, specialized in Pediatric Cardiology, and completed his training at the Hôpital Necker Enfants Malades in Paris, France, with Prof. Kachaner and Prof. Bonhoeffer and obtained a postgraduate degree in Pediatric Cardiology from the Universitè René Descartes, Paris V. He is a fellow of the GISE, SICP, AEPC, SCAI, and COMET and serves on the executive council of two charity associations. He has served as chairman of the Working Group of interventional cardiology of the AEPC. He has been a visiting professor at various universities including Harvard, Bordeaux, and Padova, and his clinical and research interests are focused on interventional cardiology in children and adults with congenital heart diseases. In addition, Prof. Butera is a reviewer for 15 international journals and Case Report Editor for Cardiology in the Young. He has authored more than 180 papers in peer-reviewed journals and has edited books on interventional and fetal cardiology. With Springer he co-edited the book *Cardiac Catheterization for CHD* and a book on fetal and hybrid cardiac procedures.

**Massimo Chessa, MD, PhD, FSCAI, FESC** main clincial focus is on management and intervention in children and adults with congenital heart diseases; since his PhD he has been working as a Consultant in Congenital Heart Disease IRCCS-Policlinico San Donato Pediatric Cardiology Department and Adult with Congenital Heart Defect Unit, practicing more than 500 catheterizations per year (80% of which interventional). He is leader of the ACHD Program devoted to adults with congenital heart defects, senior researcher, and involved in many projects for International and Humanitarian Cooperation. He has been visiting fellow in France and in the UK and is President of AICCA (Italian GUCH Association www.aicca.it). He has authored more than 150 publications in peer-reviewed journals, 8 chapters in books, has been editor of two volumes for Springer Verlag on *Adult Congenital Heart Disease* and *The Right Ventricle in Adults with Tetralogy of Fallot*, and is editor of the book series devoted to ACHD endorsed by the ESC and AEPC ACHD/GUCH Working Groups.

**Andreas Eicken, MD, PhD, FESC** is a consultant for patients with congenital heart disease (from infant to adult age) at the German Heart Centre, Munich, Germany. He studied medicine in Münster and Lübeck, specialized in pediatrics in Munich, and completed his cardiology training at the German Heart Centre in Munich with Konrad Bühlmeyer and John Hess as teachers. Andreas Eicken attended the ESC nucleus working group on Adults with Congenital Heart Disease for eight years and served as a president of that working group from 2014 to 2016. Currently, he is a council member of the AEPC board. Andreas Eicken published over 130 papers in peer-reviewed journals and is currently the head of the catheterization laboratory for children and GUCH patients at the DHM in Munich.

**John D. Thomson, MD, FRCP, FSCAI** is a consultant cardiologist specializing in the interventional treatment of children and adults with congenital heart disease. He studied in the UK and Australia and has worked in consultant positions in Guy's and St Thomas Hospital in London and the Yorkshire Heart Centre in Leeds where he is the current head of department. He wrote his MD thesis on the neurological benefits of interventional catheterization compared with standard surgical therapy. He has been the honorary secretary of the British congenital cardiac association and is the current President of the AEPC intervention working group having previously been the secretary of that association. He leads a high volume interventional unit and pioneered a number of treatment modalities including ductal stenting.

# Part I

# General Issues

# Angiography: Radiation Exposure and Standard Projections

1

F. Gutierrez-Larraya, C. Abelleira, C. Balbacid, and A. Sanchez-Recalde

## 1.1 Introduction

### 1.1.1 Radiation Exposure in Catheterization Laboratory

The large amount of collective effective dose is related to diagnostic and interventional catheterization. Many factors contribute to a relatively higher level of exposure in pediatric patients. These factors include age, body size, distance between hands and body and X-ray generator, configuration of the X-ray equipment, number of cases per day, and length of study. Both patients and working staff are at a potential risk to radiation. In particular children are at higher risk after exposure to medical radiation. In fact, 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. See Figures 1.1–1.14 and Table 1.1 for better understanding.

### 1.1.2 How to Reduce Radiation Exposure During Invasive Cardiac Procedures

*Pre-procedural Planning*:

1. Use dosimeters and shielding.
2. Know radioprotection principles.
3. Know your equipment.
4. Plan your study.

*Procedure*:

1. Follow the "as low as reasonably achievable" (ALARA) principle.
2. Limit fluoro and cine.
3. Store fluoro use as much as possible.
4. Use the lowest frame rate.
5. Keep the intensifier or flat panel detector as close to the patient as possible.
6. Use the lowest degree of magnification required for accurate interpretation.
7. Minimize radiographic beam time ("cine" acquisition creates 12- to 20-fold higher dose intensities than fluoroscopy mode).
8. Use collimation.
9. There are less-irradiating angulations: 20° right anterior oblique gets the lowest patient DAP and cranial and caudal angulations raise the doses significantly and are maximum in left lateral angulations.
10. Working for more than 6 h increases radiation exposure to 28%.
11. Remember an adequate use of filters, especially for small (<15 kg), and the simpler rule than doubling the source-to-operator distance will decrease the operator dose to approximately one quarter.
12. Operator and personnel exposure are directly related to the dose area product: when operating in a biplane cine-acquisition mode, scattered radiation multiplies by a factor between 5 and 21.

The original version of this chapter was revised. A correction to this chapter can be found at https://doi.org/10.1007/978-3-319-72443-0_46

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_1) contains supplementary material, which is available to authorized users.

F. Gutierrez-Larraya (✉) · C. Abelleira · C. Balbacid
A. Sanchez-Recalde
Pediatric Cardiology Department, Hospital Universitario La Paz, Madrid, Spain
e-mail: Federico.gutierrezlarraya@salud.madrid.org; recalde@secardiologia.es



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_1

## 1.2 Angiographic Projections

1. Plan your case in advance, and use informations coming from other imaging modalities (echocardiography, CT, MRI).
2. 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.
3. 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.
4. Useful "rules of thumb": (a) achieve the correct degree of steepness or shallowness; (b) choose the degree of cranial or caudal tilt.
5. Very important rule: get useful and not only fine pictures!

Exam Protocol

Patient Info:
Name: [redacted] Sex:: F ID: [redacted]

Pos. d. paciente: HFS 03-Sep-10 09:19:58

| | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|
| 2 | CARD | | FIXED | LV 3040 | | | 6s | 30F/s | 03-Sep-10 | 10:32:33 |
| B | 73kV | 66mA | 3.2ms | 0.0CL small | 25cm | 26.1µGym² | 5.0mGy | 90LAO | 1CAU | 182F |
| 2 | CARD | | FIXED | LV 3040 | | | 6s | 30F/s | 03-Sep-10 | 10:32:33 |
| A | 73kV | 55mA | 3.3ms | 0.0CL small | 22cm | 42.7µGym² | 4.8mGy | 13LAO | 4CRA | 182F |
| 3 | CARD | | FIXED | LV 3040 | | | 5s | 30F/s | 03-Sep-10 | 11:15:40 |
| A | 73kV | 60mA | 3.3ms | 0.0CL small | 22cm | 38.8µGym² | 4.4mGy | 13LAO | 4CRA | 154F |
| 3 | CARD | | FIXED | LV 3040 | | | 5s | 30F/s | 03-Sep-10 | 11:15:40 |
| B | 73kV | 78mA | 3.2ms | 0.0CL small | 25cm | 25.2µGym² | 4.8mGy | 90LAO | 1CAU | 154F |

***Datos de expos. acumulada*** 03-Sep-10 12:07:26

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| Fis.: Dr[redacted] | | Expos.: 4 | Fluoro: | 10.5min | Total: | 256.2µGym² | 35.4mGy |
| A | Fluoro: | 5.5min | 85.5µGym² | 9.2mGy | Total: | 167.1µGym² | 18.4mGy |
| B | Fluoro: | 4.9min | 37.8µGym² | 7.1mGy | Total: | 89.1µGym² | 16.9mGy |

**Fig. 1.1** Terminology. Total *air kerma (K, Gy units)* is the procedural cumulative X-ray energy delivered to air at the interventional reference point and is associated with threshold-dependent deterministic skin effects. *Peak skin dose* is the maximum dose received by any local area of the skin; it is not measured but is derived from total air kerma and also reflects deterministic effects. 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. So, it is used to express detriment to whole body if only a part of the body is exposed. The E characterizes stochastic cancer risk. 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. Published effective doses for pediatric catheterization range from 2.2 mSv to 12 mSv. *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. It is a marker of stochastic and no deterministic risk. Coronary angiography and interventions produce DAPs in the range 20–106 Gy $cm^2$ and 44–143 Gy $cm^2$, respectively. In order to estimate the risk of radiation-induced sequelae, the dose area product (Gy $cm^2$) must be converted to the effective dose (mSv). In computed tomography (CT), *dose-length product* (DLP) is the standard dose measurement reported, expressed in mGy·cm. The scanner-derived DLP and the catheterization-derived DAP do not allow comparisons

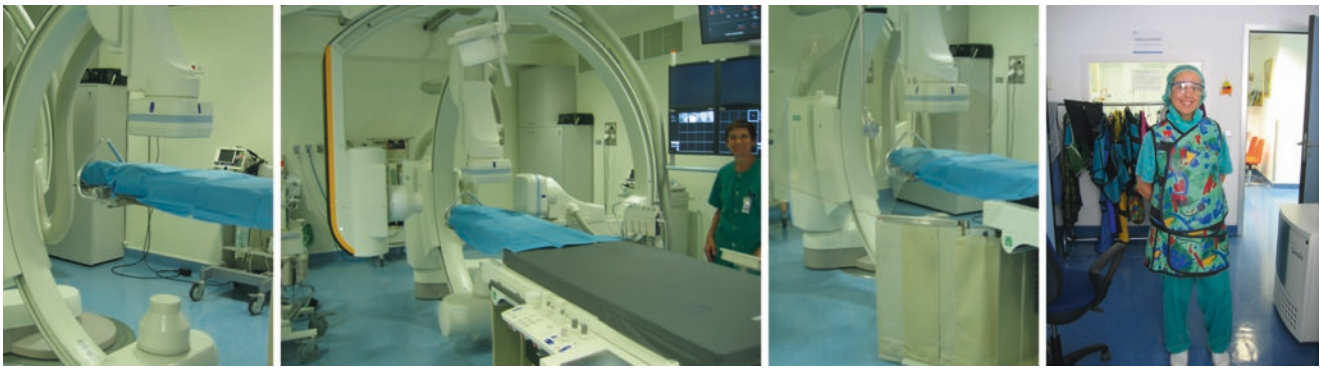


**Fig. 1.2** X-ray protection. Protect both the patient and staff. Use aprons, collars, glasses, curtains, and shields. Distance between X-ray tube and the patient should be maximized keeping the intensifier or flat panel detector as close to the patient as possible. Use the lowest degree of magnification required for accurate interpretation. Remember an adequate use of filters, and the simpler rule than doubling the source-to-operator distance will decrease the operator dose to approximately one quarter. Use the lowest degree of magnification required for accurate interpretation. Minimize radiographic beam time ("cine" acquisition creates 12- to 20-fold higher dose intensities than fluoroscopy mode). Collimation is an efficient radiation-reducing factor. There are less-irradiating angulations: 20° right anterior oblique gets the lowest patient DAP and cranial and caudal angulations raise the doses significantly and are maximum in left lateral angulations

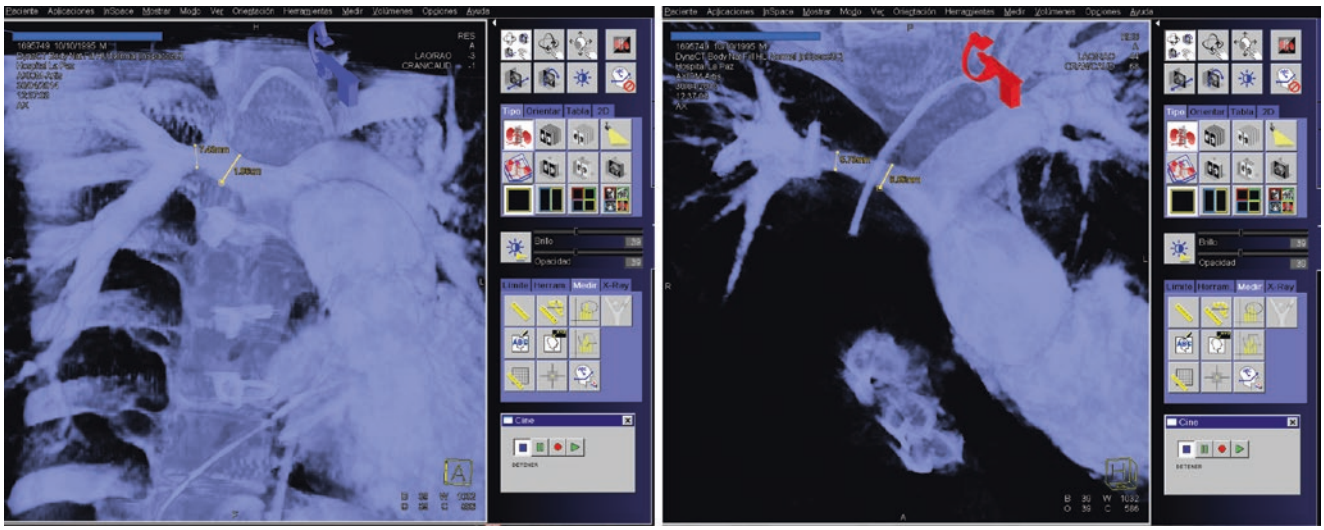


**Fig. 1.3** Camera angles. Angiographic computed tomography helps to choose best angles to take measures, but these angles are not always achievable with actual equipment, and alternative angles are necessary. It is worthless to get an angulation such that the image generator position will preclude to work with catheters, sheaths, wires, etc.

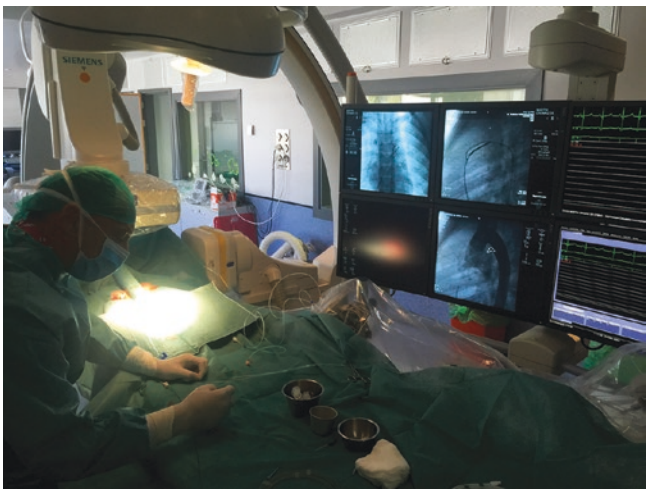


**Fig. 1.4** Although there is no general agreement, biplane (Fig. 1.4) equipment both reduces total contrast dose (not an insignificant problem) and helps to figure out the area of interest but not always with a significant total X-ray dose reduction

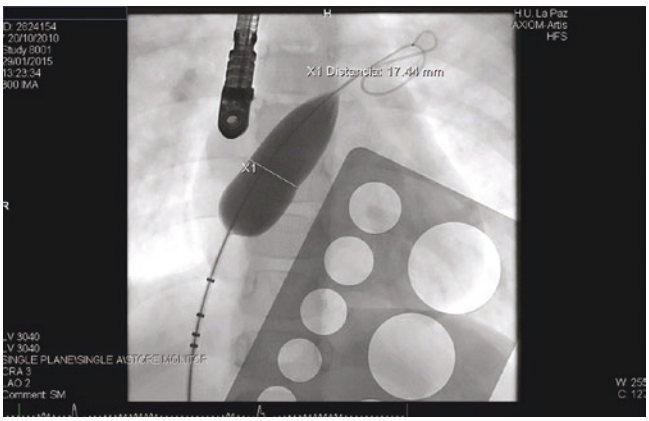
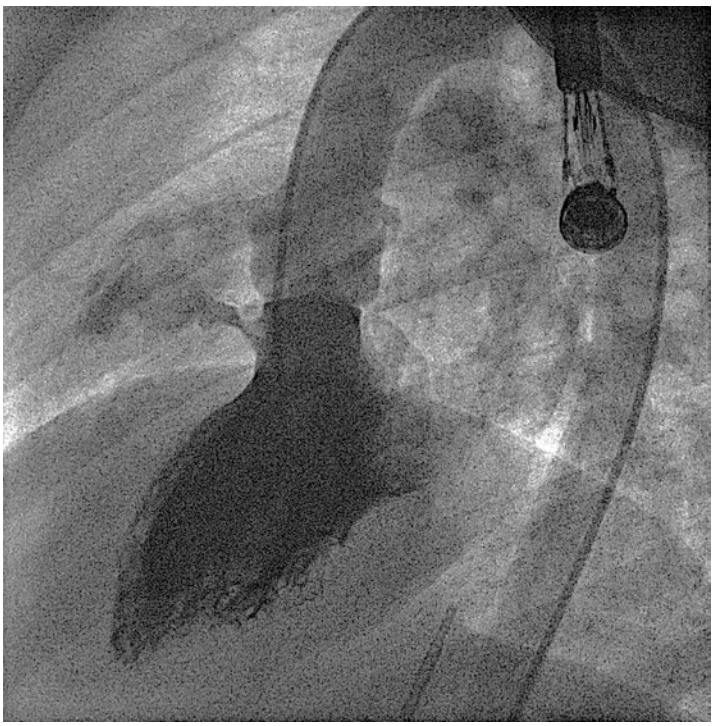



**Fig. 1.5** Secundum atrial septal defect. A good profile of sizing balloon can be done in AP projection. A good alternative is a 30° LAO + 30° cranial

**Fig. 1.6** Perimembranous septal defect: mid-cranial LAO projection at about 50–60° LAO and as much cranial tilt as the conditions allow are the best. Simultaneous orthogonal RAO if biplane system is available would help to profile the defect. RAO view will outline the high anterior and infundibular (outlet) defects



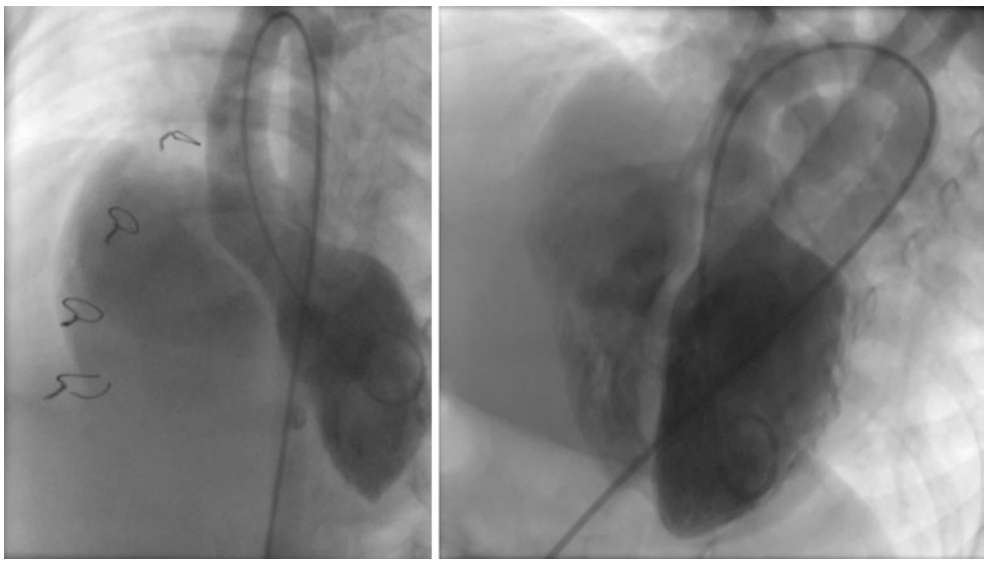


**Fig. 1.7** For patients with Gerbode (left), muscular (right), and posterior septal defects, the best option is to begin with a four-chamber view, but more projections could be necessary to profile the defect and to look to more holes

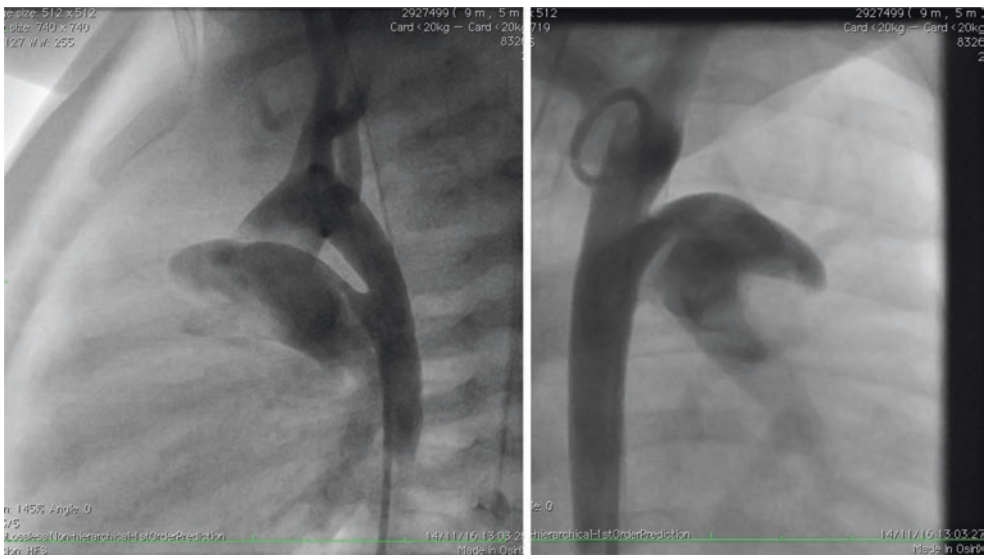


**Fig. 1.8** Patent ductus arteriosus. In most of the cases, closure can be straightly performed just with the lateral plane (left). If not well defined, a simultaneous or added shallow RAO will nicely demonstrate the ductus (right). Who ductal arch and aortic arch are overlapped, some caudal tilt on the plane B will help

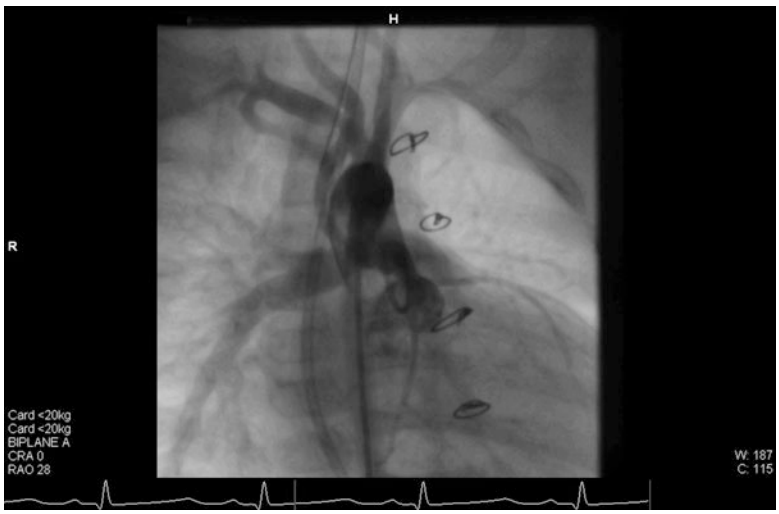



**Fig. 1.9** Modified Blalock-Taussig shunt on the right. A shallow RAO is necessary

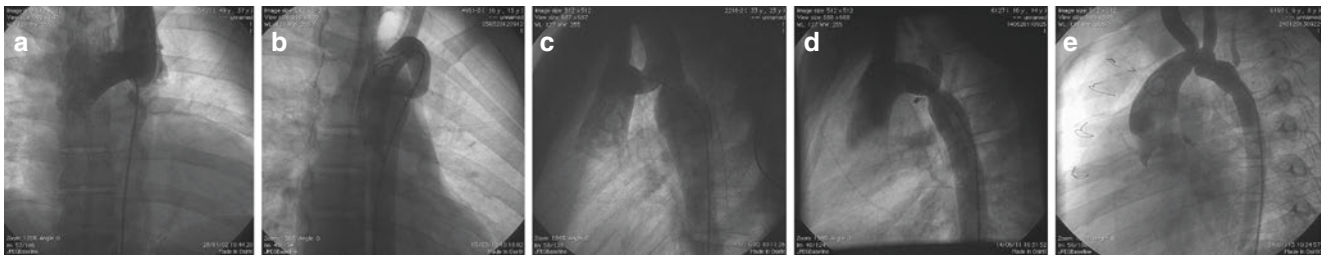



**Fig. 1.10** Aortic coarctation. PA only is not enough (**a**, **b**), so lateral to best profile the minimum diameter (**c**, **d**) and various degrees of LAO to study transverse arch (**e**) are necessary

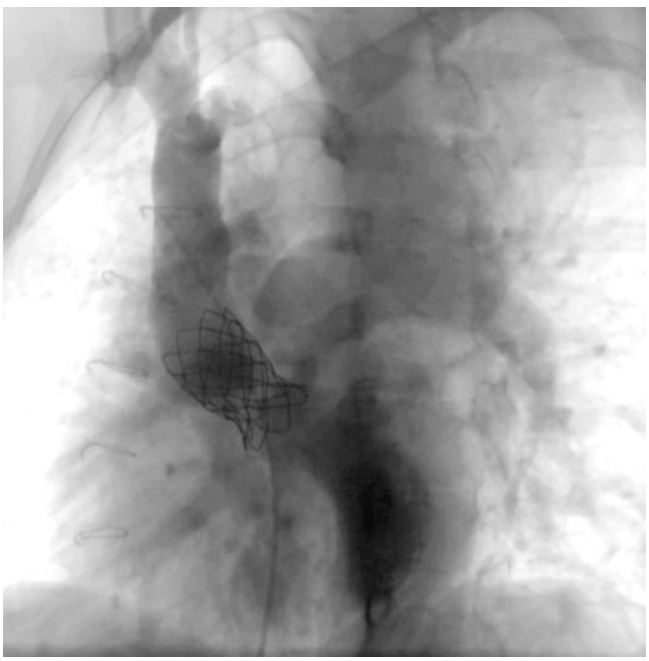


**Fig. 1.11** Mustard baffle. Superior baffle obstruction is best viewed in 30° LAO, with or without 30° cranial angulation depending on the case. Rotational angiography is an unvaluable help to choose the best projection in this setting

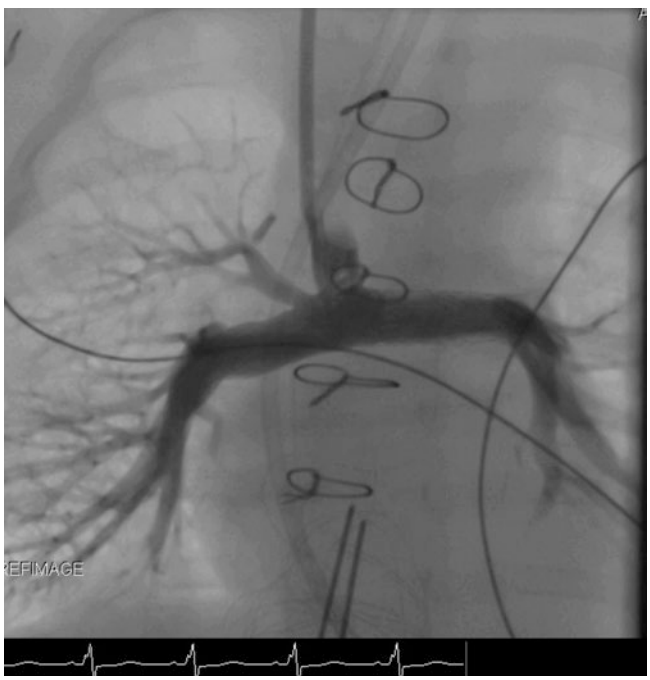


**Fig. 1.12** Bidirectional cavopulmonary connection. PA projection does not usually profile anastomosis well, so some degree of caudal (plus or not shallow LAO) should be added

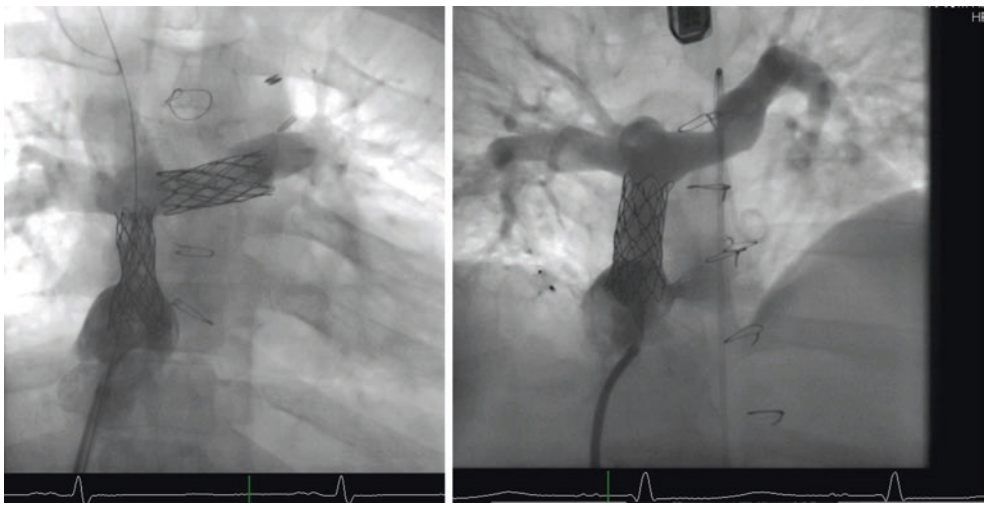


**Fig. 1.13** Fontan operation. PA projection is first obtained. Depending of the aim of the study, many particular variations will be needed

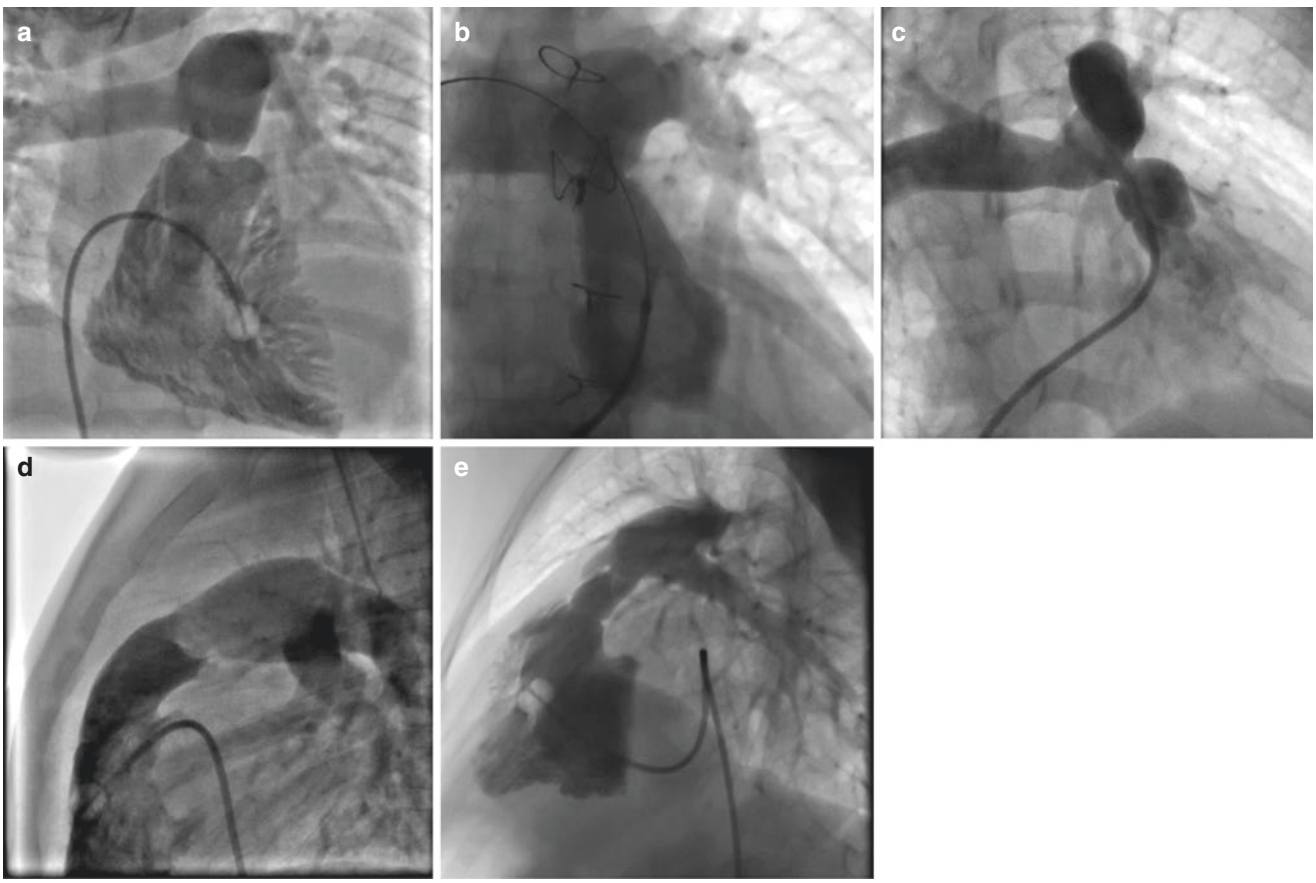



**Fig. 1.14** Pulmonary stenosis, pulmonary valve atresia, Fallot's tetralogy, etc. PA projection will produce significant foreshortening (**a**), so some degree of cranial (**b**) and/or OAD (**c**) must be introduced. Best measurements are attained in lateral view (**d**, **e**)

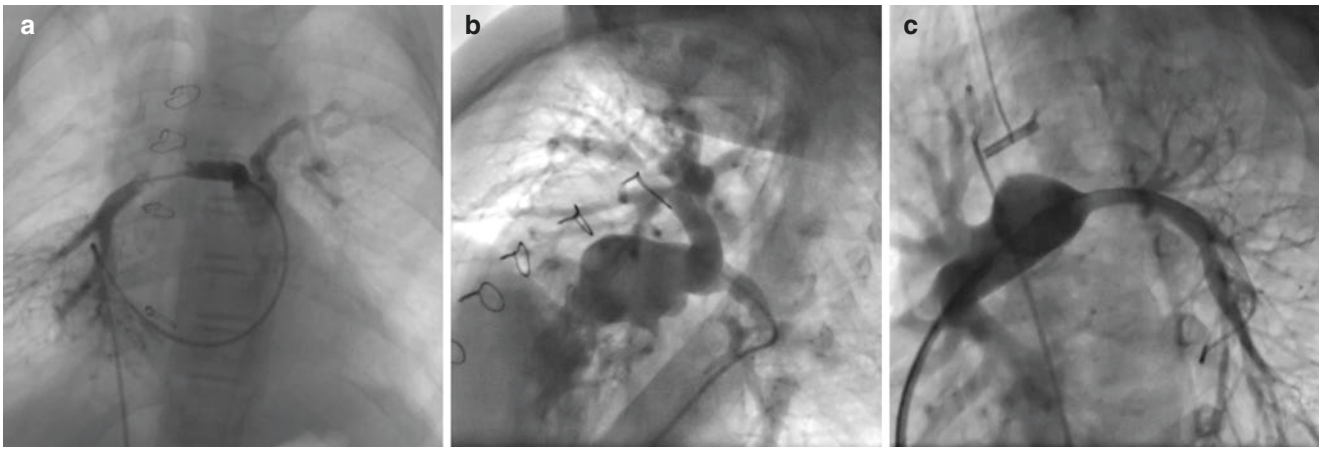



**Fig. 1.15** Branch pulmonary stenosis. Rotational angiography is particularly useful (**a**), four-chamber view (40°LAO + 40°cranial) is very useful at the beginning of the study (**b**), as a rule left pulmonary branch is best studied in left oblique projections (**c**), and conversely the right pulmonary artery is best studied in right oblique projections

**Table 1.1** Recommended projections in practice in various clinical settings

| Projection | Angles, plane A | Angles, plane B |
|---|---|---|
| Conventional RAO | 40° RAO | |
| Frontal | 0° | |
| 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 lateral | 0° | Left lateral |
| Long axis oblique | 30° RAO | 60° LAO +20–30° cranial |
| Hepatoclavicular view | 45° LAO + 30° cranial | 120° LAO + 15° cranial |
| **Specific lesions** | **Angles, plane A** | **Angles, plane B** |
| Pulmonary stenosis | 0° + 30° cranial | Left lateral |
| RVOT-MPA (sitting up) | 10° LAO + 40° cranial | Left lateral |
| Long axial for LPA biplane | 30° RAO | 60° LAO + 30° cranial |
| LPA long axis | | 60° + 20° cranial |
| ASD | 30° LAO + 30° cranial | |
| PA bifurcation and branches | 30° caudal + 10° RAO | 20° caudal |
| Left ventricular outflow tract obstruction | RAO | Long axis oblique |
| Aortic coarctation | 0°/shallow RAO/shallow LAO | Left lateral/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 | |

**Video 1** Rotational angiography requires particular attention at everything as collision is very easy (MP4 59340 kb)

**Video 2** Modern equipment allows to work with 3D images superposed to actual angiography (MP4 16732 kb)

# Catheters and Wires

2

Adam Koleśnik and Grażyna Brzezińska-Rajszys

## 2.1 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.

### 2.1.1 Anatomy of the Catheter

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

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

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

Catheter manipulation requires application of torque to its part outside the patient. This torque has to be transmitted to the tip. Besides, as mentioned before, the catheter has to be kink-resistant and provide some support while passing through the vessels and/or chambers. This is why shafts of the catheters are usually composed of a plastic material (nylon, polyethylene, polyurethane, PTFE) braided with thin metal meshwork. Depending on the manufacturer, the catheter size and the distal ends of catheters can be made of braided or unbraided material. The tip itself usually lacks reinforcement to assure its softness and minimize the risk of vascular wall injury. The distal tip of the catheter may have an additional radiopaque marker to improve its visualization. Some of the catheters have a single end hole for injection of the contrast medium, for pressure measurements, and for advancing guidewire, while other catheters such as angiographic catheters have multiple side holes for even contrast distribution. It is recommended to avoid any pressure injections of contrast through a catheter with end hole only. Balloon-tip catheters have a $CO_2$ inflatable balloon at their tips. This balloon is supposed to allow free floating with the bloodstream and prevent tangling between the chordae tendineae in the cardiac chambers. Other catheters have hydrophilic coating that makes them slippery and facilitate their gliding through tortuous vessels. Sizing catheters have additional radiopaque markers embedded in their shafts at known distances, for precise calibration and measurements.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_2) contains supplementary material, which is available to authorized users.

A. Koleśnik · G. Brzezińska-Rajszys (✉)
Cardiovascular Interventions Laboratory, The Children's Memorial Health Institute, Warsaw, Poland
e-mail: a.kolesnik@ipczd.pl; gbrzezinska@hoga.pl


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_2

## 2.1.2 Types of the Catheters

### 2.1.2.1 Angiographic Catheters

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

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

Berman angiographic catheter is a balloon-tipped catheter without the end hole (Fig. 2.3a). 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. However, in the presence of interatrial or interventricular communications, it can be used to catheterize left heart structures as

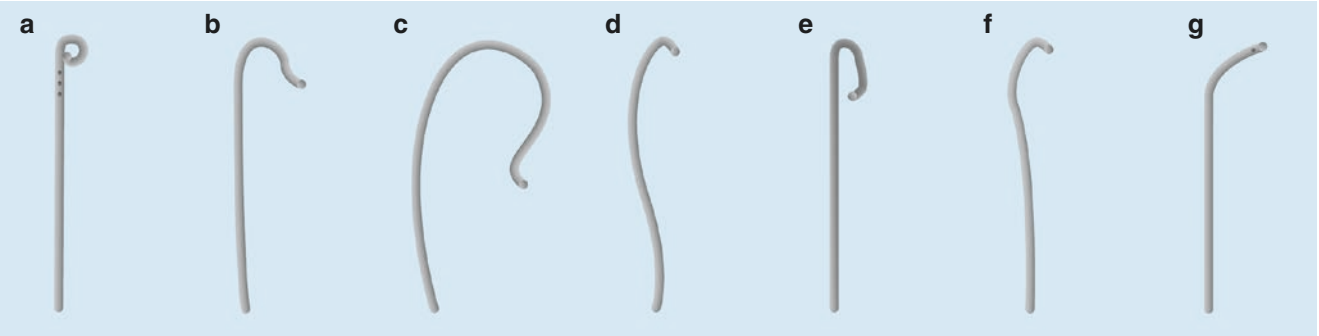



**Fig. 2.1** Shapes of selected torque-controlled catheters (see text). (**a**) Pigtail catheter, (**b**) Amplatz left coronary catheter, (**c**) Amplatz right coronary catheter, (**d**) internal mammary catheter, (**e**) Judkins right coronary catheter, (**f**) Judkins left coronary catheter, (**g**) multipurpose catheter

**Video 1** Three-dimensional rotational angiography in a patient with tight coarctation of the aorta. The contrast is injected to the left ventricle with the pigtail catheter (see Figs. 2.1a and 2.2). Injection to the ventricle allows for visualization of the aorta and its branches without rapid ventricular pacing, because the contrast mixes in the ventricle and the aorta remains contrasted throughout the injection without any contrast medium washout effect (AVI 1999 kb)

**Video 2** Closure of the perimembranous VSD using PFM Nit-Occlud Lê coil. The pigtail catheter is advanced with the guidewire inside. Pigtail catheters are thin-walled especially at their distal ends that 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 (AVI 4450 kb)

**Video 3** Closure of the perimembranous VSD using PFM Nit-Occlud Lê coil. Injection of the contrast medium to the left ventricle in order to assess position of the coil. 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 (AVI 1506 kb)

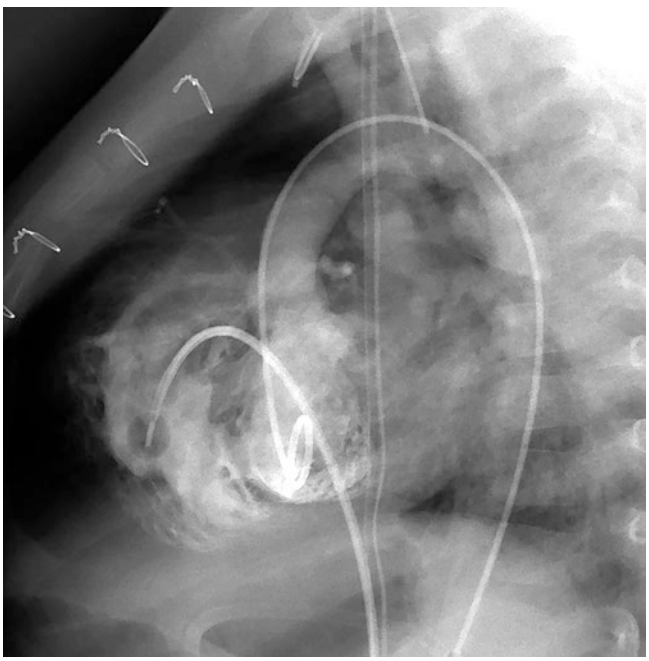


**Fig. 2.2** Left ventriculography (retrograde approach) with pigtail catheter—lateral projection. Multiple ventricular septal defects in a 2-year-old patient after pulmonary artery banding and surgical repair of aortic arch hypoplasia and coarctation. Please notice the Berman angiographic catheter with tip balloon inflated with $CO_2$

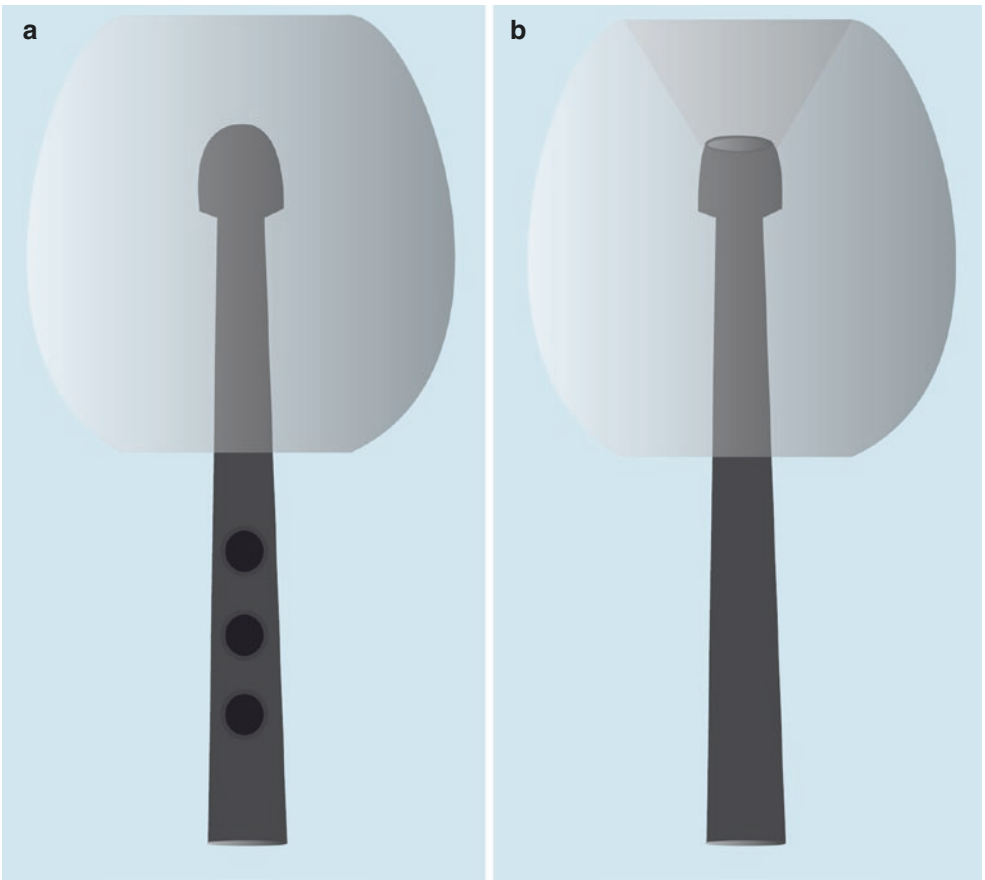



**Fig. 2.3** Tips of floating catheters. (**a**) Berman angiographic balloon catheter. (**b**) Pulmonary wedge balloon catheter

**Video 4** Three-dimensional rotational angiography. Injection of contrast medium to the right ventricle through the Berman catheter (notice the gas-filled balloon at the tip of the catheter) (AVI 2091 kb)

**Video 5** The patient with tricuspid atresia and transposition of the great arteries. The Berman catheter has been advanced to the aorta using the antegrade approach. Balloon at the tip has been inflated with $CO_2$.The catheter is moving back and forth with the bloodstream (AVI 1598 kb)

**Video 6** The patient with tricuspid atresia and transposition of the great arteries (see Video 4). The Berman catheter advanced antegradely through the atrial septal defect to the left atrium, the left ventricle, and the pulmonary trunk. Pulmonary angiography shows banded pulmonary trunk (AVI 2459 kb)

well. Antegrade approach to the aorta is feasible also in patients with transposition of the great arteries, double outlet right ventricle, or functionally univentricular hearts or in the presence of large ventricular septal defects (Fig. 2.4).

Moreover, the balloon catheter can be used to occlude the distal parts of the vessels and perform occlusion arteriography. Balloon occlusion descending aortography helps to force blood flow through aortopulmonary collateral arteries in the tetralogy of Fallot and other congenital cardiac malformations with pulmonary stenosis or atresia (Fig. 2.5).

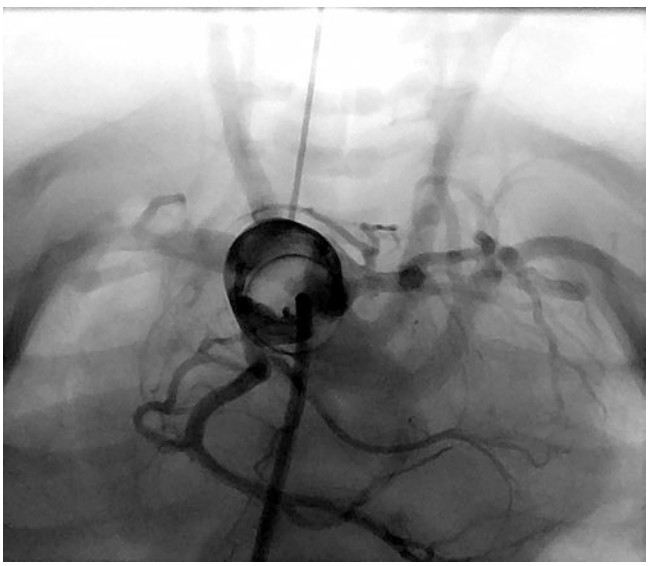


**Fig. 2.4** Balloon occlusion ascending aortography (antegrade approach) with Berman angiographic balloon catheter. Antegrade balloon occlusion aortography with 35° caudal angulation is used to visualize coronary arteries in cases of transposition of the great arteries. The angiogram shows origin of the left circumflex artery from the right coronary artery in a 2-day-old patient with transposition of the great arteries

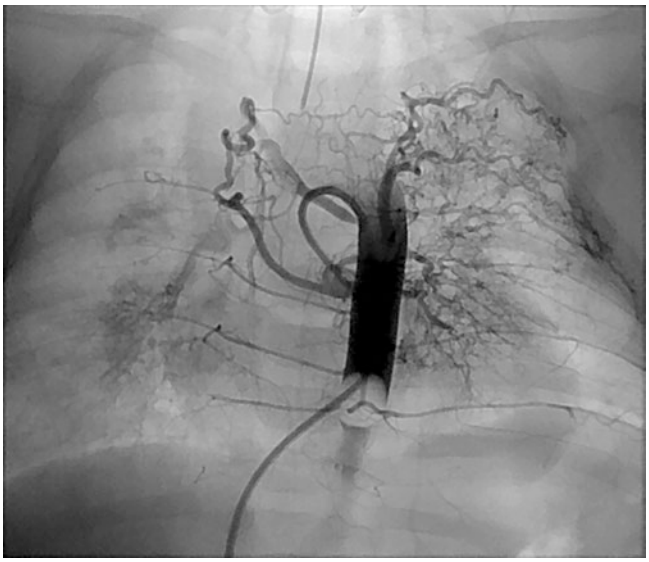


**Fig. 2.5** Balloon occlusion descending aortography (antegrade approach) with Berman angiographic catheter—anteroposterior projection. Aortopulmonary collaterals in a 6-month-old patient with double-inlet left ventricle after pulmonary artery banding

**Video 7** Balloon occlusion descending aortography in patient with hypoplastic left heart syndrome shows the aortic arch and severely hypoplastic ascending aorta filling retrogradely (AVI 1846 kb)

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.

### 2.1.2.2 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. 2.3b). 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 vein, one can measure the pressure in the pulmonary arterial bed, based on exactly the same principle as the antegrade pulmonary wedge measurement. However, in the hands of interventional cardiologist, the pulmonary balloon wedge catheter becomes more widely used for advancing the guidewire for interventional procedures, selective pulmonary arteriography, simulation of vessel occlusion, and many others. With the balloon inflated at its tip, it should cross the tricuspid valve safely and minimize the risk of its injury during the following interventions, such as balloon valvuloplasty, pulmonary artery angioplasty, or stent placement. In case of extreme pulmonary artery hypoplasia, injection of the contrast medium through the catheter wedged in the peripheral pulmonary vein with consecutive flush with saline results in retrograde visualization of the pulmonary arterial vessels. Antegrade placement of the Swan-Ganz catheter in the Blalock-Taussig shunt is used, after inflation of the balloon, to simulate the shunt occlusion and monitor pressure changes in the pulmonary arteries. Undoubtedly, the pulmonary wedge catheter should always be available for use in the catheterization laboratory shelf.

### 2.1.2.3 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. 2.1. 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 (Fig. 2.1b–e). Among them, Judkins right coronary catheter (JR) is one of the most widely used in the cardiac catheterization laboratory. The distal part of the catheter is gently rotated to find support in the ascending aorta, and the tip bends at almost a right angle to reach the orifice of the right coronary artery.

In pediatric catheterization laboratory, this shape has proved to be useful in the selective catheterization of Blalock-Taussig shunts and collateral vessels, entering the right ventricle outflow tract, and many other procedures (Fig. 2.6).

**Video 8** Right coronary artery angiography in transplanted heart. Judkins right coronary catheter, 30° right anterior view. Please notice an abnormal collateral vessel between the right conal branch and the right pulmonary artery (AVI 738 kb)

**Video 9** Hand injection of contrast medium to a major aortopulmonary collateral artery in a patient with pulmonary atresia and intact interventricular septum. The artery was engaged with Judkins right coronary catheter (AVI 1758 kb)

**Video 10** Judkins right coronary catheter advanced to right ventricle outflow tract through the femoral venous access in a patient with pulmonary atresia and intact interventricular septum undergoing radiofrequency perforation of atretic pulmonary valve. Multipurpose catheter has been simultaneously inserted to the pulmonary trunk via ductus arteriosus using femoral arterial approach (AVI 778 kb)

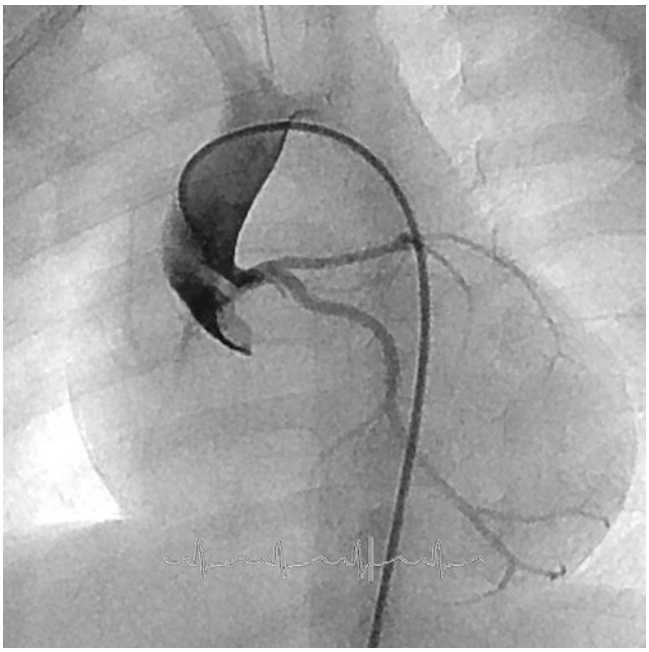


**Fig. 2.6** Ascending aortography (retrograde approach) with Judkins right coronary catheter—left anterior oblique projection. Catheter positioned in front of the opening of critically stenosed aortic valve in a 3-day-old patient allowing for easy insertion of the guidewire to the left ventricle and balloon valvuloplasty

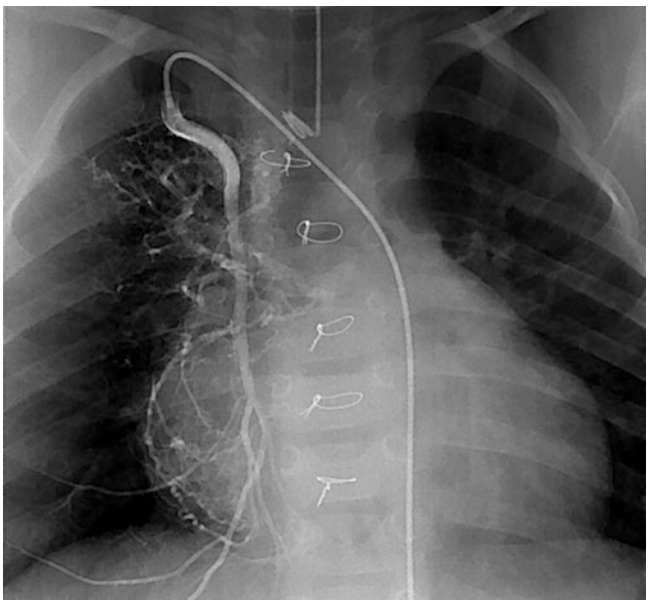


**Fig. 2.7** Selective angiography of the right internal thoracic artery (posterior-anterior projection) with right internal mammary catheter. Small systemic-to-pulmonary collaterals in a 2-year-old patient with hypoplastic left heart syndrome after Norwood-Sano operation

**Video 11** Right internal thoracic artery angiography done using multipurpose catheter advanced antegradely over the hydrophilic guidewire in patient after Norwood-Sano procedure for hypoplastic left heart syndrome. Dilated internal thoracic artery provides collateral inflow to the right pulmonary artery (AVI 565 kb)

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

Internal mammary catheters with their C-shaped tips can be used to enter vessels having origins at acute angles. Their applications include selective catheterization of Blalock-Taussig shunts and collateral vessels (Fig. 2.7).

Multipurpose (MP) catheters have their distal ends curved at an obtuse angle (Fig. 2.1g). 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 atrium, and place the guidewire in the pulmonary vein before atrial septal defect device closure.

### 2.1.2.4 Special Catheter Types

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

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

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

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

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

#### 2.1.3.1 Floating Catheters

Floating catheters (Berman, Swan-Ganz) are very soft, and their response to torque is limited. They should float freely in the direction of the blood flow. Nevertheless, especially in difficult anatomy or valvar insufficiency, it may be difficult to manipulate them. Additionally, floating catheters are packaged in a curved manner, and it may be impossible to straighten them completely. The problem can start just after crossing the introducer sheath. Without the balloon inflated, they can enter side branches. Inflation of the balloon makes the catheter float to the right atrium. Once the right atrium has been reached, it is possible to enter the superior vena cava. Sometimes it is possible to direct the tip posteriorly with a gentle torque and push the catheter up to the superior vena cava. In case of failure, insertion of a straight guidewire may solve the problem. Without a guidewire inside and without the balloon inflated, the catheter is likely to enter the left atrium via an atrial septal defect or patent foramen ovale, if present. To enter the left pulmonary veins, the catheter should be directed posteriorly. Otherwise, it will enter the left atrial appendage. Inflation of the balloon of catheter placed in the left atrium, close to the interatrial septum followed by a clockwise torque, may help to cross the mitral valve and enter the left ventricle. If this maneuver does not work, one can use the stiff end of the guidewire bent in a U-shape to angle the distal end of the catheter. Guidewire will also help to transmit the torque. In the left ventricle, the catheter will float toward the apex. Again, a curved stiff end of the guidewire may help to bend the catheter (with the balloon inflated) toward the interventricular septum and push it up into the aorta. To avoid the tension onto the ventricular wall, the guidewire should be partly withdrawn from the catheter, to permit free floatation of the balloon.

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

There are some issues to be kept in mind:

1. The balloon is able to accommodate more $CO_2$ than just one syringe; the more the balloon is inflated, the easier the floating is, but caution is needed as the balloon can rupture with too much $CO_2$.
2. The catheter can be straightened or bent using the guidewire; the guidewire helps to transmit the torque.
3. Creating a loop can help to manipulate with the catheter; it is better to straighten the catheter as soon as its final destination has been reached.
4. Especially in blood vessels, the balloon can obstruct them and alter the blood pressure; it should be deflated during measurements of pressure.
5. In selected applications, obstruction of the vessel with the balloon tip can help to make a selective contrast injection or perform an occlusion test; always remember where the hole(s) is(are).
6. When performing balloon occlusion angiography, the vessel occlusion time should be kept to a minimum; after inflation of the balloon, the catheter will float downstream—pull it back to the desired position, and deflate the balloon as soon as the angiography has been performed.

#### 2.1.3.2 Torque-Controlled Catheters

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

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

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

## 2.2 Guidewires

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

**Video 12** The perimembranous ventricular septal defect is crossed with the hydrophilic guidewire. The Judkins right coronary catheter remains in the ascending aorta, while the guidewire is advanced alone through the aortic valve and the ventricular septal defect (AVI 4456 kb)

**Video 13** Patient after Norwood-Sano procedure for hypoplastic left heart syndrome. Three-dimensional road map based on 3D rotational angiography (the contrast medium has been injected to the right ventricle). Multipurpose catheter and hydrophilic guidewire are used to engage the Sano conduit (AVI 7318 kb)

Guidewires with hydrophilic coating are slippery when wet. They glide through tortuous vessels easily. It can be difficult to manipulate them, so a special plastic torque device is very useful. Hydrophilic wires have to be wet all the time, because they become sticky when dried.

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

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

Size of a guidewire is given in fraction of inch. The length is measured in centimeters. Especially long (260–300 cm) exchange wires are used to exchange long catheters. 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 softness of hydrophilic catheter tip, it can easily perforate the heart or vessel wall, especially while exiting the catheter. That is why rather standard guidewires and not hydrophilic nor thin coronary wires are recommended to cross a critically stenotic aortic valve. The latter are more likely to perforate valvar leaflets.
4. Length of a soft tip: long, soft tips can enter the planned location but appear too extensive to support interventional equipment. On the other hand, guidewires with short tips are more traumatic.
5. Stiffness: it is safer and easier to advance catheters or sheaths using a stiff wire because of the better support. Softer wires can kink and lose their position or just make advancement of equipment impossible.
6. Shape of a tip: J-tipped guidewires are considered to advance more easily without entering side branches or tributaries. Nevertheless, the vessel diameter has to be large enough to accommodate the tip. Curved tips of some guidewires help to manipulate with the torque to reach a chosen location. Steel-core wire tips can be formed by an operator to get the best shape she/he needs.
7. Torque transmission: wires with a core continuous from proximal to distal end transmit the torque better than those lacking the core at their tips. The longer the floppy tip is, the more is the torque transmission limited.
8. Trackability and steerability: the shaft of the wire should be able to follow its tip through the tortuosities or narrowings in accord with operator's maneuvers.

**Video 14** A 13-year-old patient with calcified and stenotic aortic homograft in the pulmonary position (primarily pulmonary stenosis with dysplastic pulmonary valve replaced with Contegra followed by pulmonary homograft and, finally, aortic homograft). Balloon interrogation test to check the proximity of coronary arteries. The balloon catheter (BiB, NuMED) has been advanced to the graft over extra-stiff Lunderquist guidewire through 12F Mullins long sheath and inflated. At the same time, 3D rotational aortography has been performed to visualize coronary arteries (AVI 2111 kb)

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.

## 2.3 Introducer Sheaths

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

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

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 the ability to pass through especially tortuous vessel without kinking or collapsing are the features of long Flexor introducer sheath. Details of long sheath 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.

**Video 15** Presenting of the calcified aortic homograft prior to implantation of the Melody percutaneous pulmonary valve. The covered CP stent crimped on 18 mm BiB balloon is advanced over the Lunderquist guidewire through 12F Mullins sheath. Notice two kinks of the sheath the stent has to pass (AVI 4461 kb)

**Video 16** Percutaneous closure of the perimembranous ventricular septal defect. Kink-resistant Flexor sheath is advanced using "kissing" technique over the guidewire arteriovenous loop (AVI 4456 kb)

# Balloons

3

Caroline Ovaert and Duarte Martins

## 3.1 Introduction

Balloons are generally used for direct dilation of vessels or stents. Specific balloons may be used for other purposes, such as atrial septostomy, sizing of defects, temporary occlusion of shunts or collateral vessels in order to stabilize occlusion devices or to perform hemodynamic assessments.

Balloons are currently available in various lengths and diameters and differ with regard to pressure characteristics, compliance, profile, balloon morphology and inner lumen diameter (Table 3.1). A broad spectrum of balloon catheters must be available in each catheterization laboratory when performing procedures in patients with congenital heart defects, in order to be able to adapt to different weights, anatomies and procedures.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_3) contains supplementary material, which is available to authorized users.

C. Ovaert (✉)
Pediatric Cardiology, Timone enfants, Assistance Publique—Hôpitaux de Marseille, Marseille, France

D. Martins
Pediatric Cardiology, Timone enfants, Assistance Publique—Hôpitaux de Marseille, Marseille, France

Pediatric Cardiology, Hospital de Santa Cruz, Centro Hospitalar de Lisboa Ocidental, Lisbon, Portugal


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_3

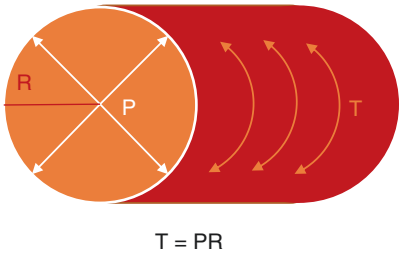
**Table 3.1** Low-pressure, medium-pressure, high-pressure and ultra-high-pressure balloons

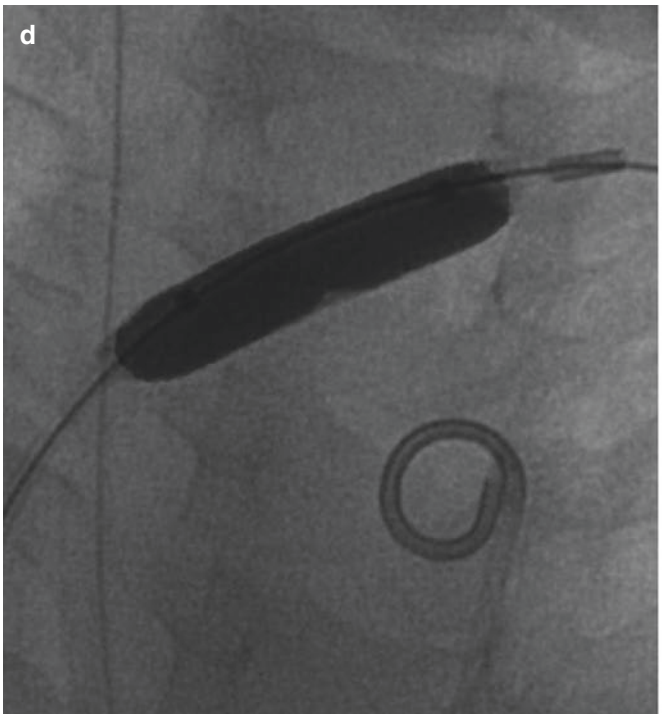
| Pressure | Name | Company | Diameter (mm) | Introducer (F) | NP (atm) | RBP (atm) | Wire |
|---|---|---|---|---|---|---|---|
| *Low-pressure* | | | | | | | |
| – Compliant, low burst pressure (<10 atm)<br>– Small profiles and flexible shafts<br>– Useful for pulmonary or aortic valves, arterial or venous stenosis in the young child<br>– Unsuitable for dilation of 'pressure-resistant' stenosis and for stent placement<br>– Indeflator recommended to avoid balloon rupture. | Tyshak Mini®<br>Tyshak II® | Numed<br>Numed | 4–10<br>4–12 | 3–4<br>4–6 | 3–4.5<br>3–4.5 | 3.5–6<br>3.5–6 | 0.014"<br>0.021–0.035" |
| *Medium- and high-pressure* | | | | | | | |
| – Burst pressure: 8–20 atm<br>– Stiffer shaft and higher profile<br>Useful for pressure-resistant lesions (not responding to the 'low-pressure' balloons) especially in pulmonary arteries or calcified conduits. Indicated for stent insertion.<br>– Indeflator recommended for controlling the inflation pressure. | Opta™ Pro | Cordis | 3–12 | 5–8 | | 6–10 | 0.035" |
| | Ultra-thin™ SDS | Boston | 4–10 | 5–7 | | 12 | 0.035" |
| | Z-med II™ | Sci | 4–10 | 5–7 | 6 | 13–15 | 0.025–0.0035" |
| | Z-med II-X™ | Numed | 8–30 | 7–16 | 2–6 | 3–15 | 0.035" |
| | Powerflex™ | Numed | 3–12 | 5–8 | | 8–15 | 0.035" |
| | Mustang | Cordis | 3–12 | 5–6 | | 24 | 0.035" |
| | Advance® 35LP | Boston | 3–12 | 5–7 | 5–10 | 8–15 | 0.035" |
| | Mullins-X™ | Sci | 12–25 | 9–16 | | 9–14 | 0.035" |
| | Cristal Balloon | Cook | 8–40 | 6–15 | 3–8 bar | | 0.035–0.038" |
| | Sterling Balloon™ | Numed<br>Balt<br>Boston<br>Sci | 2–10 | 4–6 | | 10–14 | 0.018" |
| *Ultra-high pressure* | | | | | | | |
| – Ultra-high molecular weight polyethylene (UHMWPE)<br>– Non-compliant , burst pressures, >20 atm<br>– Useful for in-stent stenosis, stenosis adjacent to stents, pressure-resistant stenosis, to rupture stent cells (overlapping stents).<br>– Special 'high-pressure' indeflator | Conquest®<br>Atlas® Gold | Bard<br>Bard | 5–12<br>12–26 | 6–8<br>7–12 | 8<br>4–6 | 20–30<br>12–18 | 0.035"<br>0.035" |

Four categories exist according to the maximal pressure that the balloons can sustain: low, medium, high and ultra-high pressure. The limit between medium- and high-pressure balloons is however not very well defined, and they will be described in the same group. The table gives a non-exhaustive list of commercially available and frequently used balloons in congenital heart catheterization (*atm* atmosphere, *F* French, *NP* nominal pressure, *RBP* rated burst pressure)

**a**

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

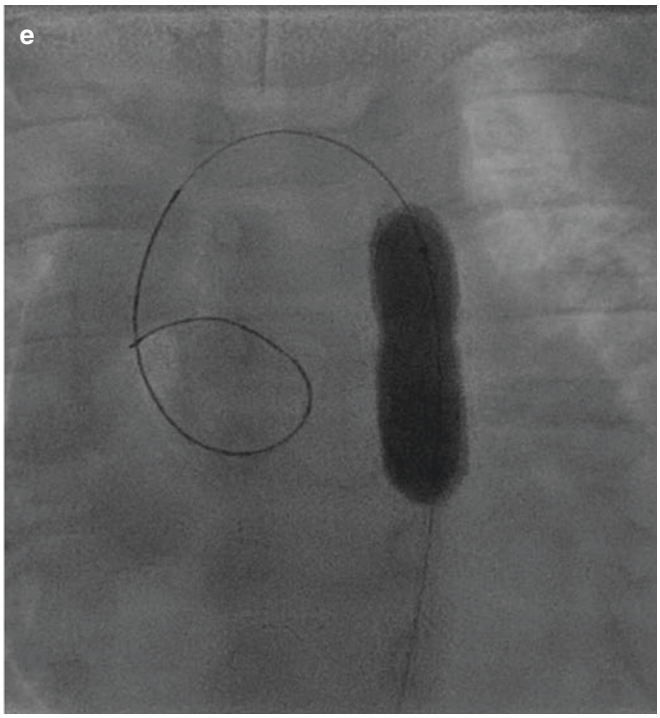




**Fig. 3.1** Balloon characteristics: diameter, pressure and compliance

- Dilation balloons are static balloons. They expand to a fixed *diameter* when inflated to a certain maximum pressure also called *nominal* pressure. The *rated burst pressure* indicates the level of pressure where the balloon is likely to rupture. Nominal and rated burst pressures are always provided by the manufacturer as shown in Fig. 3.1a (table summarizing the characteristic of some of the Atlas Gold® balloons (Bard)). The same information is sometimes mentioned on the balloon catheter itself (Fig. 3.1b).
- The maximum *pressure* tolerated varies with the material used for the balloon and with the diameter. In large balloons, lower pressure is needed when compared to small balloons, to generate identical wall tension and as a correlate, clinical efficacy (Fig. 3.1c).
- The material used determines the *compliance* of the balloons. Non-compliant balloons have a fixed diameter all along the balloon at nominal pressure and even if pressure is increased above nominal pressure. Figure 3.1d is a superposition of two fluoroscopies of the same non-compliant balloon inflated at low pressure and at high pressure. At low pressure there remains a waist. At high pressure, the waist disappears, but the diameter of the rest of the balloon does not increase. In compliant balloons, the diameter may increase with pressure, especially in the areas of the balloon facing less resistance from the surrounding structures. This is shown in Fig. 3.1e, which is again a superposition of two fluoroscopies and shows nicely the increase of the diameter of the proximal and distal parts of the balloon at higher pressure

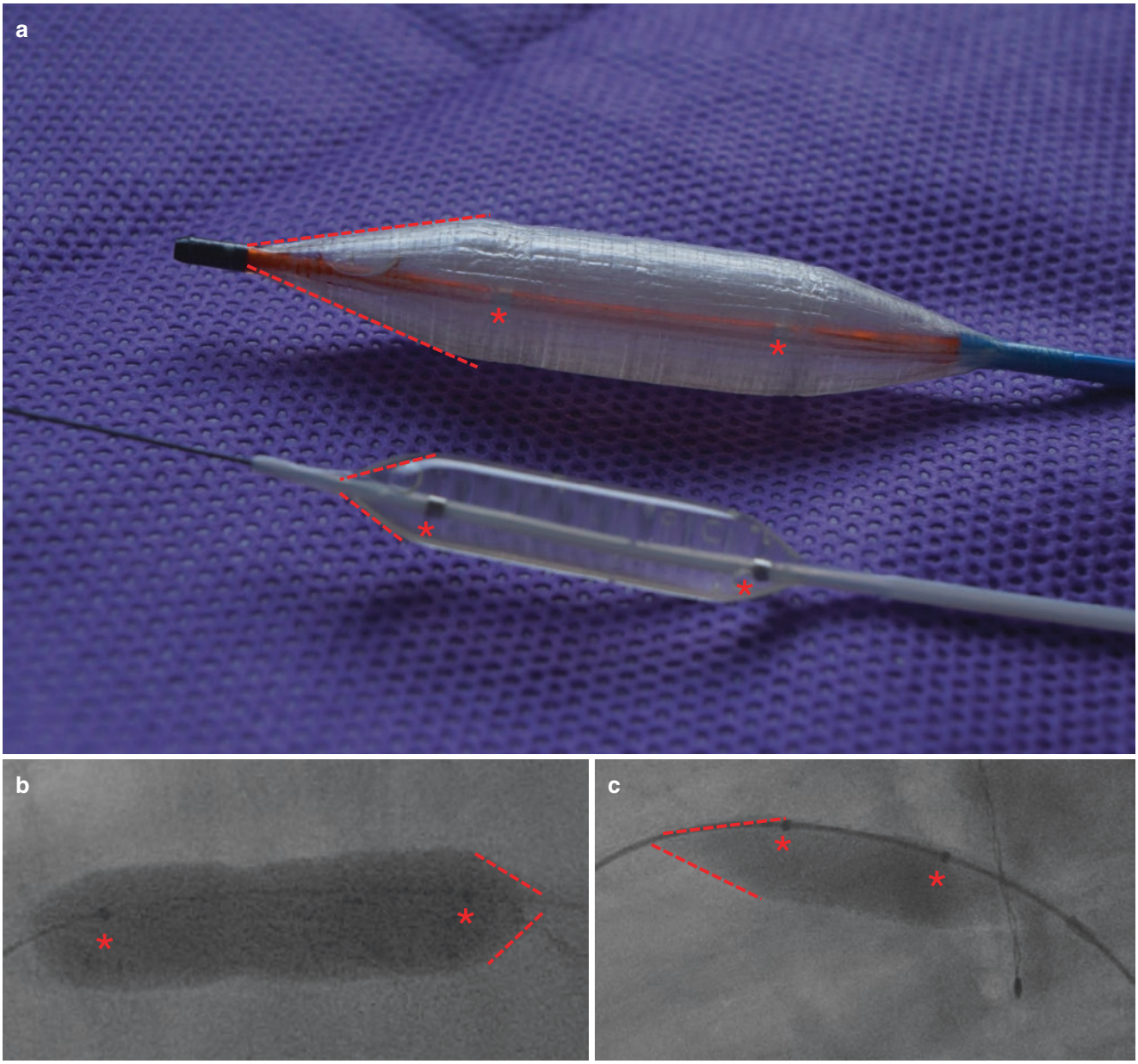



**Fig. 3.2** Balloon morphology
The length mentioned by the manufacturer relates to the working part of the balloon and does not include the shoulders of the balloon. The *shoulders* are the end parts of the balloon, where the diameter reduces to the shaft's diameter. Most balloons have *radio-opaque markers* to indicate the working part of the balloon and where the shoulders start. Figure 3.2a shows a picture and Fig. 3.2b a fluoroscopy of a Tyshak II® balloon (Numed Inc.) known to have very short and round shoulders. Figure 3.2c shows a picture and Fig. 3.2d a fluoroscopy of Atlas® balloons (Bard) with long shoulders. The red lines delineate the shoulders. The red asterisks are placed at the level of the radio-opaque markers. Short shoulders are usually preferred in paediatric heart catheterization as the shoulders tend to increase the length of the rigid part of the total balloon catheter. This rigidity makes manipulation in small heart and vessels more difficult and dangerous [1–3]

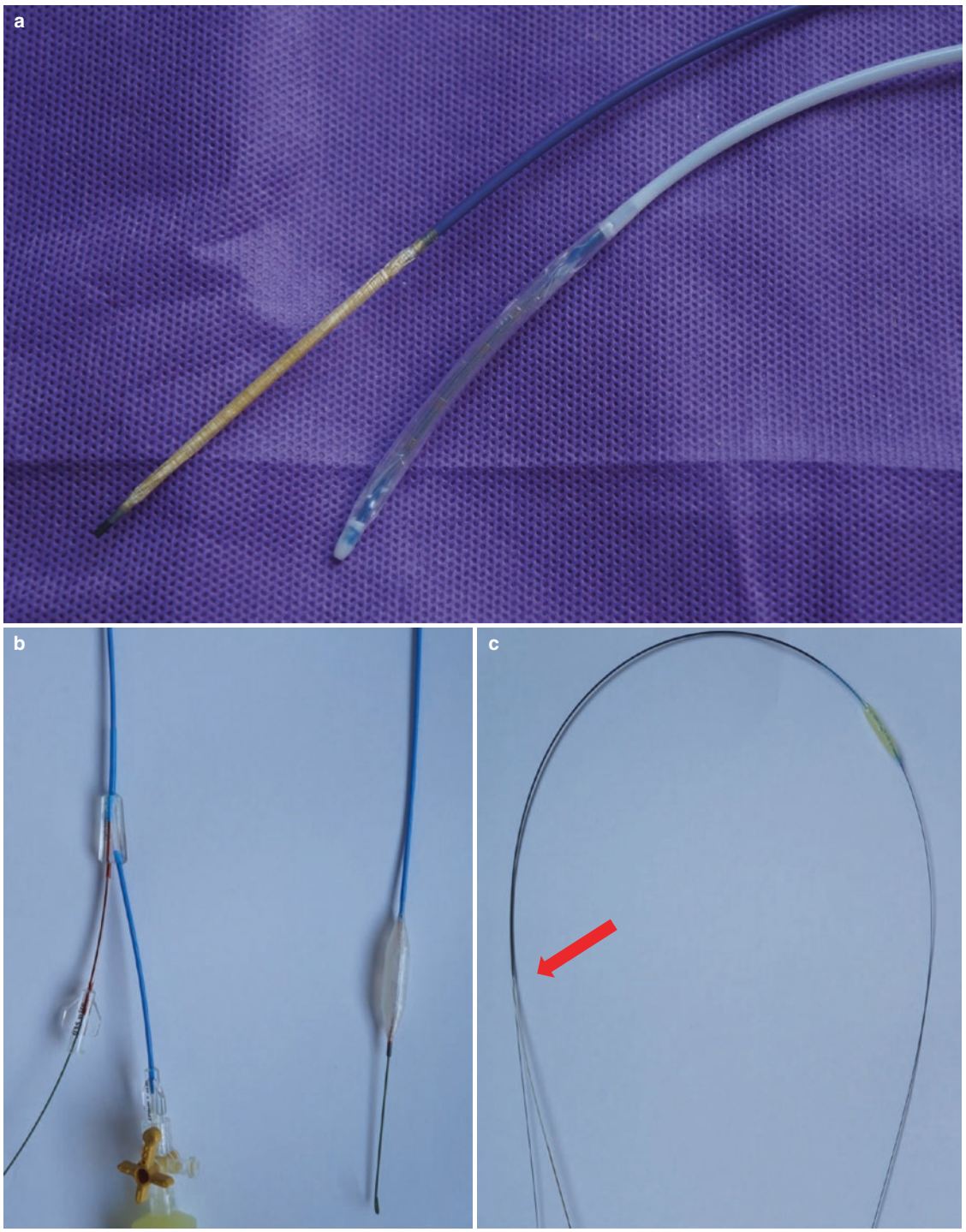



**Fig. 3.3** Miscellaneous balloon properties

- The *profile* of the balloon catheter determines the size of the introducer that is needed to insert the balloon. Balloons with smaller profile are as a consequence preferred for small children to avoid vascular complications. Lower profile is usually achieved at the cost of a smaller internal wire lumen, accepting only 0.014″ or 0.018″ wires, or a smaller balloon lumen that results in increased inflation and deflation time. Figure 3.3a shows two balloons with similar balloon diameter (18 mm) but very different profile.
- Other characteristics of balloon catheters include *flexibility*, *kink resistance*, *pushability* and stretchability that all differ between balloons.
- 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 (Fig. 3.3b). This is different from most coronary angioplasty balloons, where the wire tracking is limited to the distal part of the catheter shaft (*mono-rail*) (Fig. 3.3c, the red arrow shows where the wire exits 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 (mono-rail).

Every paediatric catheterization laboratory should be equipped with a minimum amount of balloons with different characteristics. It is however impossible and unnecessary to have the whole range of commercially available balloons. The interventional cardiologist will have to select, based on his or her experience, a few balloons with different and complementary characteristics, to respond to the various clinical situations.

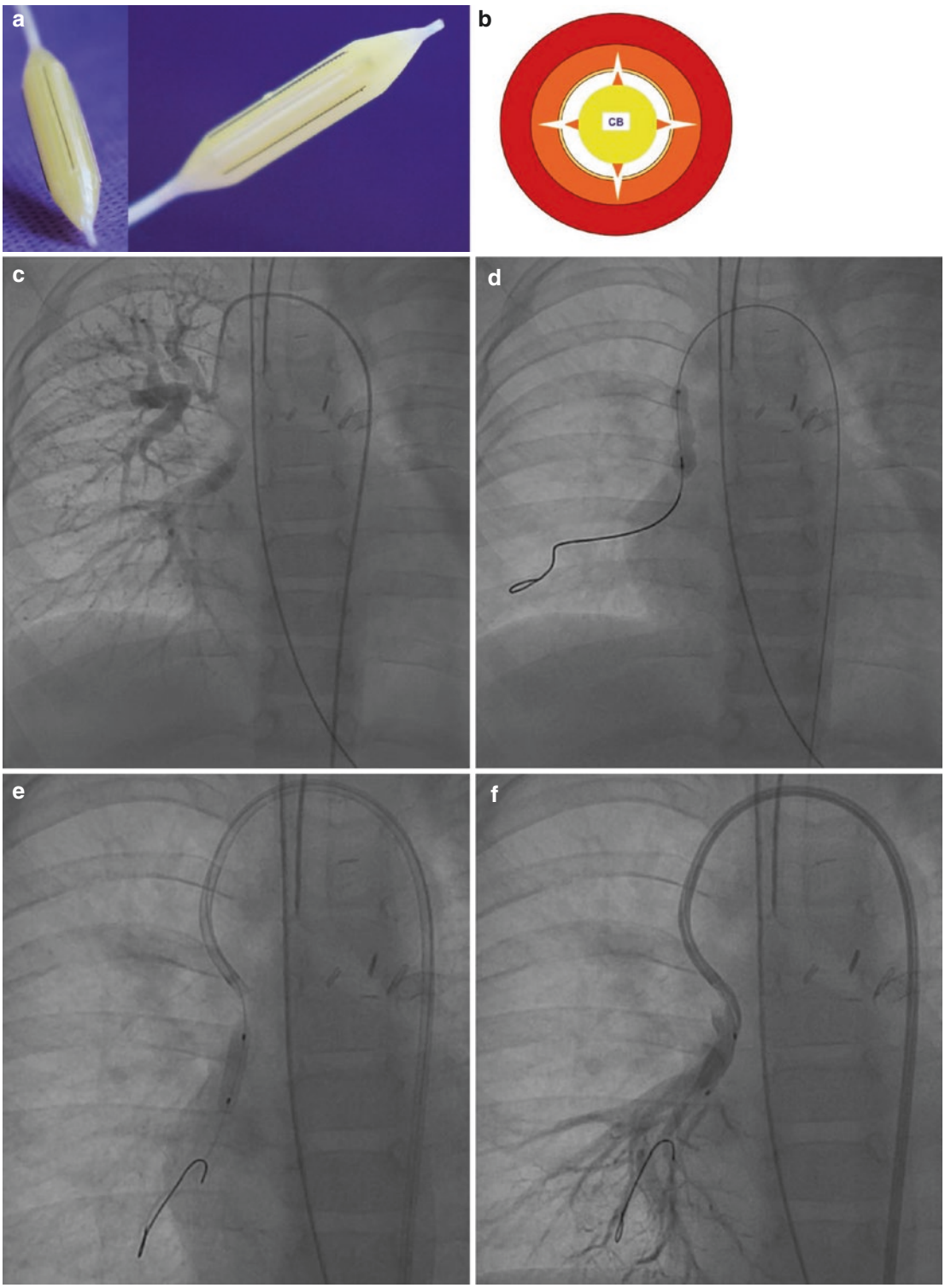



**Fig. 3.4** Special balloons: cutting balloons
The Boston Scientific Cutting Balloon® (Boston Scientific, MA, USA) is a non-compliant balloon with four sharp steel blades embedded longitudinally on its surface (Fig. 3.4a). Other bladed balloons are on the market, but their experience in paediatric or congenital interventions is scarce.

- Rationale: to create four controlled tears in a thickened intima and media, without over-dilating the vessel, avoiding non-controlled and deeper tear (Fig. 3.4b).
- Diameter: 2–8 mm, Burst pressure: 8 atm.
- A long sheath is required to avoid damaging cardiac and vascular structures with the blades.
- Slow inflation and deflation is recommended, with an indeflator, to allow proper opening and refolding of the blades.
- Use: The cutting balloons are initially designed to dilate resistant coronary stenosis. Currently the cutting balloons are also used to dilate pressure-resistant stenosis in pulmonary arteries, in-stent stenosis and restrictive atrial septal defects

Figure 3.4c shows pulmonary arteries in a patient born with pulmonary atresia, ventricular septal defect and major aorto-pulmonary collaterals, status post unifocalization. There is marked stenosis of one of the formal collaterals. A compliant balloon (Fig. 3.4d) shows multiple resistant lesions. After balloon dilation with a cutting balloon (Fig. 3.4e) the angiography shows marked enlargement of the lumen (Fig. 3.4f)

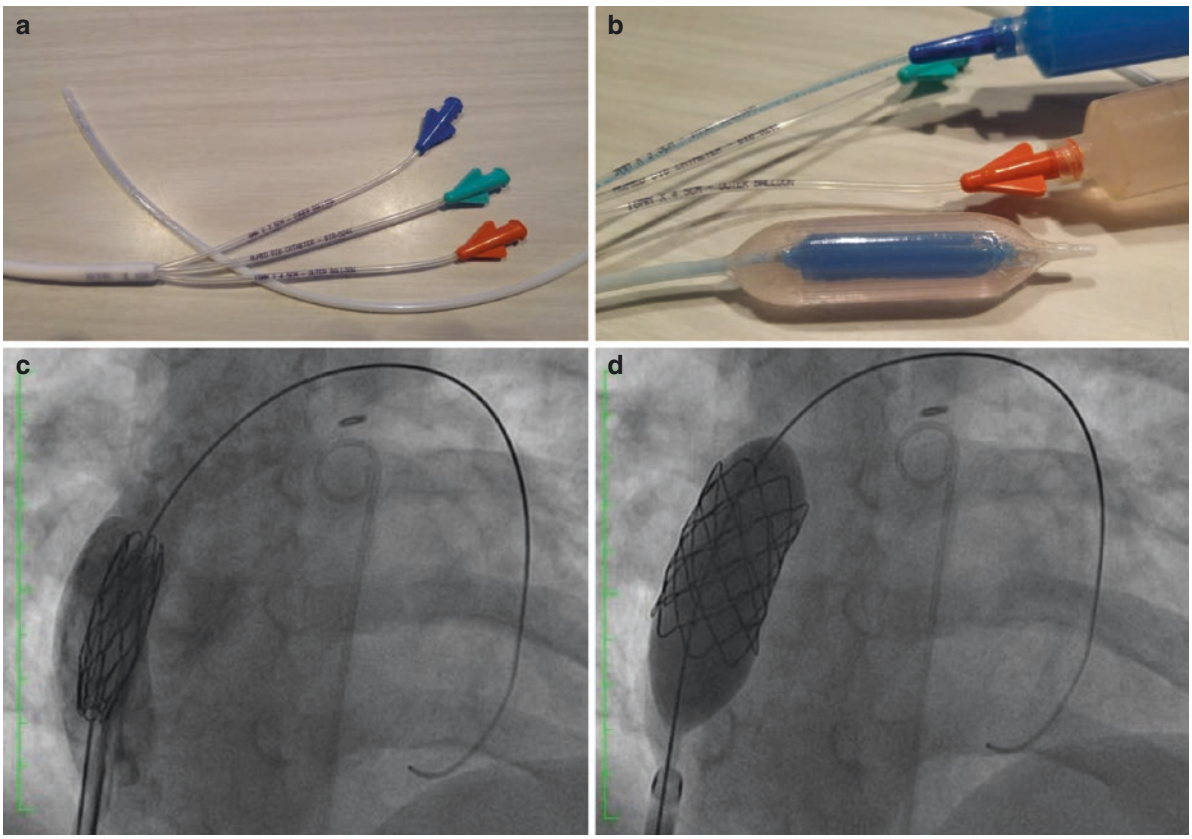



**Fig. 3.5** Special balloons: the Balloon in Balloon (BIB®) catheter (Numed Inc., NY, USA)

- Triaxial catheter: 1 lumen (green) is for tracking over a guide wire, 1 lumen (indigo) is for dilation of the small inner balloon, and 1 lumen (orange) is for inflation of the larger outer balloon. The inner balloon is completely inside the larger balloon, inflates to half the diameter of the outer balloon and is 1 cm shorter (Fig. 3.5a, b).
- Rated burst pressure: different for each size.
- Advantages: allows incremental 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 (Fig. 3.5c, covered stent implantation in an extracardiac conduit for fenestration closure). The outer balloon is then inflated securing the stent against the vessel wall (Fig. 3.5d)

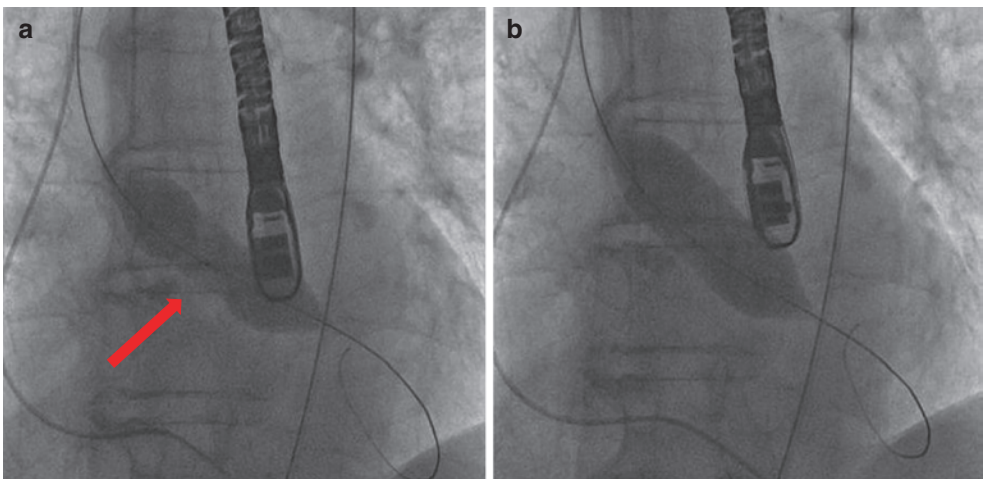



**Fig. 3.6** Special balloons: the dumbbell-shaped balloons

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

Left panel (Fig. 3.6a) shows a Nucleus™ balloon (Numed) used for aortic valve dilation. The balloon is incompletely inflated and has the dumbbell morphology; the narrowest part is at the level of the aortic annulus (red arrow). On the right panel (Fig. 3.6b), the balloon is completely inflated with loss of the dumbbell shape

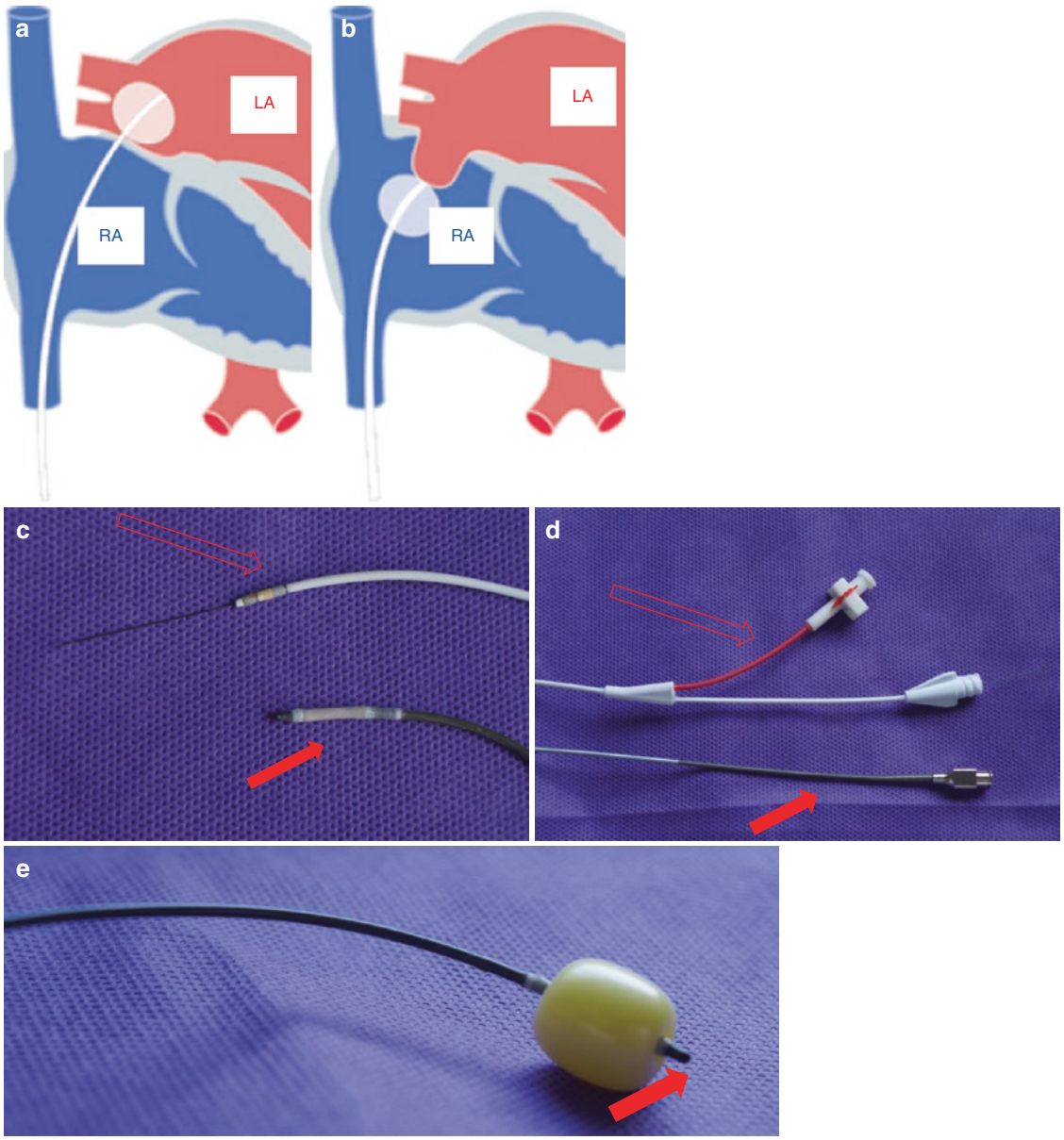



**Fig. 3.7** Special balloons: septostomy balloons

- Specifically designed to cross the *foramen ovale*, to perform the Rashkind atrial septostomy manoeuvre (Fig. 3.7a, b)
- Round, non-compliant balloons
- The currently available and most widely used are:
  - Edwards atrioseptostomy catheter (Edwards Lifesciences Corporation, CA, USA)
    5F catheter, requiring 7F introducer
    Rather stiff with curve at the end to facilitate crossing of the *foramen ovale*
    Single lumen for balloon inflation
    Balloon takes larger volumes of liquid (4 cc) than other balloons
  - Medtronic Rashkind balloon septostomy catheter (Medtronic) (Fig. 3.7c–e, full red arrow)
    6F catheter, requiring 6F introducer.
    More flexible than the Edwards catheter.
    Single lumen.
    Balloon takes 2 mL of liquid.
  - Numed Z-5TM atrioseptostomy catheter (Numed Inc., NY, USA) (Fig. 3.7c, d, open red arrow)
    Dual lumen balloon catheter
    Soft catheter, progresses easily over a wire
    Helpful when crossing restrictive *foramen ovale*
    Two sizes: 1 cc balloon–4F catheter (useful for small preterm babies), 2 cc balloon–5F catheter
    Requires that the procedure is performed under fluoroscopy

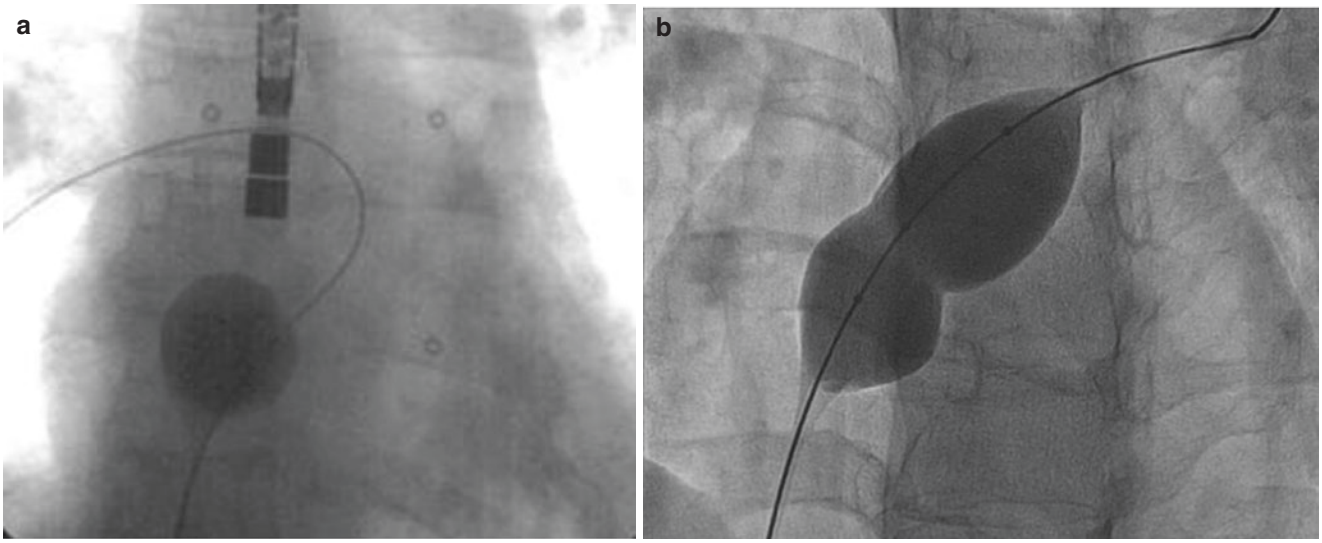



**Fig. 3.8** Special balloons: sizing balloons

- Used for measurement of atrial septal defects during percutaneous atrial septal closure
- Two types exist based on the method used for sizing:
  - 'Pull-through' technique: a soft, compliant and spherical balloon 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' (Fig. 3.8a).
  - 'Static' technique: a soft, compliant, low-pressure and elongated balloon 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 (Fig. 3.8b).
- Currently available sizing balloons:
  - 'Pull-through': EqualizerTM (Boston Scientific, MA, USA)
  - 'Static': AmplatzerTM sizing balloon II (St Jude Medical Inc., MN, USA),
    PTS® and PTS-XTM (Numed Inc., NY, USA)

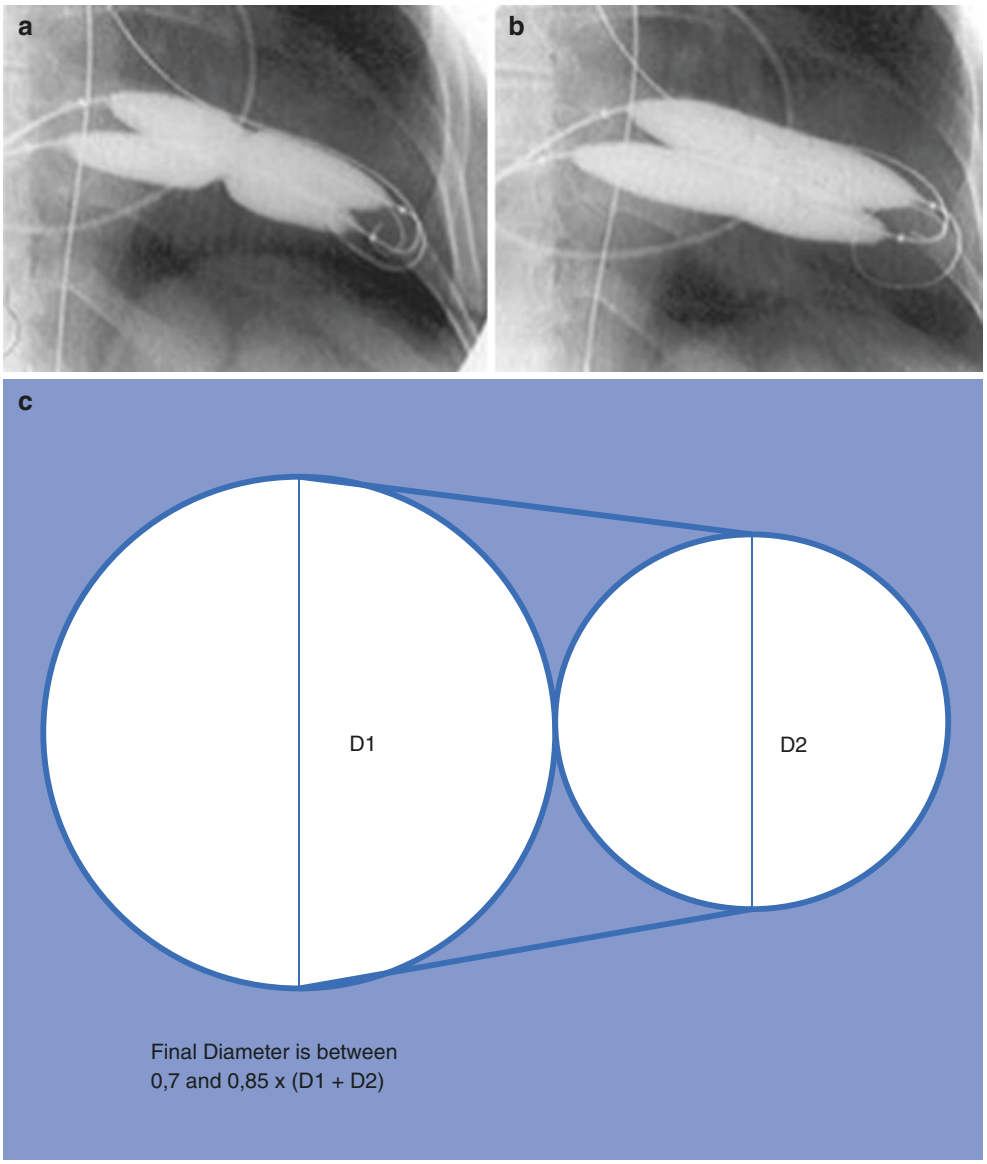



**Fig. 3.9** Special techniques: 'double balloon' technique

- Insertion and inflation of two balloons, simultaneously, at the same location
- Indications:
  - The valve annulus is too large for the available balloons (e.g. pulmonary or mitral valves in adult patients). The simultaneous use of two balloons is then required. Figure 3.9a, b shows the double balloon technique for mitral valve dilation. In Fig. 3.9a the balloons are not fully inflated and the waists are still visible; in Fig. 3.9b the waists have disappeared with full inflation.
  - To reduce the sheath size in small patients: insertion of two smaller balloons in two separate veins will require smaller sheaths than one large balloon through one access vein.
  - To improve hemodynamic tolerance: inflation and deflation time is shorter in smaller balloons, and persistence of some residual flows between the balloons even at full inflation, which may improve hemodynamic tolerance [3].
  - To increase balloon stability: the inflation of the first balloon followed by the inflation of the second balloon may increase balloon stability, for example, during aortic valve dilation.
- Several formulas exist to calculate the effective diameter of the two 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 two diameters (Fig. 3.9c) [2]

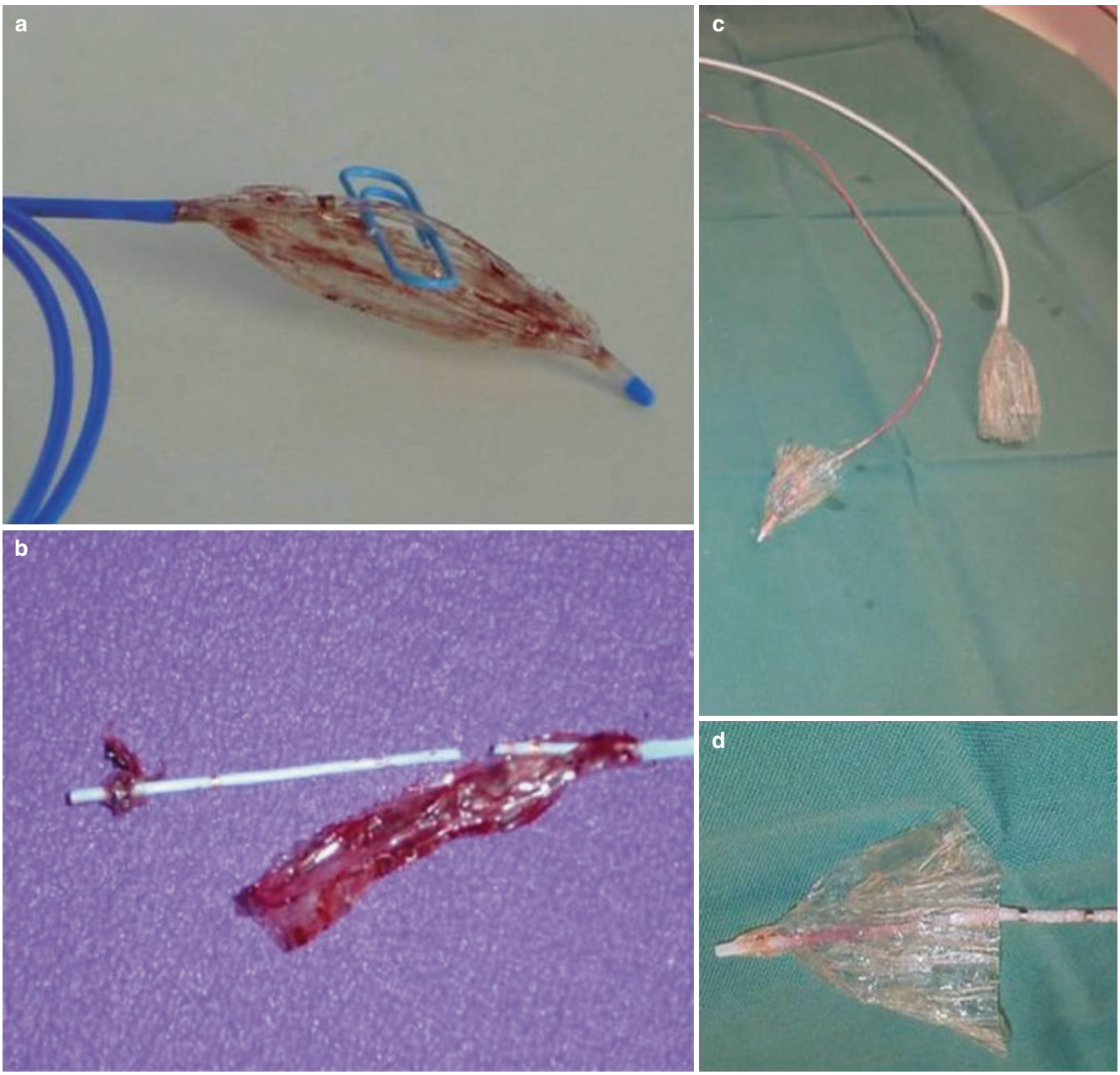



**Fig. 3.10** Complications with balloons

- *Balloon rupture*: Fluoroscopy may show extravasation of contrast. Applying negative pressure to deflate the balloon brings back blood together with contrast. The balloon can rupture in three different ways:
  - Balloon puncture: most often caused by a stent strut or a calcified vessel. The contrast leak is localized.
  - Longitudinal tear: usually the result of balloon inflation with an excessive pressure or in case of dilation of a very calcified lesion (such as a conduit). Fluoroscopy may show sudden disappearance of the contrast. Retrieval of the entire balloon is usually not problematic. Figure 3.10a shows a longitudinal tear. The blue paper clip shows the balloon shaft exposed by the longitudinal tear (Courtesy Dr. Joseph De Giovanni, Birmingham, UK).
  - Circumferentially tear (less frequent). Retrieval may be complicated by folding of the distal part over the tip of the balloon catheter and possible disruption of the distal balloon, which will then be free-floating. The use of a snare may be needed to stabilize the distal end and retrieve the complete balloon (see Fig. 3.11). Figure 3.10b shows a 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). Figure 3.10c, d shows a 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 (10d) and the proximal part on the external shaft (Courtesy Prof R. Berger, Groningen, Netherlands).
- *Inability to deflate a balloon*: Usually the result of a localized rupture or puncture of the balloon at the proximal end, with the proximal deflated end of the balloon obstructing the communication between the balloon and the balloon lumen.
- *Fracture of the shaft of the balloon*: Very rare and unlikely with single-used balloons but may happen with reused and re-sterilized balloons.
- *Inability to expand a vessel or stent as a result of balloon puncture*: Connecting the balloon to the dye injector and injecting dye under high pressure may improve balloon and stent expansion and allow safe retrieval of the balloon

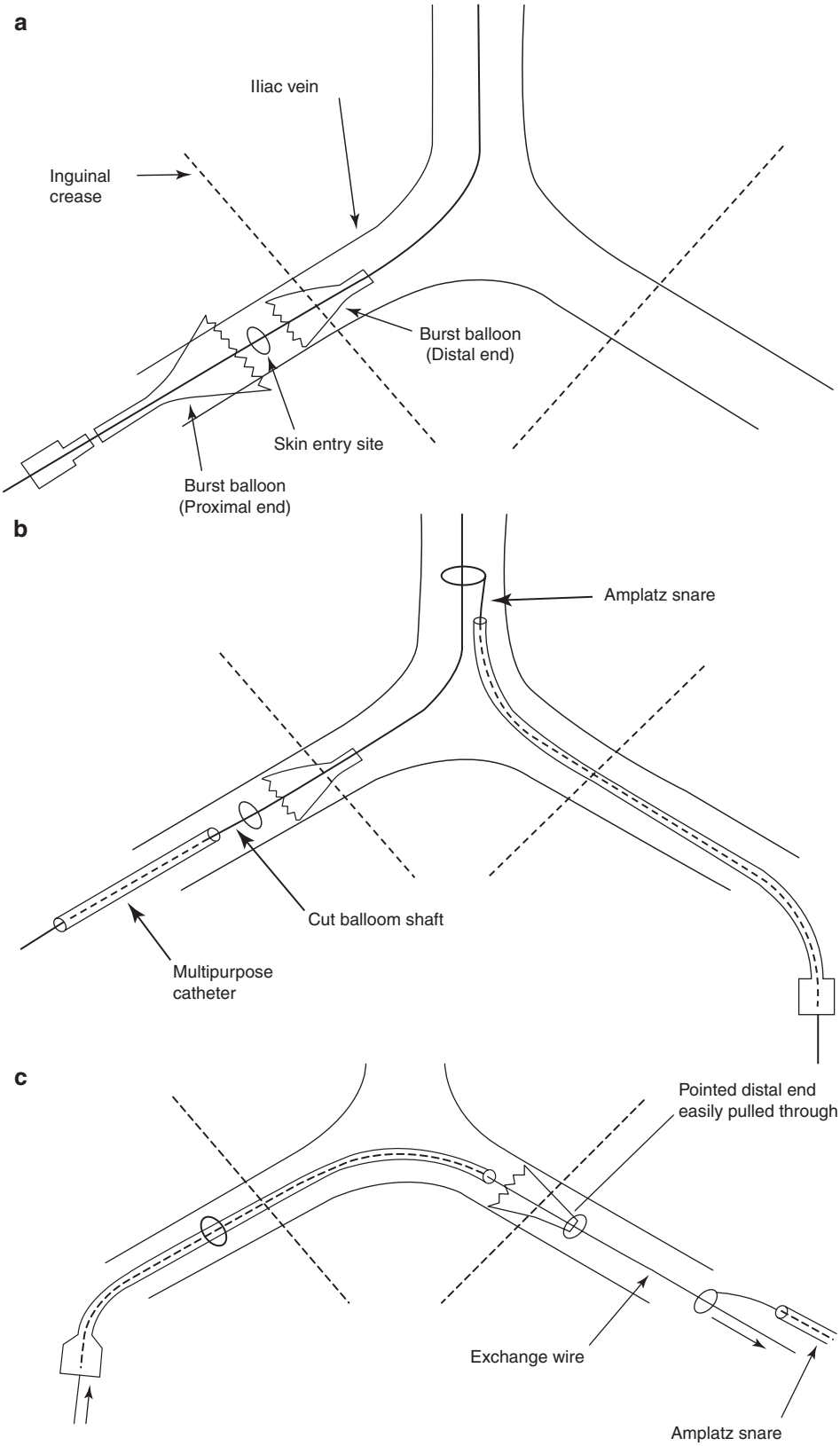



**Fig. 3.11** Technique for retrieval of circumferentially burst balloons: (**a**) wire position has to be conserved. (**b**) The wire is snared distally, using the contralateral vein. The proximal part of the balloon will be taken out of the body, and the balloon shaft will be cut. A multipurpose catheter 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. Joseph De Giovanni, Birmingham, UK)

## References

1. Abele JE. Balloon catheters and transluminal dilatation: technical considerations. Am J Roentgenol. 1980;135:901–6.
2. Lock JE, Keane JF, Perry SB, editors. Diagnostic and interventional catheterization in congenital heart disease. 2nd ed. Boston, MA: Kluwer Academic Publishers; 2000.
3. Mullins C. Balloon dilation procedures—general. In: Mullins C, editor. Cardiac catheterization in congenital heart disease: pediatric and adult. Masachusetts: Blackwell Futura; 2006. p. 410–29.

# Stents

# 4

Sebastian Goreczny and Eric Rosenthal

For over three decades, stents have been used in congenital heart disease with increasing experience and application in more diverse lesions at all ages. Stent implantation enables overcoming of several limitations of isolated balloon dilation leading to a superior relief of stenosis both acutely and in the long term. Especially elastic lesions, long-segment stenoses, hypoplastic vessels, stenoses related to kinking or tension on a vessel respond better to stent implantation. However, several issues unique to the paediatric population like small vessel access, difficulty in advancing the rigid stent through a tortuous vascular route or somatic growth requiring stent redilation warrant careful selection of stent and implementation of modified interventional techniques.

Given the diversity of lesions and patient size range, a single type of stent does not suit all situations. Technological advances have led to availability of a wide range of stents to match particular lesions. Modern stents (Figs. 4.1, 4.2 and 4.3) can be classified in several groups according to:

- Stent size:
  - Small size (up to 5–6 mm)
  - Medium size (6–12 mm)
  - Large size (12–20 mm)
  - Extra large (more than 20 mm)
- Cell design:
  - Closed-cell design
  - Open-cell design
  - Hybrid design
- Mounting:
  - Unmounted
  - Premounted
- Mode of implantation:
  - Self-expanding stents
  - Balloon-expandable stents
- Covering:
  - Uncovered stents
  - Covered stents
- Other types:
  - Growth and biodegradable stents
  - Stent grafts

The basic principles of stent implantation are common to most lesions (Fig. 4.4). These include:

- Obtaining access
- Haemodynamic assessment and angiography
- Predilation (optional)
- Stent choice
- Guide wire and sheath placement
- Mounting (for unmounted stents)
- Stent introduction
- Stent positioning
- Stent deployment
- Final haemodynamic assessment and angiography

S. Goreczny (✉)
Department of Cardiology, Polish Mother's Memorial Hospital, Research Institute, Lodz, Poland

E. Rosenthal
Paediatric Cardiology, Evelina London Children's Hospital, St Thomas' Hospital, London, UK


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_4

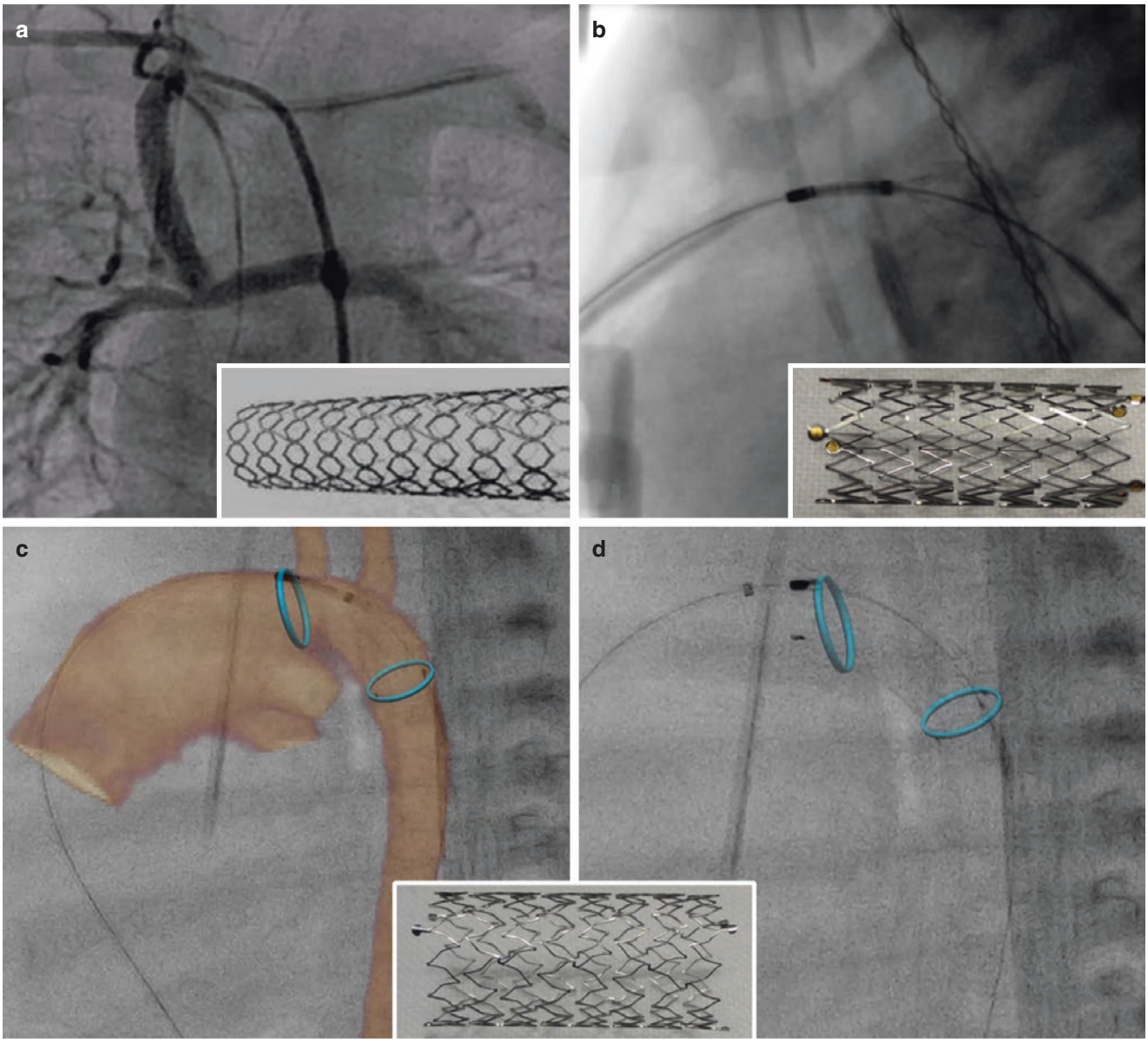



**Fig. 4.1** Examples of selected small size stents. (**a**) Balloon-expandable stainless steel coronary stent implanted in a systemic-to-pulmonary artery shunt. (**b**) Hybrid deployment of a self-expanding, nitinol sinus-SuperFlex-DS to the ductus arteriosus. (**c**) Percutaneous three-dimensional guided implantation of a self-expanding, nitinol Zilver Flex to the ductus arteriosus. (**d**) The latter stent after deployment. Radiopaque markers at both ends of the sent align with marking rings indicating pulmonary and aortic insertions of the duct

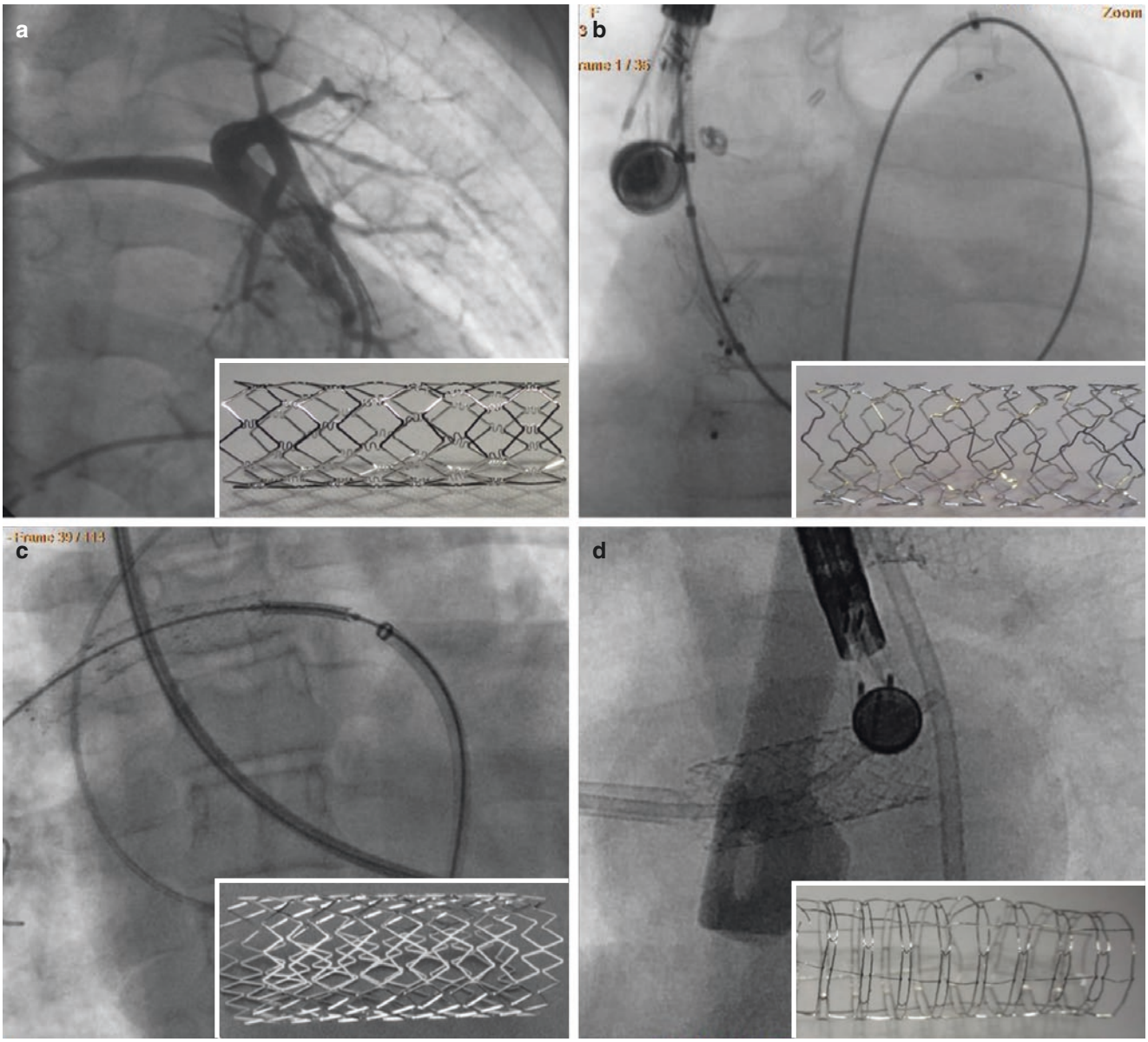



**Fig. 4.2** Examples of selected medium size stents. (**a**) Stainless steel, closed cell Palmaz Genesis stent implanted in the right ventricular outflow tract. (**b**) Stainless steel, open-cell Valeo Lifestent, placed in fenestration in a patient after Fontan completion. (**c**) Stainless steel, open-cell Visi-Pro is being positioned in the right pulmonary artery. (**d**) Stainless steel, open-cell Formula stent after hybrid implantation to restrictive interatrial communication

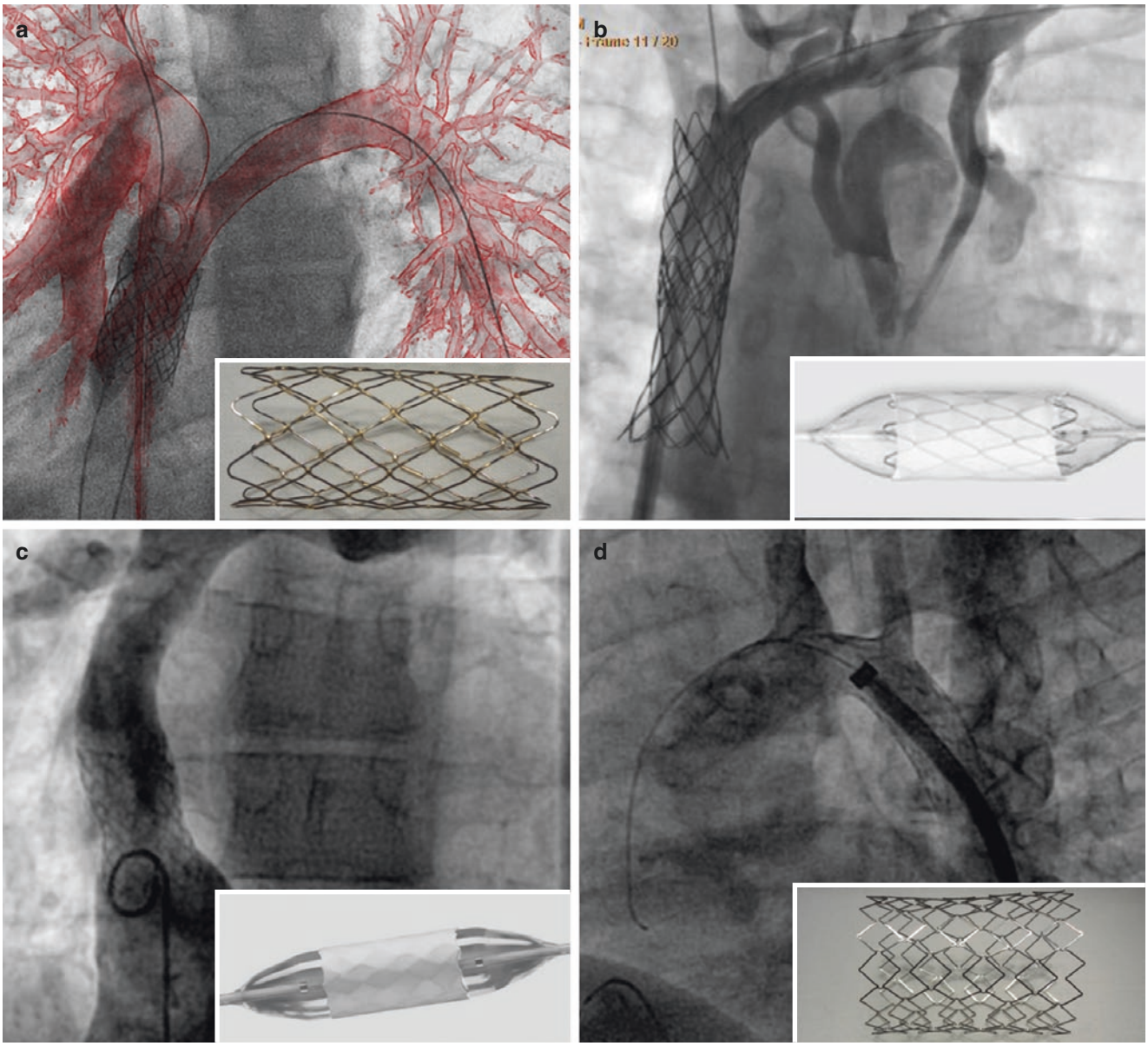



**Fig. 4.3** Examples of selected medium to large size stents. (**a**) Balloon-expandable platinum-iridium closed-cell design Cheatham-Platinum stent is being deployed on two balloons to the extracardiac tunnel in a patient after Fontan completion. (**b**) Angiography after reconnection of the superior caval vein to the right atrium with implantation of two covered Cheatham-Platinum stents. (**c**) A stainless steel, open-cell design, covered Advanta V12 stent was implanted to the extracardiac tunnel in a patient after fenestrated Fontan operation. (**d**) A chromium cobalt, hybrid cell design AndraStent was implanted to aortic coarctation

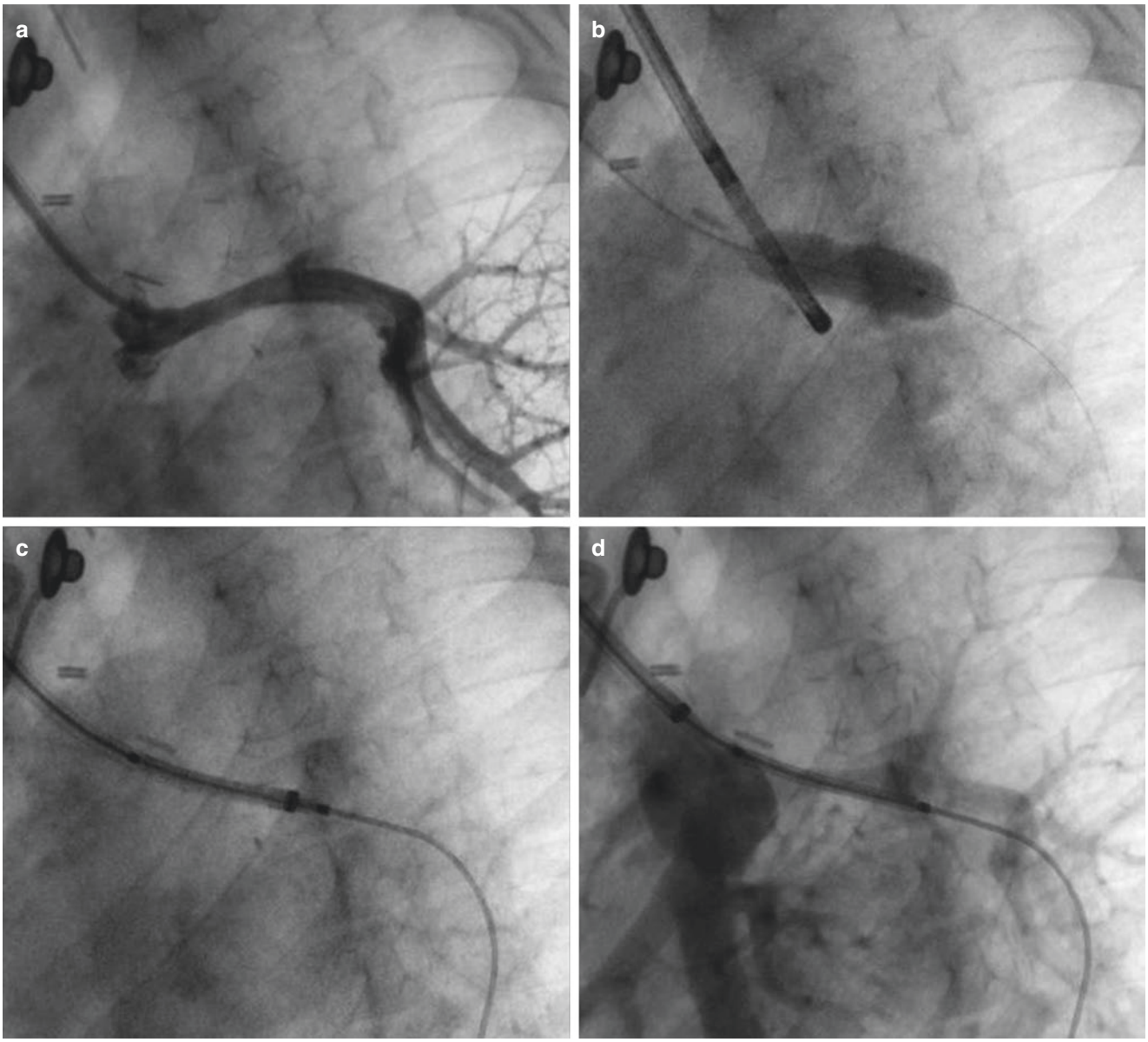



**Fig. 4.4** Step-by-step stent implantation. (**a**) A patient with hypoplastic left heart syndrome after hemi-Fontan operation. Angiography from SVC shows hypoplastic left pulmonary artery with poor contrast flow to the upper lobe. (**b**) The pulmonary artery was predilated for evaluation of compliance and potential compression on the left main bronchus. Simultaneous bronchoscopy was performed to assess consequence of balloon inflation on the bronchus. (**c**) An open-cell Valeo Lifestent was delivered through a long sheath introduced distally to the lesion. (**d**) Check angiography during positioning of the stent shows the implant placed between the origin of the left upper lobe branch artery and proximally the hemi-Fontan anastomosis. (**e**) The dedicated balloon was fully inflated with nominal pressure. (**f**) Angiography shows improved diameter of the left pulmonary artery with proximal end of the stent "hanging" in the hemi-Fontan connection. The distal end of the stent is placed just proximal to the first branching of the left pulmonary artery. (**g**) The proximal end of the stent was flared with a compliant oversized balloon. (**h**) Final angiography shows satisfactory result

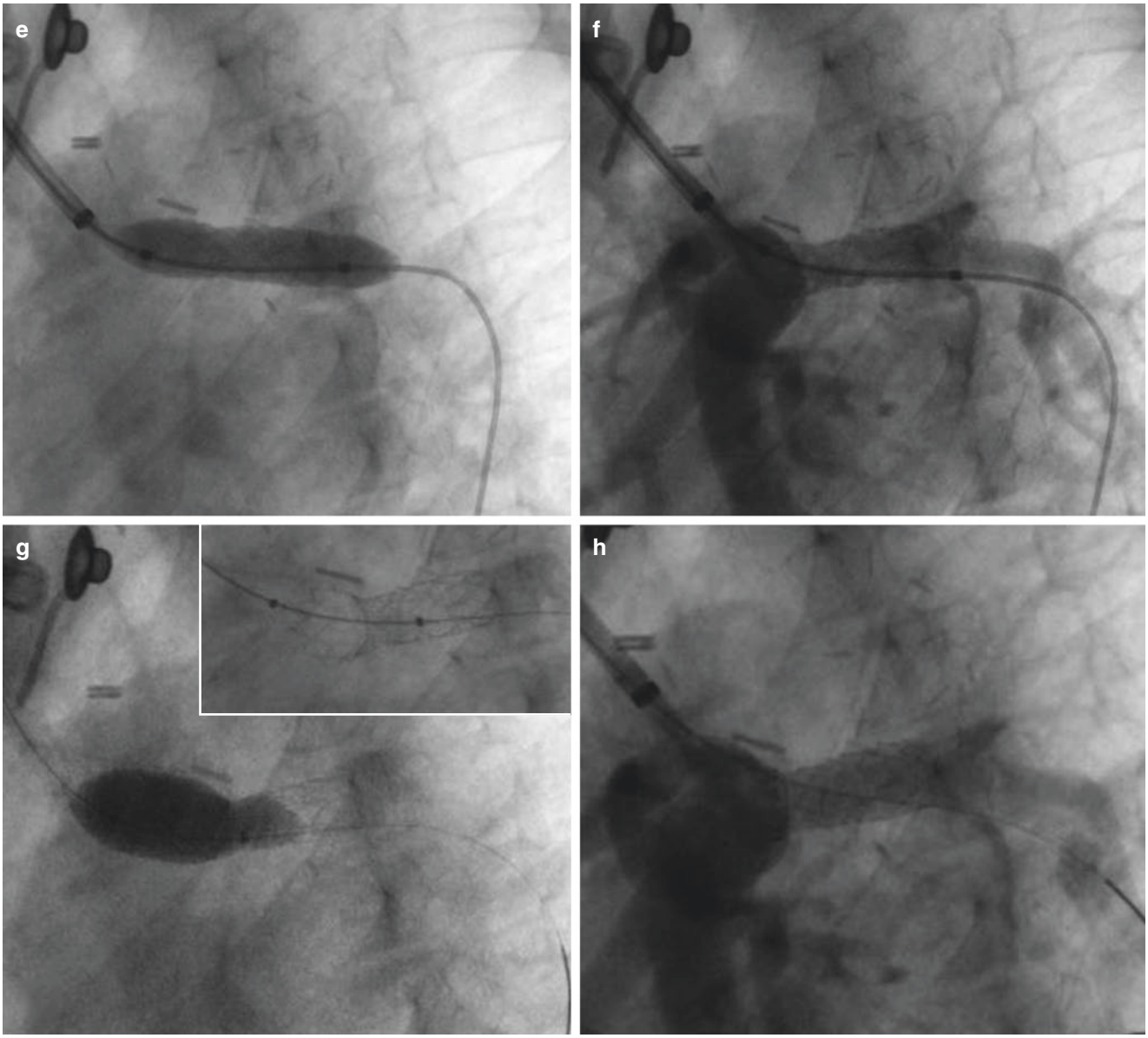



**Fig. 4.4** (continued)

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. Complications after stent implantation include:

- Stent malposition or migration (Fig. 4.5)
- Side-branch compromise
- Vessel dissection and rupture (Fig. 4.5)
- Balloon rupture
- Stent fracture (Fig. 4.6)
- Restenosis (Fig. 4.7)

The vast majority of acute stent-related complications can be prevented by meticulous step-by-step approach with attention to details. When they occur, however, it is vital to maintain guide-wire position for remedial action with the stent and vessel still accessible.

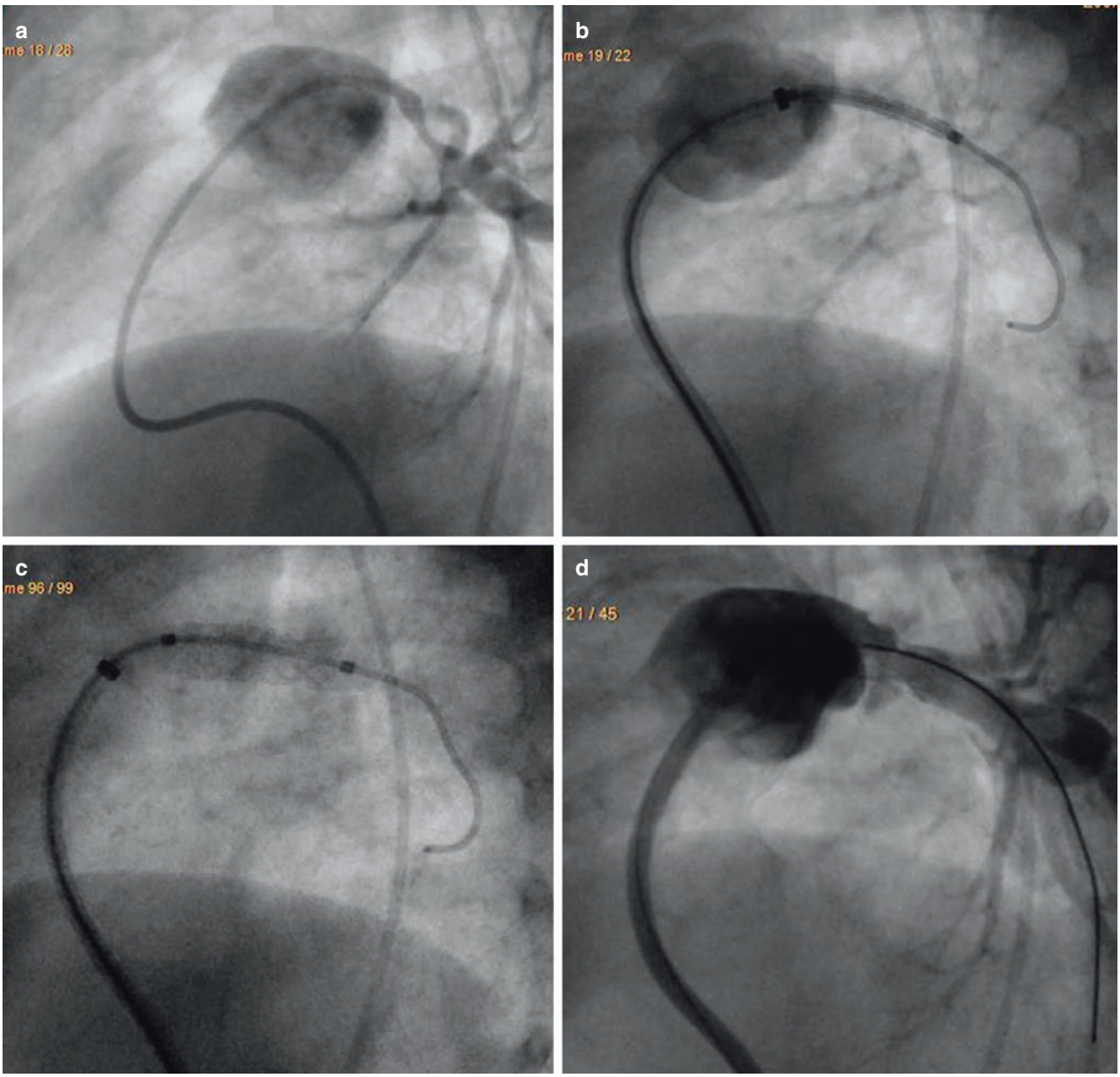



**Fig. 4.5** Step-by-step stent implantation. Complications— stent malposition and vessel dissection. (**a**) Initial angiography shows severely hypoplastic left pulmonary artery with discrete stenosis just proximal to the first branch. (**b**) A balloon-expandable Valeo Lifestent stent was introduced through a long vascular sheath placed in the main pulmonary artery. The nasogastric tube was used as a marking point for the distal end of the stent. (**c**) During balloon inflation, the stent milked back towards the main pulmonary artery. (**d**) Final angiography shows significant improvement of the left pulmonary artery diameter with dissection of the bottom wall and contrast extravasation

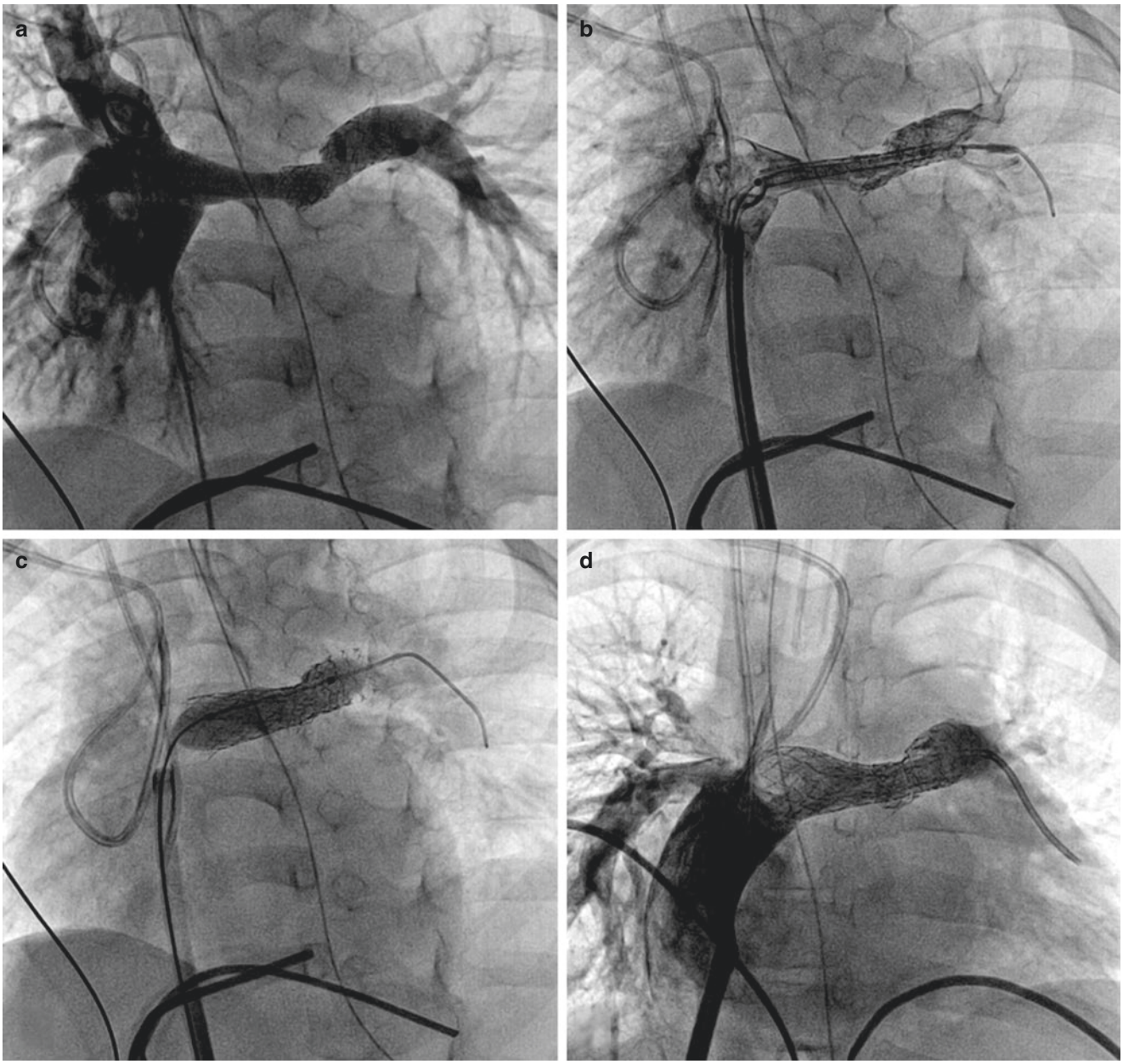



**Fig. 4.6** Complications—stent fracture. (**a**) A patient with hypoplastic left heart syndrome early after Fontan operation. Angiography shows fractured and fragmented previously implanted Palmaz Genesis stent in the left pulmonary artery. Additional tubular stenosis of the proximal right pulmonary artery may be seen. (**b**) A covered Advanta V12 stent was introduced through a long sheath placed in the extracardiac tunnel. Contrast injection was performed to position the stent so as to cover the proximal right pulmonary artery. (**c**) Both stents were postdilated with a high-pressure balloon to achieve larger final diameter. On full balloon inflation, a residual narrowing at the site of the first stent fracture may be seen. (**d**) Final angiography shows stabilised segments of the Palmaz Genesis stent, improved diameter of the proximal right pulmonary artery and residual stenosis at the level of the previous stent fracture

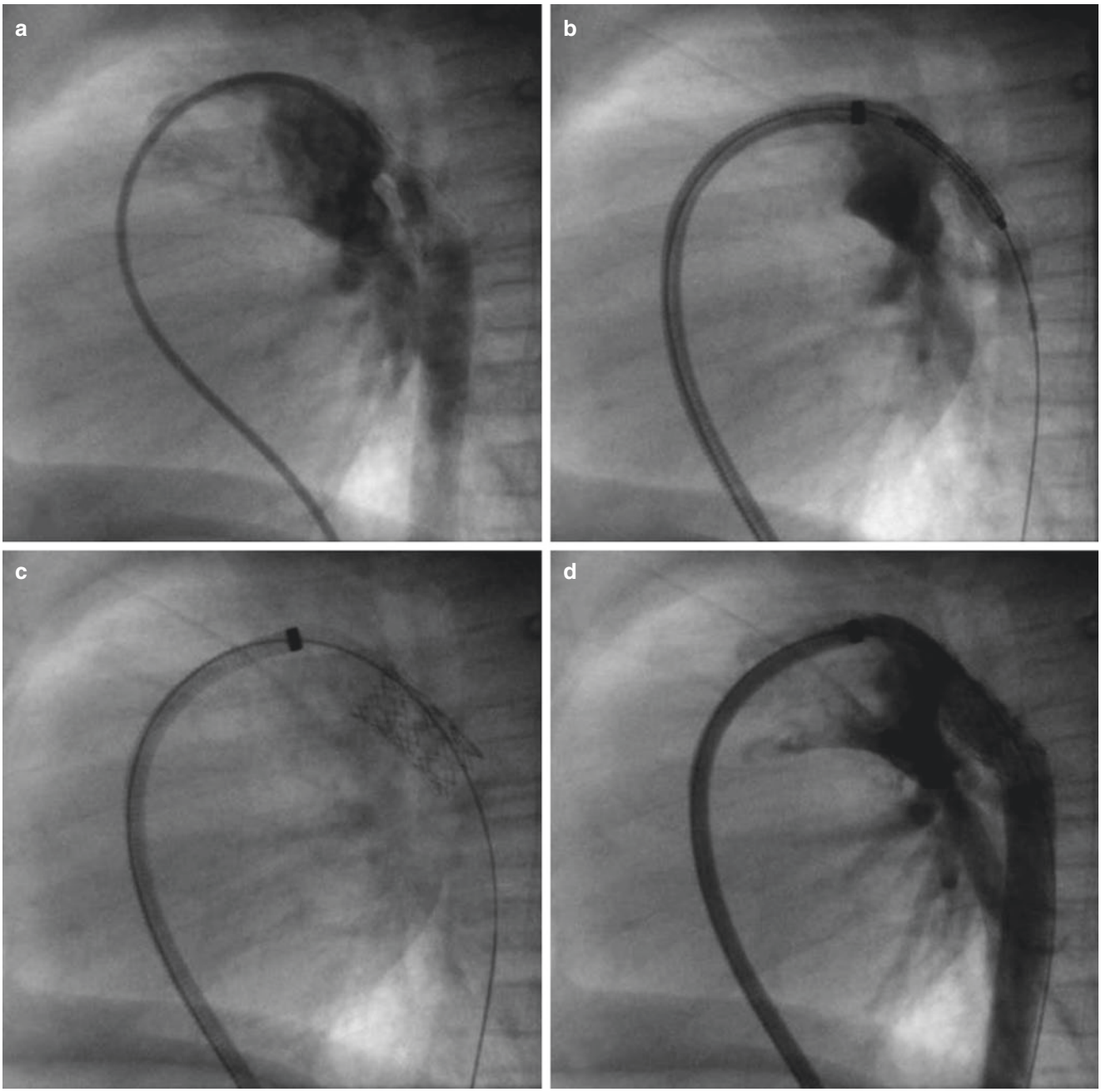



**Fig. 4.7** Complications—restenosis. (**a**) Patient with interrupted aortic arch after first-stage hybrid palliation. Angiography shows a self-expanding sinus-SuperFlex-DS stent in the arterial duct with significant stenosis due to neointimal hyperplasia. (**b**) Balloon-expandable Palmaz Blue stent was chosen to treat the stenosis. The stent was delivered through a long sheath from femoral vein access. (**c**) Still frame shows the stainless steel Palmaz Blue stent implanted inside the nitinol sinus-SuperFlex-DS stent. (**d**) Final angiography shows unobstructed flow through the stents placed in the arterial duct

# Part II

# Vascular Access

# The Usual Vascular Access

5

Daniel Tanase and Jochen Weil

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

Accurate vessel puncture requires either orientation on the surface anatomy or direct visualisation of the target vessel by ultrasound.

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

In general, forcing the guidewire almost never results in success [1].

## 5.1 Femoral Venous and Arterial Access

### 5.1.1 Positioning and Anatomic Landmark Guidance

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

*Technique:*

- Landmark is the inguinal ligament (Fig. 5.1).
- Puncturing level is at the inguinal skin crease or just below.
- For arterial puncture, the needle should be advanced targeting the umbilicus; the vein runs parallel to the artery.

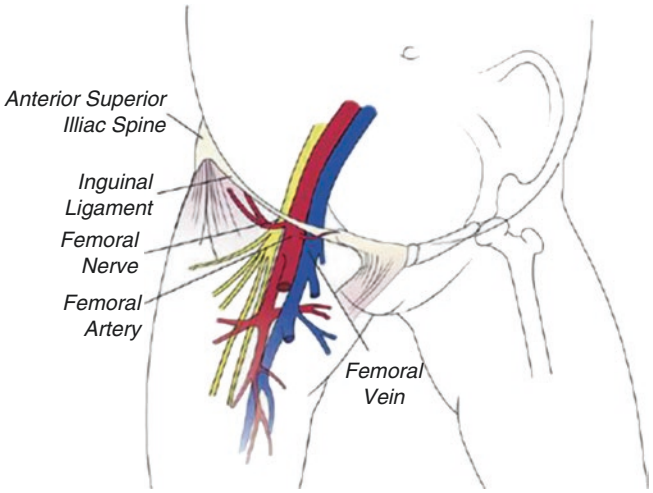



**Fig. 5.1** Landmarks of the femoral artery and vein access. The inguinal ligament runs between the superior iliac spine and the pubic tubercle. The femoral artery crosses the inguinal ligament at approximately its midpoint and should be palpated there. The femoral vein runs closely medial and parallel to the artery (from Bergersen [2], with permission)

- The angle is flat in the newborn (10°–20°) and steepens with age (45°–60° in the adults).
- Introduce gently the guidewire; it should pass with minimal resistance.
- Once the guidewire is positioned, the needle should be removed and the sheath-dilator assembly advanced with a rotatory motion.

The direction of the artery can also be checked by simultaneous palpation of the pulse with two or three fingers. For venous puncture, it is helpful to advance the needle with a syringe attached under continuous suction. Some operators prefer to puncture the artery without a syringe attached. Free pulsatile flow might be easily identifiable.

If the guidewire cannot be advanced gently but a backflow of blood is still visible, it is probable that the vessel was not hit appropriately. Then a slight change of the direc-

D. Tanase (✉) · J. Weil
Department of Paediatric Cardiology and Congenital Heart Defects, German Heart Centre Munich, Munich, Germany
e-mail: tanase@dhm.mhn.de; weil@dhm.mhn.de


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_5

tion, or the angle of the needle might facilitate the advancement of the guidewire. If this is still not possible, the needle and guidewire should be removed and a new attempt performed.

### 5.1.2 Ultrasound-Guided Puncture

Routine use of real-time ultrasound guidance reduces the number of attempts, time to access, and risk of vascular complications [3].

Possible transducers:

- A hockey stick or a 12 MHz for newborns and small children
- A 6 MHz or linear transducer for adults

Real-time ultrasound guidance can be performed either in a long-axis or a short-axis approach. Especially in small children, visualisation of the vessels in the long axis might be advantageous because needle and vessel can be visualised at the same time.

*Technique:*

- Transducer and cord should be inserted into a sterile sheath (Fig. 5.2).
- Distinguish the vessels—the vein is compressible and located medial to the artery; colour flow or Doppler imaging can distinguish the vessels, too.
- Optimise image quality (maximise the transducer frequency and the sector field focus; adjust the gain setting, able to detect low-velocity flow)
- Once the entire course of the target vessel is visualised, the position of the transducer should not be changed.

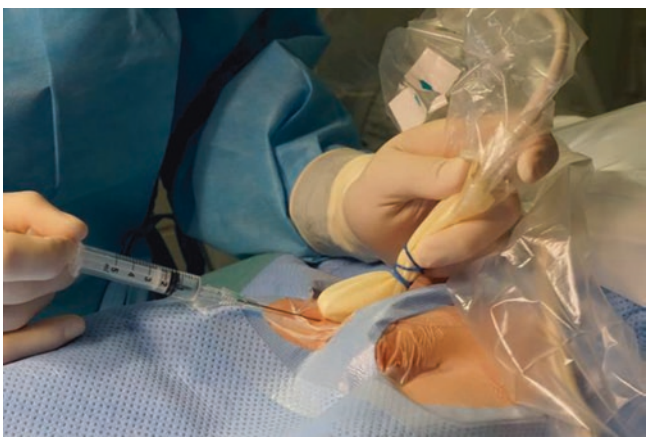


**Fig. 5.2** A hockey stick array placed inside a sterile sheath. The needle is placed directly to the contact face of the transducer

- Introduce the needle closely to the contact face of the transducer and advance it in the same direction until it penetrates the vessel wall.

If the vessel and needle cannot be visualised at the same time, it is mostly because the tip of the needle is outside the ultrasound beam of the array. Changing the position of the transducer to pick up the needle is mostly not helpful. It is better to adjust the needle to the long axis of the transducer.

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

## 5.2 Internal Jugular Vein Access

### 5.2.1 Positioning and Anatomic Landmark Guidance

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

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.
- There is less risk of damaging the thoracic duct.

### 5.2.2 Ultrasound-Guided Puncture

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

- A gentle probe pressure will compress the vein and will help to avoid arterial puncture according to the close relationship of both vessels, but both vessels can be differentiated sufficiently either by pulsation of the artery or by colour flow Doppler.
- The short axis will visualise both vessels at the same time, helping to distinguish them, while the long axis will help to rule out the direction in which the needle has to be advanced.
- Puncturing technique is the same as for femoral vessel cannulation.

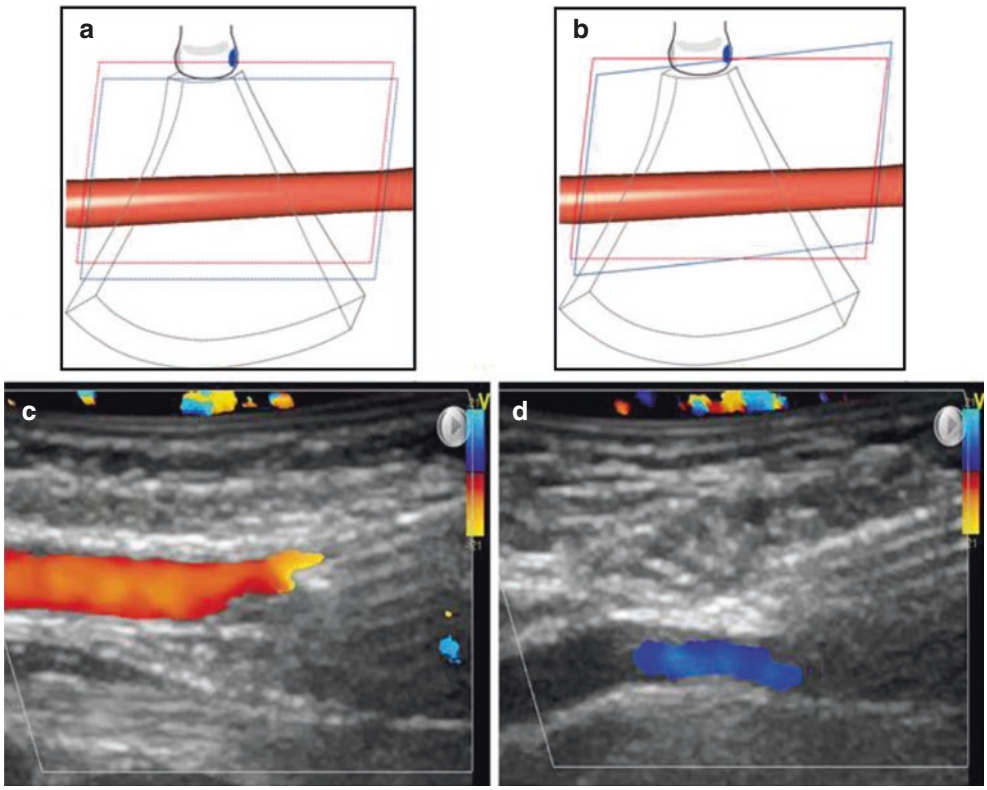



**Fig. 5.3** Technique of ultrasound-guided puncture. The transducer should be aligned to the course of the vessel—red plane in (**a**) and (**b**). If the needle is advanced in the same plane—blue plane in (**a**) the tip and distal parts of the needle become visible (**c**). Notice aliasing in the colour flow image in (**c**), as the tip of the needle penetrates the vessel wall. In (**b**) the plane in which the needle is introduced—blue plane is not aligned to the red plane of the transducer and the vessel. The vessel cannot be hit appropriately. In the corresponding ultrasound image (**d**) just mid parts of the needle are visible. The tip of the needle is near the vessel—as the vessel course is distorted—but not visible

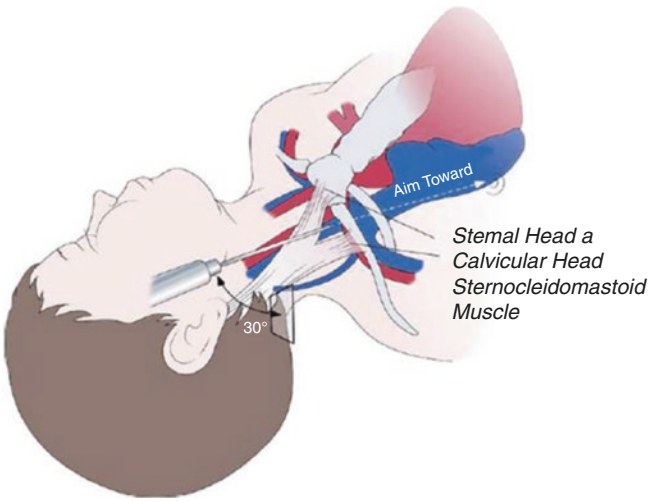



**Fig. 5.4** Landmarks of the internal jugular vein access. The essential surface anatomy is a triangle comprised of the sternal and clavicular heads of the sternocleidomastoid muscle medially and laterally and the medial third of the clavicle inferiorly, named the Sedillot's triangle. The internal jugular vein lies underneath this triangle and is located laterally to the carotid artery (from Bergersen [2], with permission)

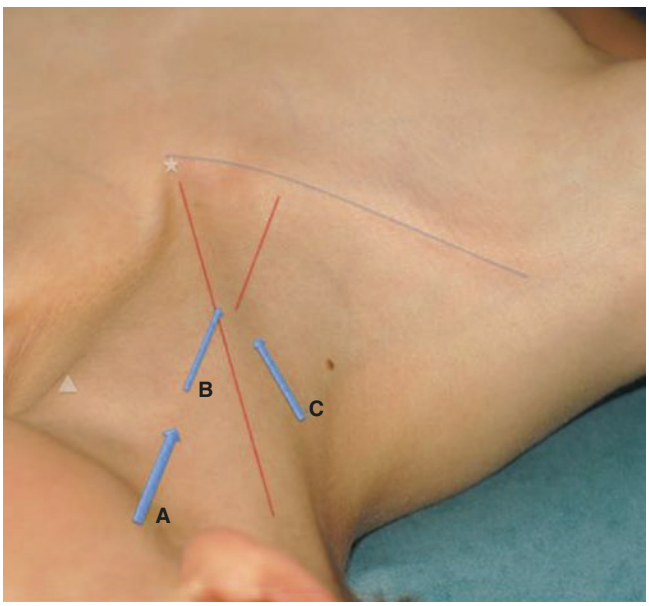



**Fig. 5.5** Three approaches to the internal jugular vein. The blue line represents the course of the clavicle. The red lines show both heads of the sternocleidomastoid muscle. Star—suprasternal notch. Triangle—thyroid cartilage. (*A)* For the *anterior approach*, the needle is introduced at the medial margin of the sternocleidomastoid muscle approximately at the level of the thyroid cartilage and directed towards the ipsilateral nipple; (*B)* for the *middle route*, the needle enters the apex of the triangle formed by both heads of the sternocleidomastoid muscle and the clavicle and also directed towards the ipsilateral nipple; (*C*) in the *posterior route,* the needle should be introduced along the lateral margin of the sternocleidomastoid muscle cephalad to the apex of the Sedillot's triangle. In this case, it should be directed towards the suprasternal notch

## 5.3 Subclavian Vein Access

Because of attachments of the subclavian vein to surrounding tissues, the vein remains patent even in hypovolemic patients.

- A slight Trendelenburg position of the patient will not increase the diameter of the vessel but will avoid complications of air embolism.
- The head should be kept neutral, as an excessive rotation to the contralateral side will increase the angle between the internal jugular and the subclavian vein, facilitating the advance of the guidewire into the IJV or the anonymous vein.
- The needle should be advanced parallel to the floor targeting the suprasternal notch.
- A close puncture to the clavicle is disadvantageous because the needle has then to be directed posteriorly to negotiate with the clavicle—this will increase the risk for pneumothorax.

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

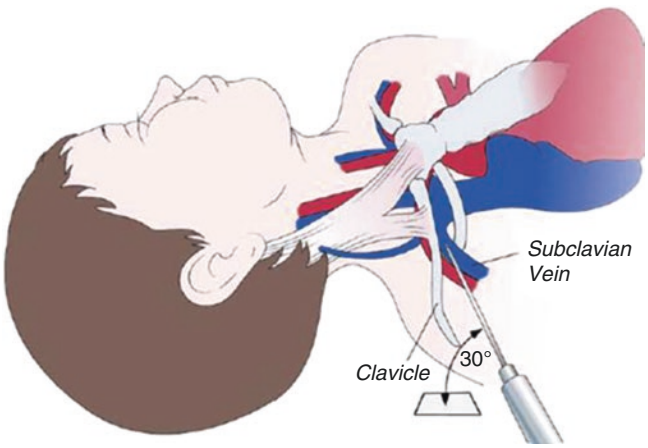



**Fig. 5.6** Landmarks of the subclavian vein access. The subclavian vein lies between the first rib and the clavicular bone and is bordered posterior by the subclavian artery and brachial plexus. The anatomic landmark is the junction of the medial one third and lateral two thirds of the clavicular bone. From medial, this corresponds to the first slope of the bone when the anterior convexity goes on to the anterior concavity. The cutaneous puncture should be inferiorly and laterally to this point (from Bergersen [2], with permission)

## 5.4 Umbilical Venous Access

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

- Catheterisation of the vessels is performed with a 3.5 or 5 F umbilical catheter.
- The 3.5 F catheter takes a 0.021″ and the 5 F catheter a 0.025 “guidewire.
- If these catheters are in place, they can be cut with a scalpel short above the umbilicus, and a sheath can be inserted over a guidewire.

*Problems:*
Umbilical vein

- The guidewire and catheter are difficult to manoeuvre into the ductus venosus due to the entering of the portal vein.
- Withdraw the catheter into the umbilical vein and give a small injection of contrast to delineate the course of the ductus venosus into the IVC.

Umbilical artery

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

## 5.5 Radial Artery Access

Positioning and landmarks

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

Technique:

- The needle entry superficially with a 30 ° angle without a syringe attached
- Advance until jerks of pulsating blood will flow

## References

1. Perry SB. Manual techniques of cardiac catheterization: vessel entry and catheter manipulation. In: Lock JE, Keane JK, Perry SB, editors. Diagnostic and interventional catheterization in congenital heart disease. New York: Springer; 2000. p. 13–35.
2. Bergersen L. Vascular access. In: Bergersen L, Foerster S, Marshall AC, Meadows J, editors. Congenital heart disease the catheterization manual. New York: Springer; 2009. p. 21–34.
3. Seto AH, et al. Real-time ultrasound guidance facilitates femoral arterial access and reduces vascular complications: FAUST (Femoral Arterial Access With Ultrasound Trial). JACC Cardiovasc Interv. 2010;3(7):751–8.

# Hemostasis

6

Zakhia Saliba, Ramy C. Charbel, and Tarek Smayra

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

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

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

## 6.1 Manual Compression (MC)

MC remains the gold standard and the most commonly used technique around the world for achieving hemostasis after a femoral vessel puncture (Video 1). This is due to its effectiveness, tolerability, good safety profile, and short learning curve. It is also a cheap maneuver that does not leave a foreign body in or around the vessel.

A successful hemostasis using manual compression is dictated by a set of steps to follow, as early as positioning the patient and locating the vascular access site (see Chapter "Vascular Access").

Usually, the sheath is removed immediately at the end of the procedure.

When removing the sheaths, the operator should ensure arterial hemostasis first and then remove the venous sheath at the end of the procedure. This approach decreases the risk of arteriovenous fistula formation and also provides a useful means of treating a vagal reaction should the peripheral IV inadvertently be lost.

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

When pulling the sheath, first prepare the area and clear away equipment, syringes, and tubing. Aspirate and flush the sheath with saline to clear any thrombi.

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

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

Monitor vital parameters and keep track of arterial pulsation with a pulse oximeter placed on the distal limb.

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

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

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

Maintain adequate compression (enough to counter vascular pressure without compromising distal pulsation) for

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_6) contains supplementary material, which is available to authorized users.

Z. Saliba (✉)
Pediatric Cardiology, Hôtel-Dieu de France Hospital, Ashrafieh, Beirut, Lebanon

R. C. Charbel
Pediatric Department, Hotel Dieu De France Hospital, Ashrafieh, Beirut, Lebanon

T. Smayra
Radiology Department, Hotel Dieu De France Hospital, Ashrafieh, Beirut, Lebanon


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_6

10 min. Apply more gentle pressure for 2–5 min and then apply a pressure dressing while keeping light compression.

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

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

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

The protamine dose to neutralize 100 units of heparin according to the elapsed time since the heparin was given is as follows:

<30 min—1 mg
30–120 min—0.5–0.75 mg
>120 min—0.25–0.375 mg

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

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

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

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

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

**Video 1** A 3-year-old boy underwent percutaneous closure of the patent ductus arteriosus. The left femoral artery and vein were cannulated with a 4 and 5 Fr Terumo introducers, respectively. At the end of the procedure, and before the introducers are retrieved, the area is cleared from equipment, syringes, and tubing. The operator flushes both introducers with saline to clear any thrombi. Gentle pressure is applied using a sterile gauze, taking care not to crush the sheath and "strip" clot into the artery. The arterial sheath is removed first and then the venous one to reduce the risk of arteriovenous fistulae formation. While pressure is maintained, the distal pulse is closely monitored with a pulse oximeter, and pressure is calibrated accordingly to maintain adequate distal limb perfusion (MP4 27873 kb)

## 6.2 Vascular Closure Devices (VCDs)

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

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

All VCDs reduce the time to obtain hemostasis. Each device has its own unique insertion technique (Table 6.1), failure mode, and complications.

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

### 6.2.1 Passive Vascular Closure Devices

- Hemostasis pads (Fig. 6.1)
- Compression devices: FemoStop (St. Jude Medical, USA), SafeGuard (Merit Medical) (Fig. 6.2), and ClampEase (Pressure Products Inc., USA)
- Compression devices are intended to replace humans with mechanical compression, either by using an inflatable bubble (FemoStop) or a clamp (ClampEase).
- The advantages and disadvantages of the several common arterial closure devices are summarized in Table 6.2.

**Table 6.1** This table shows the different mechanisms of action of the different vascular closure devices intended for femoral vascular hemostasis

| | AngioSeal | Cordis Exoseal | MYNX | Abbot StarClose | Abbot PerClose |
|---|---|---|---|---|---|
| Bio absorbable | ✓ | ✓ | ✓ | | |
| Active/mechanical sealing | ✓ | | | ✓ | ✓ |
| Non suture-based closure | ✓ | ✓ | ✓ | ✓ | |
| Extra-vascular | | ✓ | ✓ | ✓ | |
| No manual comp needed | ✓ | | | ✓ | ✓ |

### 6.2.2 Active Vascular Closure Devices

- Cardiva Catalyst (Cardiva Medical Inc., USA) (Fig. 6.3)
- Collagen plug device: Angio-Seal (St. Jude Medical, USA) (Fig. 6.4—Video 2)
- Collagen plug device: Mynx (Access Closure, USA) (Figs. 6.5 and 6.6)
- Polyglycolic Acid (PGA) plug device: Exoseal (Cordis Corporation, USA) (Fig. 6.7—Video 3)
- FISH (Morris Innovative, USA) (Fig. 6.8)
- Clip device: Starclose (Abbott Vascular, USA) (Fig. 6.9)
- Suture devices: Perclose (Fig. 6.10), Proglide (Fig. 6.11), and Prostar (Abbott Vascular, USA) (Fig. 6.12)

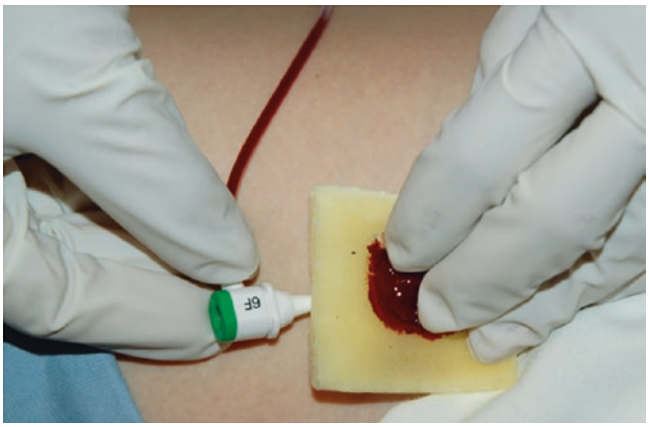


**Fig. 6.1** Hemostasis pads are covered with procoagulant material improving hemostasis and coagulation. They are used in conjunction with MC to improve both the patient and the physician's comfort, rather than to shorten the immobilization and bed rest period

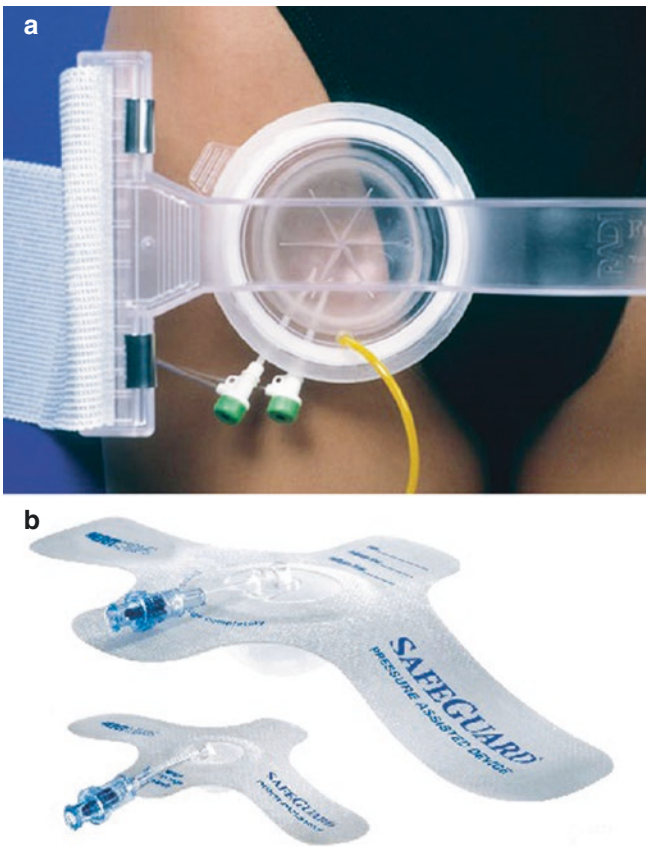
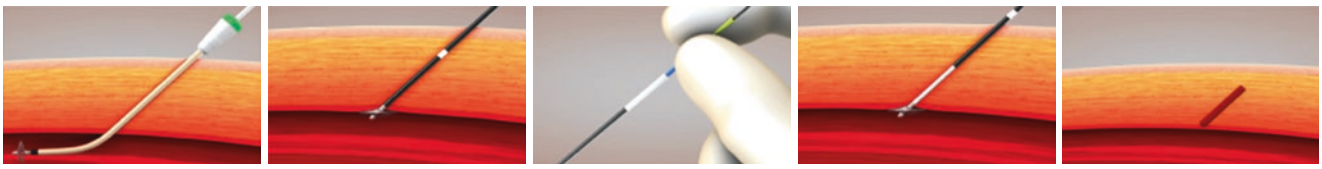



**Fig. 6.2** (**a**) The FemoStop compression device (St. Jude Medical) is an air-filled clear plastic bubble that molds to skin contours. It is held in place by straps passing around the hip. It is intended to replace humans' mechanical compression after femoral access. It is used most often for patients in whom prolonged compression is anticipated or whose bleeding persists despite prolonged manual compression. The duration of FemoStop compression and time to removal of the device varies depending on the patient and staff protocols. (**b**) The Safeguard pressure-assisted device (Merit Medical) delivers adjustable active compression and enables immediate pressure adjustment, maintains consistent pressure on the site during patient recovery as well as patient positioning and transport, and provides site management control for non-compliant patients. Additionally, it facilitates site assessment through a clear window without removing the device

**Table 6.2** This table summarizes the properties, advantages, and disadvantages of some of the active vascular closure devices on the market

| Device | On the market | Mechanism | Advantages | Disadvantages | Sheath Size | Ipsilateral access <90 days |
|---|---|---|---|---|---|---|
| AngioSeal (St Jude Medical) | 1997 to present | Collagen and suture-mediated | Secure closure, long track record | Intra-arterial component, possible thromboembolic complications, infection related to wick | 6 and 8 Fr | 1 cm higher |
| Perclose (Abbott Vascular) | 1997 to present | Suture-mediated | Secure closure | Intra-arterial component; steep learning curve; device failure may require surgical repair | 5–8 Fr | No restrictions |
| StarClose (Abbot Vascular) | 2005 to present | Nitinol clip | No intra-arterial component | Adequate skin tract needed to prevent device failure | 5–6 Fr | Not fully established |
| Mynx (AccessClosure) | 2007 to present | PEG hydrogel plug | No intra-arterial component, potential use in PVD | Possible intra- arterial injection of sealant | 5–7 Fr | No restrictions |



No need for sheath exchange: deploys through existing procedural sheath

Provides temporary hemostasis with minimal flow disruption by deploying low profile disc

Fully-integrated design with unique release mechanism allows for safe and easy collagen exposure

Exposed collagen rapidly hydrates to provide tamponade

Disc is collapsed and removed leaving nothing in intravascular space

**Fig. 6.3** The Cardiva Catalyst device is used in conjunction with manual compression. It is effective for both diagnostic and interventional procedures with sheath up to 7 Fr. It can be inserted through the existing vascular sheath. Once the tip is within the arterial lumen, the 6.5 mm umbrella-shaped disk coated with protamine sulfate is deployed. The sheath is then gently removed, and the disk is pulled against the arterial wall, where it is held in place by a tension clip. After 15 min (120 min for interventional cases) the device is withdrawn and a slight compression is applied for 5 min. The device is compatible with most patients and has been successfully used in pediatric patients

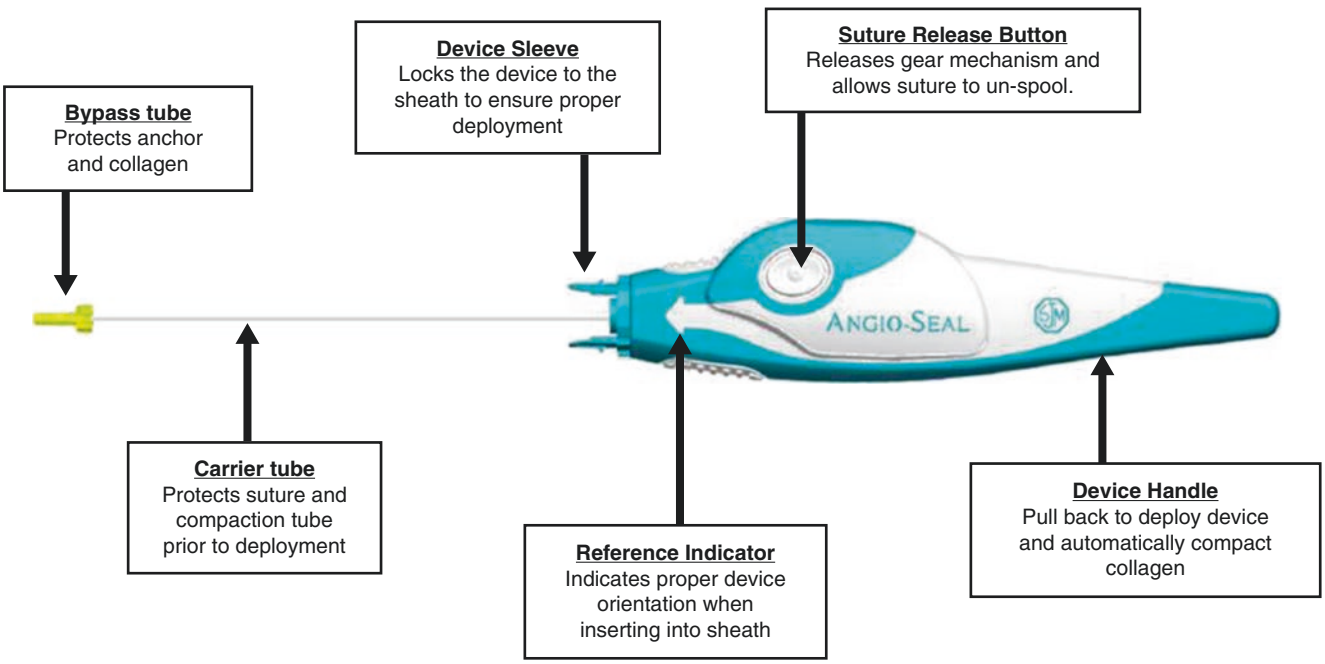



**Fig. 6.4** A three-component device, the Angio-Seal includes a small and flat anchor, a collagen plug, and a suture. It is inserted through a specially designed 6 Fr or 8 Fr sheath with an arteriotomy locator. Angio-Seal achieves hemostasis by anchoring a collagen plug to the anterior vessel wall through a sheath delivery system. The resorbable collagen plug induces platelet activation and aggregation, releases coagulation factors, and eventually results in the formation of fibrin and in thrombus generation. Once proper positioning within the arterial lumen is secured, the sheath is firmly held in place, and the guide wire is removed along with the arteriotomy locator. The device is inserted into the sheath until it snaps in place. The anchor is then deployed and pulled back against the arterial wall. This positions the collagen plug just outside the arterial wall. The suture is then cut below skin level, leaving behind the anchor, collagen plug, and suture that will all dissolve within 2–3 months

**Video 2** After percutaneous VSD closure through an 8 Fr femoral arterial introducer, the Angio-Seal device was used to achieve hemostasis. The femoral introducer is exchanged with the dedicated sheath and its arteriotomy locator using a guide wire. Once securely positioned in the vascular lumen, the sheath is firmly held in place, and the guide wire is removed along with the locator. The device is inserted into the sheath until it snaps in place. The anchor is then deployed and pulled back against the arterial wall. This positions the collagen plug just outside the arterial wall. The suture is then cut below skin level, leaving behind the anchor; collagen plug and suture will all dissolve within 2–3 months (MP4 6630 kb)

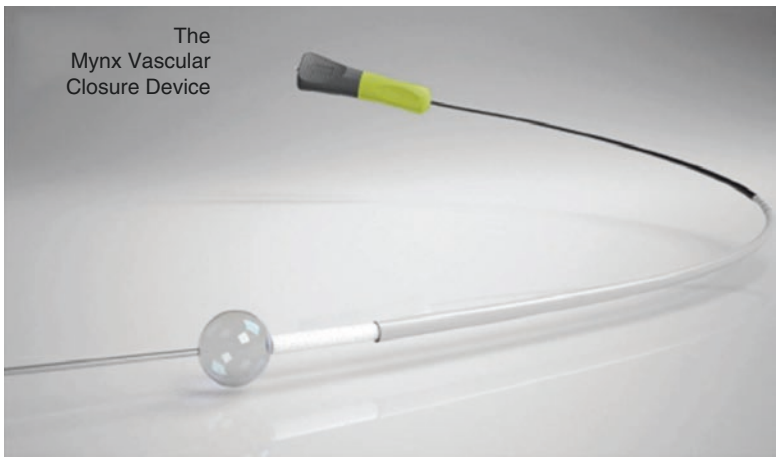



**Fig. 6.5** The Mynx Vascular Closure Device (AccessClosure, Mountain View, CA) features a polyethylene glycol sealant ("hydrogel") that deploys outside the artery while a balloon occludes the arteriotomy site within the artery (MYNX 2). The Mynx device is inserted through the existing procedural sheath, and a small, semicompliant balloon is inflated within the artery and pulled back to the arterial wall, serving as an anchor to ensure proper placement. The sealant is then delivered just outside the arterial wall where it expands to achieve hemostasis. Finally, the balloon is deflated and removed through the tract, leaving behind only the expanded, conformable sealant

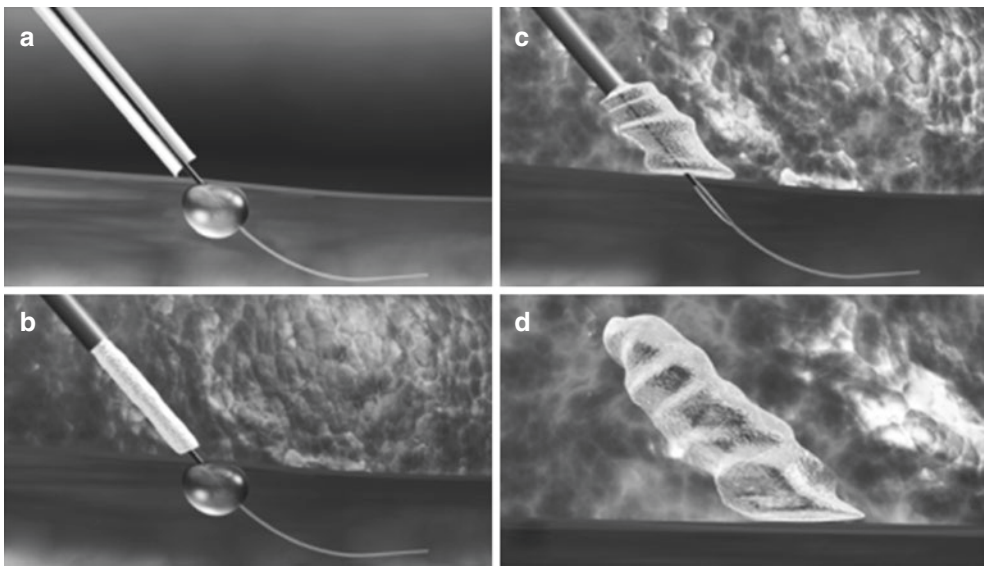



**Fig. 6.6** Once the balloon is inflated within the arterial lumen and pulled against the arterial wall ensuring proper positioning (**a**), the sealant is placed in the extravascular tissue track (**b**, **c**). The sealant expands to maintain hemostasis after the device is withdrawn (**d**)


**Fig. 6.7** The Exoseal device (Cordis Corporation, Miami Lakes, Florida) delivers a synthetic, bioabsorbable plug to the extravascular space adjacent to the arteriotomy using visual guidance. No anchor is left inside the artery. The Plug exhibits partial to advanced absorption at 30 days, with complete absorption between 60 and 90 days post-implant

**Video 3** After percutaneous closure of a coronary fistulae through a 7 Fr femoral arterial introducer, the Exoseal device was used to achieve hemostasis. The Exoseal Vascular Closure Device (VCD) consists of a plug applier and an absorbable plug. The plug applier consists of a handle assembly and a delivery shaft. The absorbable plug is fully enclosed in the distal portion of the delivery shaft. The plug applier positions and deploys the absorbable plug to the extravascular surface of the femoral artery access site through the existing 7 French procedural vascular sheath introducer without the need for a vascular sheath introducer exchange before device deployment. Insert the distal end of the delivery shaft into the vascular sheath introducer hub using the right hand to support and guide the Exoseal VCD. The indwelling sheath introducer must allow the bleed-back port to extend beyond the distal tip of the sheath introducer. Orient the Exoseal VCD such that the indicator window on the handle assembly is facing upward. Advance the delivery shaft into the proximal end of the vascular sheath introducer to the marker band, keeping the Exoseal VCD oriented upward and advancing at the angle of the tissue tract. Without advancing the Exoseal VCD, use the left hand to retract the vascular sheath introducer proximally toward the handle assembly. While still holding the Exoseal VCD stationary, use the left hand to continue retracting the vascular sheath introducer proximally. Once the hub of the sheath introducer engages with the indicator wire cowling, retract them together using one fluid motion until they lock into position against the handle assembly and an audible "click" signifies proper connection. The indicator wire will automatically deploy at this point. Observe pulsatile flow from the bleed-back indicator. Using the left hand, slowly retract the Exoseal VCD and vascular sheath introducer at the angle of the tissue tract until pulsatile flow has significantly slowed or stopped from the bleed-back indicator. While holding the Exoseal VCD in the right hand, making sure the thumb is not placed on the plug deployment button, continue retracting the Exoseal VCD and vascular sheath introducer very slowly (controlling retraction with the left hand) until the graphic pattern in the indicator window changes to a solid black color, at which point the plug is correctly positioned for deployment. Use the left hand to anchor the vascular sheath introducer and the Exoseal VCD in a stationary position. Press the deployment button to deploy the plug, ensuring that the deployment button is fully depressed and flushed against the handle assembly. The indicator wire will automatically withdraw and the plug will be deployed. Approximately 1–2 s after depressing the deployment button, retract the Exoseal VCD and vascular sheath introducer as a unit until the system is fully removed from the patient and apply light, nonocclusive pressure to the wound site. Wipe the entry site and evaluate for hemostasis. If hemostasis has not occurred, continue applying light manual pressure to achieve hemostasis (MP4 34620 kb)

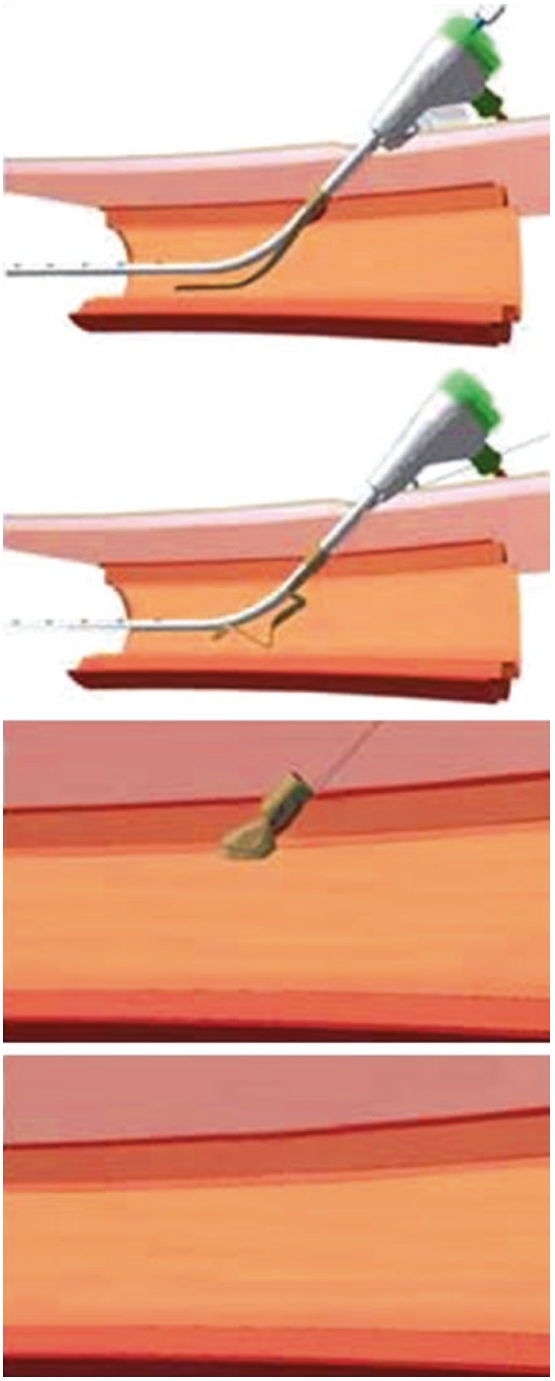


**Fig. 6.8** FISH uses an absorbable extracellular matrix "patch" made from porcine intestinal submucosa. It is indicated for procedures using 5–8 Fr sheaths. (1) Position the SIS: Place the FISH device into the vessel over the guide wire. A release wire that travels from within the proximal hub to the tip of the patch will hold the patch until it is placed in the vessel wall. Upon removal of the release wire, the sheath is advanced into working position. (2) Advance the SIS: With the SIS positioned at the arteriotomy, the compression tab, located at the base of the hub, can be removed and pulled until resistance is felt. This allows the ribbon of SIS to form a plug at the vessel wall. (3) Sheath removal: The FISH sheath is removed the same way a standard sheath is removed. Place firm downward pressure at the base of the hub and remove the sheath. The plug seeds itself into the artery and provides effective hemostasis. (4) Remodels host tissue: Within 30 days, the SIS completely remodels the host tissue—resisting infection inflammation and scar tissue formation

## 6.3 Hemostasis in Non-femoral Access Sites

### 6.3.1 Jugular Access

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

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

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

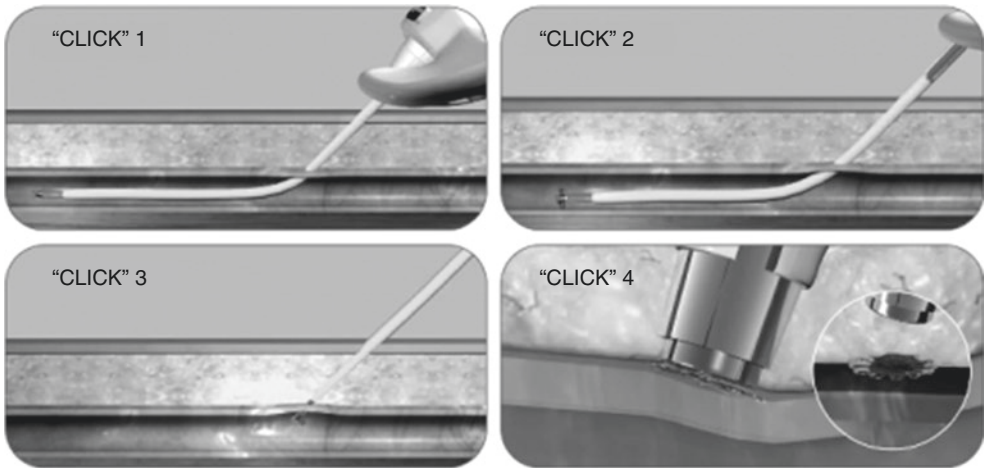



**Fig. 6.9** Starclose uses a 4 mm nitinol clip implant to reach hemostasis. It is designed for invasive procedures using 5–8 Fr arteriotomies. Click 1: The StarClose device is in the common femoral artery. Click 2: The locator "wings" are deployed. Click 3: The device is withdrawn until the "wings" abut the arterial wall. Click 4: The nitinol clip is deployed, which grasps the arteriotomy edges and pulls the vessel wall together to achieve hemostasis

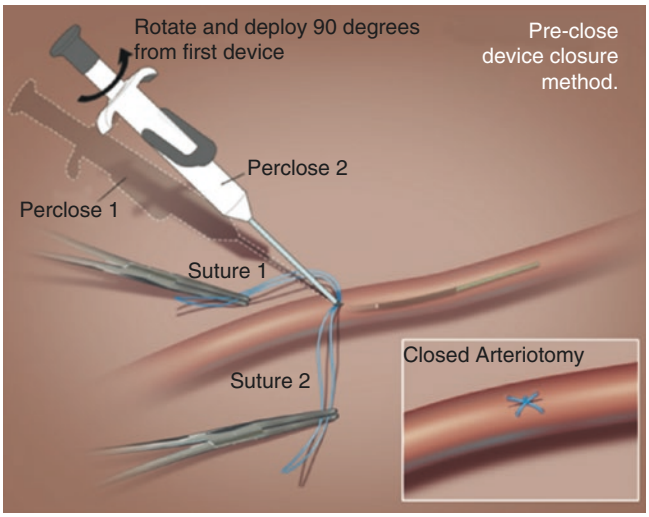



**Fig. 6.10** Perclose offers suture-mediated VCD and has been successfully used in pediatric cases. The hemostasis procedure requires several steps: positioning the device, needle deployment, suture capture, and needle removal. Vessel closure starts by replacing the vascular introducing sheath with the Perclose device. Guide wire 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 guide wire are then removed as tension is maintained on the knot to achieve hemostasis

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

### 6.3.2 Umbilical Access

In newborns of 3–5 days of life, umbilical arteries and vein are usually available for vascular access.

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

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

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

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

### 6.3.3 The Transhepatic Access

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

The transhepatic vessels can accommodate relatively large sheaths.

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

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

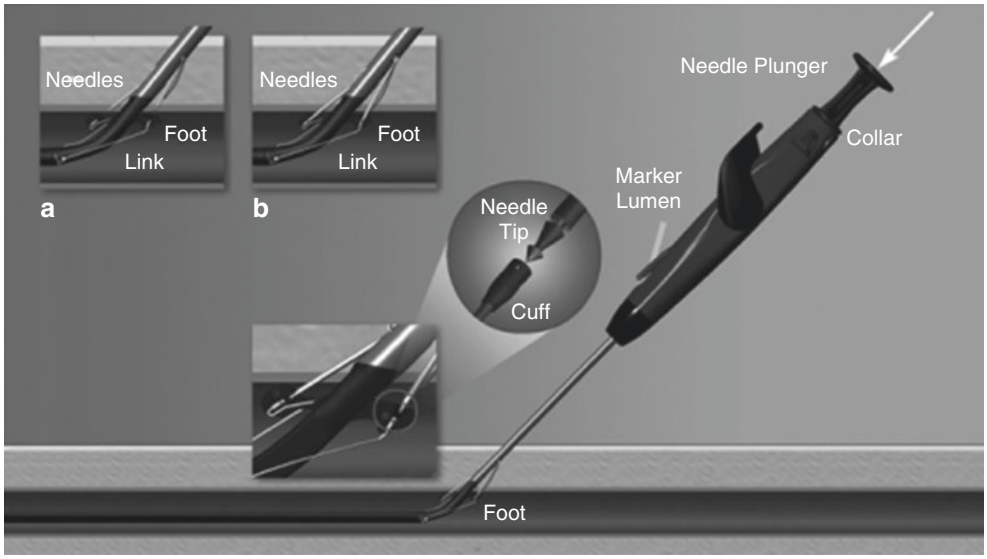



**Fig. 6.11** Blood return in the marker lumen indicates that Proglide device is in the artery. Then, the "feet" are deployed, and the device is withdrawn until the "feet" abut the arterial wall and blood return will stop. The needle plunger is depressed, which deploys the needles (**a**) through the arterial walls and into cuffs on the "feet" (**b**) which completes the suture loop. The device is then withdrawn and a knot is tied in the suture and pushed down to the arteriotomy site for hemostasis

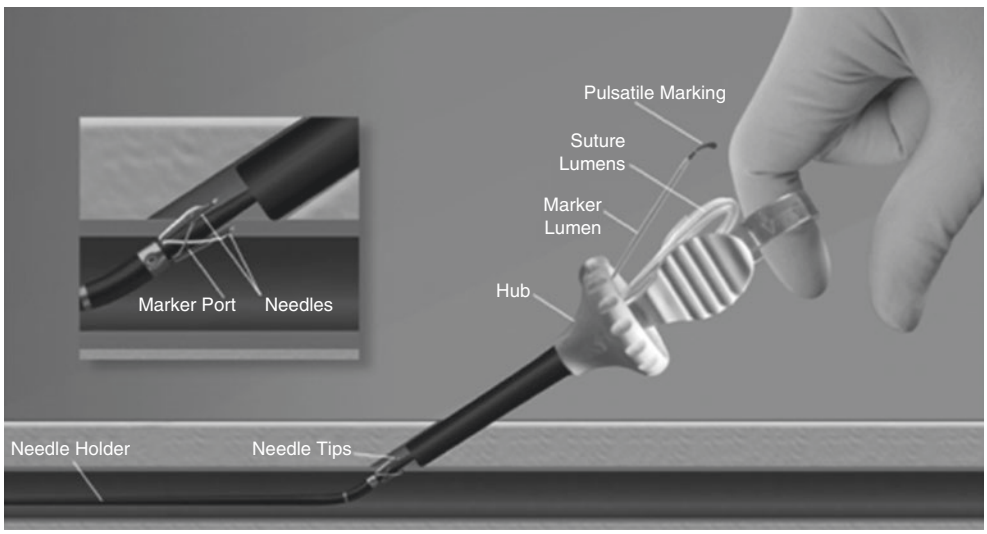



**Fig. 6.12** Once proper placement is confirmed visually, the right hand rotates and pulls the handle to deploy the needles, which are pulled through the arterial wall and collected through the device hub. A knot is tied in each suture and pushed to the arteriotomy site, forming two suture loops for hemostasis

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.

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

The Gelfoam plugs are formed from small Gelfoam strips rolled to fit into the delivery sheath. The plug is advanced to the end of the sheath, which is then withdrawn while deploying the Gelfoam plug. Alternatively, an MReye Embolization Coil (Cook Inc., USA) (Fig. 6.13) or an Amplatzer Vascular Plug (AGA Medical co., USA) can be used for this purpose.

### 6.3.4 Surgical Vascular Access (Video 4)

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

Sheaths are inserted through the artery or vein and secured with 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).

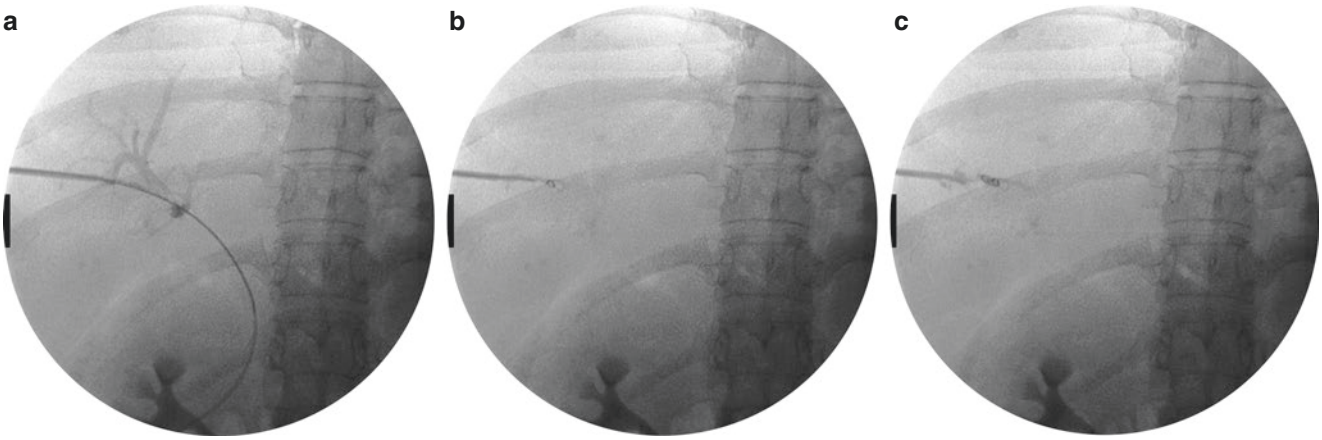



**Fig. 6.13** Hemostasis after transhepatic catheterization: (**a**) A small volume (1–2 mL) of the contrast is injected to visualize the hepatic veins. (**b**) An MReye Embolization Coil (0.035 in × 4 cm × 3 mm) is advanced to the end of the sheath and deployed between the hepatic vein and the liver capsule to minimize the risk of bleeding. (**c**) The intraparenchymal position of the sheath tip is documented by contrast injection. 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

### 6.3.5 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.

### 6.3.6 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.

**Video 4** A 15-year-old boy (65 kg) underwent percutaneous stenting for a native aortic coarctation. He was already on warfarin for a mechanical aortic valve. Due to the large caliber of the required sheath, vascular access was obtained surgically. A small incision was made at the level of the right groin. The subcutaneous tissue was dissected and the femoral vessels exposed. We performed a purse-string suture using a Prolene 6/0 on the right femoral artery and secured it with a snugger. After that, an 18G cannula was inserted inside the purse string and followed by a guide wire over which we advanced a 12 Fr sheath. At the end of the procedure, the surgeon removed the catheters and the sheath. Hemostasis was achieved by tying the purse-string suture. The wound is then closed in layers. Distal pulse is checked and regularly monitored (MP4 344473 kb)

# Access Complications and Management

7

Zakhia Saliba, Elie B. Sawan, and Kamal Hachem

## 7.1 Access Complications and Management

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

Arterial vessels in young patients are more fragile and prone to vasospasm or subadventitial injury if dilated extensively. Moreover, multiple sites of access may be required in structural heart disease, while repeated cardiac interventions and multiple cardiac surgeries may result in incremental loss of sites. Thus, efforts should be made for preservation. It is essential to be familiar with the predisposing factors affecting the outcome of the procedure to avoid complications. These factors may be divided in three points:

- Operator-related factors: experience and the in(ability) of using ultrasound for access
- Patient-related factors: younger age, anticoagulant medication, bleeding disorders, polyglobulia, connective tissue disorders, multiple cardiac surgeries or repeated catheterization using the same VAS
- Technical factors: the use of larger sheath in smaller patient, complex and long exposure, unplanned access, simultaneous venous and arterial ipsilateral femoral access, insufficiently immobilized patient and the use of vascular closure devices (VCD) for haemostasis

Special preventive haemostasis measures should be considered for patients with one or more of the previous conditions.

### 7.1.1 Preventive Measures

In order to prevent complications, some measures may be recommended:

- Stop blood thinners at least 4–5 days before the procedure.
- Assume a deep sedation for complete VAS immobilization.
- Use an accurate disinfectant for an aseptic environment.
- Avoid ipsilateral arterial and venous femoral access.
- Whenever possible, obtain vascular access using ultrasound guidance.
- Introduce the guidewire without resistance and check its position with fluoroscopy.
- When in doubt about the guidewire position, exchange the needle with a cannula and perform hand-injected angiogram to verify the vessel anatomy.
- Adapt the stepwise predilatation if introducing larger dilator/sheath.
- Check ACT regularly during a long procedure for adequate heparinization.

It is important to ensure a good haemostasis (refer to the Chap. 6) and a close patient follow-up in the hours post catheterization and for a couple of weeks later.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_7) contains supplementary material, which is available to authorized users.

Z. Saliba (✉) · E. B. Sawan
Pediatric Cardiology, Hôtel-Dieu de France Hospital, Ashrafieh, Beirut, Lebanon

K. Hachem
Radiology Department, Hotel Dieu De France Hospital, Ashrafieh, Beirut, Lebanon


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_7

### 7.1.2 Complications Related to the VAS and Techniques

#### 7.1.2.1 Femoral Access

Percutaneous puncture of the femoral artery and vein for cardiac catheterization is commonly adopted because of the larger diameter of those vessels and the better accessibility.

Any puncture below the femoral bifurcation may lead to haematoma, pseudoaneurysm, arteriovenous fistula, dissection and lymphocele (Fig. 7.1), whereas a higher puncture may perforate the inferior epigastric artery or a posterior wall and cause retroperitoneal haemorrhage. In addition, other complications may occur such as arterial or venous occlusion, femoral neuropathy and systemic or pulmonary embolism.

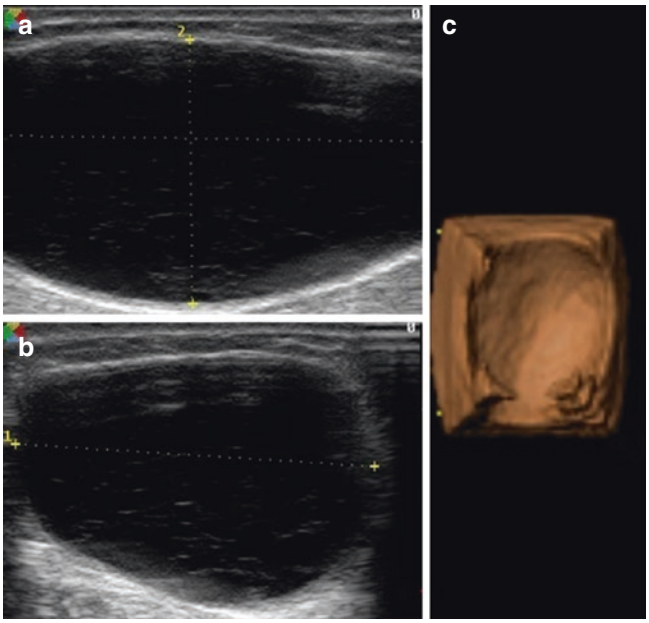



**Fig. 7.1** Groin lymphocele. Ultrasound image of a groin lymphocele diagnosed in a 6-year-old boy (22 kg) who presented for a non-pulsatile painful groin mass 1 week after right femoral artery catheterization. Grayscale image in long (**a**) and short (**b**) axis views of a 2 × 5 cm anechoic cystic collection. The lymphocele in a 3D reconstructed image (**c**)

#### Haematoma

Arterial bleeding is a common VAS complication where blood collects in the soft tissue (Fig. 7.2). When the arterial puncture is below the femoral bifurcation, the compression may be less effective without the help of the femoral head (Video 1).

Compression and immobilization of the leg are the main keys for managing a haematoma. Further actions include marking the area to assess any change in size, providing hydration, checking the haematocrit level, maintaining/prolonging bed rest and stopping antiplatelet medication and anticoagulation if possible, as well as blood transfusions in case of significant drop in haemoglobin level.

Although this is uncommon in children, the haematoma may sometimes require surgical evacuation or drainage. Haematomas usually resolve within a couple of weeks as the blood is depleted and absorbed into the tissue (Fig. 7.3).

**Video 1** Right pelvic haematoma. The right femoral artery was accessed with a 12 Fr long sheath for endovascular treatment of an aortic coarctation in a 13-year-old boy (40 kg). The patient was on warfarin for mechanical aortic valve. He presented for a non-pulsatile, painful, hardened and swollen groin. No specific treatment was needed. Cineloop in B-mode shows a right pelvic collection in short- and long-axis views. The fluid is anechoic and the mass is well defined without vascular flow inside on colour Doppler (MP4 2805 kb)

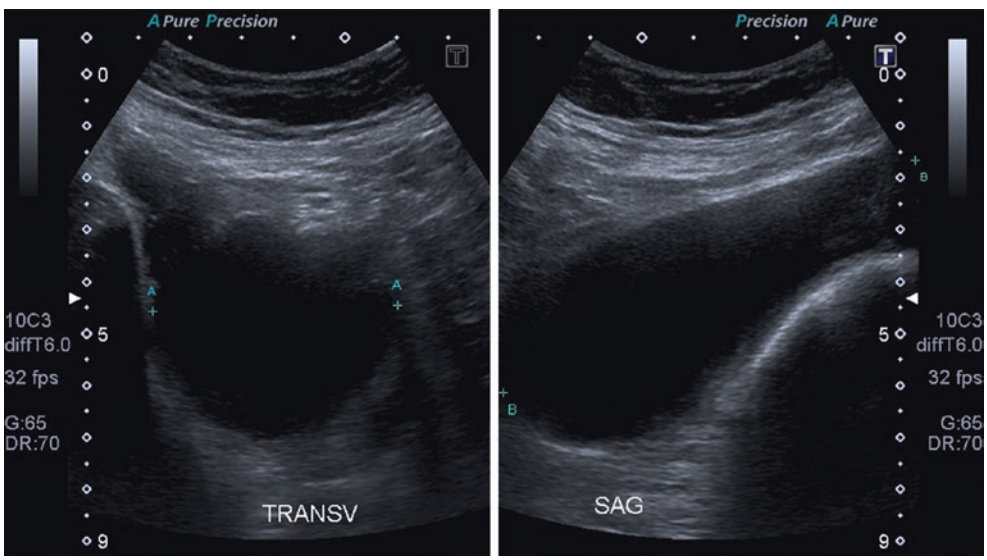



**Fig. 7.2** Right pelvic haematoma. The right femoral artery was accessed with a 12 Fr long sheath for endovascular treatment of an aortic coarctation in a 13-year-old boy (40 kg). The patient was on warfarin for mechanical aortic valve. He presented for a non-pulsatile, painful, hardened and swollen groin. No specific treatment was needed. Short (TRANSV) and long (SAG) axis 2D views in B-mode of the pelvic haematoma measuring 5 × 9 cm

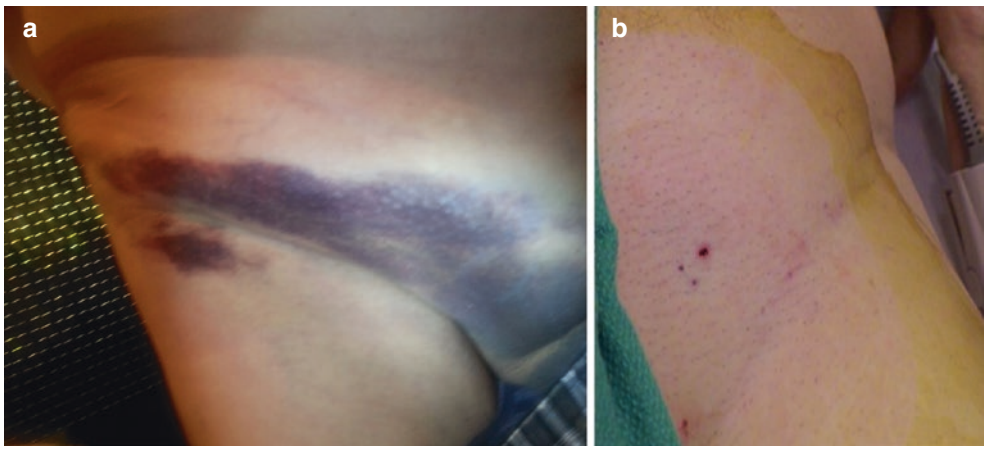



**Fig. 7.3** Groin skin bruises. (**a**) Extended ecchymosis and bruises of the right groin. Note the skin multicolouration indicating the resorption stage of the haematoma. (**b**) A good manual compression after sheath removal could avoid such a complication

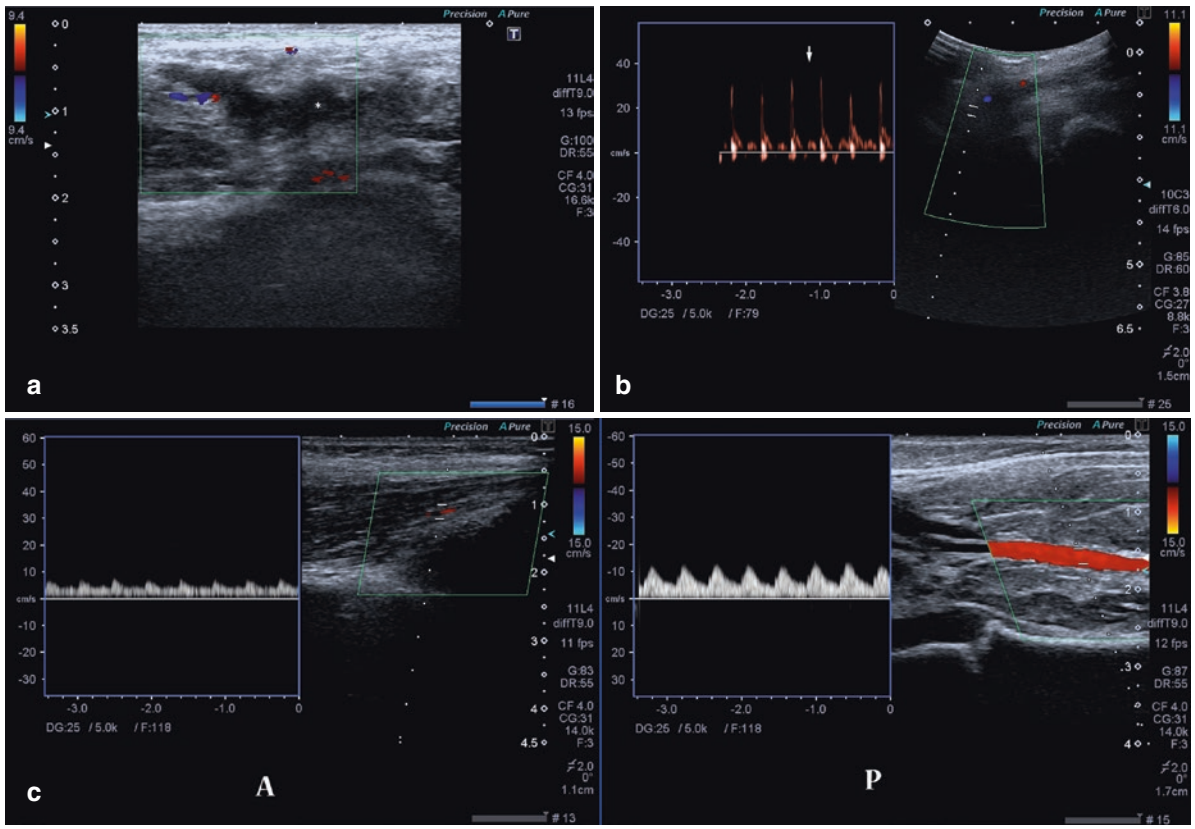



**Fig. 7.4** Ultrasound study of acute femoral artery and vein occlusion. (**a**–**c**) Acute femoral artery occlusion. A 9-year-old boy (30 kg) underwent a long and complicated percutaneous ductus arteriosus closure with an Amplatzer device. The left femoral artery and vein were cannulated with 5 and 6 Fr sheaths, respectively. At the end of the procedure the left lower limb was severely ischemic with no palpable peripheral pulses. (**a**) The colour Doppler shows absence of flow in the left common femoral artery (star). (**b**) The spectral analysis shows a weak and resistive flow (arrow) in the left external iliac artery signing an occlusion of the common femoral artery. (**c**) A tardus parvus flow in spectral analysis is detected in both anterior (A) and posterior (P) distal tibial arteries

### Acute Arterial or Venous Occlusion

After cardiac catheterization, the loss of pulse has been reported in infants weighing less than 14 kg even with prophylactic use of heparin. It is mainly related to vasospasm particularly in smaller patients (Figs. 7.4 and 7.5). The classical 6 Ps (pain, paralysis, paraesthesias, pulselessness, pallor and poikilothermia) are characteristic of impaired circulation.

Because of children's ability to develop a rich collateral network, less than 10% of patients will develop claudication or limb length discrepancy in the setting of asymptomatic persistent femoral arterial occlusion. To minimize risks, the patient's affected limb should be kept warm with close observation for 2–4 h.

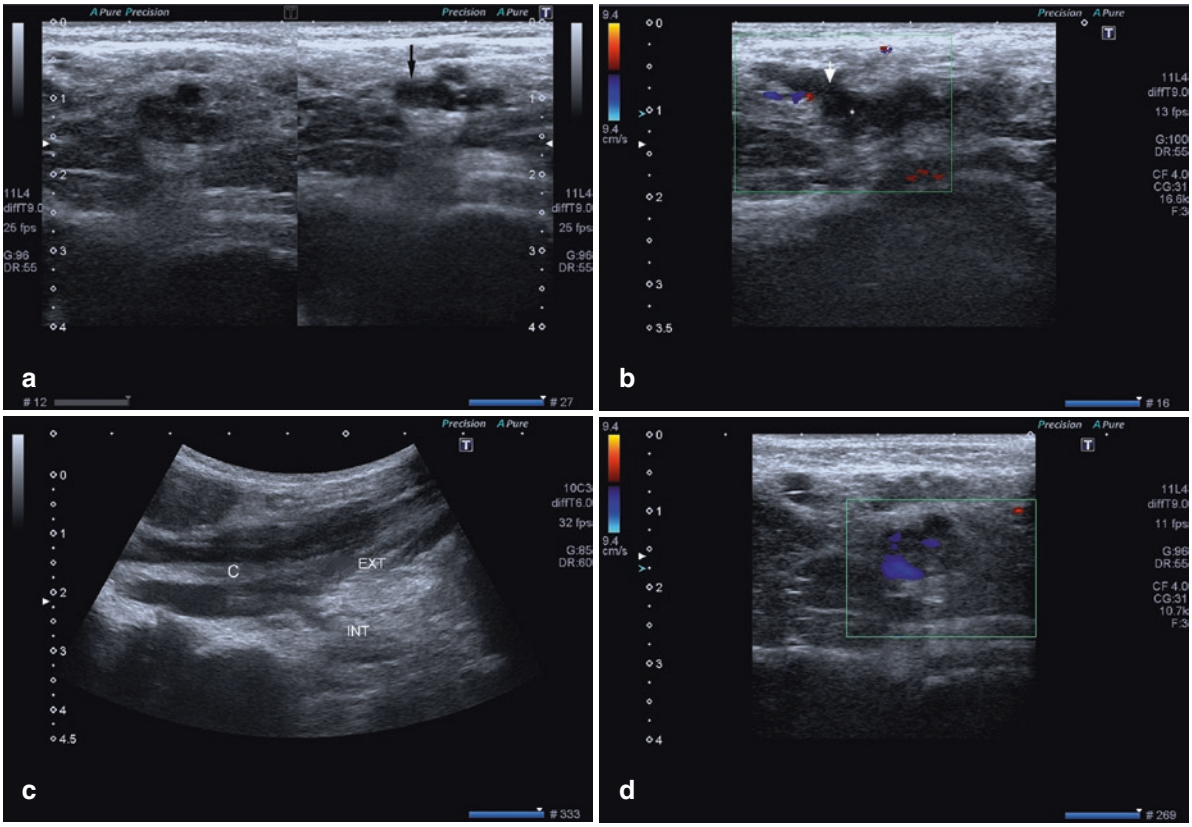



**Fig. 7.5** Ultrasound study of acute femoral artery and vein occlusion. (**a**–**d**) Acute femoral vein thrombosis. (**a**) The B-mode ultrasound shows an incomplete compression of the left common femoral vein (arrow). (**b**) The heterogeneous intraluminal thrombus (star) extends to the greater saphenous cross (arrow). (**c**) The thrombus is spread to the left internal (INT) and external iliac veins (EXT) up to the common iliac vein C. (**d**) The colour Doppler study shows a small patent residual lumen of the common femoral vein (in blue). The patient was started on heparin infusion at a rate of 500 units/kg/24 h. Four hours later, the pedal pulses remained non-palpable. Hence, thrombolysis was initiated with tissue type plasminogen activator with a bolus of 0.1 mg/kg followed by an infusion of 0.5 mg/kg/h for 2 h. We then noticed a clinical improvement and heparin was restarted for six additional hours

**Video 2a** (**a**–**d**) Recanalization of a chronic left common iliac vein occlusion (by Prof. Y. Boudjemline). A 5.5-year-old boy (22 kg) presented with severe and generalized left lower limb oedema. He had a long-lasting stay in the neonatal intensive care unit for prematurity. The opacification of the left iliac vein showed left common iliac vein occlusion with dilated collateral venous channels (MP4 1956 kb)

**Video 2b** (**a**–**d**) Recanalization of a chronic left common iliac vein occlusion (by Prof. Y. Boudjemline). A 5.5-year-old boy (22 kg) presented with severe and generalized left lower limb oedema. He had a long-lasting stay in the neonatal intensive care unit for prematurity. Normal right ilio-cavogram (MP4 1748 kb)

**Video 2c** (**a**–**d**) Recanalization of a chronic left common iliac vein occlusion (by Prof. Y. Boudjemline). A 5.5-year-old boy (22 kg) presented with severe and generalized left lower limb oedema. He had a long-lasting stay in the neonatal intensive care unit for prematurity. Repermeabilization of the left common iliac vein with a 4 mm coronary balloon dilatation through a right-left push-pull technique (MP4 2943 kb)

**Video 2d** (**a**–**d**) Recanalization of a chronic left common iliac vein occlusion (by Prof. Y. Boudjemline). A 5.5-year-old boy (22 kg) presented with severe and generalized left lower limb oedema. He had a long-lasting stay in the neonatal intensive care unit for prematurity. A final left common iliac venogram documented successful recanalization after stents implantation (five balloon-expandable stents, Valeo 9 × 36 and 8 × 36 in the proximal part and Valeo 7 × 26 in the distal segment) (MP4 863 kb)

**Video 3** Chronic inferior vena cava occlusion. A 12-month-old girl (11 kg) was admitted for redo pulmonary balloon valvuloplasty. The right femoral vein was cannulated, but there was a resistance to advance the wire. Hand contrast injection was done and showed drainage to the right atrium through a paravertebral collateral network to the right azygous vein and the right superior vena cava. The same injection through the left femoral vein showed the same venous flow interruption at the level of the iliac bifurcation. The pulmonary balloon valvuloplasty was completed through the right internal jugular vein (MP4 7324 kb)

### Chronic Arterial or Venous Occlusion

Chronic arterial occlusion has been reported in 5.5%–20% of various catheterization series, notably in children under 1 year of age, in longer procedures and studies with difficult access (Videos 2a, 2b, 2c, 2d and 3).

In asymptomatic chronic femoral artery occlusion, the follow-up should be on a regular basis. Once the symptoms appear, an operative intervention may be warranted.

In older children, 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 achieved lumen is adequate for catheterization but may not remain patent on the long run. Accordingly, stent implantation should be considered to achieve a more uniform lumen, large enough to provide long-term patency.

## Pseudoaneurysm

A pseudoaneurysm refers to a defect in the arterial wall, allowing communication of arterial blood with the adjacent extraluminal space (Videos 4a, 4b, 5 and Fig. 7.6).

**Video 4a** (**a**, **b**) Thrombin embolization of a left deep femoral artery pseudoaneurysm (by Prof. Y. Boudjemline). A 4-year-old boy (20 kg) presented with painful swelling on the left groin 24 h after coronary artery stenting for vasculitis. The ultrasound with Doppler study showed a left deep femoral artery pseudoaneurysm. A cross-over left common femoral artery angiogram confirmed a voluminous deep femoral artery pseudoaneurysm (MP4 1393 kb)

**Video 5** A pseudoaneurysm formation complicating a femoral artery catheterization. A 25-year-old man (75 kg) presented with a mass in the groin 10 days after femoral artery catheterization. A bruit was heard over the puncture site (MP4 2228 kb)

**Video 4b** (**a**, **b**) Thrombin embolization of a left deep femoral artery pseudoaneurysm (by Prof. Y. Boudjemline). A 4-year-old boy (20 kg) presented with painful swelling on the left groin 24 h after coronary artery stenting for vasculitis. The ultrasound with Doppler study showed a left deep femoral artery pseudoaneurysm. A balloon occlusion of the artery was done, and thrombin was percutaneously injected after direct ultrasound-guided puncture of the aneurysm. The selective left deep femoral artery angiogram after thrombin injection showed reduced aneurismal circulation (MP4 2607 kb)

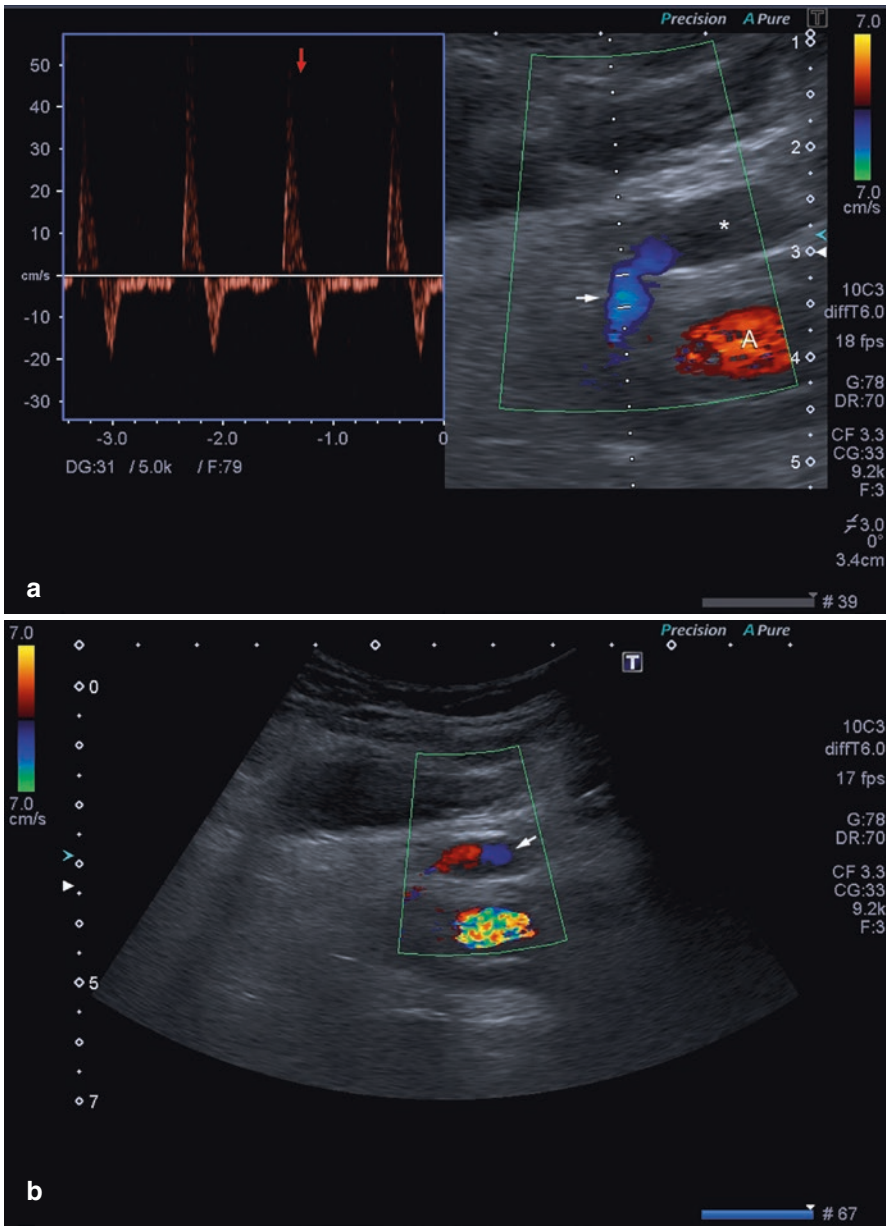



**Fig. 7.6** A 25 year-old man (75 kg) presented with a mass in the groin 10 days after femoral artery catheterization. A bruit was heard over the puncture site. (**a**) The grayscale ultrasound study of the right groin shows the pseudoaneurysm as a small hypoechoic saccular collection (star) in the region of the puncture site facing the common right femoral artery (A). Colour Doppler imaging reveals the pseudoaneurysm containing circulating blood and communicating with the lumen of the artery via a neck (arrow). Pulsed wave Doppler imaging detects the "to and fro" typical blood flow in the neck of the pseudoaneurysm. (**b**) Due to the turbulent forward and backward flow, a characteristic yin-yang sign (arrow) is seen on colour flow inside the pseudoaneurysm

Possible causes include difficulty with arterial cannulation, inadequate compression after sheath removal or inadequate haemostasis. It may occur if the arterial puncture is below the femoral bifurcation so the femoral head is not accessible to help with compression.

Clinical signs include swelling at insertion site, large and painful haematomas, ecchymosis, a 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 unusual and inversely proportional to the haematoma size. Nerve compression can result in limb weakness that takes few weeks to resolve.

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

#### Arteriovenous Fistula

Arteriovenous fistula is an abnormal connection between the arterial and venous system that bypass the normal anatomic capillary beds. Acquired AVF of the lower extremity is by far the most commonly occurring AVF due to the frequency of the groin as a site for percutaneous arterial and venous access (Videos 6a, 6b and 7).

The main causes include multiple access attempts, punctures above or below proper site level and impaired clotting.

Long-standing AVFs can lead to limb oedema, high-output cardiac failure or aneurysmal degeneration of the artery. AVF can be asymptomatic and result in bruit and/or thrill at VAS and in swollen and tender extremities (Fig. 7.7). In adults with pre-existing peripheral artery disease (PAD), it can lead to the onset or worsening of lower extremity ischaemic symptoms.

In most cases, the communication between the artery and vein will spontaneously seal. Whereas, others require ultrasound-guided compression or percutaneous device closure, but surgical repair is rarely needed especially in children.

#### Arterial Access Dissection

Iatrogenic arterial access dissections may complicate cardiac catheterization and can sometimes be a life-threatening event if not treated. It results from mechanical injury to the arterial wall during catheter or wire manipulation, passage or deployment of an interventional device, forceful injection of contrast medium or balloon dilatation or stenting.

Patients with hypertension, Marfan syndrome, congenitally unicuspid and bicuspid aortic valves, ostial coronary artery stenosis and cystic medial necrosis have been reported to be at higher risk of dissection.

Management is based on the extent of the dissection. In most focal or localized dissections, stenting should be performed as soon as possible (Videos 8a, 8b). From the technical viewpoint, soft-tip wires should be used when attempting to access the true lumen, and if the initial wire enters the false lumen, another soft-tip wire should be carefully manipulated into the true lumen (double-wire technique). On the other hand, extensive iatrogenic arterial and aortic dissection should be managed by surgical intervention.

#### Retroperitoneal Haemorrhage

Bleeding occurs behind the serous layer covering 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 diagnosed early. The computed tomography remains the gold standard diagnostic test.

Patients mostly complain of moderate to severe back pain, ipsilateral flank pain, vague abdominal or back pain and abdominal distention (usually not associated with evident hypotension and tachycardia). Late signs include ecchymosis and drop in haemoglobin and haematocrit levels.

Retroperitoneal haemorrhage is managed by hydration, performing serial blood cell counts, maintaining bed rest, interrupting anticoagulant and antiplatelet medications if necessary and performing blood transfusion if indicated. In rare cases, it may require surgical intervention. (Fig. 7.8).

### 7.1.2.2 The Internal Jugular Vein

It is generally used for access, especially in patients with interrupted inferior vena cava or after Glenn operation (Fig. 7.9).

Multiple attempts to obtain the jugular vein increase the rate of complications, and patients should be closely observed. Complications include puncture of the carotid artery, Horner's syndrome, air embolism, mediastinal haematoma, haemothorax and carotid jugular fistula.

When incidental carotid artery cannulation or haemothorax/pneumothorax has 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

**Video 6a** Percutaneous closure of iatrogenic femoral arteriovenous fistulae (by Prof. Y. Boudjemline). A 2.6-year-old boy (12 kg) presented with right femoral thrill 2 months after ductus arteriosus closure with an Amplatzer device. Doppler study documented right femoral arteriovenous fistulae. The cross-over right femoral artery angiogram showed direct fistulae between the right superficial femoral artery and the femoral vein. Using a 4-Fr short right arterial sheath, an Amplatzer Duct Occluder IIAS 3 × 2 was uneventfully placed in the fistulae (MP4 1367 kb)

**Video 6b** Percutaneous closure of iatrogenic femoral arteriovenous fistulae (by Prof. Y. Boudjemline). A 2.6-year-old boy (12 kg) presented with right femoral thrill 2 months after ductus arteriosus closure with an Amplatzer device. Doppler study documented right femoral arteriovenous fistulae. The final angiogram shows a patent femoral arterial flow with the device well placed and no residual shunt (MP4 1165 kb)

**Video 7** Arteriovenous fistula complicating transfemoral cardiac catheterization. Colour Doppler ultrasound shows communication between the common femoral artery and vein in a 7-year-old boy (22 kg), 1 month after device ductus arteriosus closure using an ipsilateral arteriovenous femoral access. The colour Doppler shows a perivascular artefact around the fistula due to high blood velocity flow through the fistula (MP4 1227 kb)

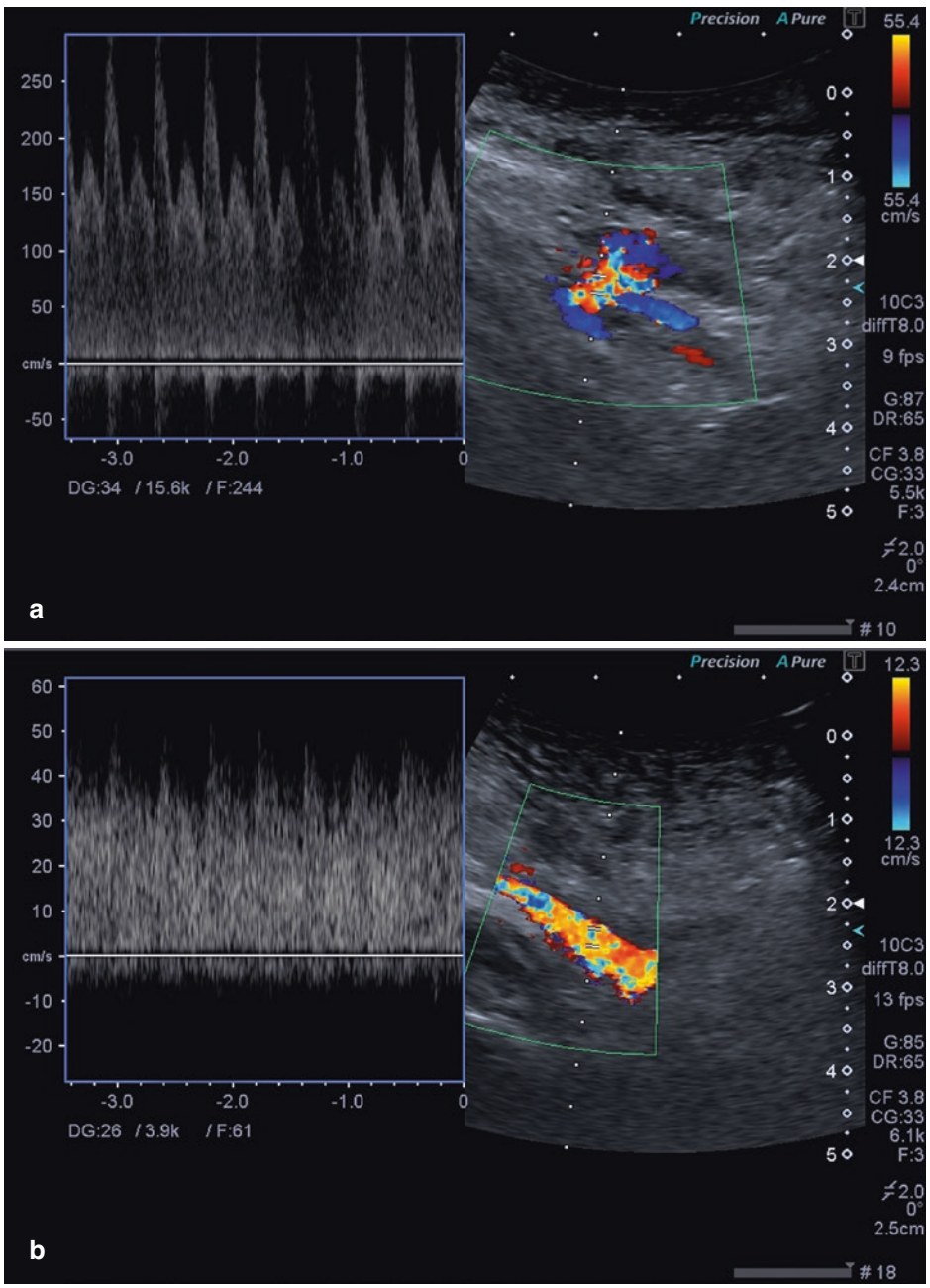
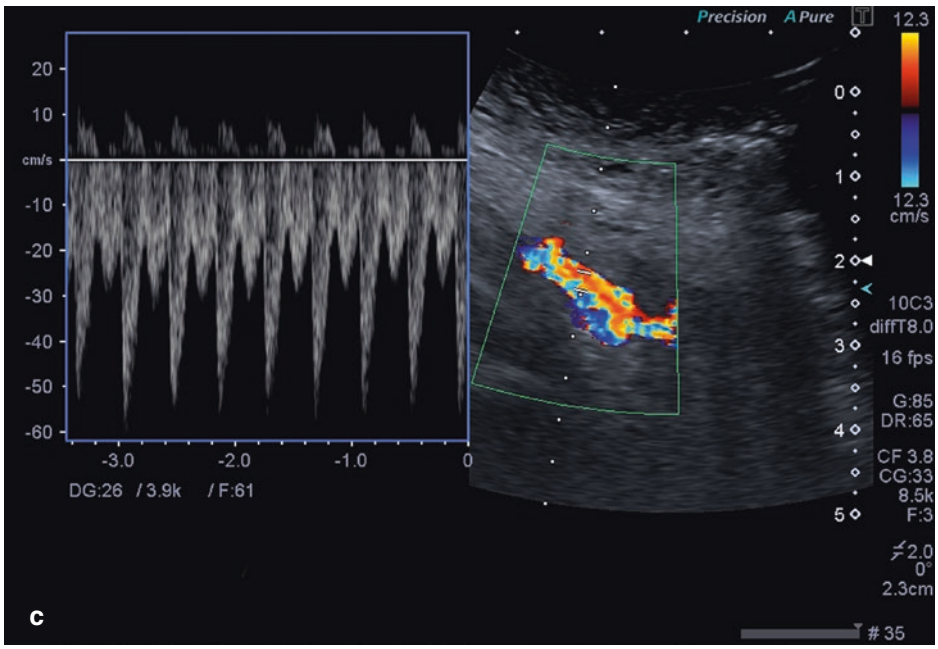



**Fig. 7.7** Arteriovenous fistula complicating transfemoral cardiac catheterization. Colour Doppler ultrasound shows communication between the common femoral artery and vein in a 7-year-old boy (22 kg), 1 month after device ductus arteriosus closure using an ipsilateral arteriovenous femoral access. (**a**) A high-velocity low resistive flow through the fistula is shown in spectral analysis. (**b**) The spectral analysis shows turbulent arterialized high-velocity flow in the efferent common femoral vein draining the fistula. (**c**) Note the high-velocity low resistive flow in the afferent common femoral artery feeding the fistula

**Fig. 7.7** (continued)




**Video 8a** Stenting of a right external iliac artery dissection (by Prof. Y. Boudjemline). An 8 month-old infant (9 kg) presented with severe right lower limb ischaemia 24 h after an arterial femoral diagnostic catheterization for cerebral artery malformation. The cross-over right iliac artery angiogram documented dissection of its proximal portion (MP4 3480 kb)

**Video 8b** Stenting of a right external iliac artery dissection (by Prof. Y. Boudjemline). An 8 month-old infant (9 kg) presented with severe right lower limb ischaemia 24 h after an arterial femoral diagnostic catheterization for cerebral artery malformation. Three Herculink 5 mm balloon premounted stents were successfully telescoped to cover the dissected segment and the post-stenting angiogram showed satisfactory recanalization of the iliac artery (MP4 2579 kb)

stop bleeding, and then monitoring the vital signs of the patient. The treatment is not specific and depends on clinical tolerance.

### 7.1.2.3 Transhepatic Access

Potential complications of transhepatic access include pneumothorax, haemorrhage, haemoperitoneum, haemobilia, cholangitis, liver abscess, sepsis and hepatic vein thrombosis. On the other hand, intraperitoneal haemorrhage may occur requiring laparotomy.

Ultrasound-guided catheterization of the hepatic veins during transhepatic access is an important progress that may decrease the risk of puncturing other structures during access

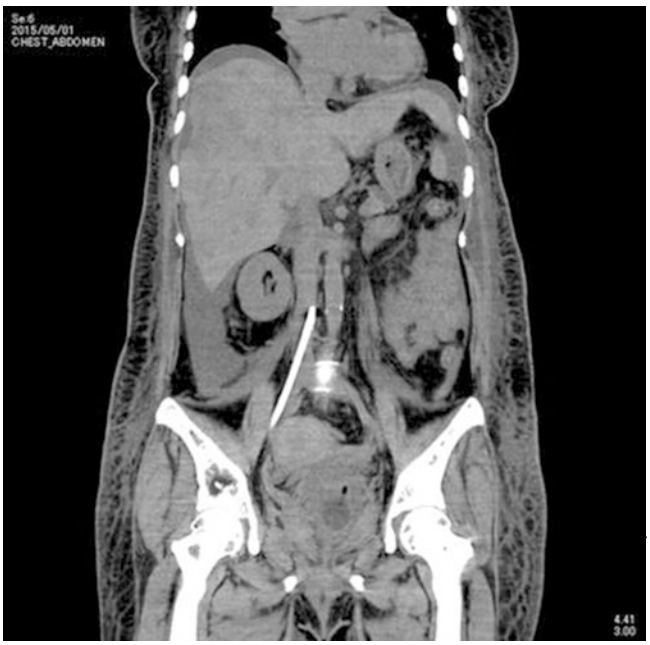



**Fig. 7.8** Retroperitoneal haemorrhage after femoral venous catheterization (3). A 60-year-old female with breast cancer was hospitalized for anuric acute renal failure caused by thrombotic microangiopathy. An experienced physician inserted a four-lumen central venous catheter (13 Fr, 25 cm length) into the right femoral vein to enable haemodialysis. The catheter was introduced using ultrasound guidance, with no specific procedural difficulties. Immediately after catheter insertion, the patient reported severe lower abdominal pain. Contrast-enhanced computed tomography demonstrated that the catheter was correctly positioned, but retroperitoneal haemorrhage had occurred. This case was the bleeding of the inferior epigastric artery caused by resistance from the hard catheter in the femoral vein. The patient underwent coil embolization of the inferior epigastric artery, and the bleeding was successfully controlled. Retroperitoneal haemorrhage and correctly positioned femoral venous catheter (computed tomography)

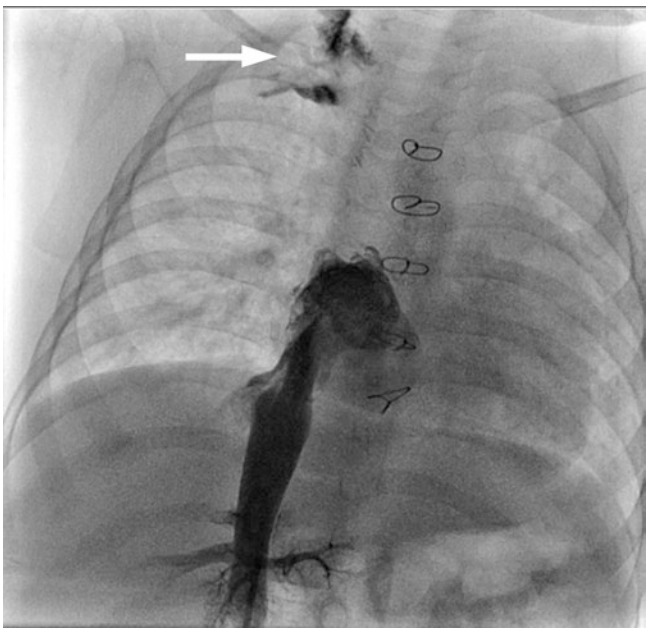


**Fig. 7.9** Subcutaneous neck haematoma after failed right internal jugular vein puncture. A 5-year-old boy (18 kg) underwent a diagnostic cardiac catheterization before totalization of his cavopulmonary connection. The right internal jugular vein was cannulated but the 5 F introducer failed to progress on the wire. A hand contrast injection through the introducer showed dye extravasation in the neck tissue (arrow). The sheath was uneventfully removed, and a gentle compression was sufficient for haemostasis

with either coil embolization (see Fig. 6.10 in Chap. 6) or Gelfoam plugs. When it occurs, parenchymal haemorrhage should be treated conservatively. If bad tolerance, exploratory laparotomy may be needed.

#### 7.1.2.4 Umbilical Access

In newborns that are under 4 days old, the umbilical vein is accessible for a quick vascular access, but this can prompt a few types of access complications including blood loss, vascular perforation and thrombosis or catheter migration (Fig. 7.10).

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.

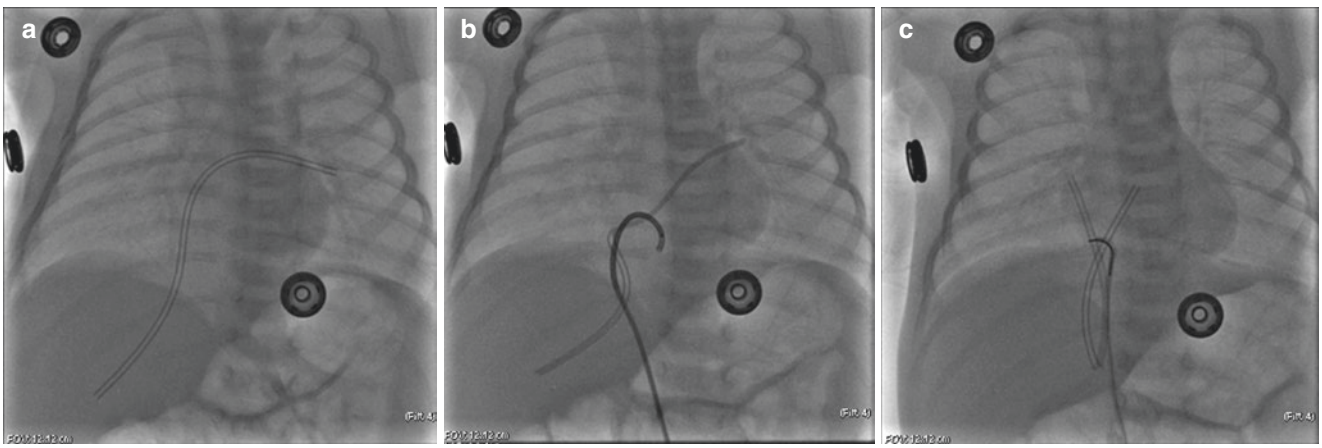



**Fig. 7.10** Indwelling umbilical venous catheter migration. A 10-day-old premature baby (2.2 kg) was transferred to our cath lab after an accidental migration of an indwelling umbilical venous catheter (UVC). On chest *X*-ray, the UVC was stuck between the hepatic vein and the right pulmonary vein through the PFO (**a**). Through a 5 F short venous femoral sheath, a 4 F pigtail catheter was used to first liberate the UVC extremities with a "snake lovers" manoeuvre (**b**). The UVC was then pulled to the IVC. After the tips were exposed, it was removed using a 4 F "gooseneck" 5 mm catheter (**c**)

# Transseptal Access

8

Tilak K. R. Pasala, Vladimir Jelnin, and Carlos E. Ruiz

## 8.1 Introduction

Transseptal (TS) access was initially described in 1959 and since has been widely utilized for congenital and structural heart interventions. A thorough understanding of the anatomy is crucial for TS access (Fig. 8.1). Perfoming contemporary TS access stresses on not only understanding the anatomy but also on the knowledge of modern imaging modalities, alternative techniques for difficult or high-risk patients, site-specific puncture based on the type of procedure, and a systematic approach to management of complications (1). Advanced imaging such as fusion imaging may improve the site specificity of the TS access. There is a steep learning curve, and trainees may benefit from collaborating with other disciplines and doing simulator training. There is a renewed interest in percutaneous interventions for congenital and structural heart disease, and TS access remains an integral part of the skillset that is needed for safe and effective procedures.

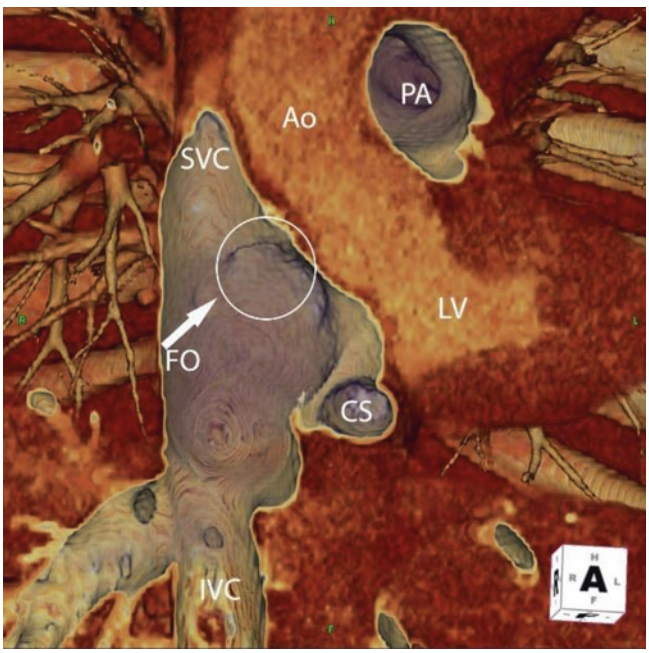



**Fig. 8.1** Understanding transseptal anatomy. A majority of the atrial septation is formed by infolding of the right and left atrial walls (interatrial groove), with puncture outside the fossa ovalis valve (FO) and adjacent margins of its muscular rims (limbus), leading to perforation. The interatrial 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 superoposterior 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

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_8) contains supplementary material, which is available to authorized users.

T. K. R. Pasala · V. Jelnin · C. E. Ruiz (✉)
Structural and Congenital Heart Center, Hackensack University Medical Center and the Joseph M. Sanzari Children's Hospital, Hackensack, NJ, USA
e-mail: CRuiz@StructuralHeartCenter.org


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_8

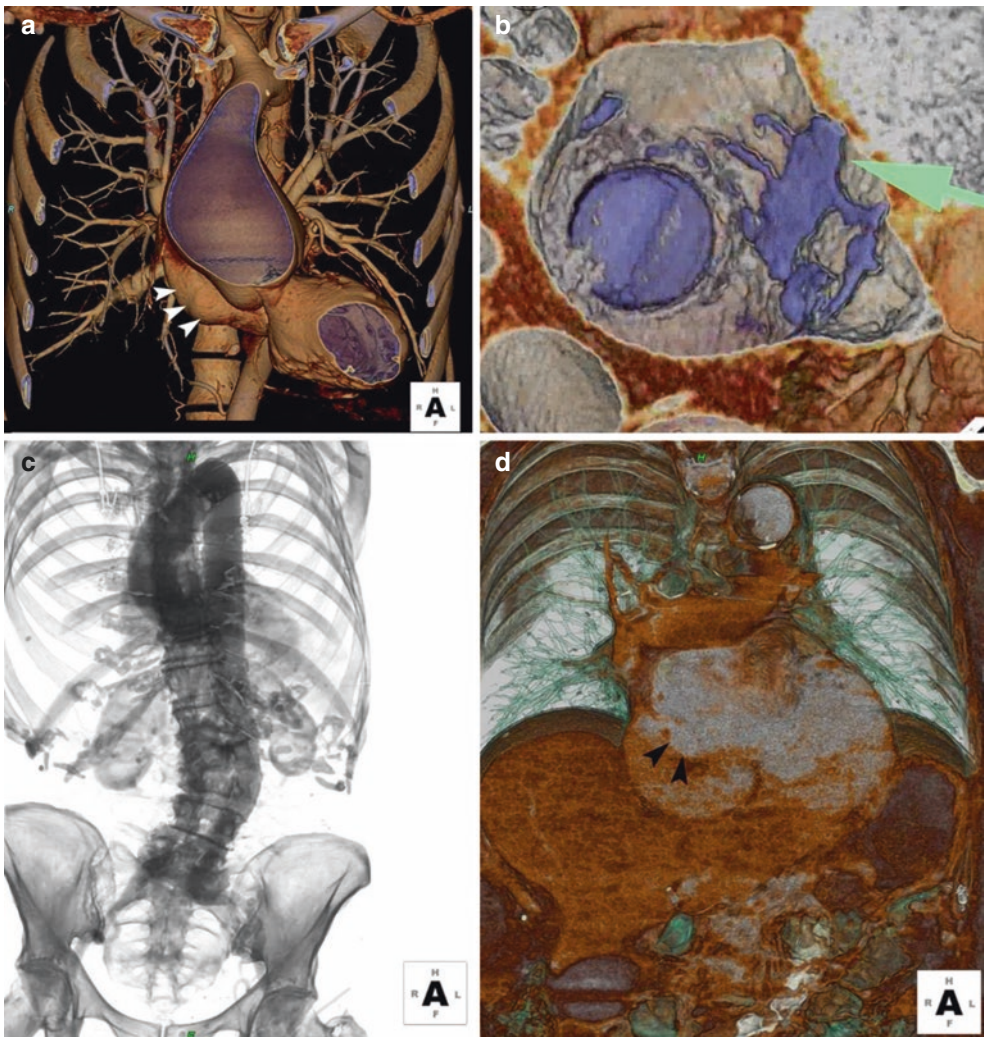



**Fig. 8.2** High-risk transseptal anatomy. (**a**) Extracardiac distortion by an ascending aortic aneurysm can cause bulging of the anterosuperior aspect of the interatrial septum (*white arrowheads*). (**b**) Extensive calcification (*green arrow*) of the IAS can be visualized in a postsurgical patient with limited location for transseptal puncture. (**c**, **d**) Patient with significant scoliosis leading to a more horizontal orientation of the IAS (*black arrowheads*)

**Table 8.1** Indications and contraindications of transseptal access for congenital and structural heart procedures

| Indications | Absolute and relative contraindications |
|---|---|
| Hemodynamic assessment | Lipomatous or thickened atrial septum |
| Patent foramen ovale closure | Hypoplastic left atrial cavity |
| Atrial septal defect closure | Presence of atrial thrombus or mass |
| Left-sided electrophysiology procedures | Organized thrombus (relative contraindication) |
| Pulmonary hypertension with low cardiac output | INR >2.5 without reversal |
| Single ventricle physiology with low cardiac output or hypoxemia | Platelet count <50,000 cell/dL |
| Protein-losing enteropathy (failing Fontan physiology) | Abnormal IVC precluding normal access to the septum |
| Percutaneous balloon mitral valvuloplasty | |
| Antegrade aortic balloon valvuloplasty | |
| Transcatheter mitral valve repair | |
| Transcatheter mitral valve/valve-in-valve implantation | |
| Percutaneous mitral paravalvular leak repair | |
| Percutaneous left ventricular assist devices | |
| Left atrial appendage closure | |

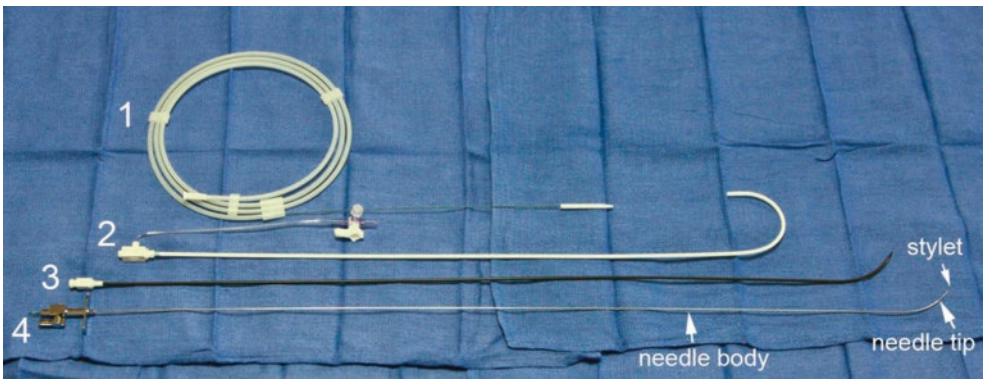



**Fig. 8.3** Standard equipment for transseptal access is shown in the above figure: (1) 0.025-in. guidewire, (2) Mullins sheath, (3) dilator, (4) and Brockenbrough needle (Medtronic, Santa Rosa, Calif). The needle body is 18G, the needle tip is 21G, and the stylet is 0.014-in. in diameter. The Brockenbrough needle is available in multiple lengths and curves. Various other equipment are used (not shown). The NRG RF transseptal needle (Baylis, Montreal, Quebec, Canada) uses RF energy and can be used to avoid forced entry with the needle or when thick septum is present. Steerable sheaths (Agillis NxT; St Jude Medical, St. Paul, Minn) allow maneuverability in complex anatomy

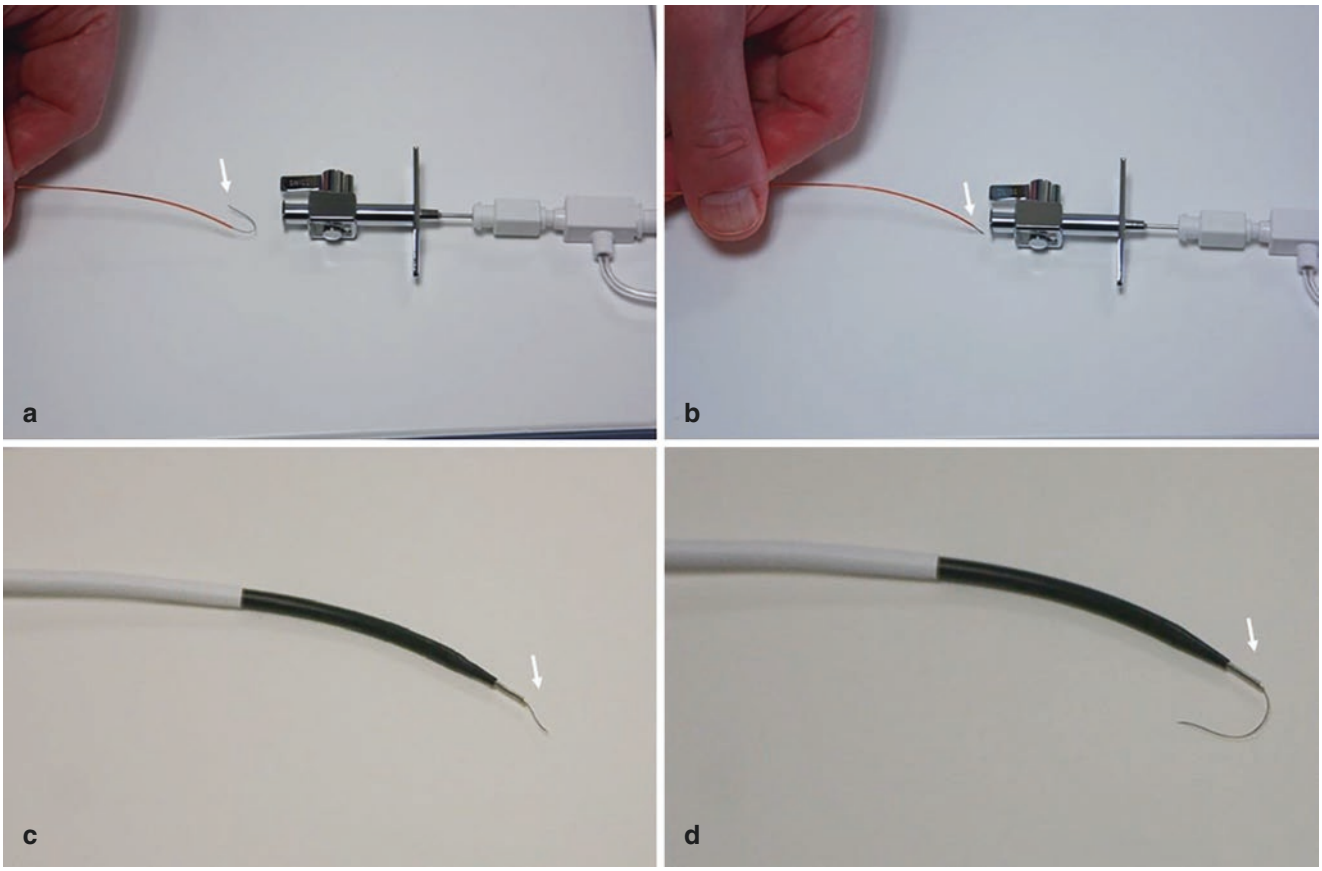



**Fig. 8.4** The SafeSept wire (Pressure Products, San Pedro, California) is a nitinol guidewire (0.014-in. diameter) that has a sharp floppy "J tip" (white arrows). The tip assumes a straight orientation (**b** and **c**, white arrow) while inside the needle and allows crossing the septum, but upon entry into the left atrium, it assumes a J shape (**a** and **d**, white arrows). This allows for a safer entry into left atrium

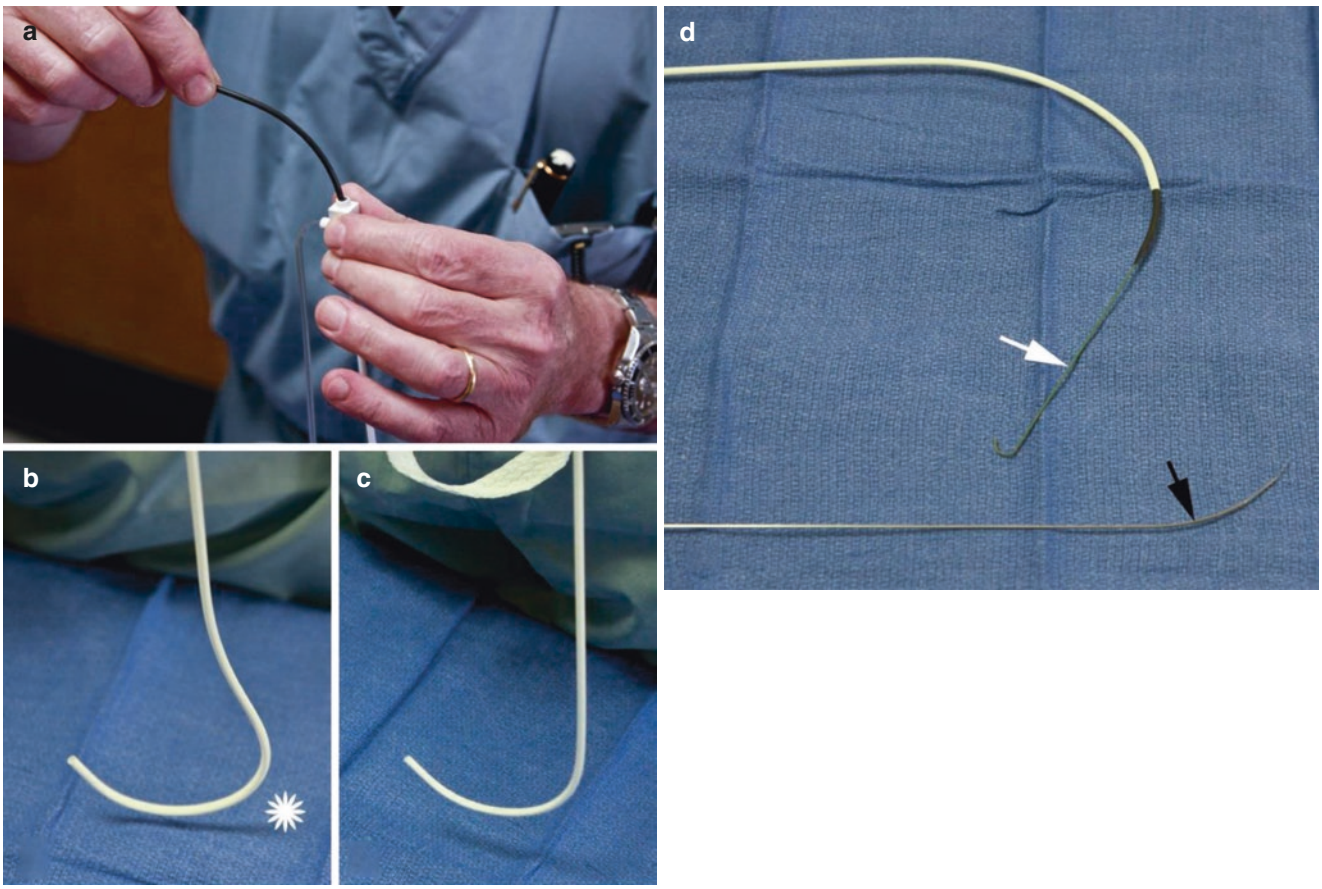



**Fig. 8.5** Prepping the standard transseptal access equipment. (**a**) Dilator is inserted into the sheath in the air to align the curves of the dilator and the Mullins sheath. (**b**) Incorrectly aligned dilator and sheath. A secondary curve is seen in the sheath (*). (**c**) Correctly aligned dilator and Mullins sheath, a single curve is noted. (**d**) With a correctly aligned dilator and sheath, wire and needle follow the curve allowing a more predictable angle when externalizing the needle for puncture

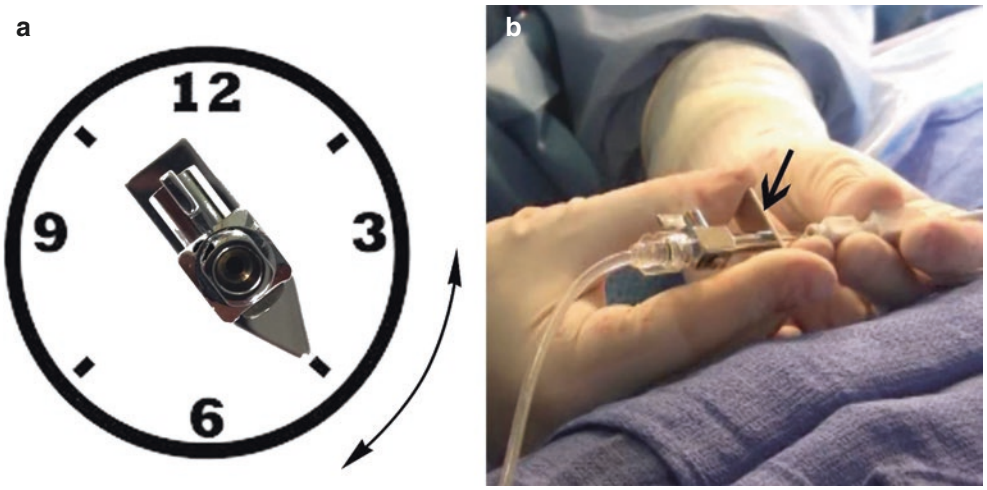



**Fig. 8.6** Transseptal needle position. (**a**) The transseptal (TS) needle indicator arrow dictates the location of the needle tip. With the patient on a horizontal plane, the sheath with side port and the indicator arrow are pointed in the same direction to the 4–5 o'clock location. This allows for reaching the fossa ovalis and interatrial septum which is located posteriorly. (**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

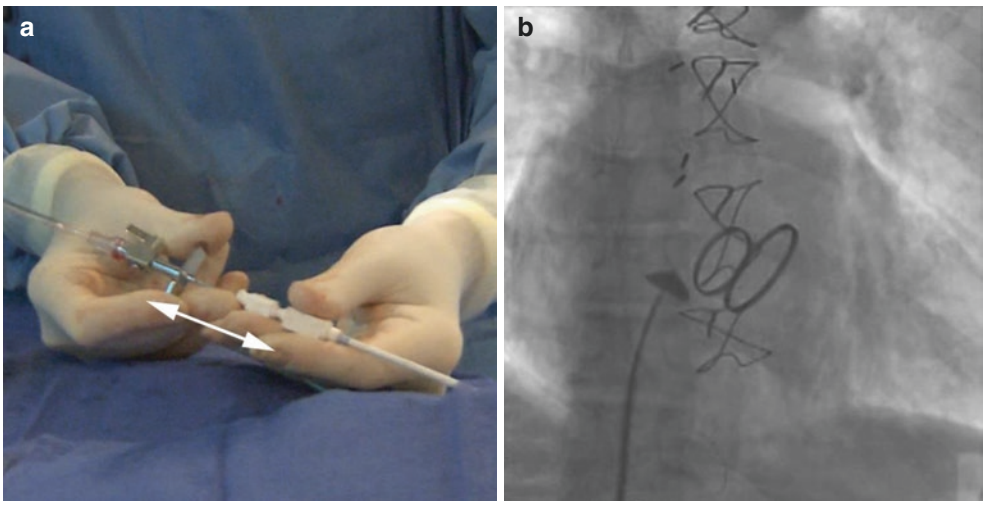



**Fig. 8.7** Technique of transseptal puncture. (**a**) The 8Fr 62 cm-long sheath is advanced over a J-tipped guidewire to the superior vena cava. The guidewire is removed a 71 cm transseptal (TS) needle is introduced under continuous flush. The needle is kept approximately 1–2 cm away from the sheath hub; the entire system is pointed at the 4–5 o'clock location and withdrawn caudally until it encounters two leftward jumps: SVC/right atrial junction and muscular interatrial septum. (**b**) TS tip subsequently engages the fossa ovalis (FO), confirmed by the contrast injection

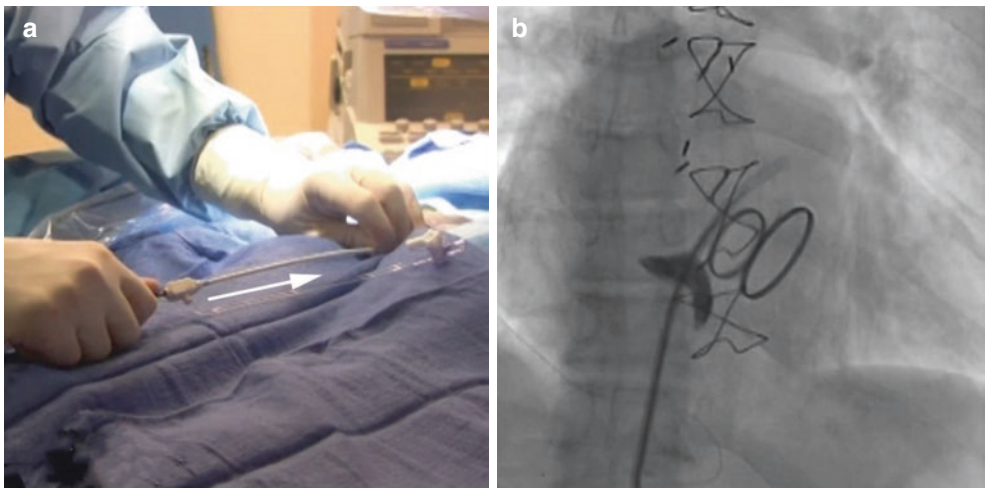



**Fig. 8.8** Technique of transseptal puncture—contd. Once the needle position is confirmed in the left atrium (LA), the entire system is advanced 1 cm (**a**). The dilator is disconnected from the sheath, and the needle/dilator is turned toward the 12–1 o'clock location (**b**). Then the dilator is held still with the right hand and the sheath is advanced over the dilator into the LA. Then the dilator/needle are removed and passive back bleeding is done to de-air the system

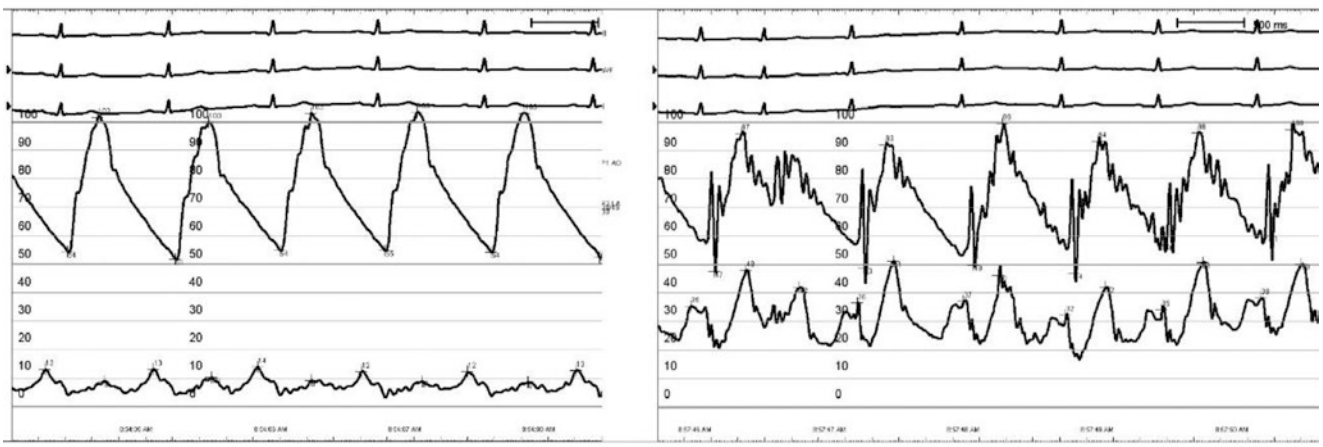


**Fig. 8.9** Pressure monitoring can be performed during the transseptal puncture to observe the pressure change from right atrial wave form (left figure, lower tracing) to left atrial wave form (right figure, lower tracing). In this case, pulmonary artery pressure is being monitored (upper tracing) with a Swan-Ganz catheter

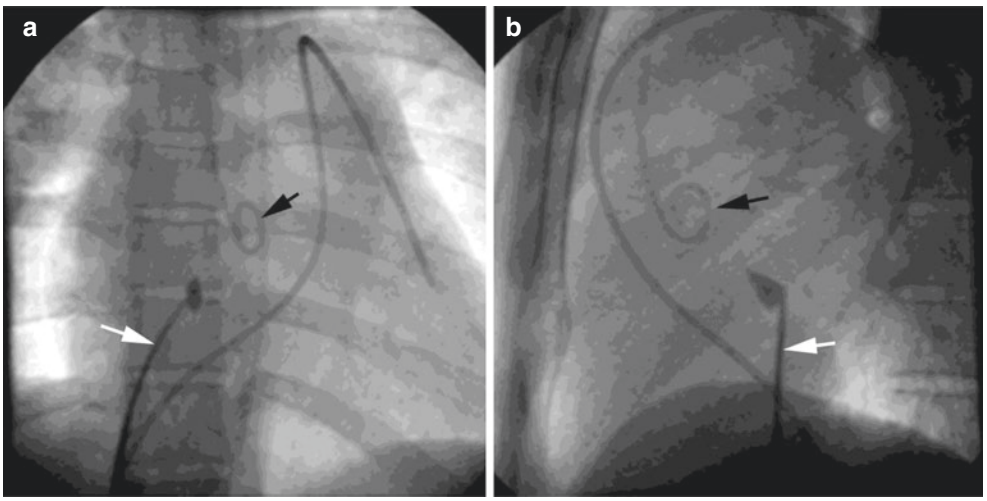



**Fig. 8.10** Fluoroscopic guidance for transseptal access. The antero-posterior view (**a**) shows a pigtail placed in the noncoronary aortic valve cusp (**a**, black arrow) identifying the posterior border of the aortic wall and aortic root. A balloon-tipped catheter is also visualized within the right ventricular outflow tract into the left branch pulmonary artery. The TS system (white arrows) and puncture site (contrast injection) is seen lower and medial to the pigtail catheter. In the left anterior oblique view (**b**) at 30–35°, the needle is directed posterior to the pigtail catheter

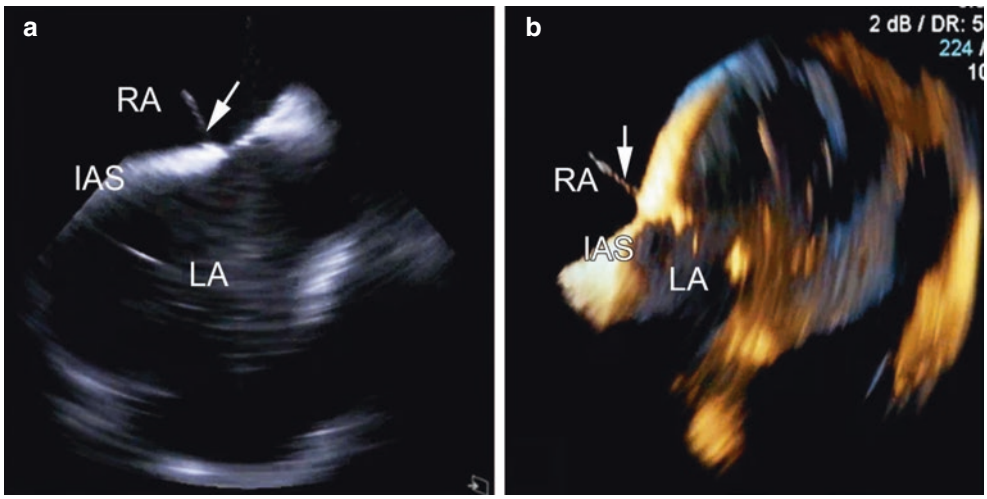



**Fig. 8.11** Transseptal access with intracardiac echocardiography guidance. Intracardiac echocardiography (ICE) provides two-dimensional (**a**) and three-dimensional (**b**) imaging with clear definition of intracardiac chambers. The transseptal system (white arrow) is seen tenting prior to the puncture. Bubbles can be visualized in the LA confirming needle position

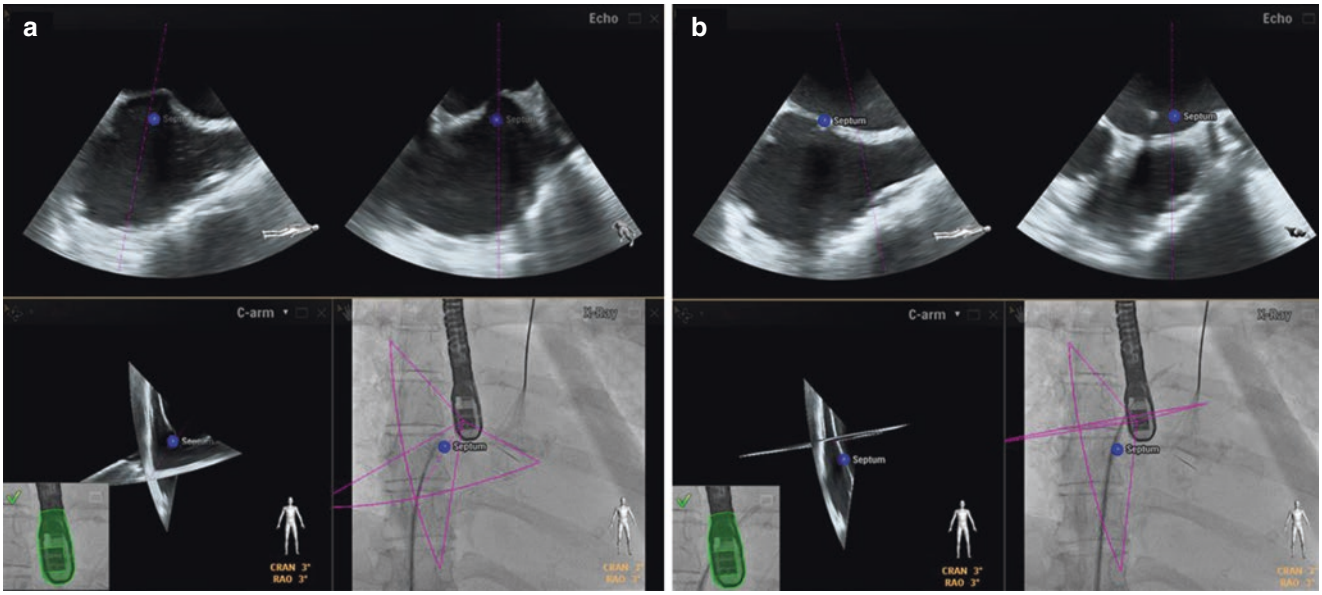



**Fig. 8.12** Transseptal access with echo-fluoroscopy fusion. (**a**) Upper window demonstrates the position of the transseptal (TS) needle tenting the interatrial septum. Upper right window demonstrates the view from EchoNavigator; the blue marker placed on the TEE, identifying the best puncture location, is overlaid on live fluoroscopy and is used for guidance of the TS puncture. (**b**) The moment after puncture. Upper windows show the tip of the needle in the left atrium. Lower right window of EchoNavigator confirms the needle position. Blue marker—the optimal location to cross the atrial septum

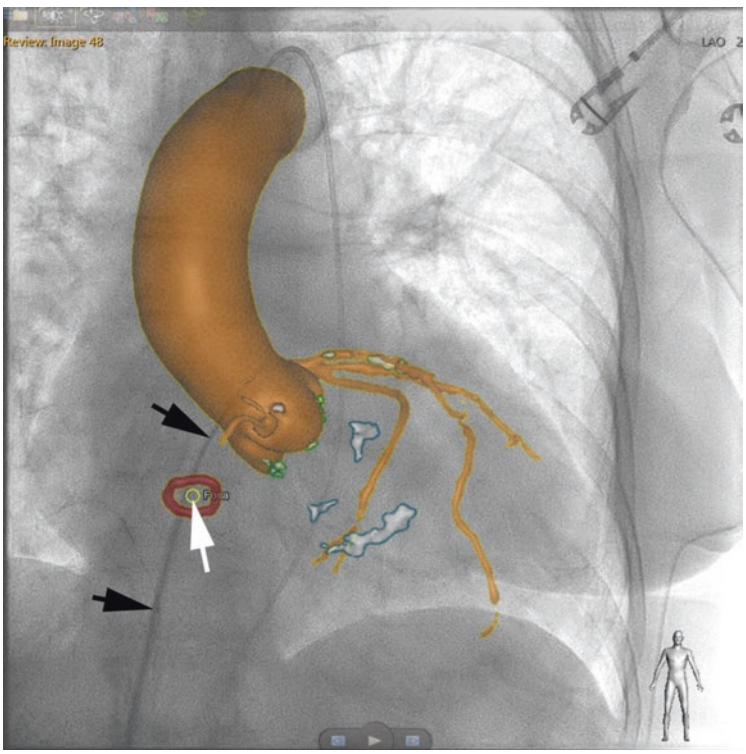



**Fig. 8.13** Transseptal access with CTA-fluoroscopy fusion. The optimal location for transseptal puncture is identified on the 3D CT and marked with yellow circle (white arrow). Fossa ovalis is marked with the large red circle. All markers are overlaid over the live fluoroscopy to guide TS access. The TS system is seen crossing the septum at the marked site into the left atrium

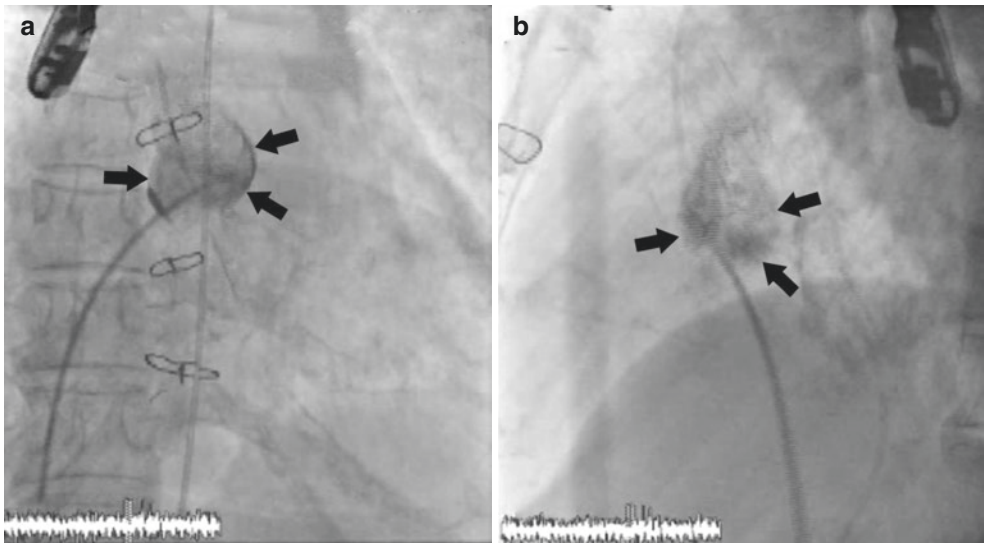



**Fig. 8.14** One of the more serious complications during transseptal access is aortic root puncture. Due to the proximity of the aortic root, care must be taken to be lower the aortic root on anteroposterior view (**a**) and posterior to it the aortic root on left anterior oblique (LAO) view (**b**). In the above figure, TS system is seen in the aortic root evidenced by contrast injection (black arrows). The TS catheter is seen directed anteriorly in the LAO view (**b**)

**Video 1** Transseptal puncture steps (MP4 234056 kb)

**Video 2** The transseptal access can be closed by using a septal occluder (MP4 148507 kb)

**Video 3** TrSept puncture assistance in performing a transseptal puncture in complex cases (MP4 44661 kb)

# Part III

# Fetal Procedures

# Fetal Cardiac Interventions

9

Carlos A. C. Pedra, Simone F. Pedra, and C. Fabio Peralta

## 9.1 Introduction

In the last 15–20 years, fetal cardiac interventions have become an accepted therapeutic modality for some complex congenital heart diseases (CHD) in utero, including 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). The purpose of a fetal cardiac intervention is to remodel cardiac morphology and function and improve pre- and postnatal outcomes. Increased survival rates, reduced morbidity, and increased likelihood of achieving a biventricular (BV) circulation may all result from a successful fetal cardiac intervention in some scenarios. The objective of this chapter is to give an overview of these procedures with emphasis on pictures to illustrate how they are performed in a step-to-step fashion.

## 9.2 Indications for Interventions and Patient Selection

### 9.2.1 Fetal Critical AS and Evolving HLHS

The diagnosis of fetal critical AS is made by fetal echocardiography upon visualization of a thickened, immobile aortic valve with turbulent or decreased color Doppler flow. It is important to remember that the Doppler-derived gradient may be misleading due to commonly associated left ventricular (LV) dysfunction and endocardial fibroelastosis (EFE). Evolving HLHS is defined by the presence of fetal critical AS associated with 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. Appropriate timing for the procedure is between 24 and 30 weeks gestational age. Imaging and size of the patient are limiting factor to perform it before 24 weeks. On the other hand, it is probably too late to achieve any cardiac remodeling after 30 weeks.

### 9.2.2 Fetal HLHS and Intact or Highly Restrictive IAS [1–3]

These fetuses have an established prenatal diagnosis of HLHS associated with either an intact IAS or a tiny (≤1 mm) atrial septal defect (ASD) or patent foramen ovale (PFO). Prominent flow reversal in the pulmonary veins is seen. Fetal atrial septostomy is indicated to avoid/minimize severe hypoxemia immediately after birth due to pulmonary venous congestion and parenchymal changes. In utero ASD creation is performed between 29 and 32 weeks gestation in order to endure until term.

C. A. C. Pedra (✉)
Catheterization Laboratory and Hybrid Room for Congenital Heart Disease, Instituto Dante Pazzanese de Cardiologia and Hospital do Coração da Associação Sanatório Sírio, Sao Paulo, SP, Brazil

S. F. Pedra
Echocardiography Laboratory for Congenital Heart Disease, Instituto Dante Pazzanese de Cardiologia and Hospital do Coração da Associação Sanatório Sírio, Sao Paulo, SP, Brazil

C. Fabio Peralta
Fetal Medicine Program, Hospital do Coração da Associação Sanatório Sirio, Sao Paulo, SP, Brazil


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_9

### 9.2.3 Fetal PA/IVS or CPS/IVS and Evolving HRHS [1–3]

Such patients have a prenatal echocardiographic diagnosis of PA/IVS or CPS/IVS with identifiable pulmonary valve (PV) leaflets, membranous atresia or pinpoint PV stenosis with no or minimal systolic opening, and no or minimal color Doppler ultrasound flow across the pulmonary valve (PV). The ventricular septum is intact. There is flow reversal across the duct (PDA) (left-to-right shunting). The right ventricle is hypoplastic with a tricuspid valve (TV) annulus *Z*-score below ≤2. On serial assessment, there is no evidence of RV growth after 2–4 weeks. Those patients with associated major coronary-to-RV fistulas should not undergo in utero pulmonary valvuloplasty due to the risk of cardiac ischemia secondary to RV decompression in the setting of RV-dependent coronary circulation. Fetal pulmonary valvuloplasty is carried out between 24 and 30 weeks gestation.

### 9.2.4 Fetal Critical AS with Massive Mitral Regurgitation (MR), Giant Left Atrium (LA), and Hydrops [1, 2]

These fetuses are very sick in utero. Although they may have normal-sized LV and function, there is reversed flow in the TAA due to the lack of antegrade flow across the aortic valve. The MV is abnormal and massively insufficient, which results in a conspicuously dilated LA that compresses the pulmonary veins and the RV. Aortic valvuloplasty and atrial septostomy should be both considered in such grave cases between 30 and 34 weeks gestation as a "salvage" procedure to diminish the risk of fetal loss.

## 9.3 Planning of the Procedure

A multidisciplinary approach is key to perform fetal cardiac interventions. The core team should include the fetal cardiologist, the fetal specialist (or perinatologist), the interventionalist, and the anesthesiologist. An obstetrics (OB) team should also be involved should any obstetric complication arise during the procedure. Initial patient screening and selection with pre-procedural echocardiographic assessment is under the domain of the fetal cardiologist. The whole team discusses the peculiarities of the case, and the pictures are extensively reviewed. During the procedure, the mother is taken care by the anesthesiologist and the OB team. The role of the fetal medicine specialist is crucial. He or she conducts fetal positioning and anesthesia and simultaneously controls the puncture needle and the ultrasound probe. The interventionalists (usually two) handle the catheters and wires and lead the intervention to achieve optimal positioning of the balloon/wires/system inside the heart working hand in hand with the fetal specialist. Because the whole procedure relies solely on ultrasound/echo pictures, optimal imaging quality should be pursued at all costs. The immediate post-procedural care is conducted by the OB team. Follow-up outcomes are assessed by the fetal cardiologist, who performs serial echo evaluations and plans further procedures after birth with the neonatologist, intensivist, cardiac surgeon, and interventionalist.

## 9.4 Fetal Cardiac Interventions: Step-by-Step Technique

The mother is anesthetized (regional spinal blockade) and slightly sedated in the operation room. Antibiotics are given prophylactically in three doses. Nifedipine 20 mg TID for 48–72 h is given for uterine relaxation starting 12–24 h before the procedure. The uterus is slightly displaced to the left side to avoid inferior vena cava compression. Polyhydramnios is drained using a 15-cm-long 21-G Chiba needle (Cook Inc., Bloomington, IN, USA) if deemed significant. Optimal fetal position is achieved by external version. We do not proceed if fetal lie is not optimal, which is exceptionally rare. An open procedure with uterus exposure is eschewed in our practice. Once optimal fetal lie is attained, the fetus is anesthetized using a mixture of fentanyl (5–15 μg/kg), pancuronium (10–20 μg/kg), and atropine (20 μg/kg) given intramuscularly (more commonly) or in the umbilical cord using a 21–22-G Chiba needle [1, 2].

Cardiac access is achieved through direct needle puncture of the fetal heart. Under continuous two-dimensional ultrasound guidance, the needle initially crosses the maternal abdominal wall, the uterine wall, and the fetal chest wall. In some procedures, the needle may have to traverse the placenta and/or fetal liver to enter the left or right ventricular apex. We use a 15-cm-long 17–18-gauge Chiba needle with a stylet, which is advanced to the target fetal cardiac structure (LV, RV, or right atrium) (Fig. 9.1). The imaging plane is adjusted to accommodate the entire needle length and the target cardiac chamber in the field of view.

For dilation itself, we use a simple pre-marked system consisting of a rapid exchange 10-mm-long coronary balloon pre-mounted over a cutoff 0.014″ floppy tip guidewire (Fig. 9.2). This system is advanced through the needle as a single unit to the desired location. The needle, guidewire, and balloon shafts are premeasured and marked with glue tapes outside the body so that positioning within the fetal heart is known more from these external measurements rather than the echo pictures 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

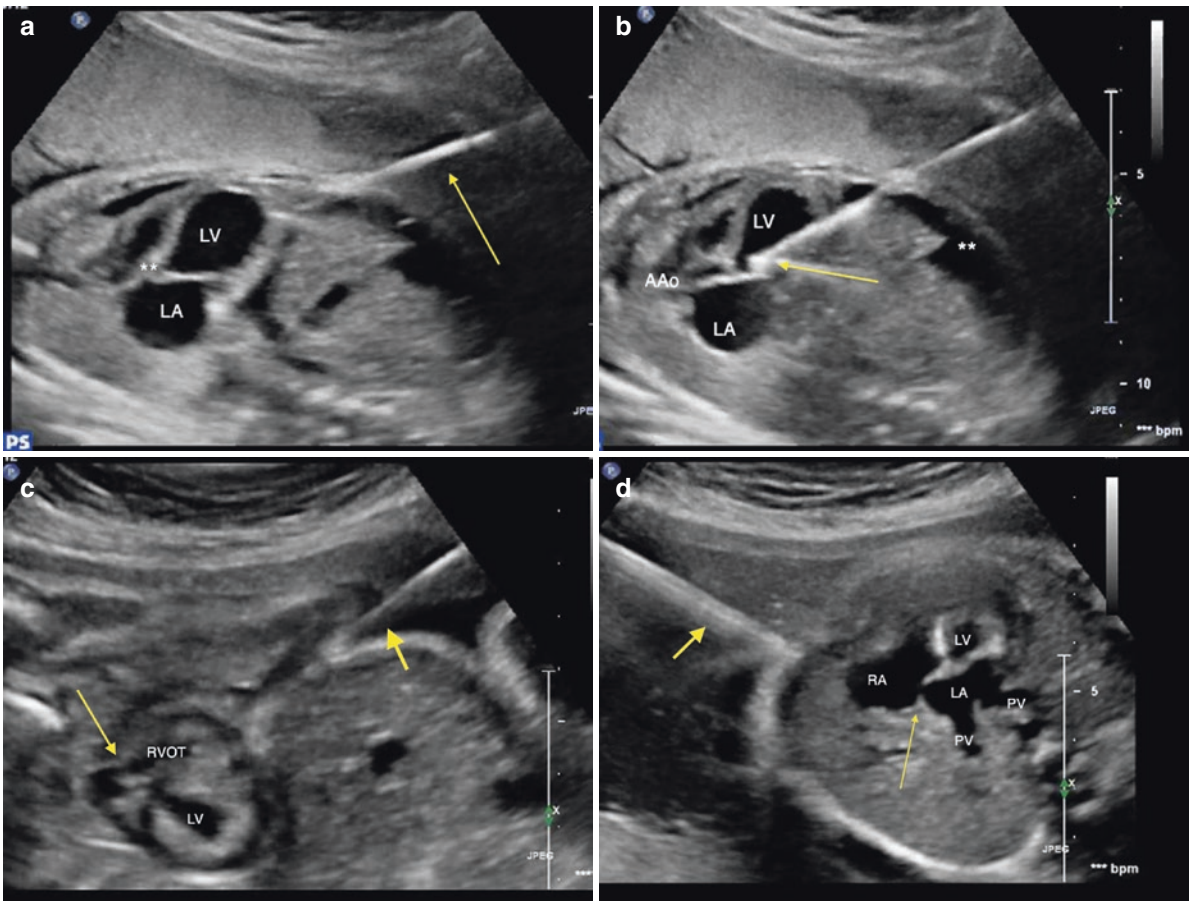



**Fig. 9.1** Proper needle course, angulation, and positioning during fetal cardiac procedures. (**a**) For fetal aortic valvuloplasty, the tip of the needle (indicated by an arrow) is aimed at the aortic valve (marked with asterisks) in an imaginary straight line before entering the left ventricular apex. (**b**) After left ventricular entry, the tip of the needle (indicated by an arrow) is kept below the aortic valve so that the valve can be crossed with minimal wire manipulation. The asterisks indicate ascites in this hydropic fetus. (**c**) For 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 straight line coming from the right ventricular apex toward the right ventricular outflow tract. (**d**) For fetal atrial septostomy, the needle (indicated by a short and broad arrow) should traverse the interatrial septum (indicated by a long and narrow arrow) in a perpendicular course. Abbreviations: *LV* left ventricle, *LA* left atrium, *RV* right ventricle, *RA* right atrium, *AAo* ascending aorta, *RVOT* right ventricular outflow tract, *PV* pulmonary vein

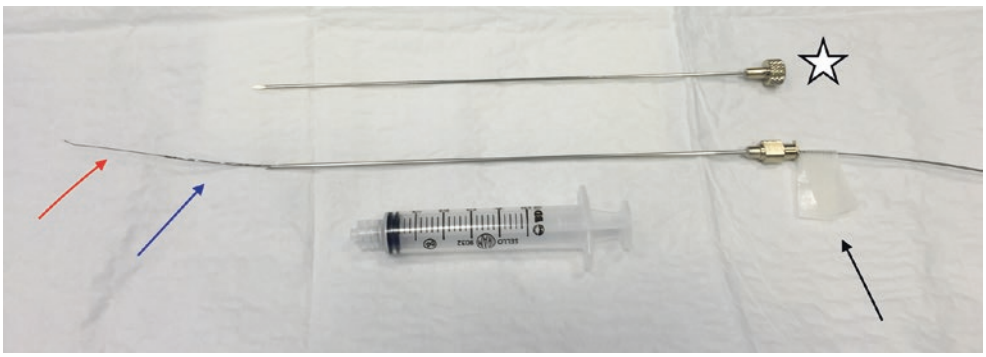


**Fig. 9.2** Pre-marked system (needle + balloon + guidewire): A 15-cm-long 17- or 18-gauge Chiba needle is used for the procedure. The needle, guidewire, and balloon shafts are premeasured and marked with glue tapes outside the body. The stylet (marked by an arrow) of the needle is taken out, and a rapid exchange 10-mm-long coronary balloon pre-mounted over a cutoff 0.014″ floppy tip guidewire is passed through the needle. The balloon shaft is marked with sterile tapes (marked by a black arrow) so that no more than the full length of the balloon is extruded out of the Chiba needle tip when fully advanced (marked by a blue arrow). 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 (marked by a red arrow). The syringe is displayed for dimensional purposes

that no more than 3–4 cm of the distal flexible wire straight tip extruded out from the balloon tip.

For aortic or pulmonary dilation, the needle perforates the apex of the LV or RV, respectively, aiming at the stenotic/atretic semilunar valve. Once the ventricular chamber is entered, the needle is kept in the outflow tract, the stylet is withdrawn, and the catheter system is introduced and advanced as a unit (Figs. 9.3 and 9.4). Usually, the wire can be seen when it is coming out the tip of the needle. Generally, the valves should be crossed with minimal wire and catheter manipulation. The needle is kept in place, and the system is further advanced so that the catheter shaft mark reaches the proximal hub of the needle, indicating that the balloon is outside the tip of the needle and straddling the valve annulus. Balloon positioning for inflation is based on these external measurements and optimal ultrasound imaging. As a practical tip, 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) is an excellent sign that the balloon is likely well positioned or requires minimal adjustment. For PV perforation, the same needle that was used for apex entry is advanced through the atretic PV. Alternatively, coronary wires designed for total chronic occlusion may be used in the pre-mounted system to perforate the PV when the tip of the needle is parked in the RVOT below the PV. Balloons are inflated with pressure gauges to reach the desired final diameters. Balloon

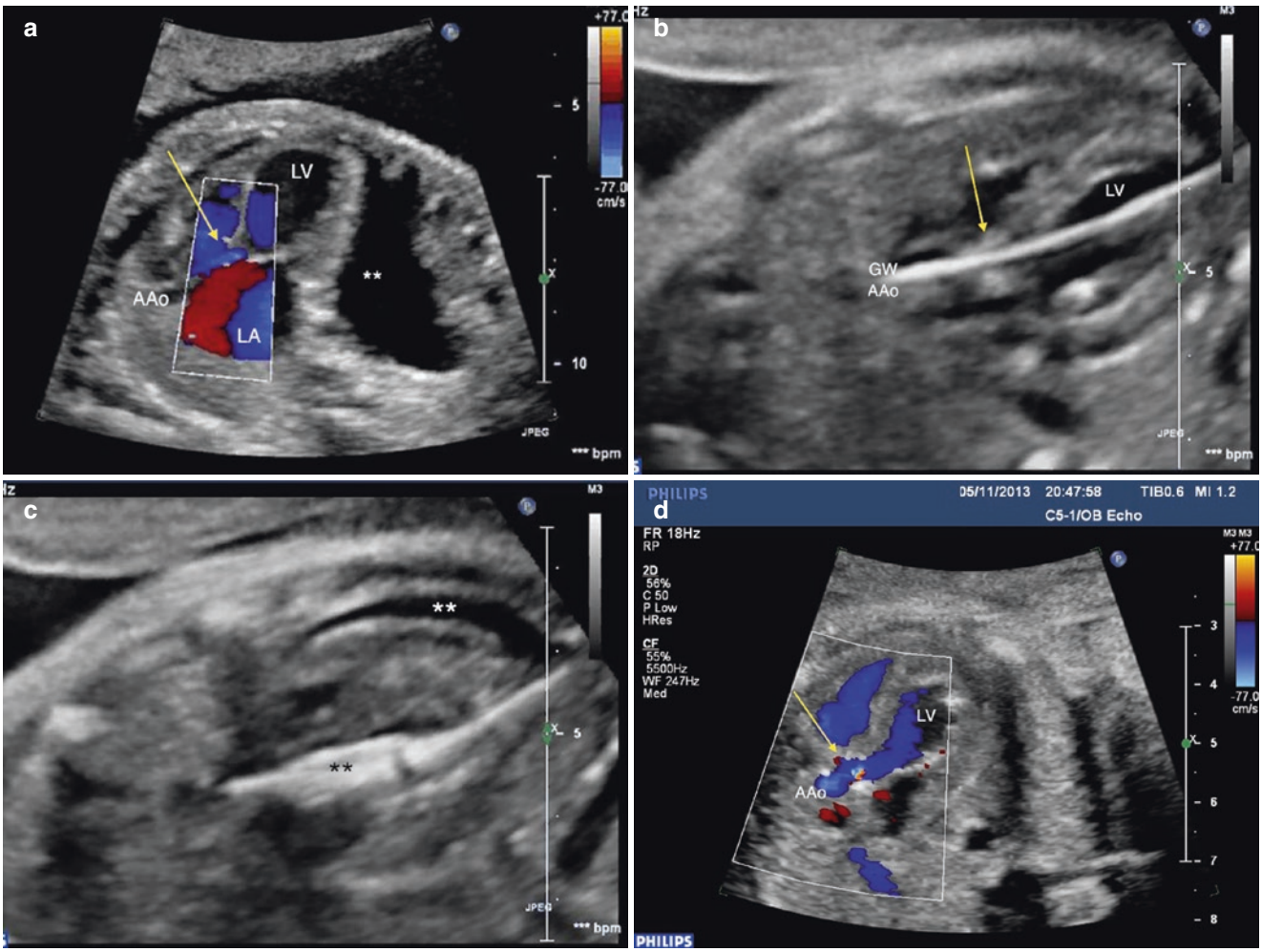



**Fig. 9.3** Critical aortic stenosis in a 26 weeks gestational age hydropic fetus. (**a**) Echocardiographic evaluation pre-intervention. There is a tiny forward flow (blue color) across the aortic valve (indicated by an arrow) as shown by color flow mapping. The LV is conspicuously dilated and dysfunctional. Hydrops results from severe in utero heart failure. Asterisks show ascites. (**b**) After crossing the aortic valve (indicated by an arrow), the guidewire is readily seen in the ascending aorta. (**c**) The balloon (black asterisks) is inflated across the aortic valve. There was some pericardial effusion (white asterisks) that required drainage after dilation. (**d**) After valvuloplasty, there is significant improvement in antegrade flow across the aortic valve (indicated by an arrow) as shown by color flow mapping (the blue color depicts a much wider vena contracta). Abbreviations: *LA* left atrium, *LV* left ventricle, *AAo* ascending aorta, *GW* guidewire

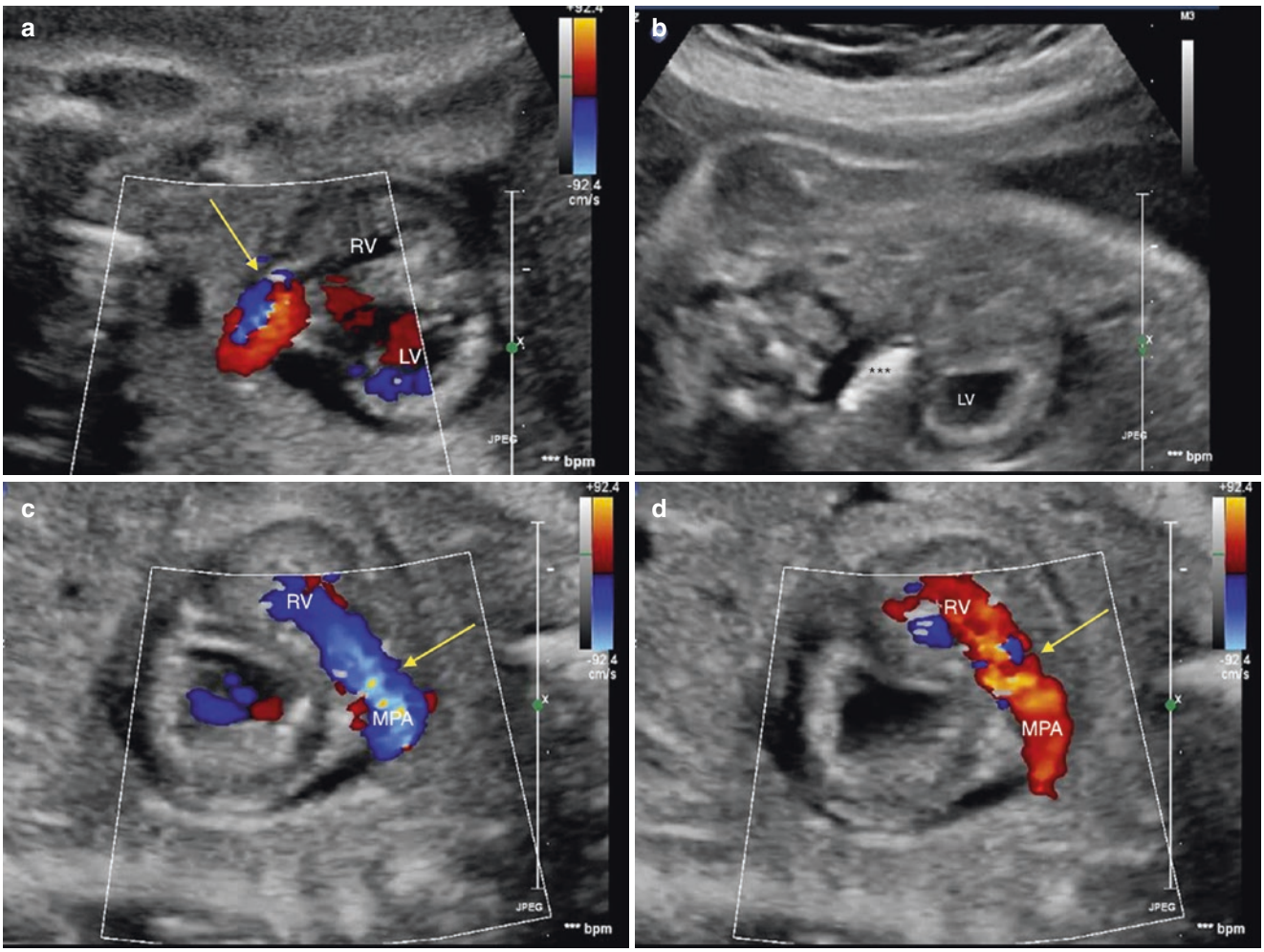



**Fig. 9.4** Critical pulmonary valve stenosis in a 28 weeks gestational age fetus. (**a**) Echocardiographic evaluation before intervention. There is a tiny forward flow (in blue color) across the pulmonary valve (indicated by an arrow) as shown by color flow mapping. Retrograde flow across the ductus is the depicted by red color on color flow mapping. The pulmonary valve annulus measured 4.0 mm. (**b**) A 10-mm-long coronary balloon (marked with black asterisks) is inflated across the pulmonary valve up to the burst pressure reaching 4.7 mm in diameter. (**c**) After valvuloplasty there is significant improvement in forward flow across the valve (in blue color) as shown by color flow mapping. The pulmonary valve is indicated by an arrow. (**d**) After effective dilation, significant pulmonary insufficiency ensues and is shown by color flow mapping (in red color) across the pulmonary valve (indicated by an arrow). Abbreviations: *RV* right ventricle, *LV* left ventricle, *MPA* main pulmonary artery

diameters 10–30% larger than the aortic or pulmonary valve annulus are selected for valve dilation. Two to four inflations are performed depending on the fetal clinical status.

We use a 17-G Chiba needle for atrial septostomy, because its greater inner lumen diameter is necessary to accommodate a larger dilating balloon (the largest possible; usually 4 mm, expandable to 4.7 mm). The needle is advanced through the right atrium (RA) and perforates the IAS in a perpendicular fashion, gaining the LA (Fig. 9.5). Once the tip of the needle is seen in the body of the dilated LA, the stylet is removed, and the system is advanced until the tape mark on shaft reaches the hub of the needle. At this point, it is known that the balloon is extruded out of the needle and the whole system (needle + catheter inside) needs to be pulled back until the balloon straddles the IAS. The balloon is then inflated up to the maximal diameter. If the created orifice is judged to be too small to decompress the LA, a second hole can be created using a similar technique. Although in our experience we attempted to implant a stent in the IAS in a single case, the limited internal lumen of the Chiba needles precluded the use of a larger diameter stent. Although stenting the IAS in fetal life may be more effective to relieve LA hypertension, it is technically challenging, with significant rates of failure or malpositioning, even using special catheters specifically designed by the Boston group for this purpose.

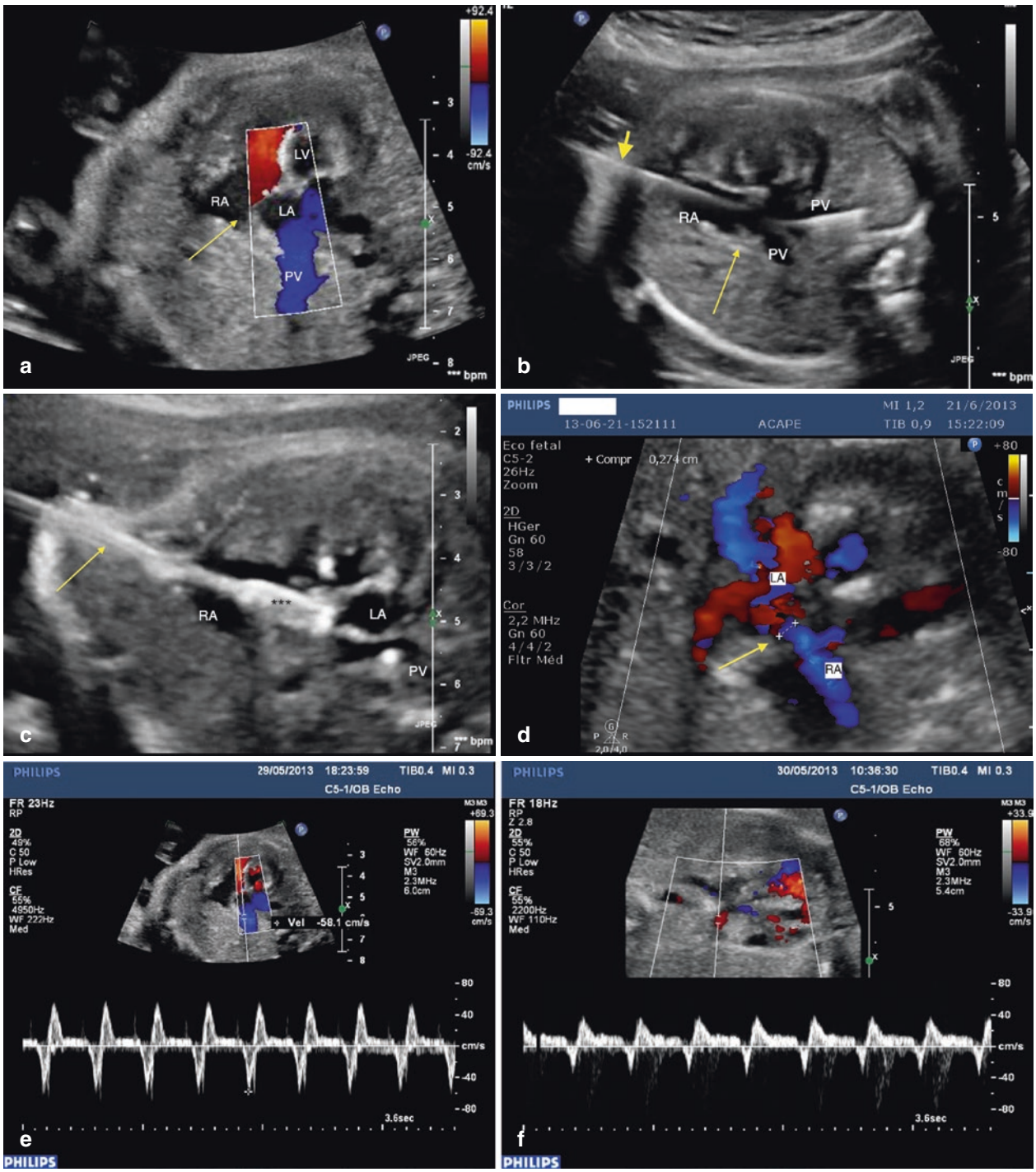



**Fig. 9.5** Fetal atrial septostomy in a 29 weeks gestational age fetus with established hypoplastic left heart syndrome. (**a**). Echocardiographic evaluation prior to the procedure. The interatrial septum (indicated by an arrow) is almost intact, and the LA is conspicuously dilated. Flow reversal in the pulmonary veins is detected by color flow mapping (blue color). The LV is small, and severe endocardial fibroelastosis can be seen as bright and diffuse 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 10-mm-long coronary balloon (marked with black asterisks) is inflated across the interatrial septum up to the burst pressure reaching 4.7 mm in diameter. The Chiba needle (indicated by an arrow) is kept perpendicular to the plane of the interatrial septum. (**d**) Echocardiographic assessment on the following day after fetal atrial septostomy shows a 2.8-mm atrial septal defect within the atrial septum (indicated by an arrow). (**e**) The Doppler tracing of the pulmonary veins before fetal septostomy shows bidirectional flow in the pulmonary vein with high reversal flow velocity (58 cm/s) during atrial contractions (negative wave below the baseline). (**f**) On the following day, there is triphasic flow pattern in the pulmonary veins (better diastolic filling) with improvement of the reversal flow velocity (less than 40 cm/s). Abbreviations: *LA* left atrium, *RA* right atrium, *PV* pulmonary vein, *LV* left ventricle

After the valves or the IAS is dilated, the whole system (needle + balloon + wire) is taken out outside the body as a unit. The balloon should not be retracted into the needle to avoid shearing off the balloon from the catheter shaft. The whole system passes through the fetal cardiac wall and fetal and maternal structures.

Mothers are hospitalized overnight, and the fetus is assessed by ultrasound later on the same day and/or the following morning before maternal discharge. Echocardiography is performed at intervals determined by the fetal cardiologist to assess in utero outcomes.

## 9.5 Complications

Extensive data on cardiac and extra-cardiac fetal procedures show that significant morbidity to the mothers is exceptional. On the other hand, fetal cardiac interventions are commonly associated with hemodynamic instability due to fetal bradycardia and hemopericardium, especially in procedures that involve ventricular access. Fetal loss may occur secondary not only to hemodynamic instability and hemopericardium but also to fetal and maternal anesthetic issues and mechanical stimuli. Inadvertent cardiac perforation in an unplanned site with uncontrolled in utero bleeding may rarely occur. It has been recently estimated that fetal demise occurs in about 10% of all cardiac interventions. Premature labor is another potential complication.

Because significant hemopericardium and bradycardia (<80–100 bpm for 3–5 min) are relatively common in such interventions, prophylactic atropine administration upon fetal anesthesia is mandatory. Moreover, the operators should have technical capability of prompt pericardial drainage and intracardiac therapeutic injections of epinephrine and atropine to carry out such interventions. Small-volume-unit doses of epinephrine (1–10 mcg/kg) and atropine are available on the table, as well as a new 21–22-G Chiba needle for pericardial drainage. Interestingly, fetal bradycardia generally occurs due to mechanical ventricular compression secondary to the hemopericardium. As such, draining 1–4 mL from the pericardial space might be enough to resume adequate heart rate before administration of intracardiac epinephrine.

## 9.6 Immediate Fetal Outcomes

Technically, fetal pulmonary valvuloplasty is more challenging than aortic valvuloplasty due to the heavy trabeculated RV and may be associated with a higher rate of failed attempts, especially at the beginning of the learning curve. Generally, technically successful fetal cardiac procedures are seen in >90% of patients.

Successful procedures are defined by echocardiography. After successful dilation of the semilunar valves, there is unequivocal evidence of antegrade flow across the aortic or pulmonary valve and/or new aortic/pulmonary regurgitation (AR or PR) as assessed by color Doppler echocardiography (Figs. 9.3 and 9.4). Post-procedural AR and PI are probably a marker of effective dilatation of the aortic and pulmonary valves. 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 for some unknown reason. A successful atrial septoplasty is achieved when there is unequivocal echocardiographic evidence of a newly created ASD measured by the width of the color jet (Fig. 9.5) associated with reduction in LA size and improvement in the pulmonary vein Doppler pattern (Fig. 9.5).

## 9.7 Late Fetal, Neonatal, and Infancy Outcomes

A BV circulation is observed in about 30–50% of fetuses with critical AS and impending HLHS who had undergone fetal aortic valvuloplasty. This includes patients in the neonatal period or late in infancy. Usually patients with a 2-ventricle circulation have a LV long-axis *Z*-score >0, a LV short-axis *Z*-score >0, an aortic annulus *Z*-score >3.5, a MV annulus *Z*-score >2, and a high-pressure LV defined by the presence of MR or AS with a maximum systolic gradient of ≥20 mmHg and milder degrees of EFE. Interestingly, progressive growth of the left heart structures during fetal life and over infancy resulting in an eventual BV repair has been observed in our and other experiences [1, 2]. We have employed a staged strategy for such patients with fetal aortic valvuloplasty followed by a neonatal hybrid procedure ± balloon aortic valvuloplasty (Fig. 9.6). This approach works as a bridge to LV overhaul and BV repair later in infancy. Although postnatal LV diastolic dysfunction may be an issue in these patients, we still think that this is a lesser evil than the immediate and long-term morbidity and mortality of a univentricular pathway [1].

Fetuses with critical AS who already have smaller LVs (LV diastolic length *Z*-score between −2 and −3) may occasionally benefit from in utero aortic valve dilation. The rationale is to promote forward flow across the aortic valve resulting in improved coronary and TAA perfusion. This may help to preserve myocardial function and minimize the neurodevelopmental abnormalities secondary to retrograde TAA perfusion. Whether these theoretical considerations have a positive impact on neonatal outcomes, regardless of the surgical strategy (Norwood vs Hybrid), is debatable. Such cases should be individualized according to the center experience and family counseling [1, 2]. It is our impression

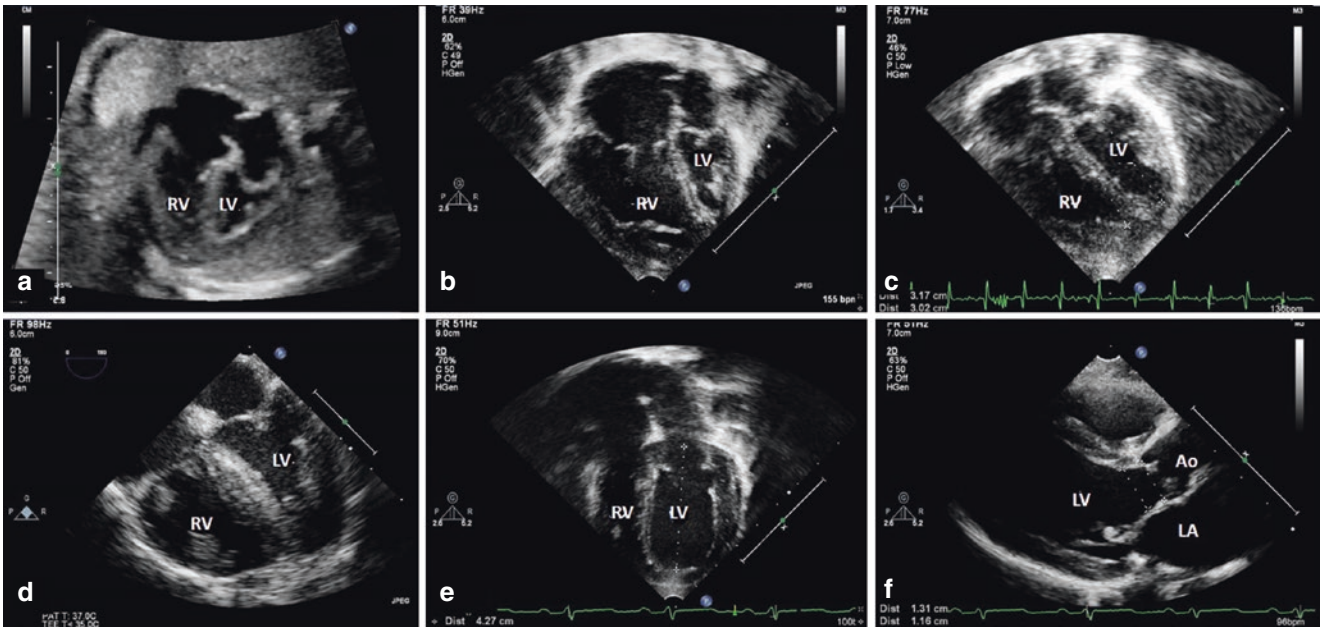



**Fig. 9.6** Staged rehabilitation of the left ventricle in a fetus with critical aortic stenosis. Echocardiographic progression. (**a**) Echocardiographic assessment at 27 weeks gestational age before fetal aortic valvuloplasty. The LV is already small (LV length *Z*-score −2.7) and dysfunctional. Maternal oxygen was given to the mother for 2 weeks after valvuloplasty. (**b**) Significant improvement of the LV size is seen after birth. The neonate underwent a hybrid procedure with bilateral pulmonary artery banding and stent implantation in the arterial duct followed by atrial septostomy. (**c**) At 6 months of age, there is some further increase in the LV size. (**d**) At 10 months of age, the infant underwent LV overhaul with resection of endocardial fibroelastosis and aortic valve plasty. A small (4 mm) atrial septal defect was left open for left atrial decompression early in the postoperative period. Intraoperative transesophageal echocardiogram shows significant improvement in the LV size (LV length *Z*-score −1.2) and normal function. (**e**) and (**f**) Late appearance of the LV at 5 years of age with normalization of LV dimensions and normal systolic and diastolic function. Abbreviations: *LA* left atrium, *RV* right ventricle, *LV* left ventricle, *Ao* ascending aorta

that such patients do better postnatally even if the LV is not recruited for a BV circulation.

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.

Although fetuses with established HLHS who underwent successful in utero ASD creation or enlargement are born with better clinical conditions and higher saturations, surgical mortality after the Norwood operation remains higher than in HLHS patients who did not require in utero ASD interventions [3]. Finding new ways to create unrestricted holes at an earlier gestational age in these fetuses is still the unmet clinical need. Creating small holes (even with stent placement) at late gestation is not efficacious to prevent the development of secondary pulmonary vascular, lymphatic and parenchymal changes.

Although in utero pulmonary valvuloplasty is more challenging from the technical standpoint, the late outcomes of fetuses with PA/IVS or CPS/IVS who underwent a successful procedure are more rewarding. Such patients show 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. We have advocated repeat pulmonary valvuloplasty and ductal stenting soon after birth as part of a staged and longer strategy to rehabilitate the RV. This enables further time for satisfactory growth of the RV during infancy. Spontaneous closure of the duct is usually seen late in infancy with effective pulmonary blood flow provided solely by the RV. A BV outcome is the rule rather than exception in such patients [2].

## References

1. Pedra SF, Peralta CF, Pedra CAC. Future directions of fetal interventions in congenital heart disease. Interv Cardiol Clin. 2013;2:1–10.
2. Pedra SR, Peralta CF, Crema L, Jatene IB, da Costa RN, Pedra CA. Fetal interventions for congenital heart disease in Brazil. Pediatr Cardiol. 2014;35:399–405.
3. Moon-Grady AJ, Morris SA, Belfort M, Chmait R, Dangel J, Devlieger R, Emery S, Frommelt M, Galindo A, Gelehrter S, Gembruch U, Grinenco S, Habli M, Herberg U, Jaeggi E, Kilby M, Kontopoulos E, Marantz P, Miller O, Otaño L, Pedra C, Pedra S, Pruetz J, Quintero R, Ryan G, Sharland G, Simpson J, Vlastos E, Tworetzky W, Wilkins-Haug L, Oepkes D. International fetal cardiac intervention registry: a worldwide collaborative description and preliminary outcomes. J Am Coll Cardiol. 2015;66(4):388–99.

# Part IV

# Step-by-Step Procedures: Valve Dilatation

# Aortic Valvular Stenosis

# 10

Xiangbin Pan

Congenital aortic stenosis (CAS) is a relatively common congenital heart disease (1.1–4.3 per 10,000 live births). The aortic annulus of patients with CAS is usually hypoplastic to some extent; the leaflets are thickened, and the commissures, to different degrees, are fused. Usually newborns with critical CAS suffer from low cardiac output and shock secondary to poor left ventricular function. In older children and adolescents with severe CAS, 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. Currently, percutaneous balloon valvuloplasty has become an important procedure for the treatment of CAS.

## 10.1 Indication for Treatment

- Regardless of valve gradient, the newborn with isolated critical CAS is ductal dependent or children with isolated CAS have depressed left ventricular systolic function.
- Patients with isolated CAS have a resting peak systolic valve gradient of >50 mmHg by catheter or ≧75 mmHg.
- Patients with isolated CAS have a resting peak systolic valve gradient (by catheter) of >40 mmHg if there are symptoms of angina or syncope or ischemic ST-T-wave changes on electrocardiography at rest or with exercise.

## 10.2 Preprocedure Imaging and Preparation

- Echocardiography can provide the following information: the morphology of the aortic valve, peak instantaneous and mean aortic valve gradient by Doppler, aortic valve annulus diameter (Fig. 10.1) and *z*-score, left ventricular dimensions, left ventricular shortening fraction and ejection fraction, severity of aortic valve regurgitation, and other lesions (Fig. 10.2a, b)
- Hemodynamic assessments: pressure in the left ventricle and ascending aorta (Fig. 10.3)
- Aortic angiography: depicting the details of the valve anatomy and assessing the aortic annulus diameter between the hinge points of the valve leaflets (Fig. 10.4)

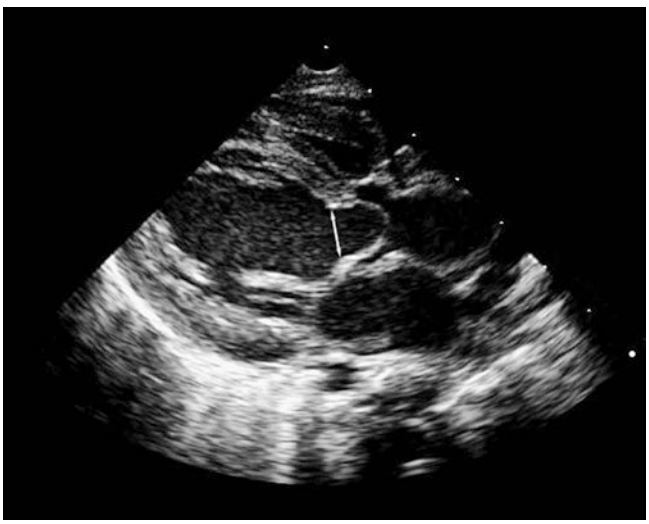


**Fig. 10.1** Echocardiographic measurement of the aortic valve annulus between the hinge points of the valve leaflets (double-headed arrows) in the parasternal long-axis view

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_10) contains supplementary material, which is available to authorized users.

X. Pan (✉)
Department of Cardiovascular Surgery, National Center for Cardiovascular Disease, China & Fuwai Hospital, Beijing, China


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_10

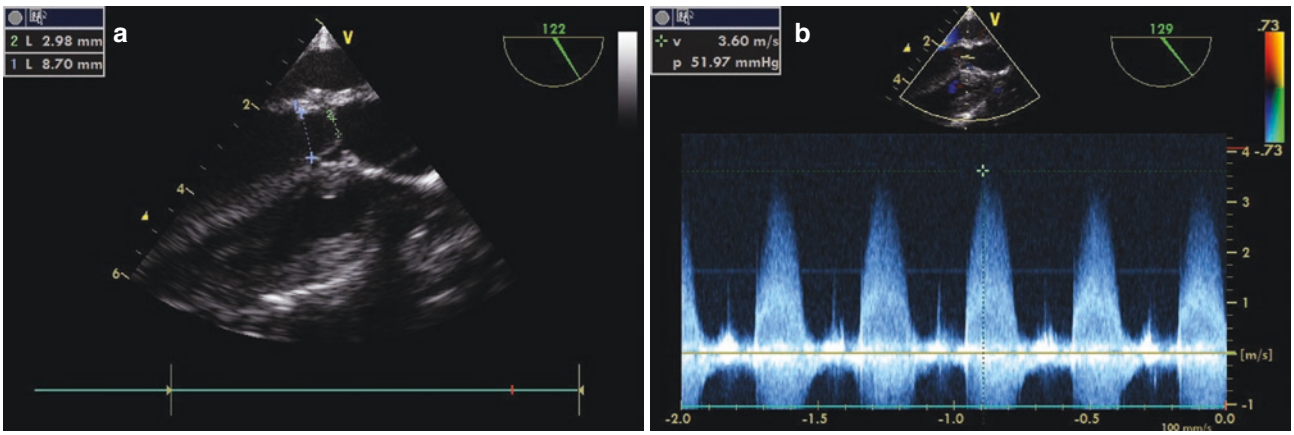



**Fig. 10.2** Transesophageal echocardiography demonstrates a 2-month-old girl (patient 1) with CAS. The aortic valve annulus is 8.7 mm (**a**). Although the peak systolic aortic valve gradient is about 52 mmHg, the ejection fraction is 31% with poor left ventricular function (**b**)

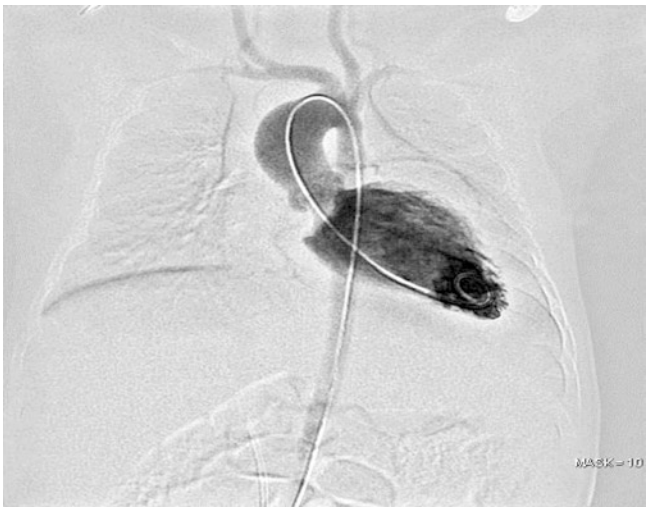


**Fig. 10.3** Patient 1: Left ventricular angiography in posterior-anterior projection shows CAS, and the ascending aorta is significantly expanded

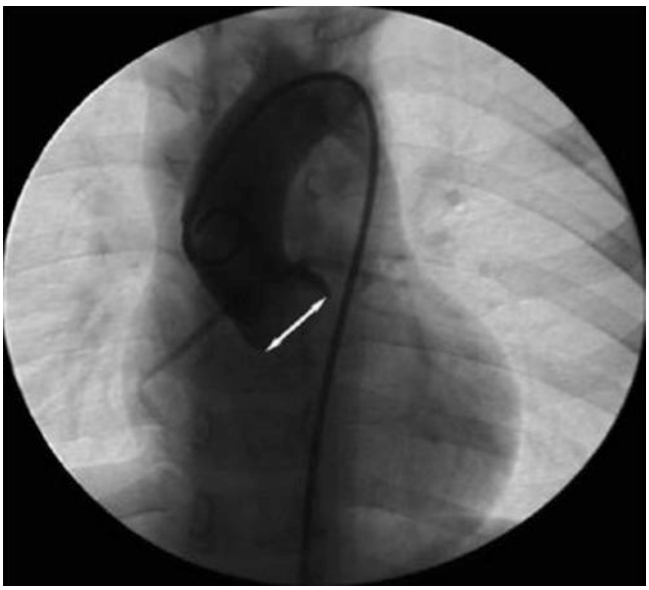


**Fig. 10.4** The aortic valve annulus is measured by aortography in posterior-anterior projection. The double-headed arrow indicates the aortic annulus diameter between the hinge points of the valve leaflets

## 10.3 Catheter Intervention

- Wires and balloon catheter (Fig. 10.5) 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 or the umbilical artery in newborns. The antegrade approach is an alternative way by using a femoral venous access or an umbilical vein when the guidewire cannot cross a severely stenotic aortic valve from the retrograde approach.
- The techniques of valvuloplasty include single- (Fig. 10.6) and double-balloon valvuloplasty. Compared with single-balloon (Fig. 10.7) valvuloplasty, double-balloon valvuloplasty has the advantages of improved gradient relief, less vessel trauma, and smaller risk of completely occluding left ventricular outflow.
- In single-balloon valvuloplasty, 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. But in double-balloon valvuloplasty, each balloon has a similar diameter and length, so the ratio of the sum to the valve annulus diameter is about 1:3.

**Fig. 10.5** Tyshak and Tyshak II (NuMED, Inc.) provide a wide range of balloon diameters, from 4 to 30 mm, with 1-mm increments up to 25 mm (except 21 and 24 mm). Tyshak can better resist the left ventricular ejection power and is therefore preferred in older children. Tyshak II is preferable in infants in whom the lowest possible introducer profile is important. Tyshak Mini usually applies to neonates

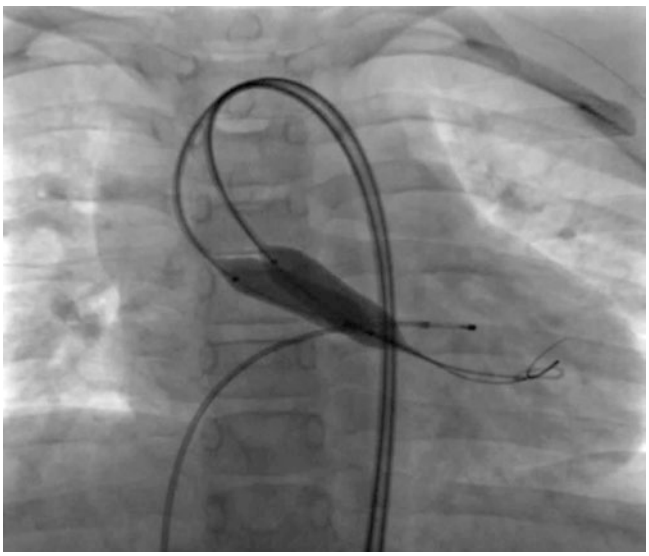


**Fig. 10.7** Patient 2: Double-balloon technique in posterior-anterior projections applied in a 1-year-old patient. The aortic valve was dilated by simultaneous inflation of two 6 mm × 20 mm Tyshak II balloon valvotomy catheters introduced into the valve from both the femoral arteries

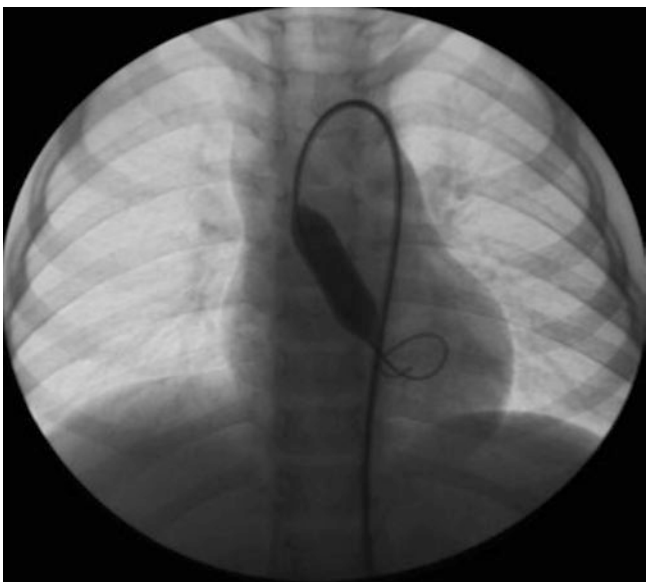


**Fig. 10.6** Patient 1: Advance an 8 mm × 20 mm Tyshak II balloon valvotomy catheter over the guidewire, straddle the valve into the correct position by retrograde approach, and inflate with a pressure of 4–7 ATM until the balloon waist disappears

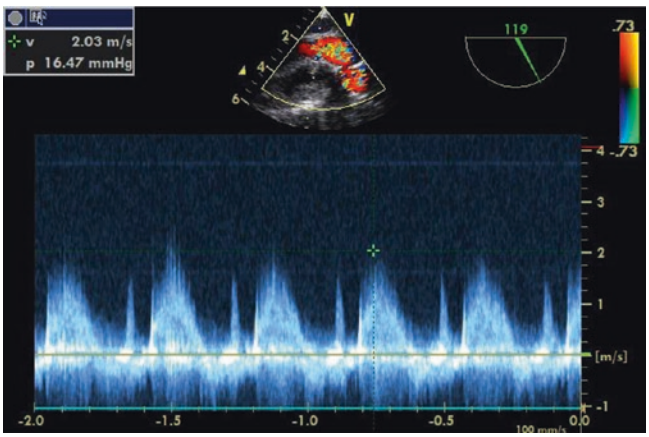



**Fig. 10.8** Patient 1: Postoperative transesophageal echocardiography demonstrates the peak systolic aortic valve gradient is about 16.5 mmHg. The ejection fraction increased significantly to 62% immediately

apex and using VVI pacing at a rate of 220–240 impulses per minute during the balloon inflation.

– 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. Each inflation-deflation period lasts no more than 5–10 s. Temporary rapid pacing is performed by putting a bipolar pacing catheter in the right ventricular

## 10.4 Assessment of Success (Fig. 10.8)

– More than 50% decrease in pressure gradient across the aortic valve and no significant aortic regurgitation.
– A residual gradient of <30 mmHg is usually aimed.

## 10.5 Complications

- Aortic regurgitation is a potentially serious complication. About 15% of patients experience moderate or severe aortic regurgitation after balloon valvuloplasty.
- Vascular complications: especially in newborns in which femoral artery access is used.
- Arrhythmia: transient bradycardia and left bundle-branch block and premature beats.
- Cardiac structural damage: mitral valve tears, annulus tears, and heart perforation.
- Death: a dilated balloon may completely block the blood flow in the aorta and induce fatal ventricular fibrillation and asystole. The early mortality rate is about 4%.

**Video 1** Angiography in the left anterior oblique (LAO) in the ascending aorta from a right internal carotid artery approach. Blood accelerating through the stenotic aortic valve is seen as a negative jet within the contrast dye injected in the ascending aorta (WMV 2368 kb)

**Video 2** Left ventricular angiography in LAO showing a hypertrophic left ventricle and dysplastic and thick aortic valve. Mitral valve is seen as a negative scado on the right of the aortic valve (WMV 2240 kb)

**Video 3** Fluoroscopy showing balloon inflation. The guidewire is looped within the left ventricle. During balloon inflation, waist appears at the level of the aortic valve. When the balloon is fully inflated and valve well opened, the waist disappears (WMV 2304 kb)

**Video 4** Left ventricular angiography in LAO after balloon dilation. The left ventricle is hyperdynamic and hypertrophic. The apex of the left ventricle is almost completely filled by the muscle during systole. The aortic valve shows a significantly improved opening. No signs of extravasation are seen (WMV 1632 kb)

**Video 5** Ascending aortography in LAO after valvuloplasty. The injection is performed with an over the wire approach. No aortic regurgitation is seen (WMV 1536 kb)

# Step-by-Step Procedure: Pulmonary Valve Stenosis

# 11

Tingliang Liu and Wei Gao

Pulmonary valve stenosis is one of the most common congenital heart defects, in which the pulmonary valves opening from the right ventricle are restricted. Two clinical scenarios may occur: (1) critical pulmonary stenosis in neonates and (2) pulmonary valve stenosis in infants, children, and adolescents. Neonates with critical pulmonary stenosis are cyanotic and require prostaglandin infusion to maintain pulmonary blood flow. And patients with pulmonary valve stenosis beyond neonatal period are mostly asymptomatic, unless severe stenosis is present. With growth, moderate stenosis may cause symptoms, including fatigue, chest pain, arrhythmias, limited exercise tolerance, and cyanosis.

Indication for treatment:

- Critical valvar pulmonary stenosis:
  This is an urgent procedure; sometimes a low gradient associated with a right ventricular dysfunction may be found.
- Pulmonary valve stenosis beyond neonatal period:
  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 RV dysfunction. It is reasonable to perform pulmonary valvuloplasty on a patient with valve stenosis who meets the above criteria in the setting of a mild-to-moderate dysplastic pulmonary valve.

Diagnostic cardiac catheterization:

- Complete hemodynamic assessment (pressures in the right atrium, RV, pulmonary arteries, and pullback gradient assessment)
- Angiographic depiction of RV and pulmonary arterial anatomy (Fig. 11.1a, b. RV angiography is performed usually using Berman angiographic catheter or pigtail catheter. In the severe case with significant tricuspid valve insufficiency, RV angiography can be done using end-hole catheter by manual control, especially in neonates.)
- Measure the pulmonary valve annulus to localize the stenotic cite precisely.

Balloon dilation:

- Positioning of the guidewire: Make sure that the guidewire does not pass through the tendinous chords and papillary muscles of tricuspid valve. Once the balloon catheter has been advanced across chords and papillary muscle of the tricuspid valve, the new premounted balloon might be advanced across the chords, but the inflated-deflated balloon after the procedure can cause chordae rupture and severe tricuspid valve insufficiency (Figs. 11.2 and 11.3).
- Single balloon dilation (Figs. 11.4 and 11.5): Usually a balloon diameter 1.0–1.2 times the pulmonary valve annulus is chosen, and the length of the balloon should be 20 mm in newborns and infants, 30 mm in children, and 40 mm in adolescents and adults.
- Special techniques: (1) Step-by-step dilation for severely stenotic pulmonary valve, especially in neonates (Videos 1, 2, 3, and 4). (2) Double balloon dilation in case of too large pulmonary valve annulus (Fig. 11.6).

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_11) contains supplementary material, which is available to authorized users.

T. Liu · W. Gao (✉)
Department of Cardiology, Shanghai Children's Medical Center, Shanghai, China



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_11

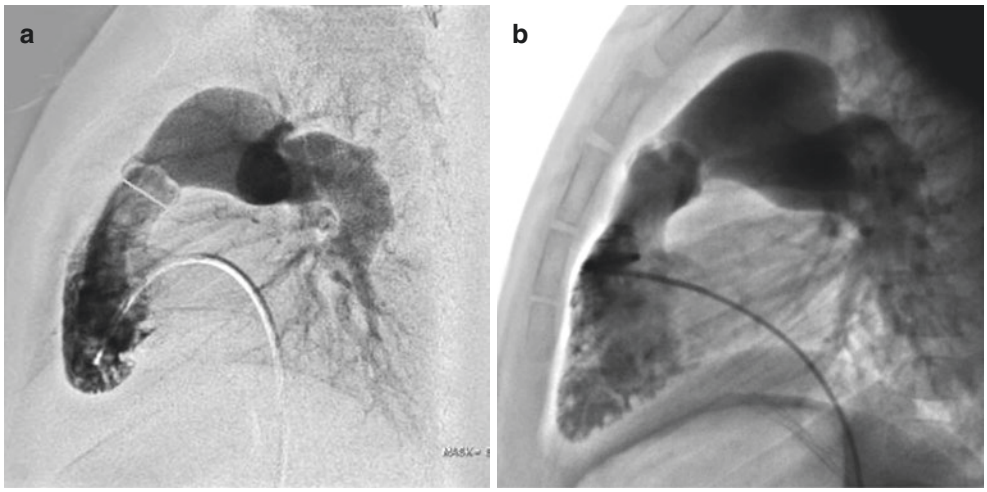



**Fig. 11.1** (**a**, **b**) Right ventriculogram in the left lateral view showing pulmonary valve stenosis: thickened, domed pulmonary valve, and poststenotic dilatation of the main pulmonary artery. The white line shows the length of the valve annulus ((**a**) using Berman angiographic catheter; (**b**) using pigtail catheter)

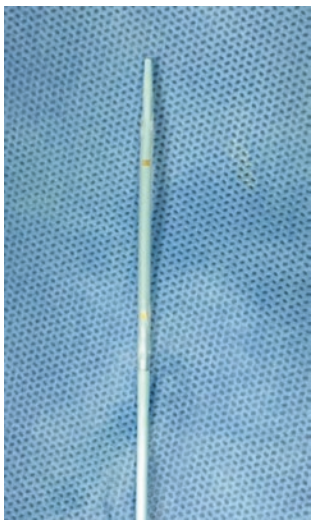


**Fig. 11.2** A new premounted balloon. The balloon is well packed and might be advanced across chords and papillary muscle of the tricuspid valve

**Fig. 11.3** An inflated-deflated balloon has much bigger size than a new premounted one. The pullback of an inflated-deflated balloon after the procedure can cause chordae rupture and severe tricuspid valve insufficiency

**Assessment of Success**

Pressure gradient across the pulmonary valve should be measured by catheter and echocardiogram after dilation. Regression of right ventricular hypertrophy on the electrocardiogram following balloon dilatation has been well documented. Echocardiography plays an essential role in follow-up of patients with pulmonary valvular stenosis. The Doppler gradient is generally reflective of the residual obstruction and is a useful and reliable noninvasive monitoring tool.

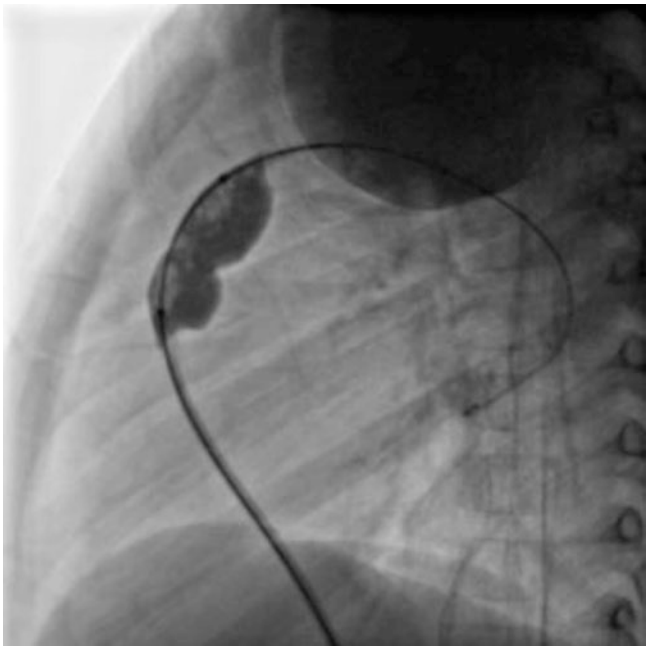


**Fig. 11.4** Left lateral view of a partially inflated balloon catheter positioned across the stenotic valve. As the balloon is inflated, a waist appears at the site of the pulmonary valve

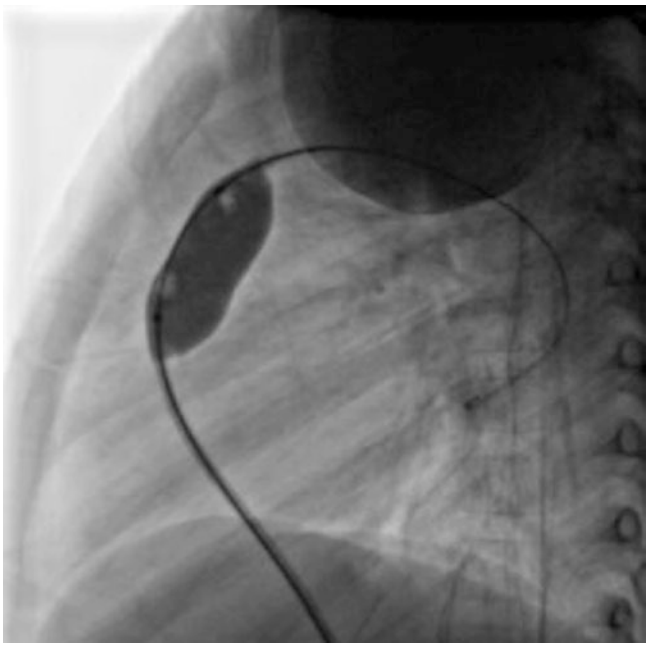


**Fig. 11.5** Left lateral view of the balloon waist completely disappeared

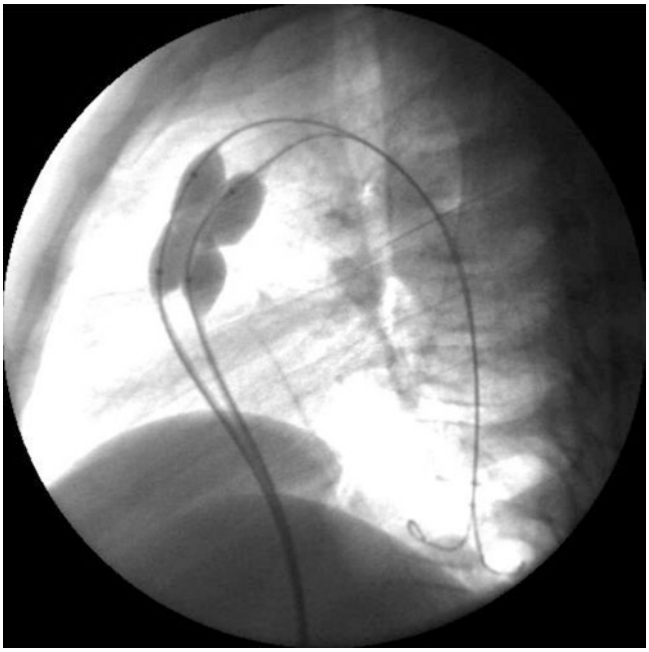


**Fig. 11.6** Left lateral view of two balloon catheters positioned across the pulmonary valve showing waisting of balloons produced by the stenotic valve

**Video 1** Right ventriculogram in the left lateral view showing critical pulmonary valve stenosis (AVI 16895 kb)

**Video 2** Small balloon has been advanced through guidewire (AVI 50126 kb)

**Video 3** Dilation with small balloon (AVI 51857 kb)

**Video 4** Dilation with bigger balloon after changing guidewire (AVI 27697 kb)

# Pulmonary Atresia and Intact Ventricular Septum

# 12

Mazeni Alwi and Zaheer Ahmad

## 12.1 Introduction

Contrary to its name that suggests a simple isolated lesion where there is complete obstruction of the right ventricular outflow to the pulmonary arteries, pulmonary atresia with intact ventricular septum (PAIVS) encompasses a wide range of anatomic abnormalities of the right heart structures from the tricuspid valve (TV), the right ventricular (RV) cavity and its three parts [the inlet, apical trabecular and infundibulum or outflow tract (RVOT)], the pulmonary valve and pulmonary arteries. Apart from complete obstruction to RV outflow which is usually in the form of an imperforate valve, there is varying degrees of hypoplasia of the right heart structures and overgrowth of RV musculature. Varying degrees of TV regurgitation are common due to dysplasia of the valve. RV-coronary connections (sinusoids) is a well-known association of this disease especially in those with diminutive RV cavity due to obliteration by muscles of all but the inlet part of the RV cavity (unipartite RV). The coronary circulation is considered "RV dependent" when myocardial perfusion is largely from the hypertensive RV and antegrade flow is absent or inadequate due to ostial atresia, proximal interruption or severe stenosis of the major coronary arteries.

For those with unipartite RV which is essentially a functional single ventricle, treatment is along the lines of the Fontan pathway. In patients where the RV has potential to function as pulmonary ventricle where the RVOT ends blindly with a membranous imperforate valve, interventional procedure with valve perforation and balloon dilation (and stenting of the patent ductus arteriosus in those with borderline RV size) has become the first-choice initial treatment. Surgical re-interventions however are commonly required, e.g. tricuspid valve repair, enlargement of the RVOT or apical part and bidirectional Glenn anastomosis in patients where the RV is inadequate as an independent pump (1 ½ ventricle circulation). Even in the most favourable anatomy, re-interventions in adult life are likely to be required in many patients due to severe tricuspid or pulmonary regurgitation.

(a) *Valve perforation and balloon dilation (±PDA stent)*
*Patient selection*
Obviously only patients with patent infundibulum that ends blindly with an imperforate valve are suitable for this procedure. However there is also a spectrum of anatomy for this subgroup of patients. In patients with the most favourable anatomy, the success rate of the procedure is high, and the complications rate is low. These are patients who have well-developed RV with wide, smooth infundibulum with equally reasonable-sized pulmonary valve annulus and an imperforate membranous valve. The main pulmonary artery and sinuses are also well developed. In some of these patients, only the inlet and RVOT are reasonably well developed, but the apical part is obliterated by muscles (non-apex forming, bipartite RV). Coronary sinusoids are not uncommon in these patients.

The less favourable anatomy for valve perforation and balloon dilation are those with smaller bipartite RV, and the infundibular lumen is small and irregular due to heavy musculature. The valve annulus is equally small, and the imperforate valve-rather than a thin mobile membrane-is a thick plate. The MPA and sinuses are also small and poorly developed. In these patients the valve may require a higher energy to perforate, and the risk of perforating outside the heart is higher in part due to the less well-developed main pulmonary artery (MPA) sinuses. Major coronary sinusoids are more common in

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_12) contains supplementary material, which is available to authorized users.

M. Alwi (✉) · Z. Ahmad
Institut Jantung Negara (National Heart Institute),
Jalan Tun Razak, Kuala Lumpur, Malaysia
e-mail: mazeni@ijn.com.my



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_12

this subgroup of patients. RV-dependent coronary circulation needs to be excluded by detailed angiography before valve perforation and balloon dilation. This may be difficult to determine in the absence of ostial atresia/severe stenosis or proximal interruptions of the major coronary arteries.

(b) *Exclusion criteria for balloon dilation*

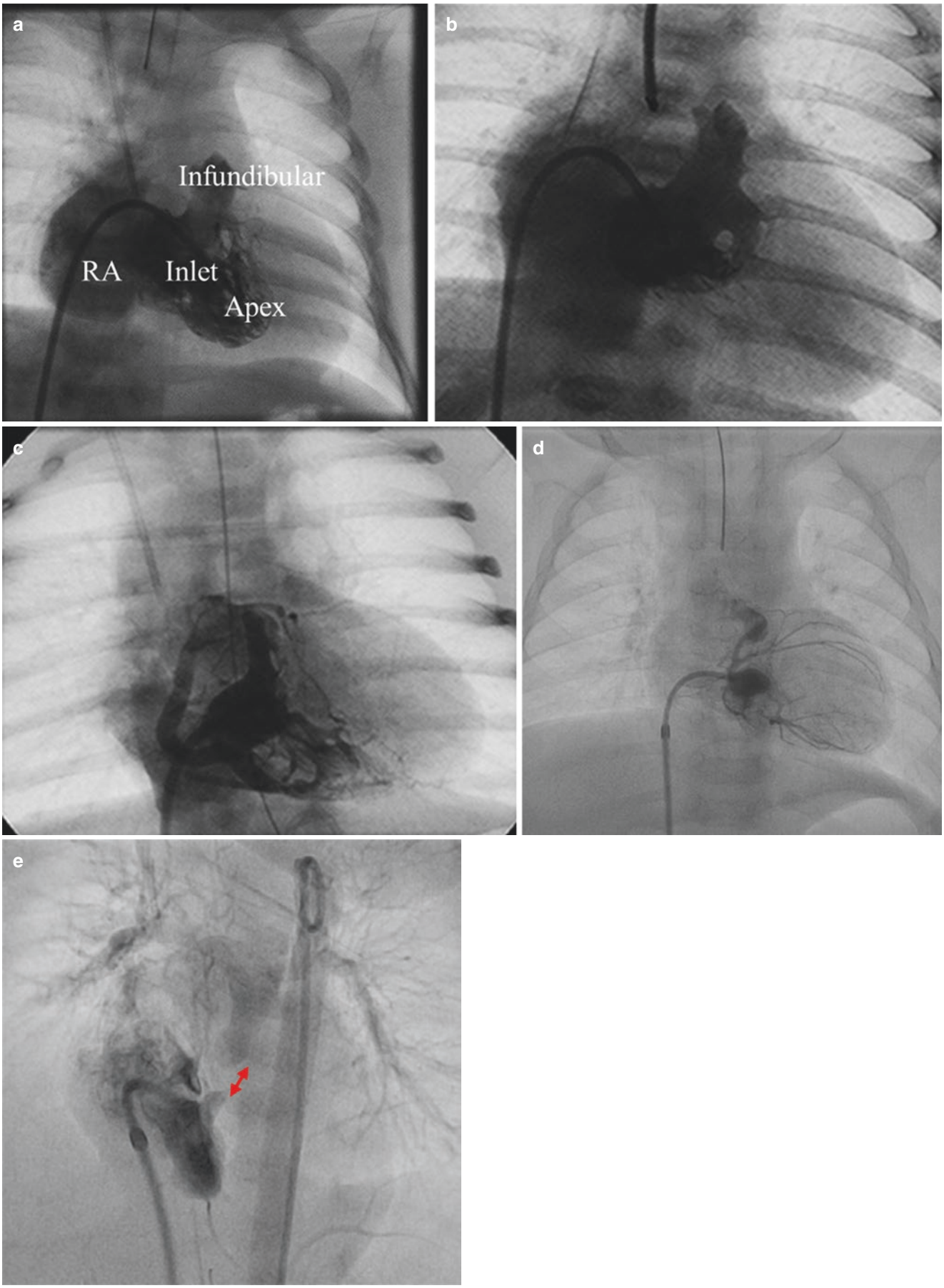
(i) **Muscular atresia of RVOT**
In patients with unipartite RV, only a severely hypoplastic inlet guarded by a very small TV constitutes the RV, with the apex and infundibulum largely obliterated by muscles. Occasionally in patients with better-sized RV, the proximal part of the infundibulum is present, but distally it is filled by muscles.

(ii) **Severe tricuspid regurgitation due to dysplastic TV or true Ebstein's anomaly**
In these patients the RA and RV are markedly dilated. The overriding clinical problem is that of severe right heart failure due to severe valve regurgitation. Balloon dilation is unlikely to alleviate the problem, and urgent surgical valve repair and pulmonary valvotomy/RVOT reconstruction is the more appropriate treatment. Furthermore the RVOT is usually markedly dilated with relatively small valve annulus, making RF valvotomy very challenging. The risk of perforating outside the heart is likely to be high.

(iii) **RV-dependent coronary circulation**
Sinusoids are more common in patients with very small RV that are more likely destined for the single ventricle pathway. However they may also be present in patients with patent infundibulum where it is feasible to open the outflow tract and decompress the RV. In a small number of patients where there is ostial atresia or severe proximal stenosis/interruption of the major coronary arteries, the diagnosis of RV-dependent coronary circulation can be readily determined. This is an absolute contraindication to balloon dilation. However without these obvious features, determining RV dependence with certainty is difficult. We speculate that very small size of a major coronary artery may also suggest RV-dependent coronary circulation even in the absence of stenosis, for when the RV is decompressed, perfusion from the antegrade flow from the aorta may be inadequate and lead to ischaemia. The risk of ischaemia/infarction vs. the long-term outcome of Fontan operation needs to be balanced in the decision-making process.

In Figs. 12.1, 12.2, 12.3, 12.4 and 12.5, all angiograms are in the anteroposterior (AP) projection or lateral unless indicated.

**Fig. 12.1** Case selection—suitable anatomy for valve perforation and balloon dilatation and those in whom this is inappropriate. Tripartite RV—well-developed inlet, apical parts and wide, smooth infundibulum that ends blindly with an imperforate valve. The RA is dilated from severe TR. There are no RV-coronary connections (**a**). "Bipartite" RV—the inlet and infundibulum are well developed but the apical part is almost obliterated by muscles. There are no RV-coronary connections. There is moderate TR (**b**). Bipartite RV with apical part as in (**b**) above. There is major RV to RCA connection but no obstruction proximally of the coronary artery. There are minor connections to the LCA. The tricuspid valve is competent (**c**). The atresia is membranous (**a**), (**b**) and (**c**), with well-developed infundibulum. These patients' anatomies are suitable for valve perforation-balloon dilation. Unipartite hypertensive RV where only a small inlet is present. The TV is competent. The left and right coronary systems are perfused from the RV. There is free reflux of contrast into the aorta via the dilated proximal LCA (**d**). Long-segment muscular atresia of the infundibulum (red arrow). Simultaneous contrast injection in the RV and aorta to opacify the MPA in LAO cranial view. A slightly better-sized RV with inlet and a short segment of RVOT present proximally. However there is a long segment of muscular atresia, the red arrow indicating the gap between the infundibulum and MPA. There are no RV-coronary connections (**e**)


a
Infundibular
RA
Inlet
Apex
b
c
d
e

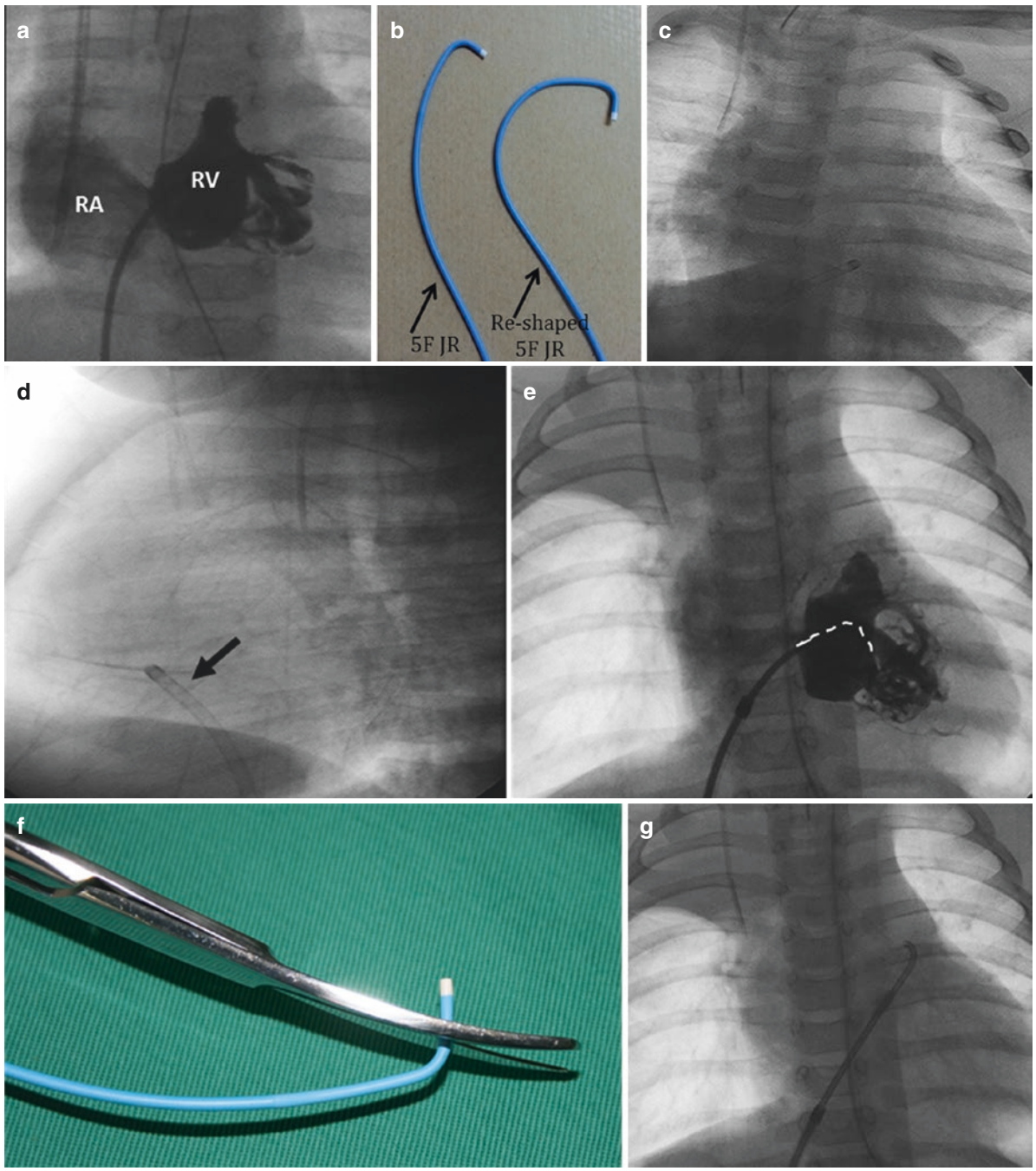



**Fig. 12.2** Catheterizing the RV and RVOT for initial angiography. Diagnostic angiography is an important step prior to intervention to measure the RV pressure, assess the RV size and morphology and determine the presence/absence of RV-coronary connections. Further anatomic details of the RVOT, the pulmonary annulus and valve tissue and the main pulmonary artery are obtained by placing a catheter tip in the RVOT underneath the atretic valve. The same catheter is used for delivering the RF wire for valve perforation. This angiogram shows that the RV mainly consists of the inlet and a well-developed infundibulum (bipartite). The apex is largely obliterated by muscles save for some inter-trabecular spaces. There is mild TR (**a**). In view of the angulation of the RVOT to the RV body, the Judkins right (JR) catheter of 4–5F is often used. A commonly encountered problem is crossing the tricuspid valve for a stable catheter position because of shape of the JR catheter. This is compounded by the dilated RA when there is significant TR, small TV annulus and shallow RV cavity due to overgrowth of muscles in the apical part. This may be facilitated by reshaping the JR catheter (**b**). Another method of overcoming this is the use of a Mullin's catheter whose tip would naturally face the TV once the dilator is removed as shown by the black arrow on the lateral view (**c**, **d**). The JR catheter is then introduced coaxially into the RV (**c**, **d**). Because of the shallow RV and heavy trabeculations, it may be difficult to manoeuvre into the RVOT and position it under the valve for further angiography and RF wire delivery (**e**). This may be facilitated by cutting the distal tip while retaining its primary curve (**f**, **g**). *Abbreviations*: *RV* right ventricle, *RVOT* RV outflow tract, *JR* Judkins right, *RF* radiofrequency, *RA* right atrium, *TR* tricuspid regurgitation, *TV* tricuspid valve

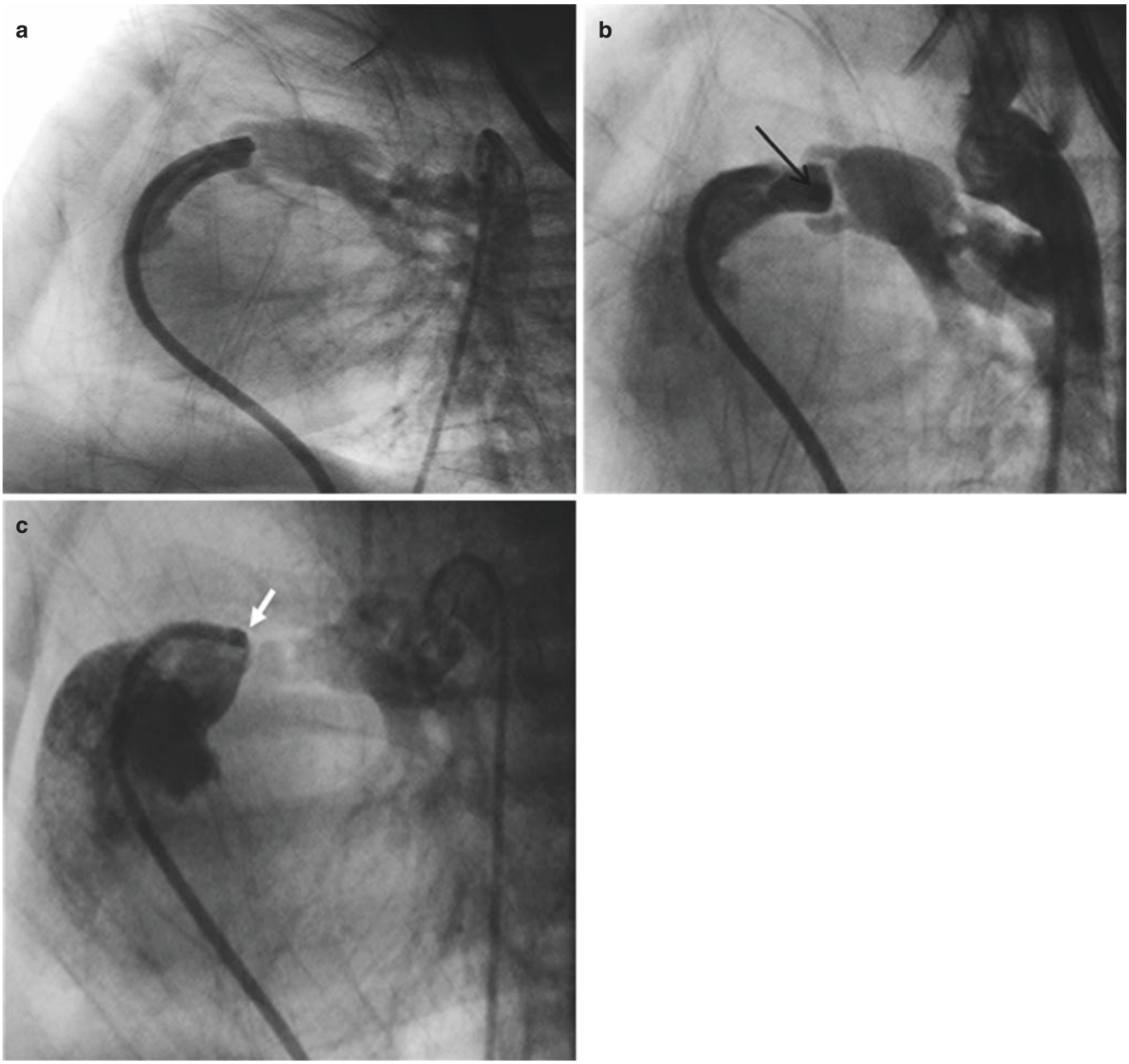



**Fig. 12.3** Profiling the RVOT, valve annulus, pulmonary valve tissue and main pulmonary artery. A wide, smooth-walled infundibulum that ends in an atretic membranous valve as indicated by a black arrow in (**b**), MPA sinuses cup over the blind-ending RVOT (**a**, **b**). In this patient with Ebstein's anomaly and severe TR, the RV and RVOT are markedly dilated. The pulmonary valve annulus and MPA are small, with poorly developed sinuses (white arrow). There is a high likelihood of perforation outside the heart with the RF wire. The overriding clinical problem is that of severe tricuspid regurgitation which needs to be treated surgically (**c**)

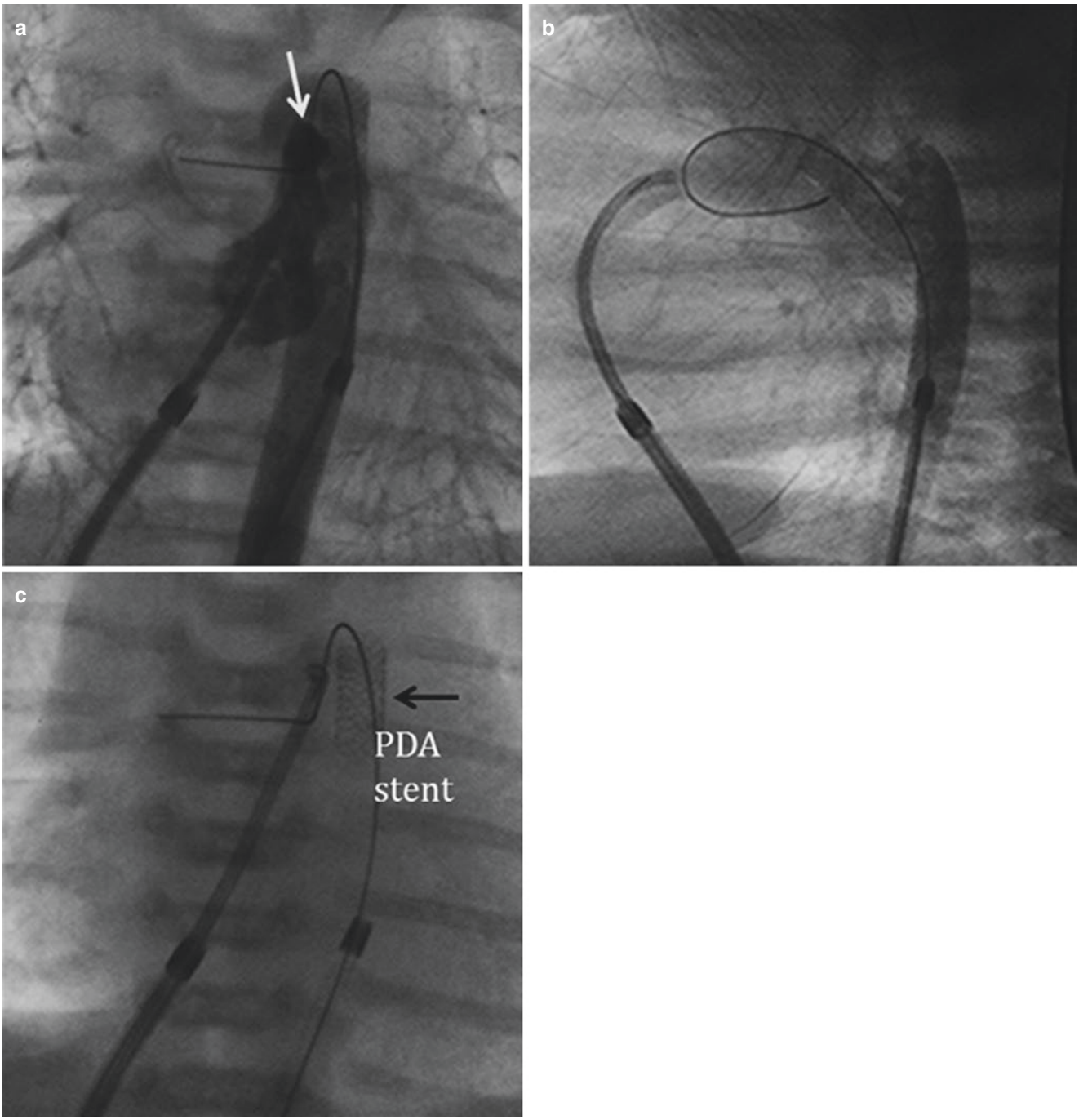



**Fig. 12.4** Use as guidewire in MPA as target for RF wire perforation. A patient with small bipartite RV but having a well-developed infundibulum and membranous atresia (**a**). She was severely hypoxic despite PGE1. The PDA was stented before valve perforation and balloon dilation. Arrow shows the membranous valve with tip of JR catheter and RF wire positioned on the central part and tenting the valve (**b**). The tip of Choice PT wire which was used for stent delivery is curved in the MPA and proximal RPA, making a useful target for directing the RF wire. The tip of JR catheter needs to be turned 5–10° rightwards to ensure that the RF wire does not perforate outside the MPA (**c**). *Abbreviations*: *PDA* patent ductus arteriosus, *RF* radiofrequency, *MPA* main pulmonary artery, *RPA* right pulmonary artery

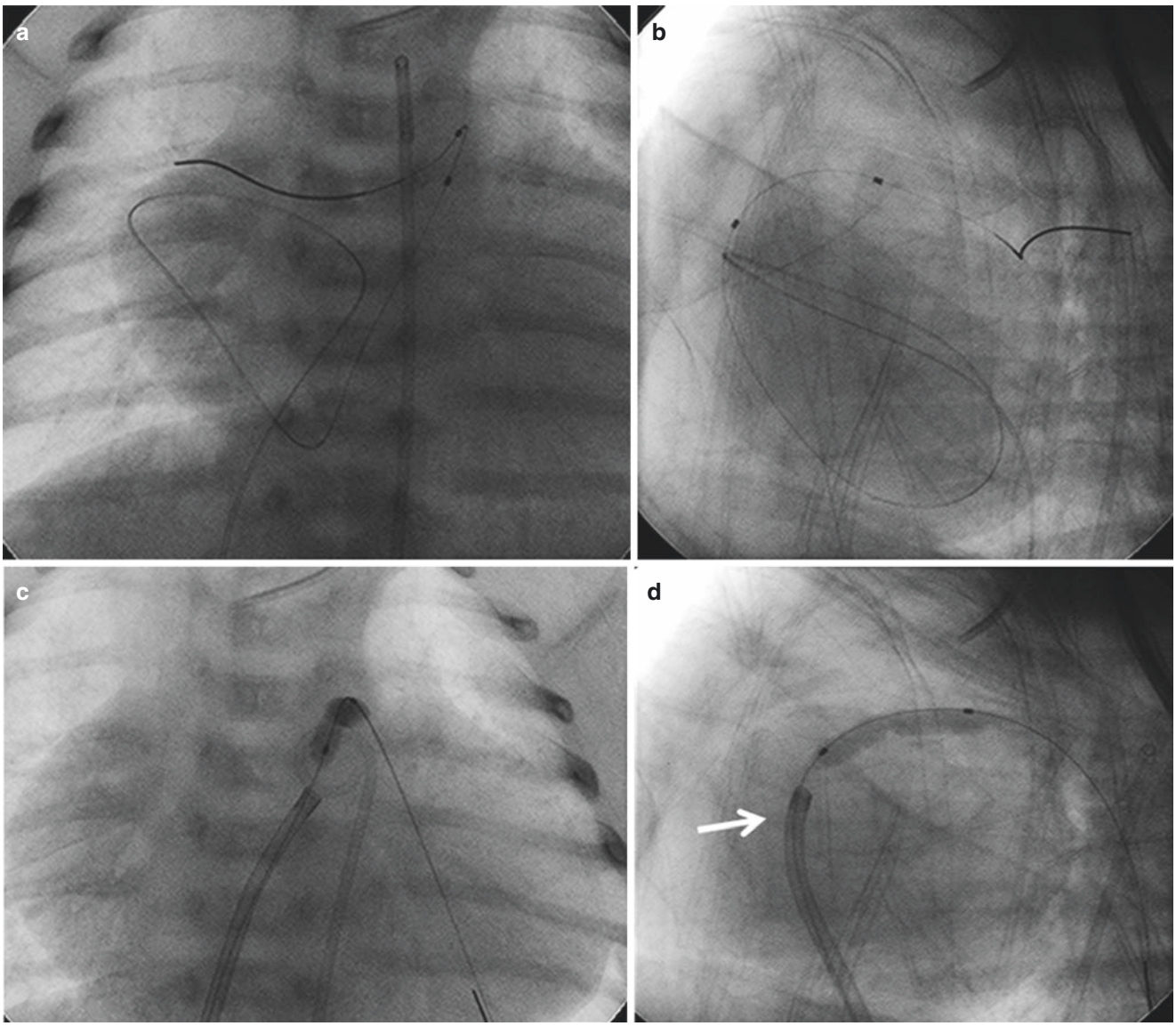



**Fig. 12.5** Tracking of coronary balloon over guidewire. In patients with relatively small RV and dilated RA due to significant TR, the wire and balloon catheter may loop in the RA, preventing satisfactory positioning of balloon across the valve. This may lead to loss of guidewire position (**a**, **b**). Changing the JR diagnostic catheter with a 5F JR guiding catheter after valve perforation is a useful method of overcoming this problem. Arrow indicates tip of 5F JR guiding catheter (**c**, **d**). When the PDA is not too constricted at the pulmonary end, the tip of coronary wire may be directed into the descending aorta to provide a stable position for tracking of balloon across the valve. During balloon dilation there is a possibility that the balloon is advanced partly into the PDA. Inadvertently dilating the PDA in this manner makes the patient unsuitable for PDA stenting should this be required. *Abbreviations*: *RV* right ventricle, *RA* right atrium, *TR* tricuspid regurgitation, *PDA* patent ductus arteriosus

**Video 1** A patient with a small but tripartite RV and membranous atresia who would otherwise be suitable for valve perforation and balloon dilation but the coronary circulation is thought to be RV dependent. The contrast in the apical part is less dense compared to the inlet and infundibulum indicating hypertrophied muscles. A dilated LAD fills retrogradely from the RV apex, seen to the left of the spine on the AP view and on the anterior border of the cardiac silhouette on the lateral view, red arrows (Video 1: shots 1 and 2). Aortic root angiogram in RAO view shows severe stenosis of LCA at the ostium (Video 1: shot 3). *Abbreviations*: *RV* right ventricle, *TV* tricuspid valve, *LCA* left coronary artery, *MPA* main pulmonary artery, *RVOT* RV outflow tract, *PAIVS* pulmonary atresia intact ventricular septum, *TR* tricuspid regurgitation, *RA* right atrium, *PDA* patent ductus arteriosus, *LAD* left anterior descending, *AP* anteroposterior, *RAO* right anterior oblique, *LAO* left anterior oblique (PPTX 620 kb)

**Video 2** Profiling the RVOT, valve annulus, pulmonary valve tissue and main pulmonary artery. In the lateral projection, with the tip of JR catheter positioned underneath the valve and a pigtail catheter opposite the PDA in the aorta, a near-simultaneous contrast injection is done to opacify the ROVT and the MPA to assess the characteristics of the infundibulum, the valve annulus size, thickness of the valve tissue and the MPA size and sinuses relative to the RVOT. The most favourable cases have wide, smooth-walled infundibulum with a thin valve membrane which is cupped over by the well-developed sinuses of the MPA as in the first video. The risk of perforating outside the heart is small. In the second video, the pulmonary valve is a thick plate, suggesting that a higher dose of energy may be required for perforation. Nevertheless the valve annulus and MPA are of good size although the sinuses are poorly developed. *Abbreviations*: *PDA* patent ductus arteriosus, *RVOT* right ventricular outflow tract, *MPA* main pulmonary artery, *TR* tricuspid regurgitation, *RV* right ventricle, *RF* radio frequency, *RCA* right coronary artery, *LCA* left coronary artery (PPTX 1150 kb)

**Video 3** Series of videos demonstrating valve perforation with radio frequency wire followed by step-wise balloon dilation. Aortic angiography opposite the PDA confirms that the tip of 5F JR catheter is abutting the centre of the imperforate valve, with the MPA and sinuses cupping over the RVOT. On the AP view, the pigtail catheter position corresponds to the central part of the MPA. This serves as a guide for directing the RF wire. The RF wire (Cereblate, Osypka AG, 79618 Rheinfelden, Germany) is pushed forwards firmly but without exerting too much force (Video 3: shot 3) perforating the valve smoothly. In most cases the wire should go slightly rightwards (Video 3: shot 4) towards the pigtail catheter suggesting that it is within the MPA lumen. This is confirmed by aortic angiography following valve perforation (Video 3: shots 1 and 2). Alongside the RF wire, a straight-tipped coronary guidewire of moderate stiffness (Choice PT Extra Support, Boston Scientific, Marlborough MA01752, USA) is passed into the pulmonary artery, again confirmed by angiography (Video 3: shot 3). The valve is dilated initially using a 3.0 mm coronary balloon followed by an 8.0 mm balloon (Tyshak mini, Numed Canada, Cornwall ON K6J 1G3) (Video 3: shots 4 and 5). *Abbreviations*: *PDA* patent ductus arteriosus, *RVOT* right ventricular outflow tract, *AP* anteroposterior, *MPA* main pulmonary artery, *RF* radiofrequency (PPTX 4601 kb)

**Video 4** Use of coronary wire with stiffer tip (for chronic total occlusion, CTO wire) for valve perforation as alternative to RF wire in favourable cases. For patients with thin membranous valve and well-developed infundibulum and MPA, a CTO wire may be used as an alternative to RF wire if this is unavailable. However it is not as effective, and the risk of perforation outside the heart is likely to be higher because of the force required. Shots 1–3 in Video 4 demonstrate a successful perforation with a CTO wire (Conquest Pro, Asahi Intecc, Aichi 489-0071, Japan). This same wire was then used for balloon dilation. Unsuccessful perforation with a CTO wire in a patient with a thin membranous valve that appeared suitable (Video 4: shot 1). RF wire at 5 W failed to perforate the valve (Video 4: shot 2). Successful perforation at 10 W (Video 4: shot 3). *Abbreviations*: *MPA* main pulmonary artery, *CTO* chronic total occlusion, *RF* radio frequency (PPTX 3778 kb)

**Video 5** Complications. Perforation outside the heart which may cause tamponade. This complication is more likely to occur in patients with less favourable anatomy, i.e. those with more severe muscular overgrowth and smaller RV cavity. They tend to have small valve annulus with thick valve plate and less well-developed MPA and sinuses. In this patient with bipartite RV, the RVOT and the pulmonary valve annulus are small with thick immobile pulmonary valve plate. The MPA and the sinuses are also relatively small. RV-coronary connections are also present (Video 5: shots 1 and 2). On the lateral view, the RF has perforated on the margin of the valve and into the pericardial space outside the MPA anteriorly (Video 5: shot 3), although on the AP view it appears to be within the MPA lumen (Video 5: shot 4). The JR catheter was repositioned with its tip facing more inferiorly to effect successful perforation into the MPA and RPA lumen (Video 5: shots 5 and 6). Balloon dilation was successfully performed. A small amount of contrast is seen in the pericardial space. *Abbreviations*: *RV* right ventricle, *RVOT* RV outflow tract, *MPA* main pulmonary artery, *RF* radio frequency, *AP* anteroposterior, *RPA* right pulmonary artery (PPTX 2344 kb)

# Part V

# Step-by-Step Procedures: Vessel Treatment

# Stent Implantation in Patients with Pulmonary Arterial Stenosis

13

Andreas Eicken and Peter Ewert

## 13.1 Introduction

Pulmonary arterial branch stenosis may occur congenitally or after surgical procedures. Newborns and adults with congenital heart disease may be affected. All sections of the pulmonary arterial tree may be involved: the main pulmonary artery or its origin, the central branch pulmonary arteries, and the segmental pulmonary arteries. In newborns and young infants with pulmonary arterial stenosis after surgical procedures involving the pulmonary arteries, balloon angioplasty may be employed as first-line treatment, if repeated surgery does not seem adequate. In these small patients, large stents dilatable to adult size vessel diameter cannot be implanted since large stents on large balloons need large introduction sheaths. However, balloon angioplasty may not always lead to an acceptable flow increase of the stenotic lung segment and even in this difficult patient group stent implantation may be the only solution. In general, stent implantation is the preferred first-line treatment in patients with pulmonary arterial stenosis, in whom a stent can be implanted, which can be expanded to an adult size diameter though.

## 13.2 Pathophysiology

A severe vessel stenosis (<50% diameter of the adjacent normal pulmonary artery) leads to a detectable systolic pressure gradient, which can be measured by a catheter pullback across the stenosis. If several lung segments are stenotic and in case of biventricular circulation a pressure rise in the subpulmonic ventricle (usually the right ventricle) is seen. Doppler Echocardiography helps to locate and quantify the flow acceleration across a stenotic vessel and in case of tricuspid regurgitation the systolic pressure in the right ventricle (RVP normal <30 mmHg) can be assessed. After noninvasive measurement of the patient's blood pressure, the pressure ratio between the right ventricle (RVP) and the aorta (AoP) can be diagnosed. In patients with univentricular hearts the source of pulmonary blood flow is important. During first-stage palliation (for example, Norwood operation in hypoplastic left heart syndrome) a right ventricular-to-pulmonary artery (Sano shunt) or an aortopulmonary shunt (modified Blalock-Taussig shunt) assures pulmonary blood flow. In impeded pulmonary blood flow, the arterial oxygen saturation may be lowered to critical values. Again, Doppler echocardiography is helpful to show a flow acceleration in the pulmonary circulation. The flow acceleration may not directly be transferred into a systolic pressure gradient in mmHg. In patients with univentricular circulation the RVP:AoP ratio is not helpful. After a cavopulmonary shunt in univentricular circulation (PCPC = partial cavopulmonary and TCPC = total cavopulmonary connection) the pressure gradient is of less importance, since there is no subpulmonary pumping chamber, and the pressure gradients are, if at all present, very low. The pre-stenotic pressures may rise and this may lead to severe symptoms (cyanosis due to veno-venous collaterals, effusions, "failing Fontan" circulation).

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_13) contains supplementary material, which is available to authorized users.

A. Eicken (✉)
Department of Pediatric Cardiology and Congenital Heart Disease
German Heart Center Munich, Technische Universität München, Munich, Germany
e-mail: eicken@dhm.mhn.de

P. Ewert
Klinik für Kinderkardiologie und angeborene Herzfehler
Deutsches Herzzentrum München, Munich, Germany


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_13

## 13.3 Indication for Treatment in Pulmonary Arterial Vessel Stenosis

Biventricular circulation treatment indication:

- RVP:AoP pressure ratio > 66% (if RV function is impaired, an intervention may be indicated at lower RVP:AoP ratios).
- Significantly abnormal (RL) pulmonary arterial flow distribution assessed by perfusion scans or by cMRI (normal is R:L = 60:40%), significantly abnormal is a perfusion distribution of 80% or more to one lung.
- Systolic pullback gradient >20 mmHg.
- Angiographic stenosis with <50% lumen diameter compared to the normal adjacent pulmonary vessel.

Univentricular circulation treatment indication:

- Arterial desaturation (<65% $SaO_2$) early after an aortopulmonary or right ventricle-to-pulmonary artery shunt operation.
- Arterial desaturation after a cavopulmonary shunt operation in the early postoperative period.
- Elevated central venous pressures with signs of congestion.
- Signs of impaired pulmonary arterial blood flow even in mild angiographic stenosis.

## 13.4 Stent Implantation

A complete cardiac catheterization with pressure assessment and catheter pullback at the site of the stenosis is performed. Following angiographic depiction of the anatomy a guidewire is advanced over the stenotic region and positioned distally into the pulmonary artery. A balloon stent assembly (either premounted or manually crimped) is chosen with a balloon diameter of the adjacent normal pulmonary vessel. A suitable sheath (coronary guide catheter in newborns or infants) or a long sheath is advanced. If possible, this sheath is advanced over the wire to pass the stenosis. The balloon/stent assembly is advanced over the wire to the target region. Then the sheath is withdrawn and small contrast injections through the sheath ascertain correct stent position. Then the balloon/stent is inflated, the balloon is withdrawn, and the result is documented by angiography. If feasible a repeated catheter pullback may be performed. The RVP:AoP ratio acquisition may be repeated to document an adequate hemodynamic effect of the procedure in patients with biventricular circulation.

## 13.5 Stents

In newborns and small infants stent implantation into the pulmonary arteries may be a lifesaving procedure, for example, after a Norwood-operation (stenotic modified BT shunt or stenotic Sano shunt). Sometimes these patients are on ECMO due to postoperative cyanosis. If the stent needs to be delivered through an aortopulmonary shunt, it may be necessary to use an arterial access (4F sheath, for example, Terumo Radifocus introducer II 25 cm; Terumo Deutschland GmbH, Eschborn, D) through which only coronary stents can be delivered. This sheath is stiff and cannot be advanced into the pulmonary arteries through the shunt; hence, the premounted coronary stent has to be advanced carefully to the target without cover. In our unit we use the cobalt chrome Coroflex blue neo premounted stent (B. Braun AG, Melsungen, Germany) for this indication. These stents can be expanded to a maximum diameter of 5 mm. Later on, the stents have to be removed surgically, cut open longitudinally and widened with a patch, or they must be cracked open with ultrahigh-pressure balloons.

In larger infants, or if a venous access can be used, stents from the Cook Formula™ family (Cook Medical; Bloomington, Indiana, USA) have proven to be very helpful recently (Fig. 13.1). The Formula™ 414 RX 6 × 12 mm stent can be implanted on a 0.014′ guidewire through a 5F long sheath or through a 6F coronary guide catheter. Shorter (65 cm) 5F and 6F coronary guide catheters are available and very helpful due to their flexibility. The short 5F Coronary guide catheter may be used to deliver a coronary stent as well. The Formula™ 414 stent may be expanded up to 12 mm before it breaks. The Formula™ 535 (8 × 12 mm stent can be expanded to 14 mm and the Formula™ 10 × 20 mm stent can be expanded to 16 mm, without significant foreshortening) stents are delivered over a 0.035′ guidewire through 6F and 7F long sheaths, respectively.

In our experience (bench tests with high-pressure balloon dilatation) performed with Formula™ 414 RX 6 × 12 mm, 535 8 × 12 mm, and 535 10 × 20 mm stents are dilatable to larger diameters mentioned above before the stent struts rupture (Fig. 13.2).

In older children and adults, stents dilatable to an adult size vessel (>20 mm) are available. The Max LD stent (Medtronic, MN, USA) is available in lengths between 16 and 36 mm and has an open cell stent design. The Mega LD stents (Medtronic, MN, USA) may be delivered on smaller balloons and through smaller sheaths, but their radial forces are less. The Andra xxl stents (Fig. 13.3) (Andramed, Reutlingen, D) have a hybrid stent design (open and closed cells) and are available in lengths from 13 to 57 mm. Another frequently used stent is the Cheatham platinum stent

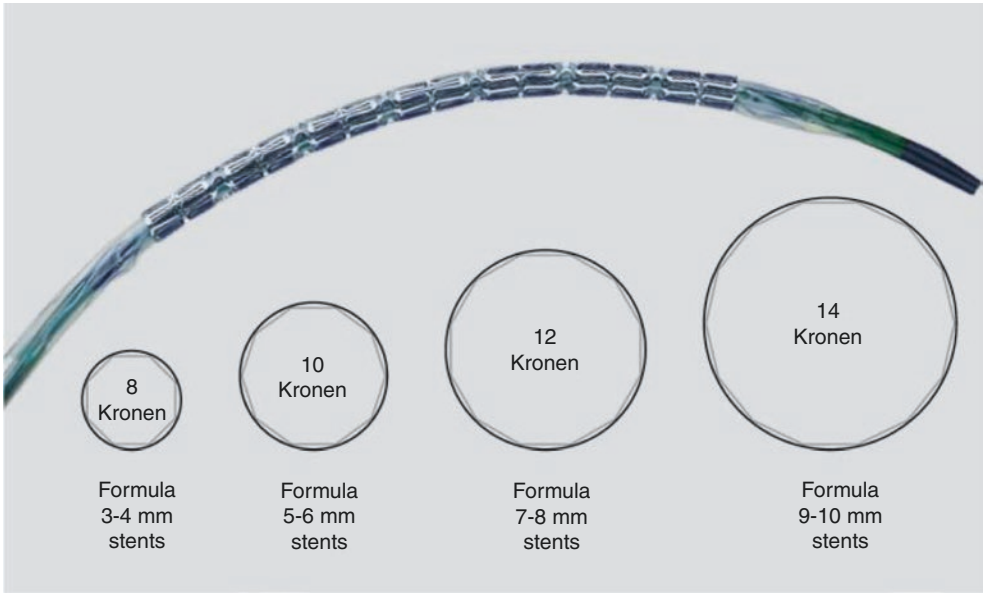



**Fig. 13.1** Premounted Cook Formula™ stents (Cook Medical; Bloomington, Indiana, USA) mounted on 6, 8, and 10 mm balloons. Kronen = stent crowns

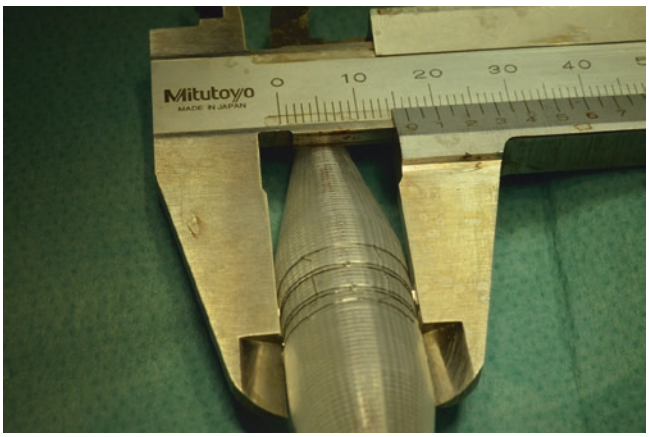



**Fig. 13.2** The 535 Cook Formula™ 8 × 12 mm stent was expanded to 14 mm diameter. At overdilatation with a 16 mm Atlas high-pressure balloon (Bard Medical, Murray Hill, New Jersey, USA), a stent strut ruptured

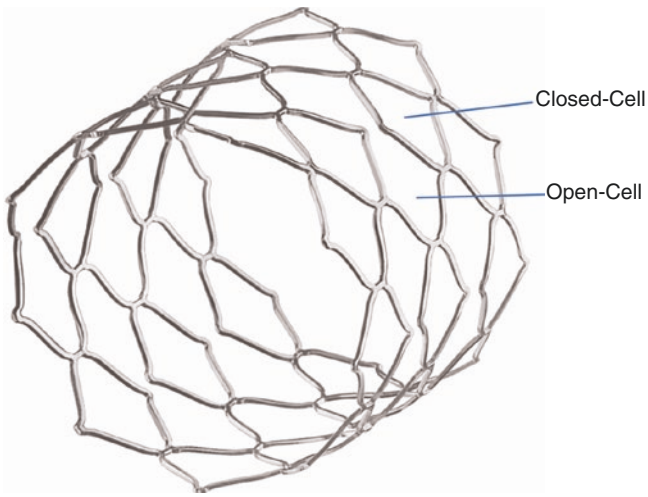



**Fig. 13.3** Andramed xxl stent (Andramed Reutlingen, D) has a hybrid cell design showing closed and open stent cells

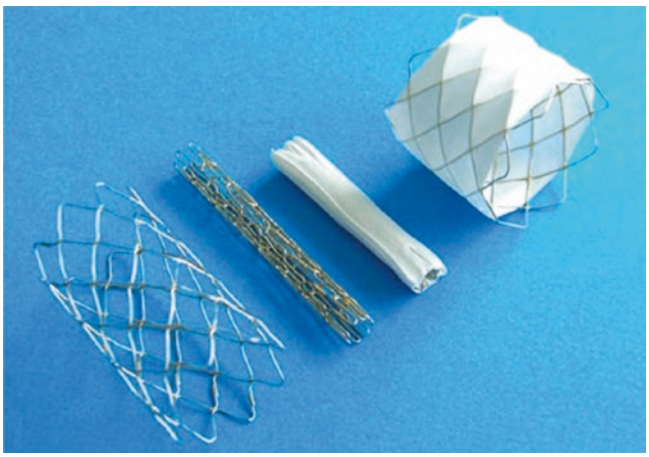
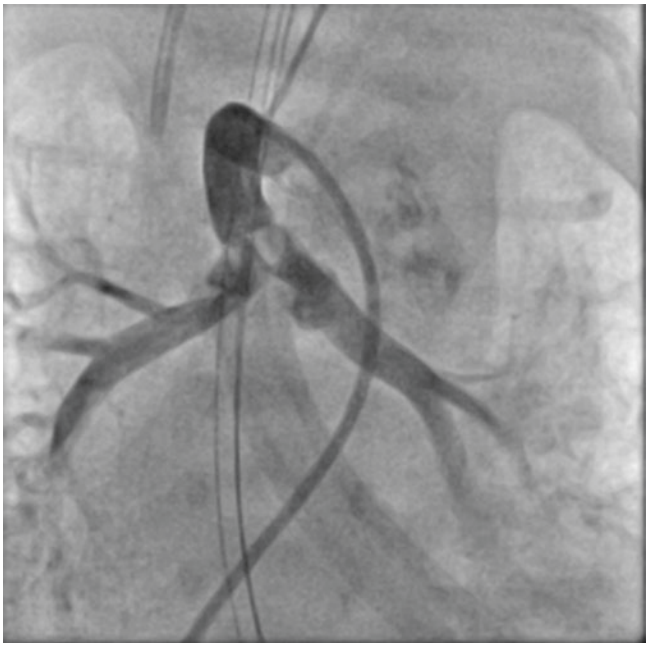


**Fig. 13.4** Cheatham 8z platinum stent, on the right an expanded bare metal and on the left an expanded covered 8z Cheatham platinum stent

(Fig. 13.4) (Numed, Hopkinton, NY, USA) which has a closed cell design and is available in lengths between 16 and 45 mm. This stent is also available as a covered stent.

### 13.5.1 Patient Examples

Example 1: Cyanotic newborn after a Norwood-Sano operation for hypoplastic left heart syndrome with severe stenosis of the distal Sano shunt anastomosis (Figs. 13.5–13.10).



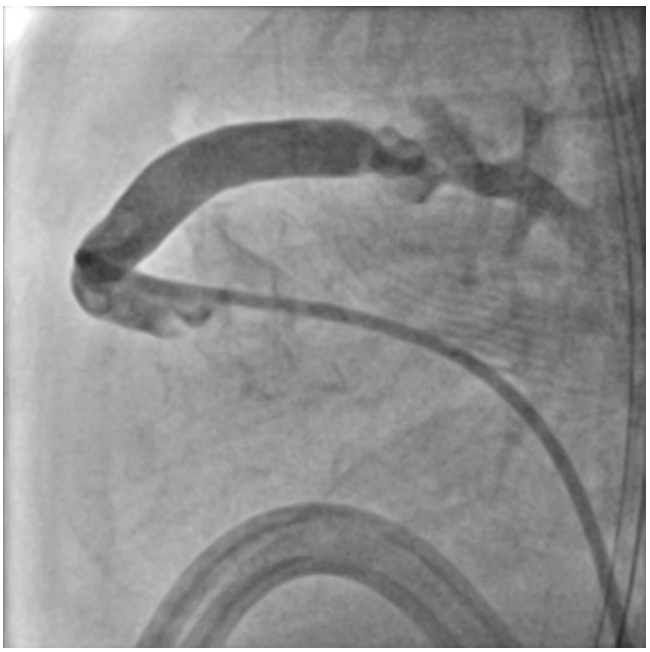


**Fig. 13.5 and 13.6** Cyanotic newborn (50 cm, 2.8 kg) 8 days after a Norwood-Sano operation (5 mm RV-PA shunt), ventilated with 100% oxygen. Arterial oxygen saturation < 65%. Angiography shows a severe distal stenosis of the Sano shunt involving both pulmonary arteries

**Video 1** Cyanotic newborn (50 cm, 2,8 kg) 8 days after a Norwood-Sano operation (5 mm RV-PA shunt), ventilated with 100% oxygen. Arterial oxygen saturation <65%. Angiography shows a severe distal stenosis of the Sano shunt involving both pulmonary arteries (WMV 2075 kb)

**Video 2** Cyanotic newborn (50 cm, 2,8 kg) 8 days after a Norwood-Sano operation (5 mm RV-PA shunt), ventilated with 100% oxygen. Arterial oxygen saturation < 65%. Angiography shows a severe distal stenosis of the Sano shunt involving both pulmonary arteries (WMV 1881 kb)

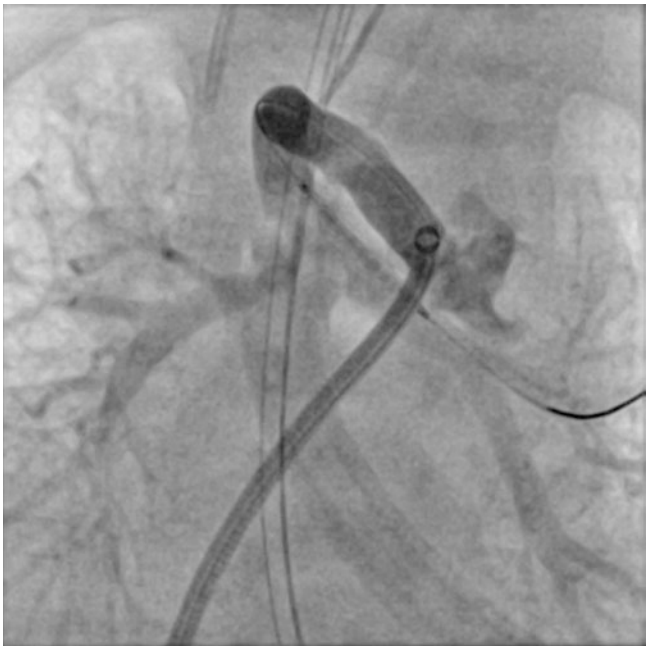


**Fig. 13.7** Through a 5F Terumo 25 cm long sheath a 0.014′ coronary guidewire was positioned over a 4F modified coronary right catheter distally into the left pulmonary artery and a 4 × 13 mm Coroflex blue (Braun Melsungen, D) stent was advanced into the LPA-Sano shunt anastomosis

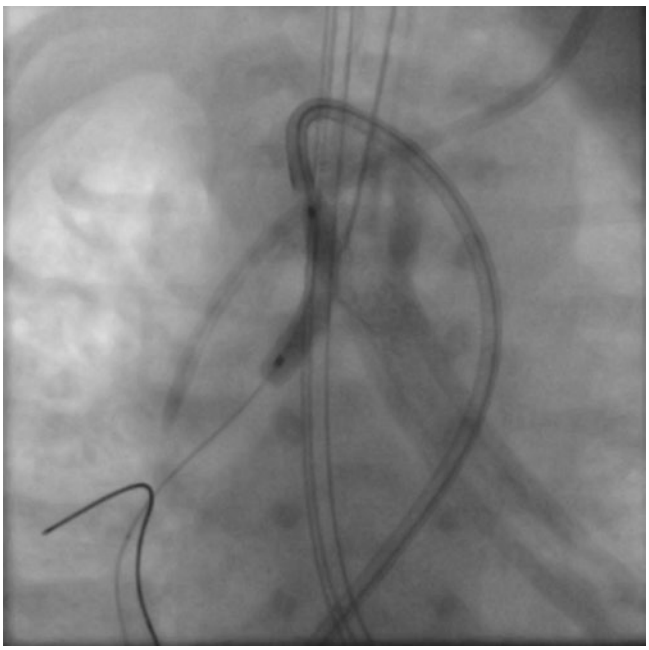


**Fig. 13.8** After inflation of the stent in the left pulmonary artery, a 5F coronary guide catheter (Medtronic Launcher 65 cm; MN, USA) was advanced into the Sano shunt and the LPA-Coroflex blue stent was crossed proximally to the right pulmonary artery with a 0.014′ coronary wire. Over the wire, a 3 × 20 mm Maverick coronary balloon (Boston Scientific Marlborough, MA, USA) was inflated in the right pulmonary artery and the stent cells were opened

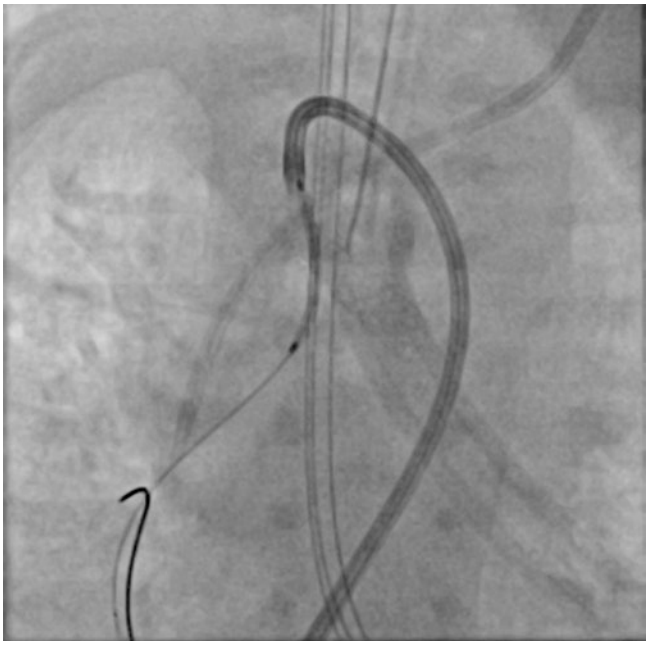


**Fig. 13.9** A second Coroflex blue 4 × 13 mm stent was positioned across the first stent into the right pulmonary artery through the 5F short coronary guide catheter

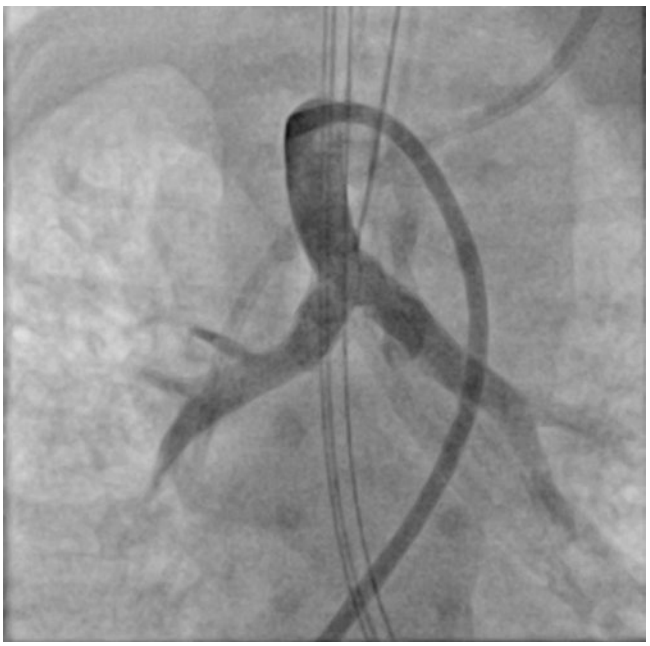


**Fig. 13.10** Final result with unobstructed flow to both pulmonary arteries. Arterial oxygen saturation rose from <65% to >75%. A Y-stent configuration of the pulmonary bifurcation was created

**Video 3** Final result with unobstructed flow to both pulmonary arteries. Arterial oxygen saturation rose from <65% to >75%. A Y-stent configuration of the pulmonary bifurcation was created (WMV 2097 kb)

**Video 4** Final result with unobstructed flow to both pulmonary arteries. Arterial oxygen saturation rose from <65% to >75%. A Y-stent configuration of the pulmonary bifurcation was created (WMV 2063 kb)

Example 2: A one year old boy (9,1 kg) with common arterial trunk after neonatal corrective surgery (VSD patch and valveless communication RV-PA), and after re-operation at the age of 5 months with a Contegra 14 mm conduit and patch enlargement of the pulmonary arteries presented with elevated right ventricular pressures (Figs. 13.11–13.13).

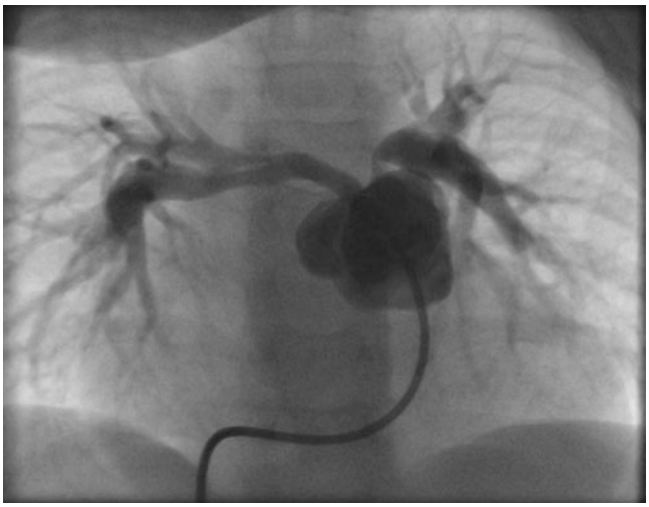


**Fig. 13.11** One year old child with common arterial trunk 6 months after implantation of a 14 mm Contegra graft (RV-PA) now with RVP 88/0/12, PAS 20/13/15, PAD 21/12/14, AoP 105/46/66 (RVP:AoP ratio 88%)

**Video 5** One year old child with common arterial trunk 6 months after implantation of a 14 mm Contegra graft (RV-PA) now with RVP 88/0/12, PAS 20/13/15, PAD 21/12/14, AoP 105/46/66 (RVP:AoP ratio 88%) (WMV 1550 kb)

**Video 6** One year old child with common arterial trunk 6 months after implantation of a 14 mm Contegra graft (RV-PA) now with RVP 88/0/12, PAS 20/13/15, PAD 21/12/14, AoP 105/46/66 (RVP:AoP ratio 88%) (WMV 1353 kb)

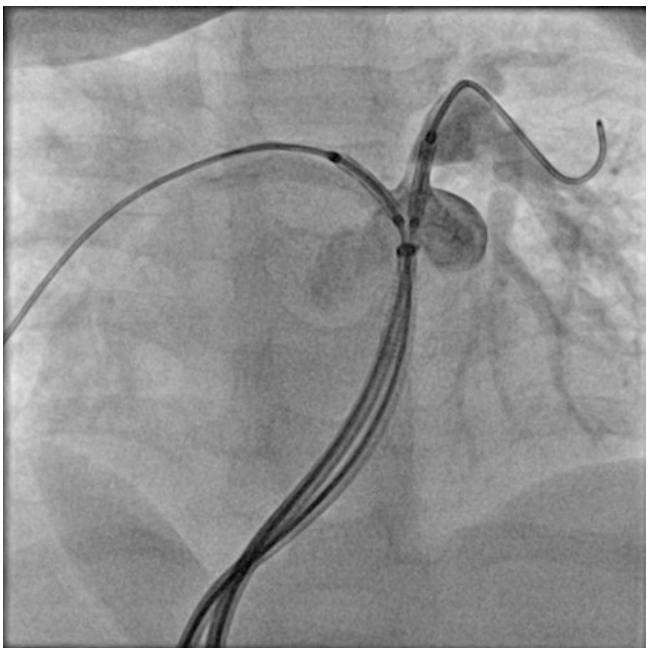


**Fig. 13.12** Two 6F Terumo 45 cm long sheaths (via both femoral veins) were advanced on 0.035′ guidewires into the right ventricular outflow tract. A 6 × 12 mm Formula™ 535 stent was positioned into the origin of the right pulmonary artery and a 8 × 12 mm Formula™ 535 into the origin of the left pulmonary artery. The stents were inflated simultaneously

**Video 7a** Two 6F Terumo 45 cm long sheaths (via both femoral veins) were advanced on 0.035′ guidewires into the right ventricular outflow tract. A 6 × 12 mm Formula™ 535 stent was positioned into the origin of the right pulmonary artery and a 8 × 12 mm Formula™ 535 into the origin of the left pulmonary artery. The stents were inflated simultaneously (WMV 972 kb)

**Video 7b** Two 6F Terumo 45 cm long sheaths (via both femoral veins) were advanced on 0.035′ guidewires into the right ventricular outflow tract. A 6 × 12 mm Formula™ 535 stent was positioned into the origin of the right pulmonary artery and a 8 × 12 mm Formula™ 535 into the origin of the left pulmonary artery. The stents were inflated simultaneously (WMV 5469 kb)

**Video 8** Two 6F Terumo 45 cm long sheaths (via both femoral veins) were advanced on 0.035′ guidewires into the right ventricular outflow tract. A 6 × 12 mm Formula™ 535 stent was positioned into the origin of the right pulmonary artery and a 8 × 12 mm Formula™ 535 into the origin of the left pulmonary artery. The stents were inflated simultaneously (WMV 1244 kb)

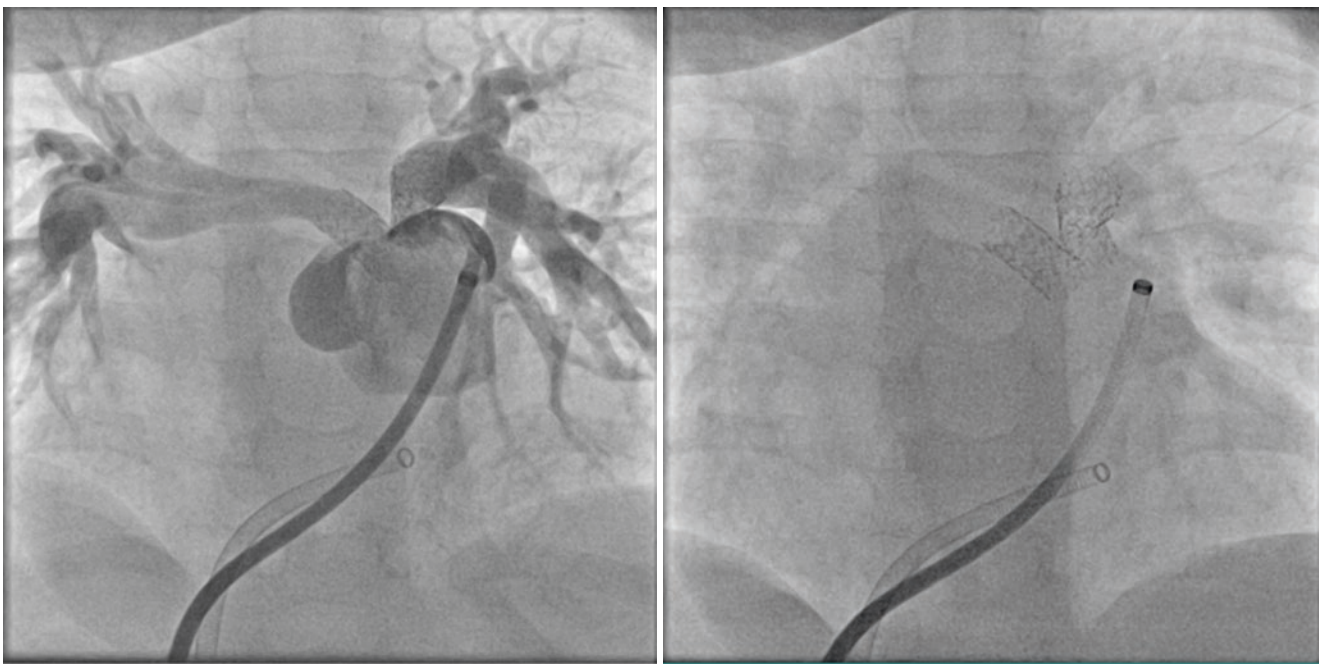


**Fig. 13.13** Result after inflation of both stents. RVP was 58/2/16, PAD 29/14/16, AoP 119/54/67 (RVP:AoP ratio 48%)

**Video 9** Result after inflation of both stents. RVP was 58/2/16, PAD 29/14/16, AoP 119/54/67 (RVP:AoP ratio 48%) (WMV 1356 kb)

Example 3: Creation of a Y-stent in preparation for percutaneous pulmonary valve implantation in a "native" right ventricular outflow tract (TOF patient) with bifurcation stenosis and severe pulmonary regurgitation (Figs. 13.14–13.20).

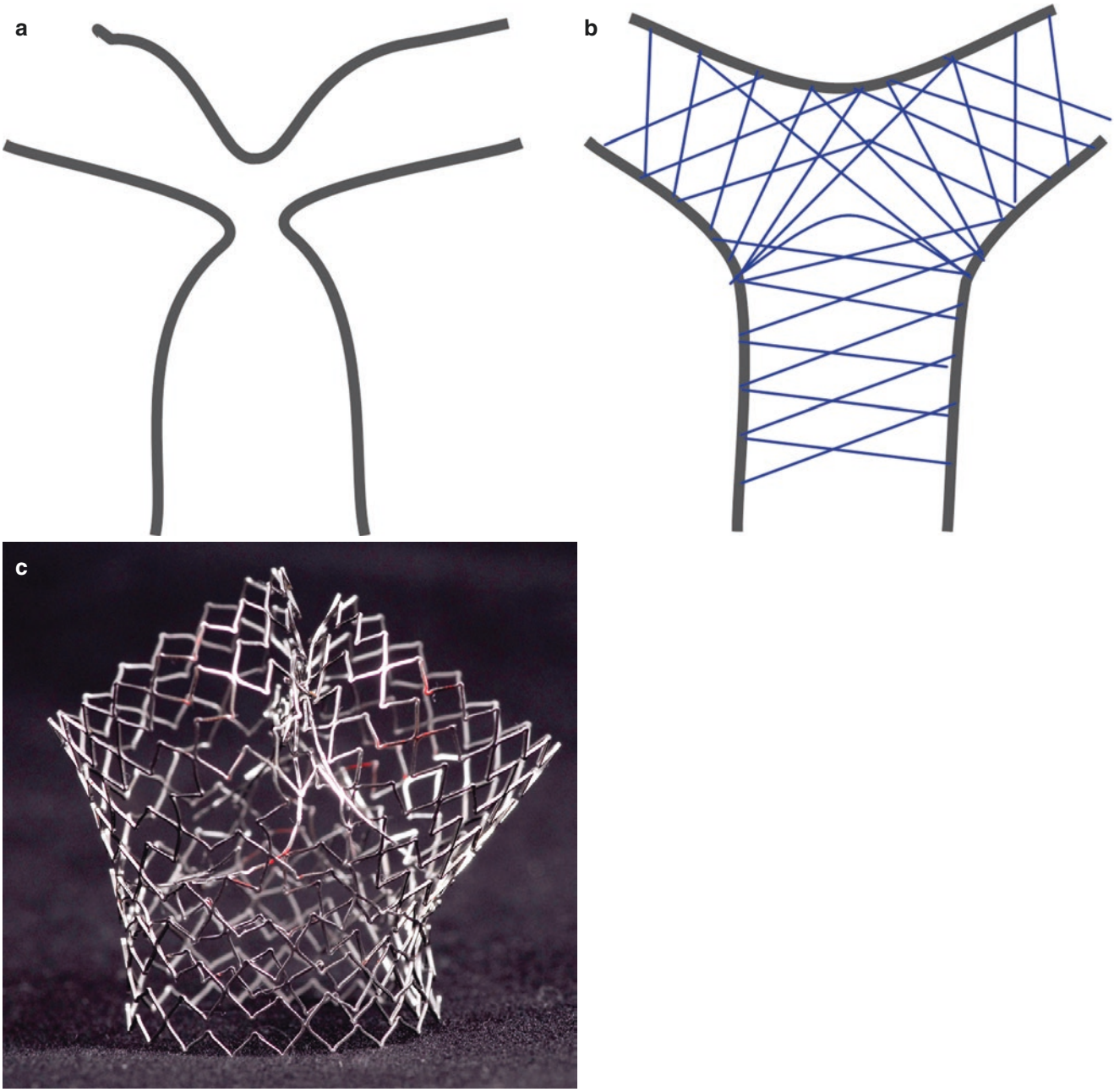



**Fig. 13.14** (**a**–**c**) Schematic drawing of a bifurcation stenosis pre and post stent dilatation (**a**, **b**). Two 26 mm Max LD stents (Medtronic, MN, USA) form the y on the bench

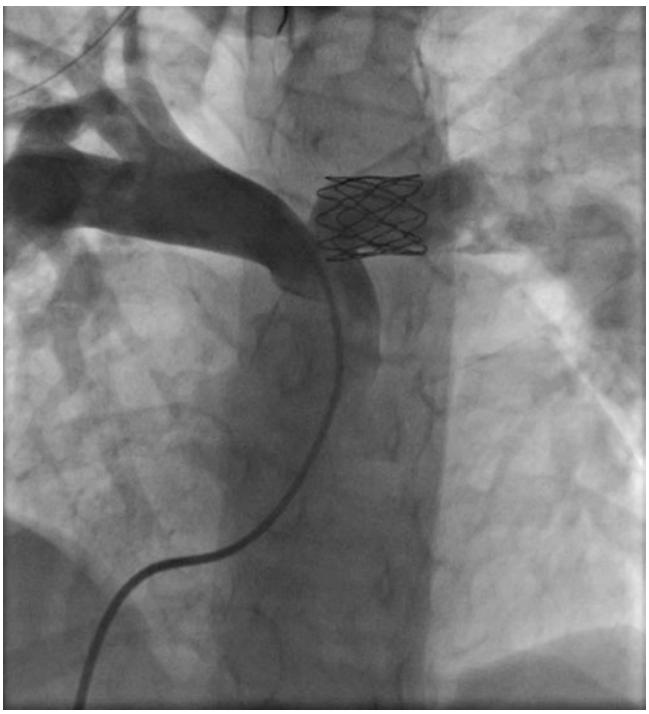


**Fig. 13.15** 15 year old boy (58 kg, 174 cm) after surgical Fallot (TOF) correction with a transannular right ventricular outflow tract (RVOT) patch at 7 months of age and after implantation of a bare metal CP stent (22 mm) into the left pulmonary artery 2 years ago for LPA stenosis. Now mild RVOT stenosis and severe pulmonary regurgitation is present (RPV 47/8/12, PAD 26/10/15, PAS 25/10/15, AoP 85/48/60; RVP:AoP ratio 55%). Although no gradient is measured at the pulmonic bifurcation bilateral stenosis are present

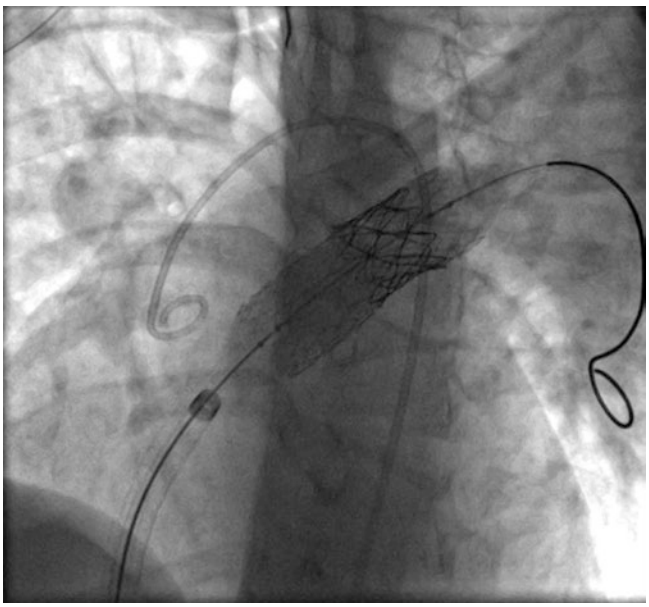


**Fig. 13.16** Through a 12F long sheath (Cook Medical; Bloomington, Indiana, USA) a 57 mm Andra xxl stent (Andramed Reutlingen, D) is deployed on a 22 mm balloon-in-balloon (Numed, Hopkinton, NY, USA) over a superstiff guidewire 0.035′ (Meyer Wire Boston Scientific, Marlborough, MA, USA)

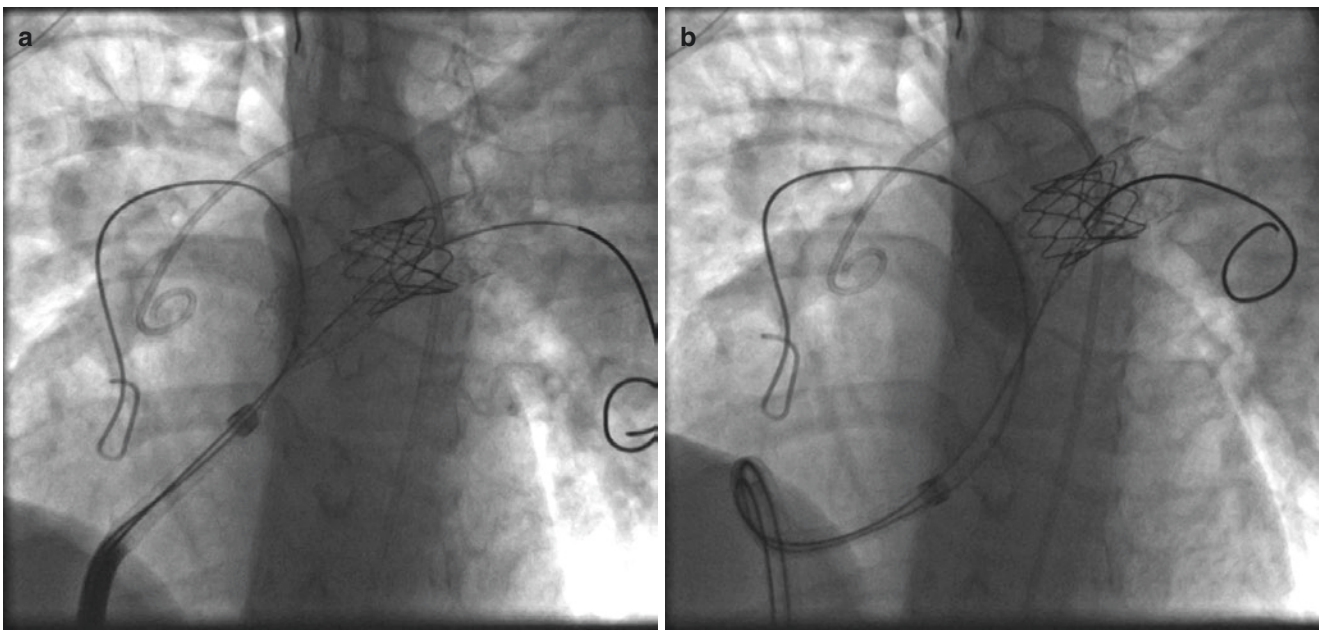



**Fig. 13.17** (**a**, **b**) A second 0.035 guidewire was positioned across the Andra stent into the right pulmonary artery and the stent was dilated with 8 mm powerflex balloon (Cordis Europe), then with an 18 mm Atlas balloon (Bard Medical, Murray Hill, New Jersey, USA)

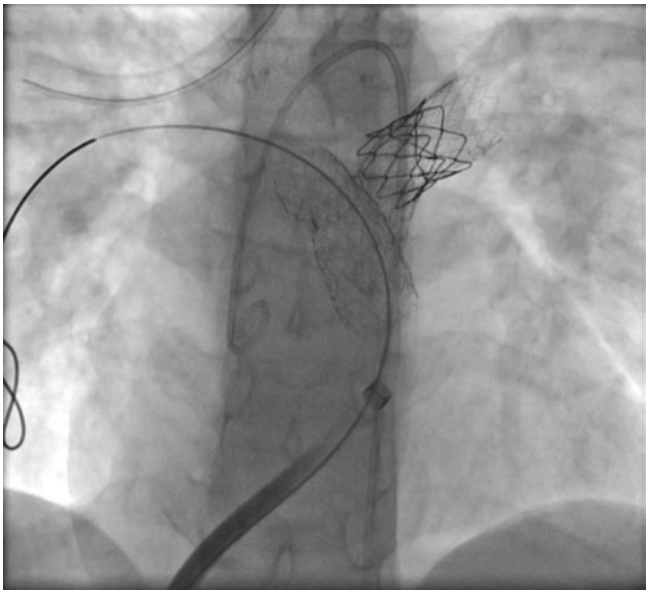


**Fig. 13.18** A 47 mm Andra xxl stent is implanted into the RPA on a 22 mm BiB

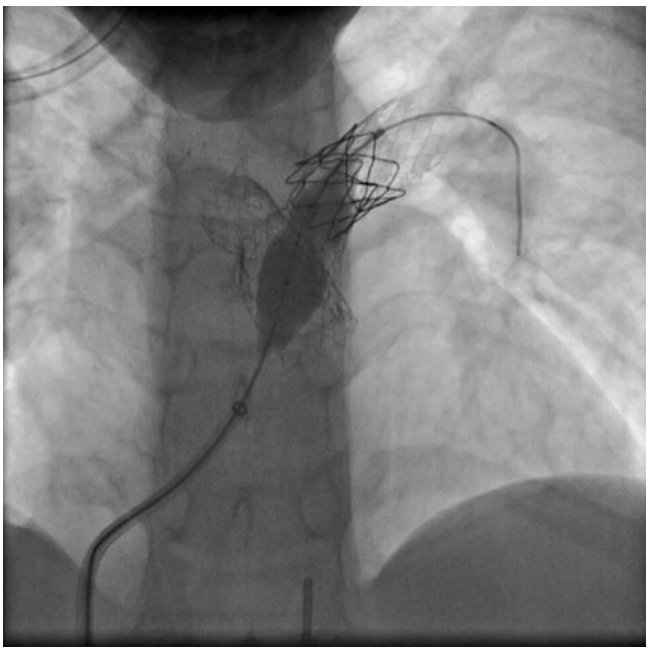


**Fig. 13.19** Then the entrance to the LPA is opened with an 18 mm Atlas balloon

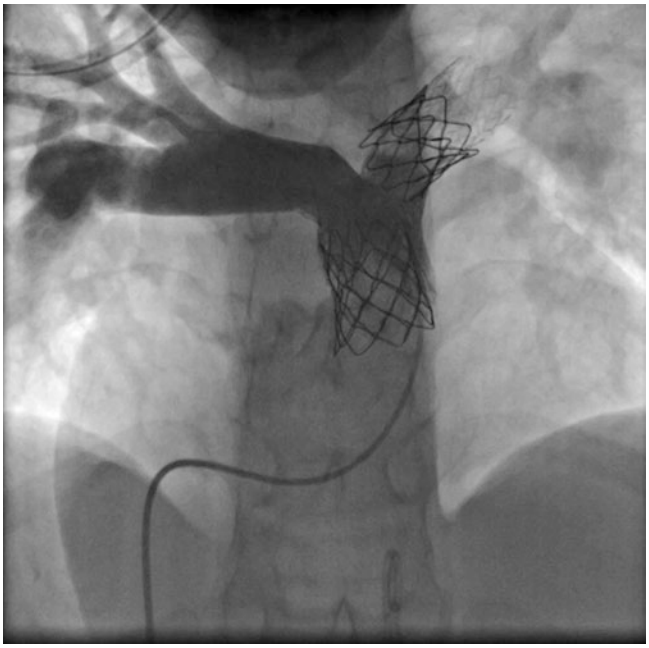


**Fig. 13.20** Finally, a Melody valve (Medtronic, Minneapolis, MN, USA) was implanted with a 22 mm delivery balloon. Final hemodynamics: RVP 45/3/10, PAD 30/17/21, AoP 128/72/91 and no residual pulmonary regurgitation in the MRI control study

# Aortic Coarctation

# 14

Raul Ivo Rossi Filho and João Luiz Langer Manica

## 14.1 Introduction

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 arteriosus. It may be associated with diffuse hypoplasia of the aortic arch and isthmus. Isolated aortic coarctation may occur in 82% of cases and is the commonest detected in adults.

Aortic coarctation is not unusually diagnosed in asymptomatic adolescents or adults in the context of investigating systemic arterial hypertension. First reported in 1991, stent implantation has been considered the gold standard treatment in patients with suitable anatomy and weighing more than 20 kg. Stents can be used both in residual and native lesions and are able to provide beneficial effects at medium-term follow-up (Fig. 14.1a, b).

Patients with less than 20 kg can be considered for stent implantation after careful evaluation of the size of the femoral arteries that should be capable to accommodate the large sheath. In smaller patients, alternative approaches are through the right common carotid or axillary arteries by surgical cutdown (Video 1a, 1b).

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).

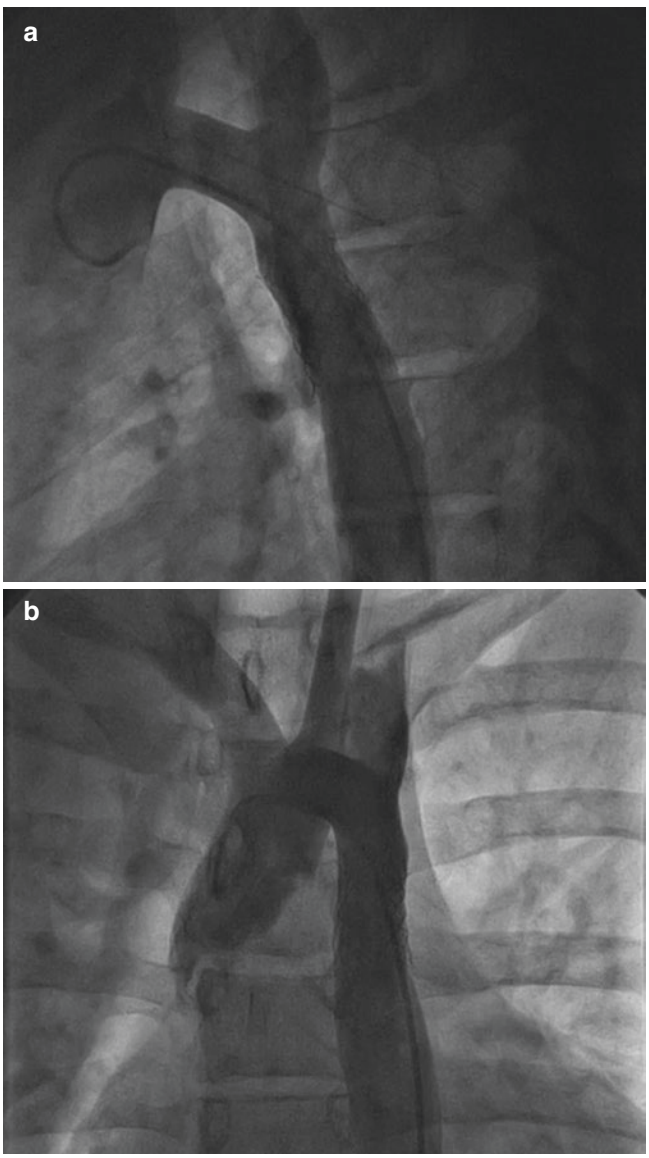



**Fig. 14.1** Stent implantation with ideal result. (**a**) Angiography depicts a bare stent implantation which is properly apposed to the aortic wall. (**b**) Posteroanterior view depicts absence of aortic wall damage

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_14) contains supplementary material, which is available to authorized users.

R. I. R. Filho (✉) · J. L. L. Manica
Instituto de Cardiologia do Rio Grande do Sul,
Porto Alegre, Brazil
e-mail: raul.rossi@cardiologia.org.br


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_14

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.

Currently, its use is also accepted in extremely severe aortic coarctation, in association with patent ductus arteriosus, previously implanted conduits, patients with inflammatory disease and long-segment stenosis, advanced age, aortic wall disease (Marfan and Turner syndromes), acute aortic rupture after primary bare stenting as a bailout situation, associated dilation of the ascending aorta, patients with an irregular aortic wall, and those previously treated with the use of surgical patches (Fig. 14.2a–e). Larger sheets are needed.

**Video 1a** Bailout stent implantation for critical restenosis in a neonate after surgical correction of interrupted aortic arch. Hand injection through the sheath demonstrates the degree of obstruction and permits the necessary measurements for stent implantation. Note the severe low output caused by the obstruction (MOV 1063 kb)

**Video 1b** Bailout stent implantation for critical restenosis in a neonate after surgical correction of interrupted aortic arch. Final aortogram depicting good stent placement and complete relief of the obstruction (MOV 904 kb)

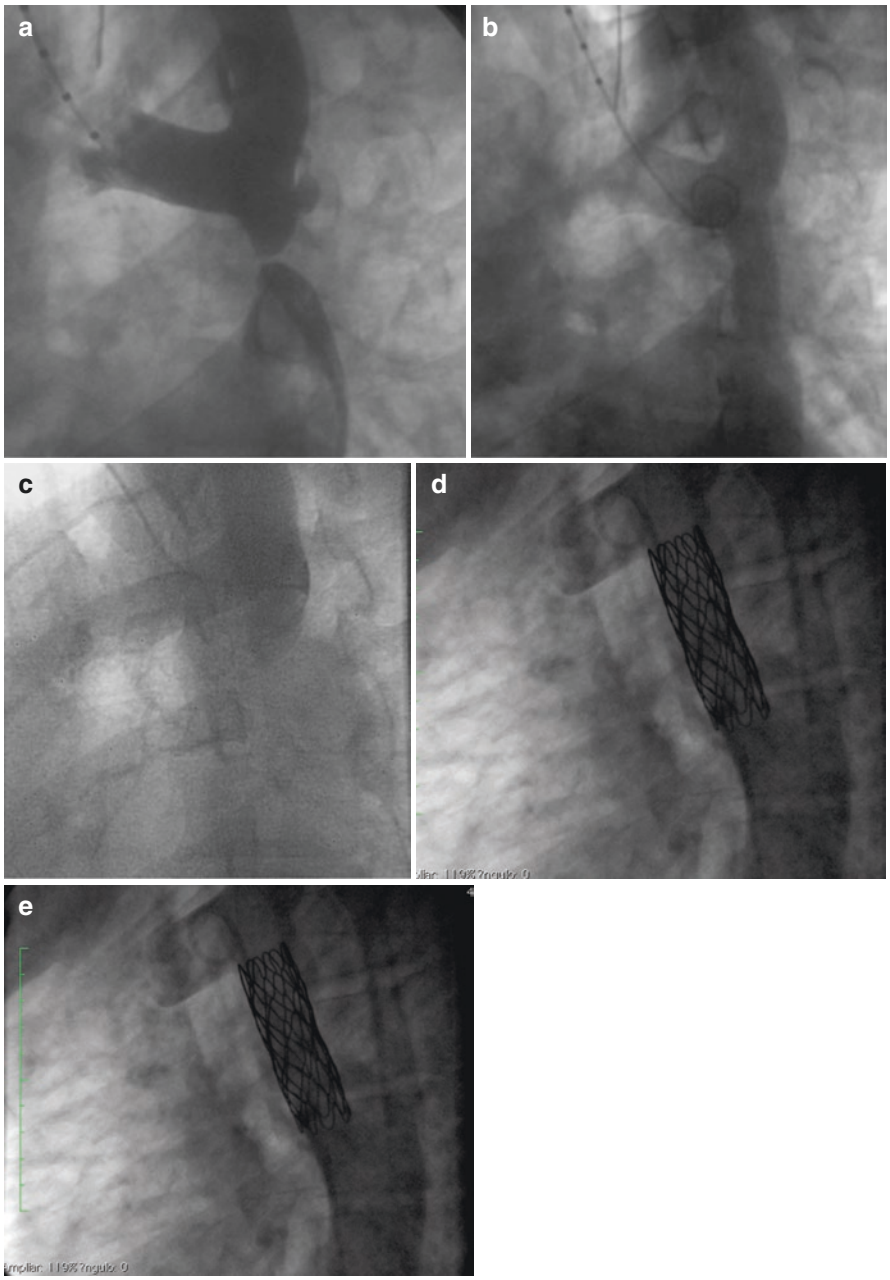



**Fig. 14.2** Examples of uses of covered stents. (**a**) Atretic coarctation with aortic wall aneurysm. (**b**) LAO aortogram showing the final result of a covered stent placement which allowed adequate relief of the obstruction and also treated the aortic wall injury. (**c**) "Acquired" aortic atresia best imaged through a catheter placed via the right radial artery. (**d**) Immediately after a covered stent placement which was (**e**) redilated at a later time with a larger balloon to best fit the aorta

Balloon angioplasty can be an option for native discrete coarctation of the aorta without associated hypoplasia of the transverse arch and/or the isthmus and recurrent coarctation of the aorta following previous surgery or intervention. It can also be used in sick neonates or infants with less than 3 months of age as palliation in the presence of severe left ventricular dysfunction or comorbidities that increase the risk of surgery.

The classic indication for treatment of aortic coarctation is systemic hypertension with resting pressure gradient between upper and lower limbs greater than 20 mmHg. Gradient lower than 20 mmHg may need intervention if associated with left ventricular systolic or diastolic dysfunction, abnormal blood pressure or gradient response to exercise, symptoms of exercise intolerance, or associated lesions such as coronary artery disease and aortic insufficiency.

## 14.2 Technique

### 14.2.1 General Anesthesia

1. Access: femoral artery via percutaneous approach. Rarely, femoral venous or carotid or brachial or axillary arterial approach may be needed.
2. In our institution, carotid, brachial, or axillary approaches are preferentially performed at the hybrid suit by surgical cutdown (Fig. 14.3).
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) (Fig. 14.4a–c).
6. Hemodynamic measurements are performed—aortic pressures: ascending and descending. The advantage of using the multitrack catheter is that the guidewire position can be maintained while repeated pullback measurements are made.
7. Aortography in left anterior oblique, right anterior oblique, and lateral projections is performed using any of the previously mentioned angiographic catheters. The projections have to image properly the structures that should be measured and depend on the anatomy of each patient. Recently, three-dimensional (3D) rotational angiography has been successfully employed in aortic

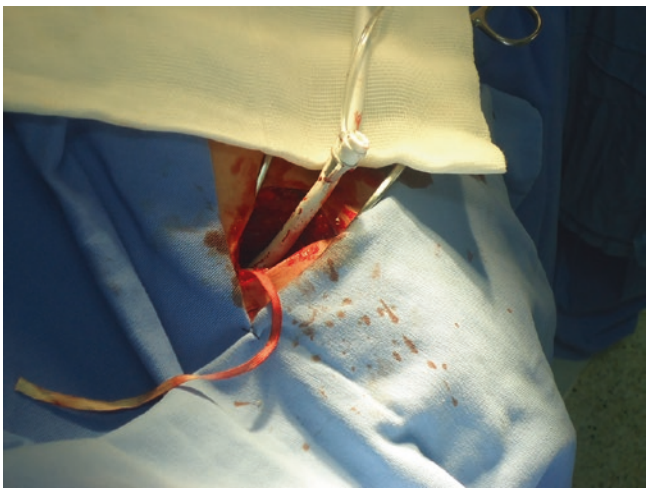


**Fig. 14.3** Axillary artery dissection in an adult with bilateral femoral artery occlusion who needed a covered stent implantation for post-dilatation aneurysm. The sheath size is 14F

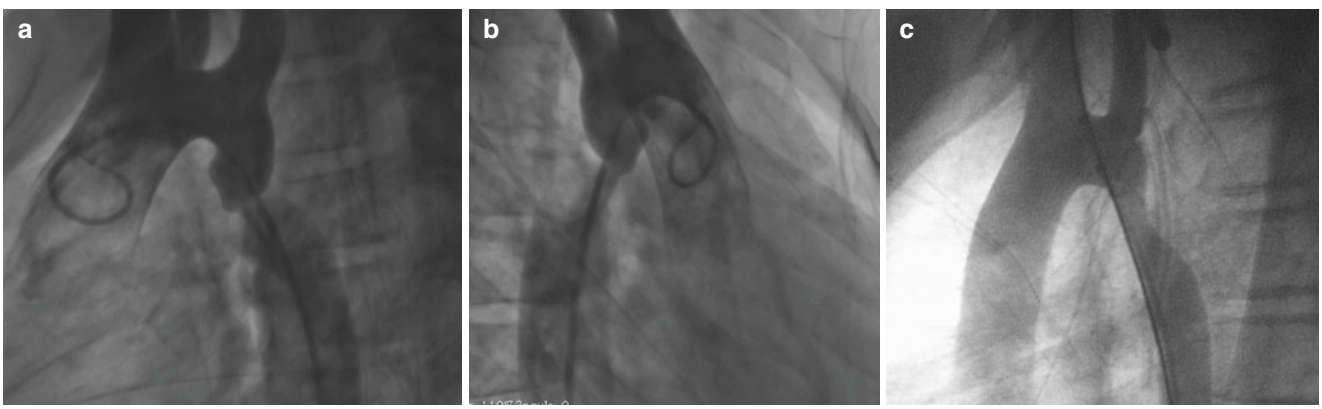



**Fig. 14.4** Techniques most frequently used: (**a**) left aortogram with a pigtail catheter allows for adequate visualization of the aortic anatomy and also measurements without the distortions caused by the stiff wire. (**b**) The same technique used to image the lesion with a 50° RAO view. (**c**) Aortography with a multitrack catheter with the wire in place (right brachial artery) that wallows fast positioning of the large sheath. Previous knowledge of the anatomy is mandatory

coarctation in order to avoid multiple injections, thus reducing radiation exposure (Fig. 14.5, Video 2).

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 the diaphragm (Fig. 14.6).

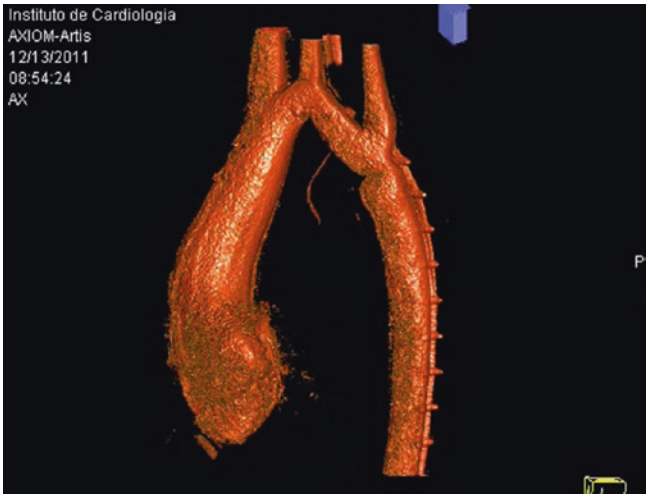



**Fig. 14.5** Rotational angiography with 3D reconstruction. The technique has gained wide acceptance in the last years and can provide additional information before percutaneous treatment. (**a**) This is an example of a gothic arch with isthmus hypoplasia that may change the technical approach to the recoarctation

**Video 2** Double aortic arch with complex coarctation that has as an additional problem which is the left vertebral artery taking off from the obstruction site (MOV 2683 kb)

9. 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.
10. 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.
11. 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 (Fig. 14.7). 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 (Fig. 14.8a, b, Video 3).
12. The same approach can be used in atretic coarctations in which a radio-frequency perforation is performed to make way for covered stent implantation (Fig. 14.9a, b).
13. 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. The sidearms of the long sheath are used to inject contrast to adequately position the stent (Fig. 14.10a, b).
14. The balloon catheter is chosen and should be longer than the stent length. 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

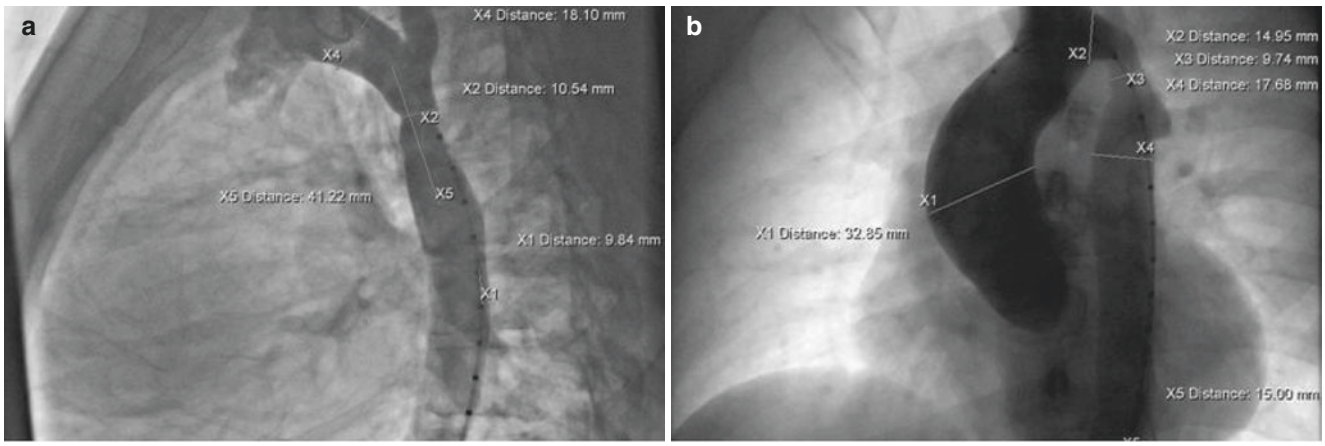



**Fig. 14.6** Examples of the measurements used for choosing balloon and stent sizes. Balloon diameter can be chosen using the diameters of the transverse arch, isthmus, or the descending aorta at the level of the diaphragm. Stent length is usually derived from a measurement that starts right below the origin of the left subclavian artery and goes down well after the obstruction site. One must make allowances for the shortening of the stent that may be as significant as 30% of the devices' length

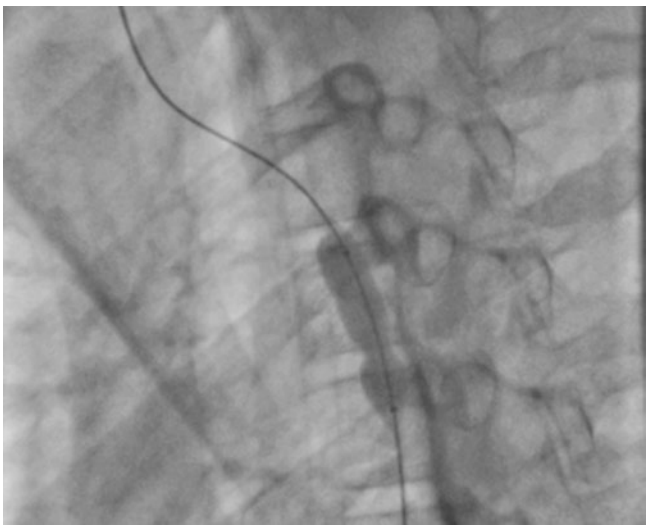


**Fig. 14.7** On occasion predilatation with a low-pressure balloon can be used to open the way for the larger sheath

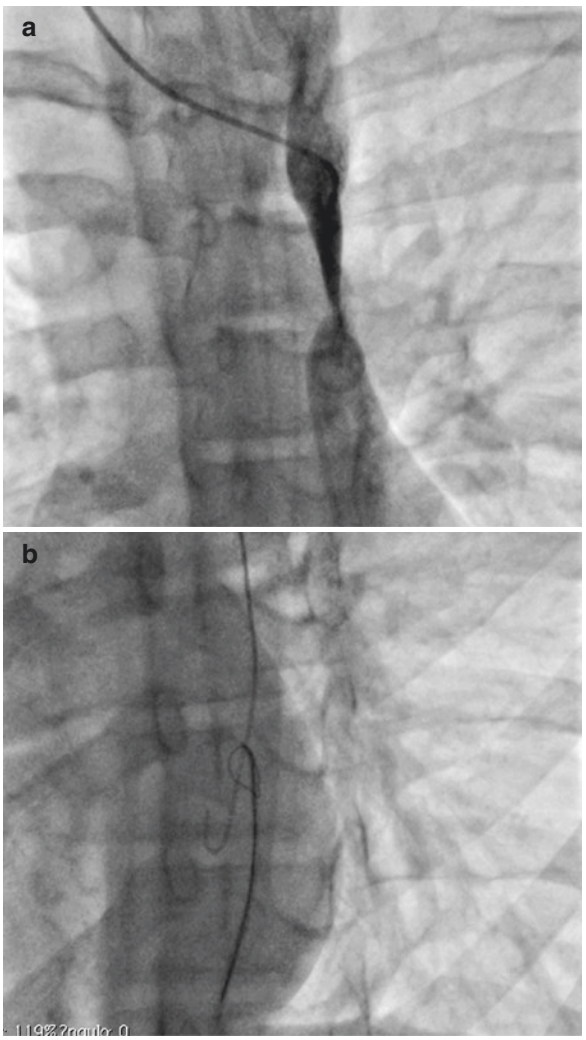



**Fig. 14.8** (**a**, **b**) Technique for subatretic aortic obstructions: this kind of lesion is best approached from above and crossed with a guidewire which must be snared at the descending aorta

**Video 3** Technique for subatretic aortic obstructions: this kind of lesion is best approached from above and crossed with a guidewire which must be snared at the descending aorta (MOV 2269 kb)

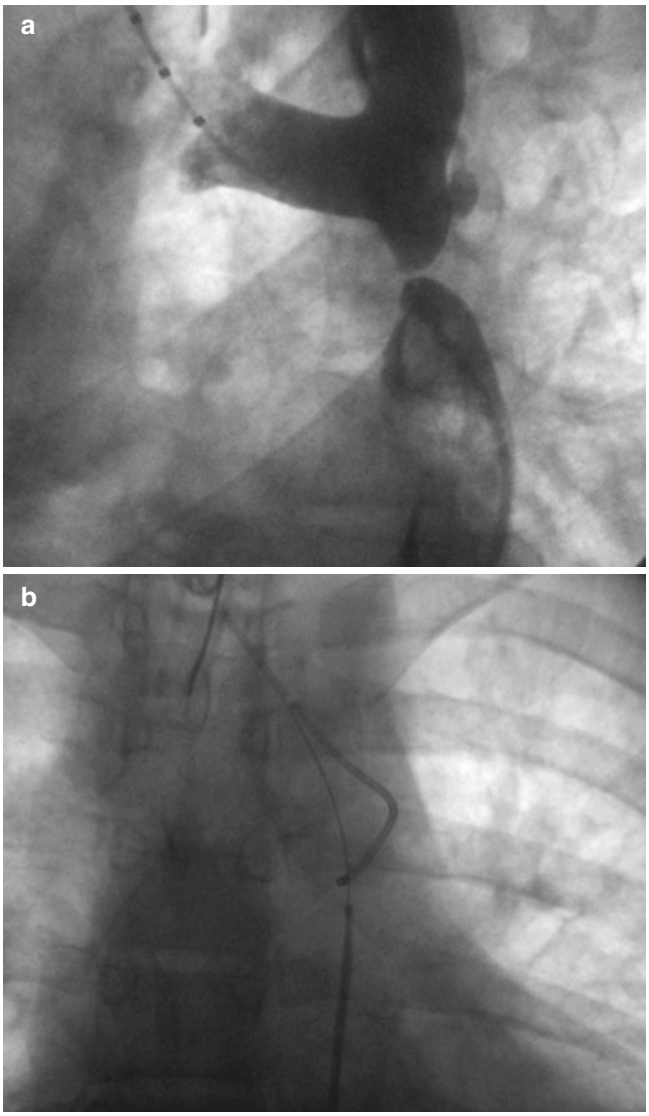



**Fig. 14.9** (**a**, **b**) The same technique used after radio-frequency perforation of an atretic segment of the aorta is needed

paramount if one wants to avoid jailing the brachiocephalic arteries, especially when using covered stents.

15. The balloon is then manually inflated up to the pressure recommended by the manufacturer, which is usually up to 4–6 atmospheres.
16. Angiography is performed during and after stent placement through the sidearm of the sheath or by using a pigtail to assess the result and rule out aortic dissection or rupture (Fig. 14.11a, b). Rotational angiography is another alternative to evaluate final result (Fig. 14.12, Video 4).

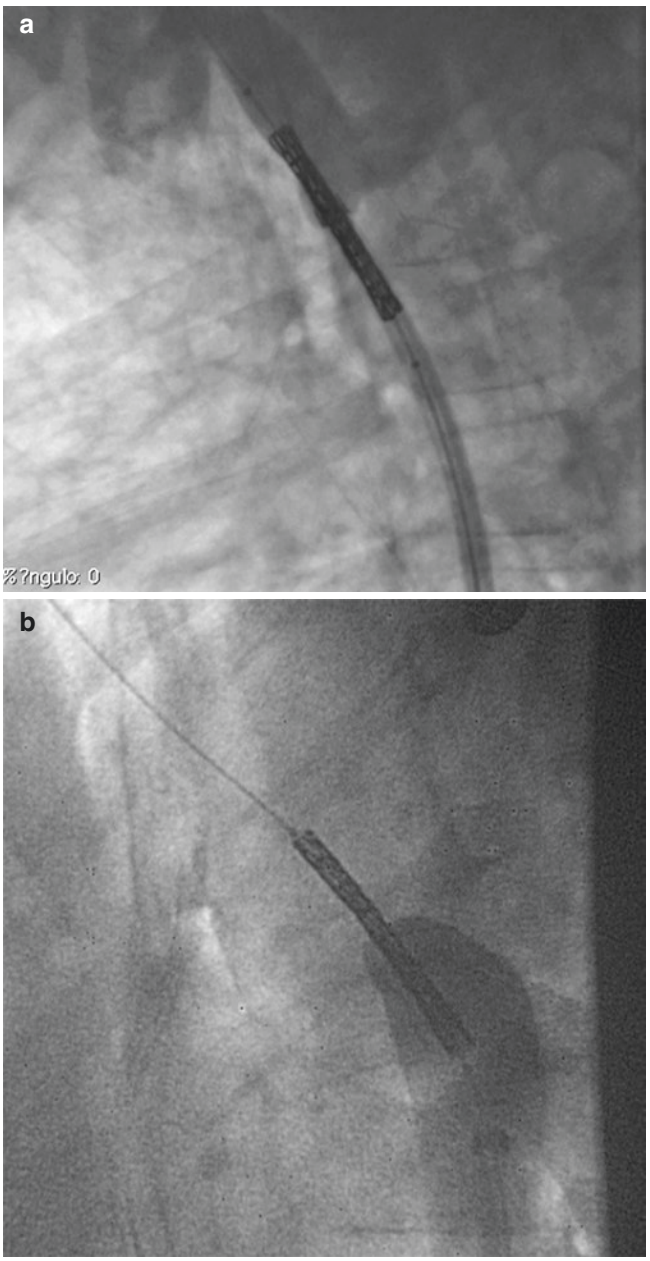



**Fig. 14.10** (**a**, **b**) Usual technique: One doesn't need a catheter positioned in the ascending aorta for stent placement. Hand angiograms through the sidearm of the sheath are more than enough for a proper stent positioning. If at all possible, one could upsize the sheath to obtain better imaging

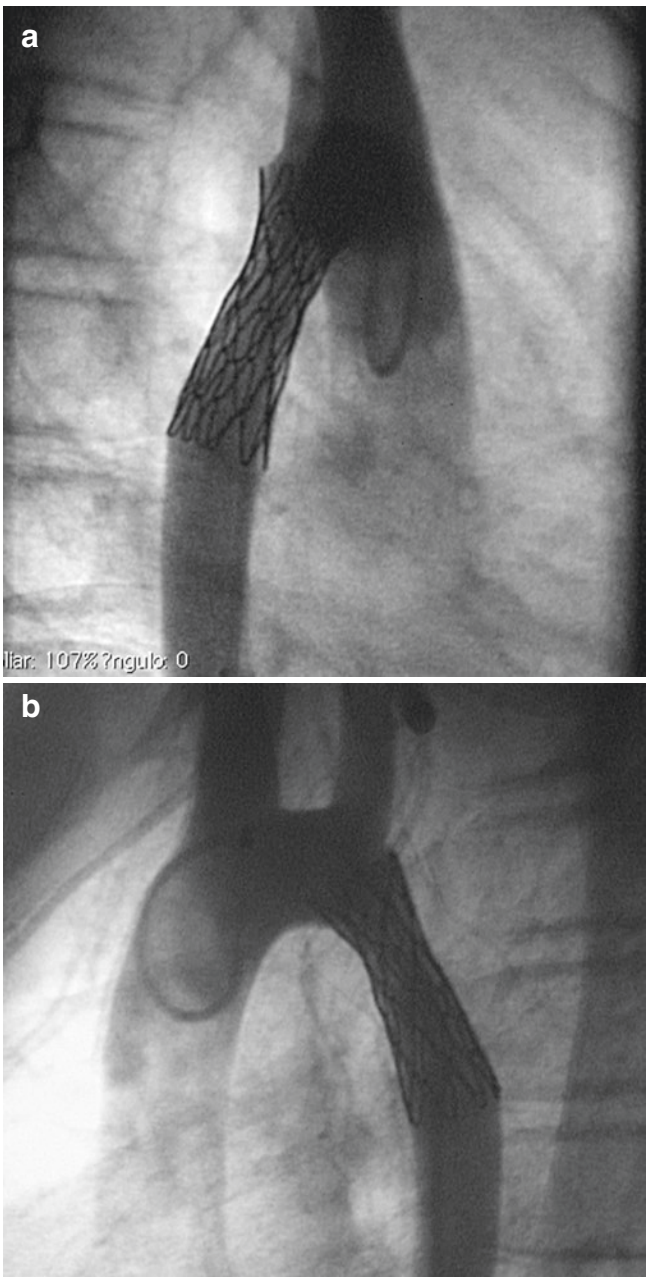



**Fig. 14.11** After stent placement, proper pump-assisted angiographies must be performed in orthogonal planes in order to make sure that the stent is properly set to the wall and also to exclude aortic wall damage. (**a**) Control angiography in lateral view and (**b**), at 50° right oblique projection

17. 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.
18. Additional dilations with large balloons can be performed in order to properly appose the stent struts to the wall or when a smaller balloon was initially used to reduce sheath size (tight lesions in small kids).
19. Hemostasis is achieved by manual compression, vascular closure devices, or even by surgical repair.
20. All patients should be on antibiotic prophylaxis (cefazolin for 24 h).
21. Redilation must be necessary to accommodate the stent in the aortic wall after somatic growth (Fig. 14.13a, b).

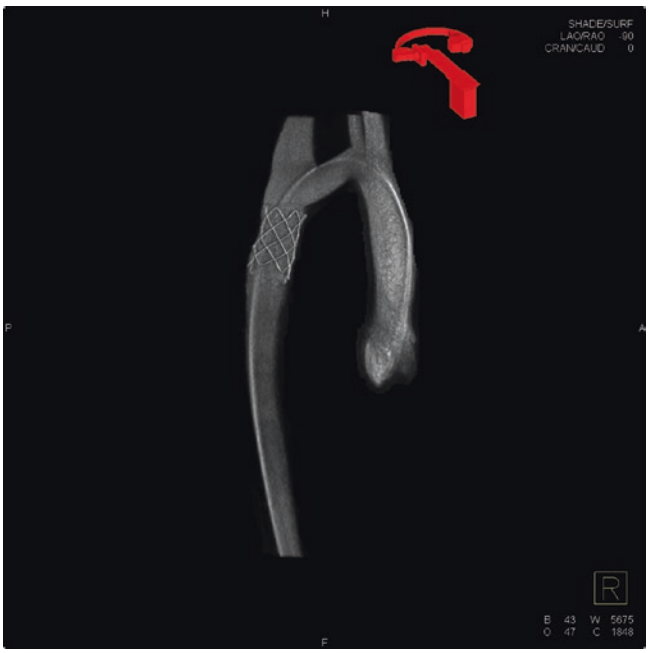



**Fig. 14.12** Or else, the same information can be obtained with just on injection using 3D reconstruction. Adequate result after stent placement

**Video 4** The same image was enlarged to access stent integrity (MOV 6629 kb)

## 14.3 Complications

Complication rate can be around 14% in the literature. Fortunately, life-threatening complications are very rare. Arterial wall damage can occur causing aortic dissection, pseudoaneurysm formation, or even overt aortic rupture (Fig. 14.14a–d). Stent migration, balloon rupture with inadequate stent expansion, the possibility of side branch occlusion during covered stent implantation, and stent fracture are other complications that can seldom occur (Fig. 14.15a, b, Video 5a, 5b, 5c, 5d and 5e). Acute arterial occlusion is a major concern when using large sheaths in patients in the first year of life. Bleeding, local hematoma, and arteriovenous fistula due to the use of large sheaths comprised other access-related complications.

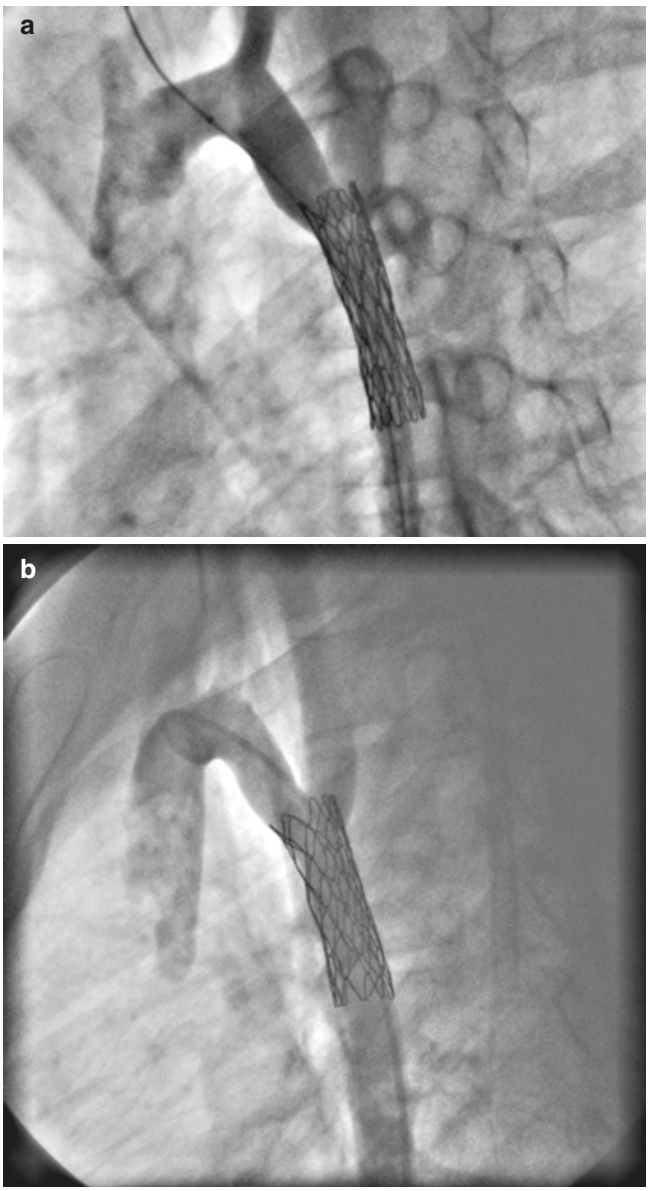



**Fig. 14.13** Sequential dilatation can be used to accommodate somatic growth of the children or as in this case when, in the absence of a covered stent, a bare stent was used in a patient with a severe long-segment obstruction which started at the origin of the left subclavian artery. (**a**) The proximal part of the stent is just above the origin of the LSA and (**b**) late redilation due to stent mismatch to the somatic growth of the patient

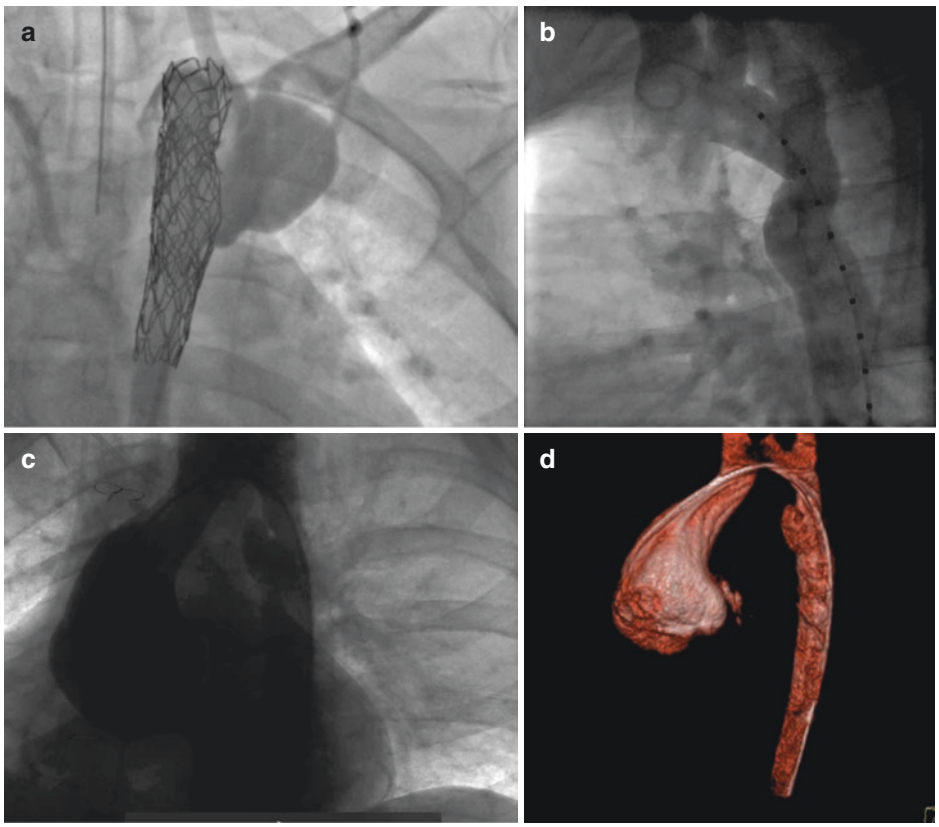



**Fig. 14.14** Examples of complications of stent implantation. (**a**) This is a very complex case of double aortic arch which demanded the implantation of multiple stents. In the last session, there was acute pseudoaneurysm formation that needed surgical resolution. (**b**) Usual plane angiograms failed to detect the aneurysm, and (**c**) rotational angiography with (**d**) 3D reconstruction allowed proper visualization of the damaged aorta. Covered stent implantation solved the problem

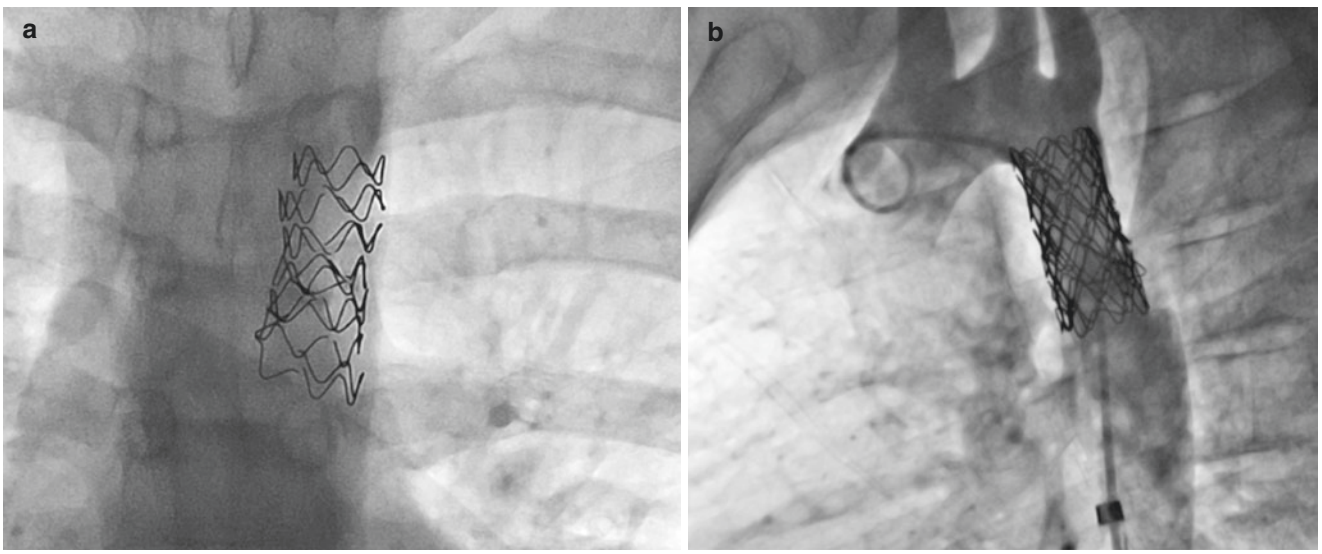



**Fig. 14.15** (**a**, **b**) Stent fractures are uncommon, and the treatment of choice is covered stent implantation as shown in these images

**Video 5a** This picture depicts a severe and eccentric native coarctation in a 22-year-old woman (MOV 3231 kb)

**Video 5b** This is the immediate result of a bare stent placement (MOV 2683 kb)

**Video 5c** Aortogram obtained 1 week after a severe car crash from which she barely survived. Notice that the stent has a grade III fracture but no aortic damage or dissection (MOV 3290 kb)

**Video 5d** This is a covered stent positioned through the fractured one (MOV 3406 kb)

**Video 5e** Final angiography depicting that the covered stent realigned the two segments of the fractured stent and maintained good aortic patency (MOV 2797 kb)

# Reopening of Peripheral and Central Arteries and Veins

# 15

Henri Justino and Athar M. Qureshi

In children and young adults, the most important cause of complete occlusion of arteries or veins, both central and peripheral, is iatrogenic catheter-induced thrombosis. In contrast, the large body of literature on treatment of chronic total occlusions in adults is most applicable to arterial atherosclerotic occlusions.

In the setting of acute thrombosis, rapid dissolution or removal of clot is needed. This may be accomplished with catheter-based thrombectomy (e.g., aspiration or rheolytic thrombectomy) and/or thrombolysis (e.g., with systemic or local infusion of tissue plasminogen activator, TPA). When indicated, chronic thrombosis should be treated by catheter means without inappropriate attempt at anticoagulation.

Indication for treatment:

- Clinical symptoms and signs depend on the location of occlusion.
- Indications for intervention on systemic venous occlusion include significant swelling of the affected territory (e.g., superior vena cava syndrome) or severe painful limb swelling (phlegmasia cerulea dolens) which may progress to venous gangrene. Another indication for intervention is when the patient requires reopening of venous channels to permit placement of central lines or to permit other transcatheter interventions. Special circumstances that warrant venous intervention also include chylothorax, which may be caused by superior vena cava or innominate vein stenosis or occlusion.
- Arterial occlusion of a limb results in 5Ps: pain, paresthesias, pallor, poikilothermia (cold), and pulselessness; paralysis (loss of motor function) is a later finding. These constitute indications for urgent intervention to restore arterial perfusion to the limb.
- Renal artery occlusion may manifest with hypertension.
- Pulmonary arterial occlusion may be thrombotic (pulmonary embolism) or acquired after surgical repair or after ductal constriction. Indications for treatment include right ventricular failure or right ventricular hypertension to greater than half-systemic pressure.
- Patients with pulmonary vein occlusion may benefit from recanalization of pulmonary veins, especially in the setting of hemoptysis and/or pulmonary hypertension.

Diagnostic catheterization:

- When feasible, having a catheter proximal and another distal to the occlusion facilitates angiography, documentation of pressure gradient across the occlusion, and intervention.
- Angiography (ideally in a biplane laboratory) is best performed upstream to the occlusion but may also be performed with a catheter located downstream; the goal is to identify a "beak-like" occlusion on one side or the other.
- Digital subtraction angiography may improve visualization, particularly in areas where there is little cardiac or respiratory movement artifact (e.g., limbs).

Catheter intervention:

- Crossing the "beak-like" occlusion is first attempted using soft 0.014", 0.018.", or 0.035" guidewires. Hydrophilic wires will aid in crossing the occlusion but also risk ending up subintimal. If the occlusion is difficult to cross, the tip load of the wire can be increased, or the wire thickness can be decreased to improve likelihood of perforating the occlusion cap.
- If the wire crosses the occlusion and regains the true lumen, a snare should be used to grasp the soft end of the wire and to exteriorize it via a sheath. The wire can then be secured by clamping it to the drapes. We suggest exteriorizing the wire whenever possible as this facilitates the

The original version of this chapter was revised. A correction to this chapter can be found at https://doi.org/10.1007/978-3-319-72443-0_47

H. Justino (✉) · A. M. Qureshi
Department of Pediatrics, Texas Children's Hospital,
Houston, TX, USA
e-mail: hjustino@bcm.edu; axqureshi@texaschildrenshospital.org



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_15

procedure by resulting in secure access for subsequent balloon angioplasty and/or stent placement. However, access from the other side of the lesion is not always possible or practical (e.g., this is generally not possible in the pulmonary circulation).

– If the wire crosses the occlusion but remains subintimal beyond it, angiography with the second catheter beyond the occlusion will confirm the subintimal location. Reentry into the true lumen may then be accomplished using a variety of techniques including a sharply curved catheter (e.g., Judkins right) with a high tip load wire designed for chronic total occlusions (CTO), the stiff end of a 0.014" wire, or radio frequency perforation (e.g., Nykanen wire, Baylis Medical, Montreal, QC, Canada). Once the true lumen is reentered, the wire is exteriorized using a snare, if possible.
– After establishing access through the occlusion, balloon angioplasty is performed, and stents are placed unless contraindicated. Balloon-expandable stents may be used within the chest and abdomen, but self-expanding stents are preferred in vessels prone to flexion and/or compression (e.g., jugular veins, femoral veins, and femoral arteries). Certain vessels are prone to stent fracture because of bony compression, such as the subclavian vein between the first rib and clavicle, in which case surgical repair may be preferable.

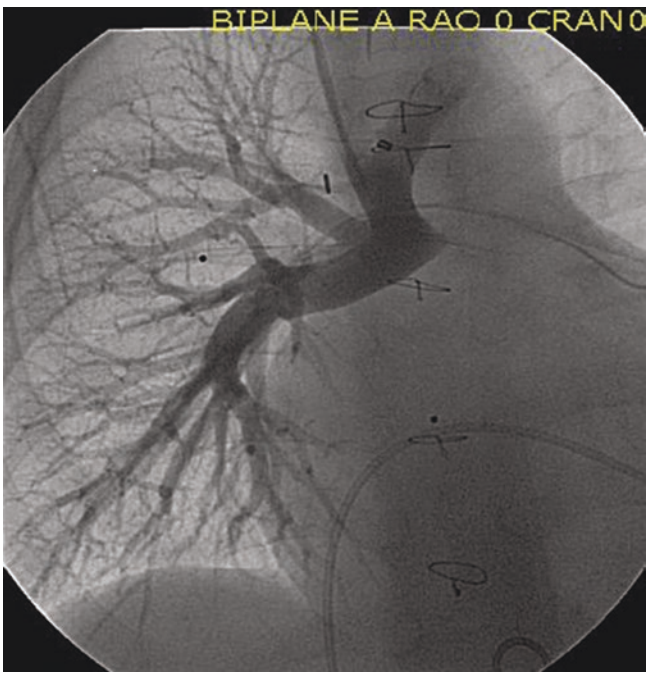



**Fig. 15.1** Patient 1. Left pulmonary artery occlusion. A 2-year-old boy with hypoplastic left heart syndrome had undergone a Norwood operation followed by a right bidirectional cavopulmonary anastomosis. He was transferred to our institution, where flow in the left pulmonary artery could not be seen by echocardiogram. He was taken to the cardiac catheterization laboratory for intervention on the left pulmonary artery. This figure demonstrates a widely patent right cavopulmonary anastomosis with flow into the right pulmonary artery and complete occlusion of the left pulmonary artery. Note that the occlusion has a prominent "beak," a favorable finding for attempting wire recanalization

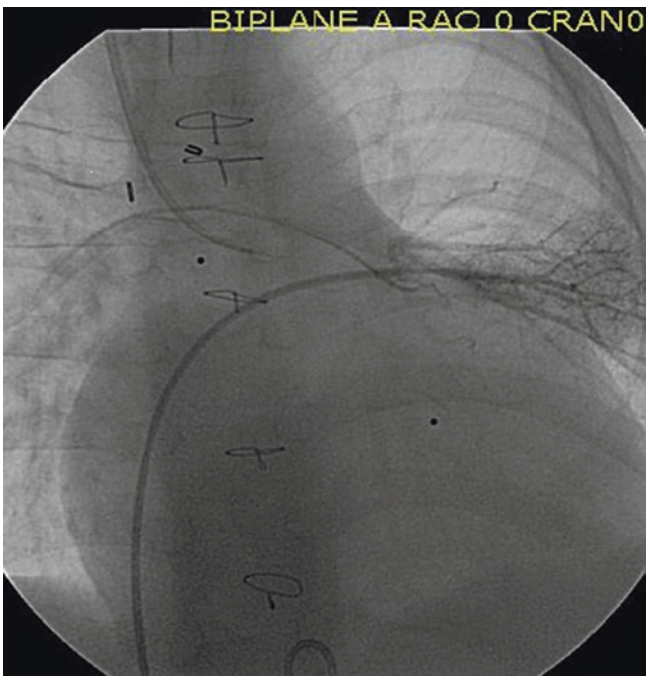



**Fig. 15.2** Patient 1. Left pulmonary artery occlusion. Pulmonary vein wedge angiogram showing contrast filling of the hilar branches of the left pulmonary artery, confirming its distal patency and providing a landmark for subsequent wire advancement across the occluded left pulmonary artery

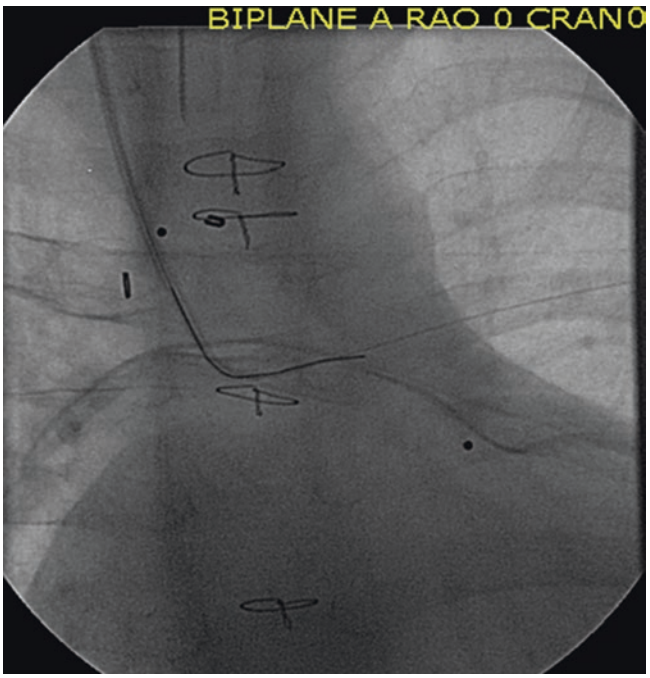



**Fig. 15.3** Patient 1. Left pulmonary artery occlusion. The occlusion has been crossed using a 0.018" straight glide wire (Terumo Corp., Tokyo, Japan) within a 5F JR3 catheter. Because the glide wire is not ideal for supporting the advancement of an angioplasty balloon, a second wire (0.014" Asahi Grand Slam, Abbott, Abbott Park, IL) was advanced alongside the first wire

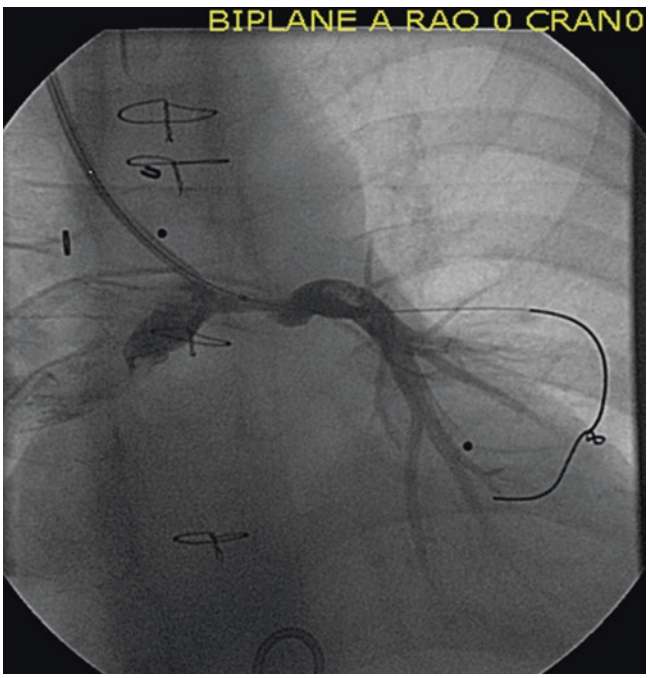



**Fig. 15.4** Patient 1. Left pulmonary artery occlusion. Left pulmonary artery angiogram after angioplasty with a 4 mm balloon, confirming no extravasation of contrast

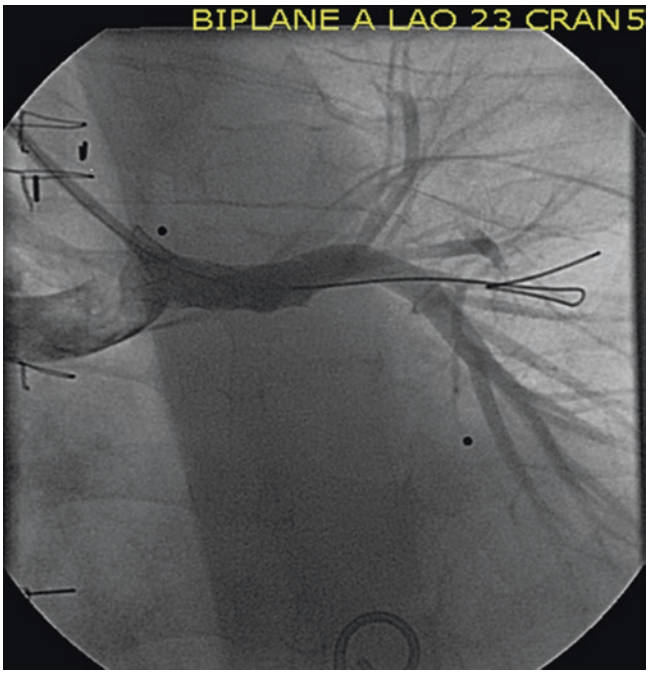



**Fig. 15.5** Patient 1. Left pulmonary artery occlusion. Final angiogram after placement of a Genesis 24 mm long stent premounted on a 6 mm balloon (Cordis, Cardinal Health, Milpitas, CA)

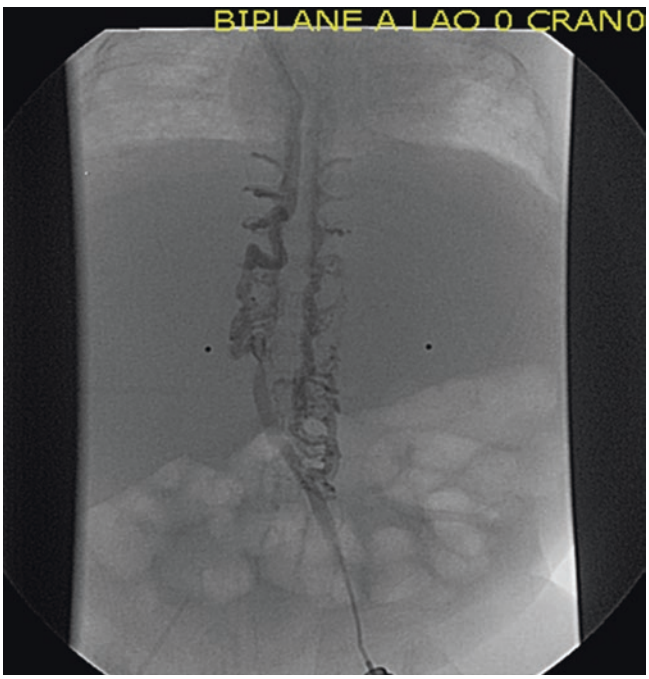



**Fig. 15.6** Patient 2. Chronic inferior vena cava thrombosis. A 5-month-old, former 27-week premature, boy with numerous medical problems including bronchopulmonary dysplasia, previous necrotizing enterocolitis with intestinal perforation requiring bowel resection, and multiple episodes of sepsis, developed complete thrombosis of the inferior vena cava (IVC) due to a central line. The line was removed, and he received several months of anticoagulation with low molecular weight heparin but had persistent IVC occlusion with some leg edema. He was taken to the cardiac catheterization laboratory weighing 4 kg for possible transcatheter recanalization. This figure shows a patent left external and common iliac vein, with complete occlusion of the suprarenal IVC and numerous paravertebral venous collaterals

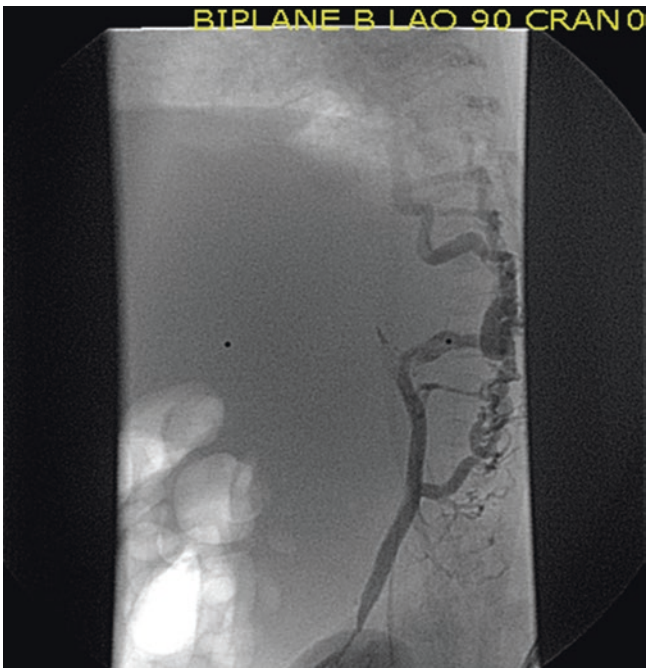



**Fig. 15.7** Patient 2. Chronic inferior vena cava thrombosis. The lateral projection of the same angiogram as Fig. 15.6 shows the presence of a "beak" anterior to the spine at the site of occlusion of the suprarenal IVC

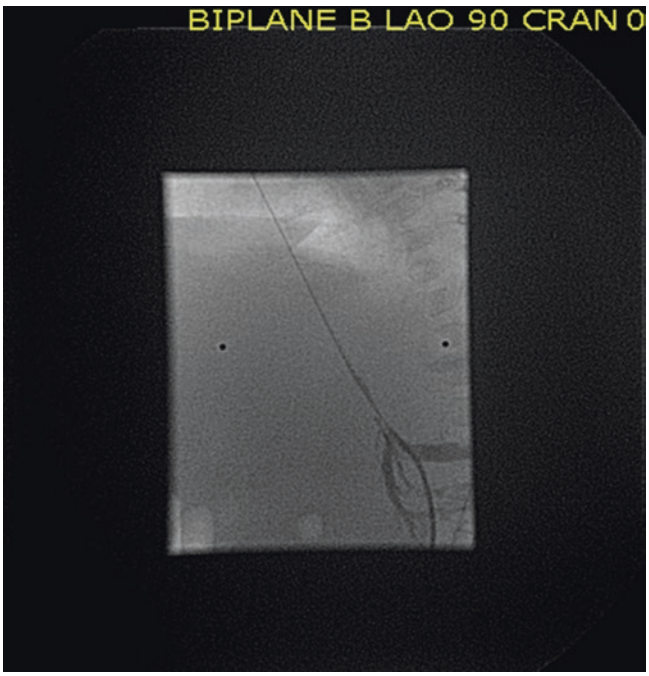



**Fig. 15.9** Patient 2. Chronic inferior vena cava thrombosis. Lateral projection of same angiogram as Fig. 15.8 shows wire position in the expected location of the IVC, anterior to the spine, with its tip in the right atrium

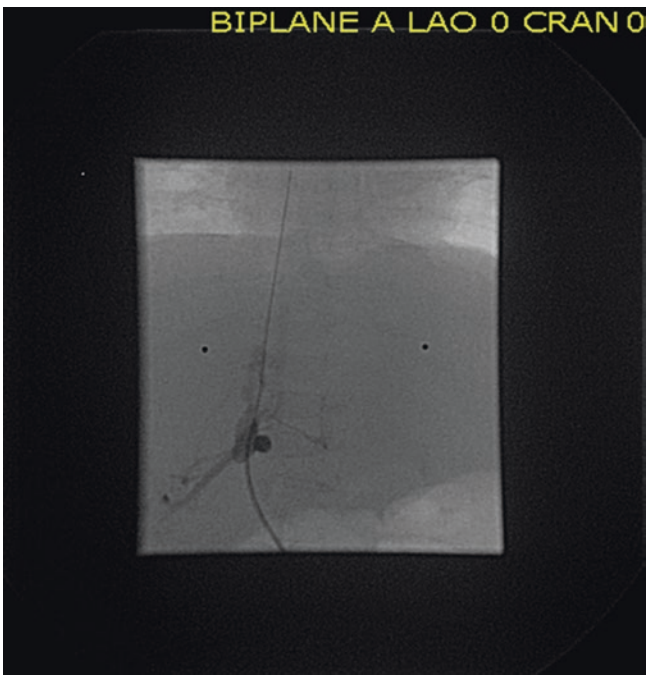



**Fig. 15.8** Patient 2. Chronic inferior vena cava thrombosis. Recanalization of the occluded IVC was performed using the stiff end of a 0.018" glide wire (Terumo) after failed attempts with soft wires, and the wire tip is located in the right atrium

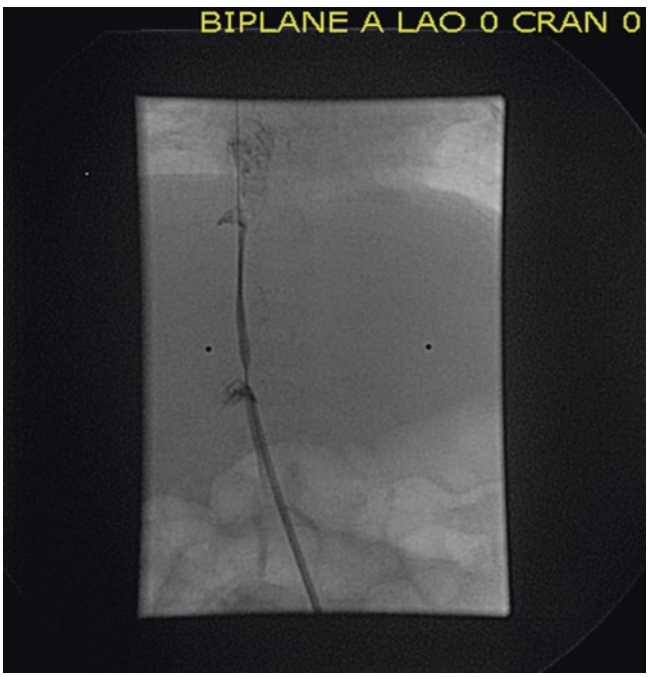



**Fig. 15.10** Patient 2. Chronic inferior vena cava thrombosis. After balloon angioplasty with a 3 mm balloon, the suprarenal and intrahepatic IVC is now visible, with a small amount of contrast refluxing into the right hepatic vein

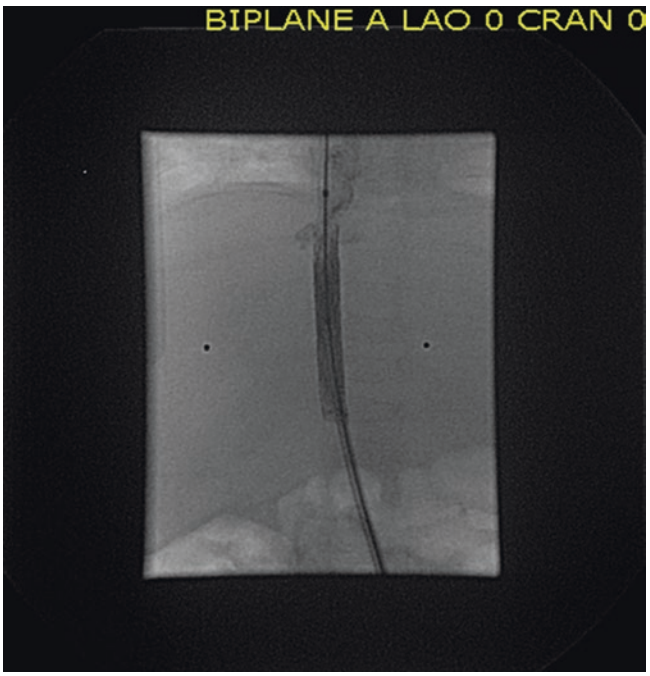



**Fig. 15.11** Patient 2. Chronic inferior vena cava thrombosis. After placement of two Genesis XD stents hand-mounted onto a 6 mm balloon, delivered via a 7F sheath, the suprarenal and intrahepatic IVC is widely patent. Note that a variety of premounted 6 mm stents are deliverable through smaller sheaths, but those stents have far lesser redilation potential (in the range of 10–12 mm) and would one day result in fixed IVC stenosis; in contrast, non-premounted Genesis XD stents can be post-dilated to 18 mm

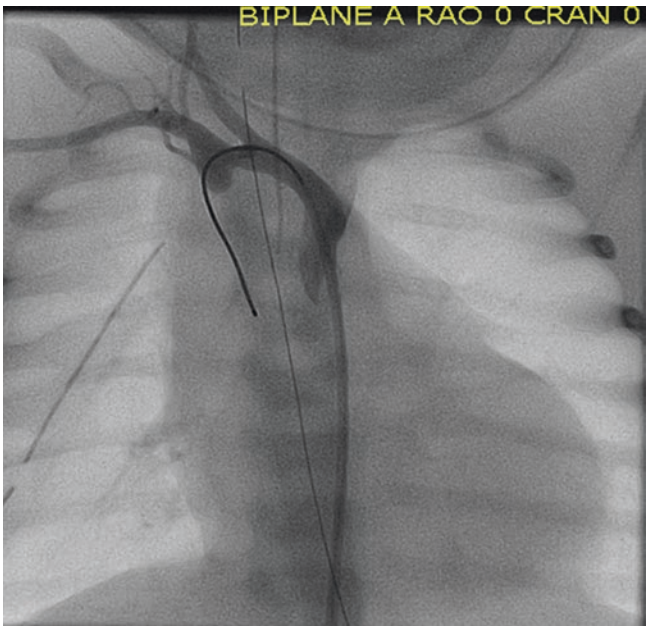



**Fig. 15.13** Patient 3. Acute modified Blalock-Taussig shunt (mBTS) occlusion. Using a 4 Fr JR 2 catheter (Cordis, Miami Lakes, Florida, USA) from a femoral arterial access route, the soft end of a 0.018" Flex T wire was used to cross the occluded mBTS, given the acute nature of the occlusion

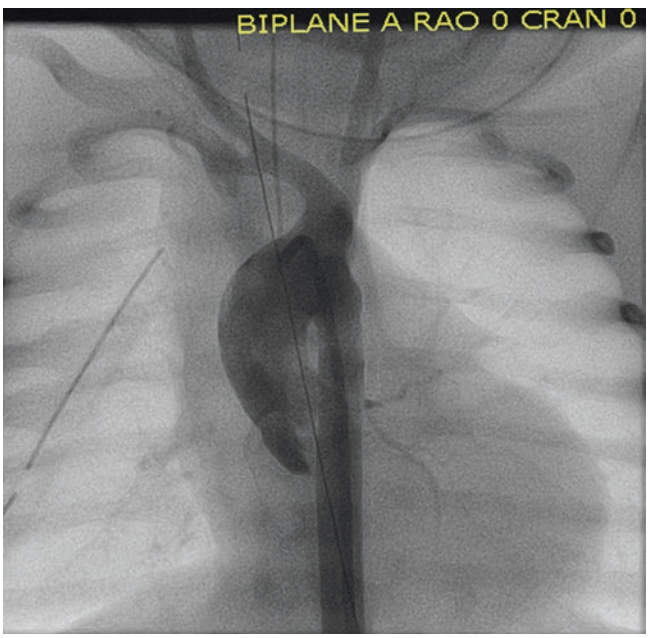



**Fig. 15.12** Patient 3. Acute modified Blalock-Taussig shunt (mBTS) occlusion. A 2-week-old 2.9 kg baby girl with hypoplastic right heart structures underwent placement of a 3.5 mm right mBTS from the right subclavian artery to the right pulmonary artery. Soon after arrival in the intensive care unit, she was noted to be desaturated with no audible shunt murmur, and no mBTS flow seen by echocardiogram. She was taken urgently to the cardiac catheterization laboratory for mBTS recanalization. The occluded mBTS is seen with an occluded beak seen from the right subclavian artery

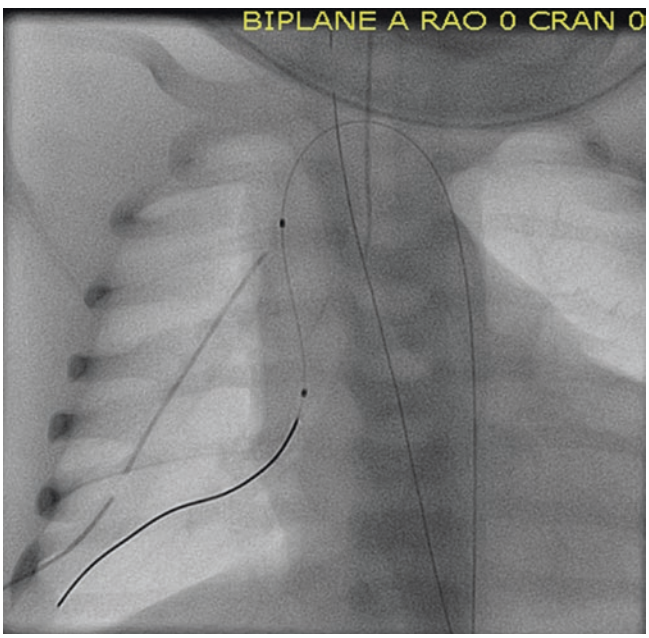



**Fig. 15.14** Patient 3. Acute modified Blalock Taussig shunt (mBTS) occlusion. The wire was then exchanged for a 0.014" Grand Slam wire which was placed in the distal right pulmonary artery, facilitating balloon dilation of the mBTS throughout its length

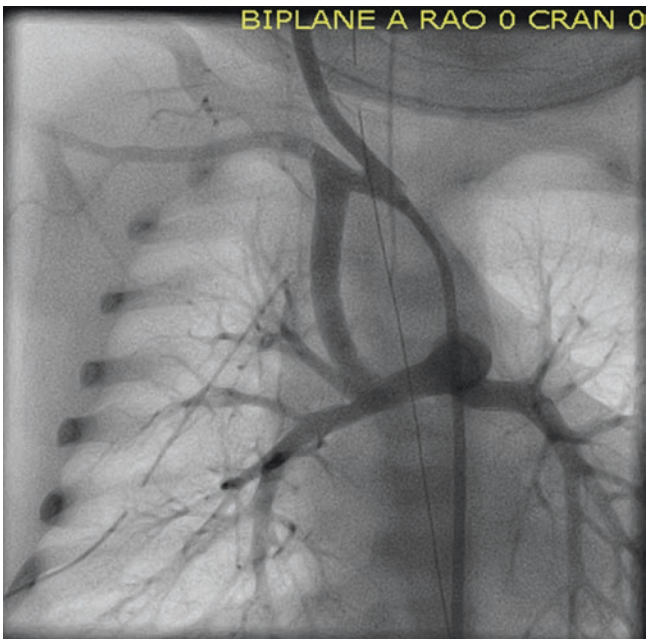



**Fig. 15.15** Patient 3. Acute modified Blalock-Taussig shunt (mBTS) occlusion. Patency of the mBTS is seen after balloon angioplasty was performed multiple times throughout the length of the mBTS with 3.5 mm balloons. The patient continued anticoagulation therapy immediately after the catheterization and was discharged home on aspirin

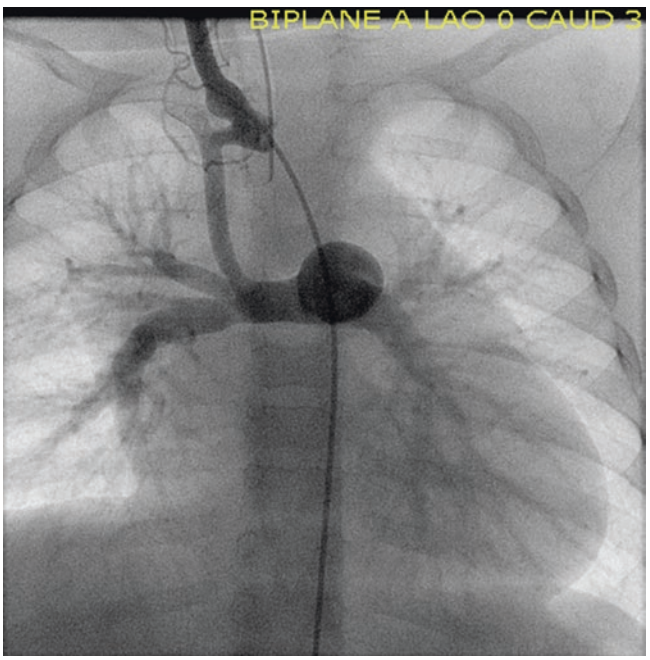



**Fig. 15.16** Patient 3. Acute modified Blalock-Taussig shunt (mBTS) occlusion. An angiogram in the right innominate artery 7 months later confirms persistent patency of the right mBTS

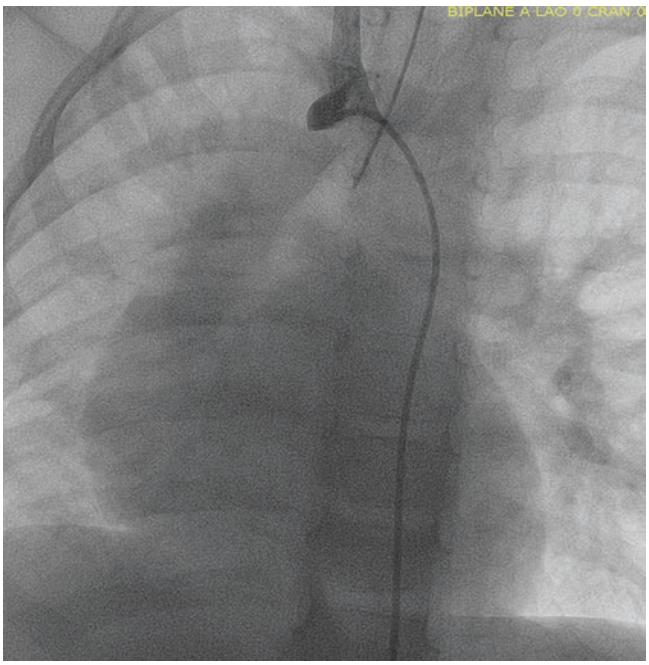



**Fig. 15.17** Patient 4. Ductal origin of a pulmonary artery. A 3-year-old female with history of so-called "absent" right pulmonary artery (RPA) was found to have an occluded ductal origin of the RPA from the right innominate artery. Via femoral arterial access, an angiogram with a 4 Fr JR 2 catheter in the ductal ampulla at the base of the right innominate artery shows an occluded ductus arteriosus

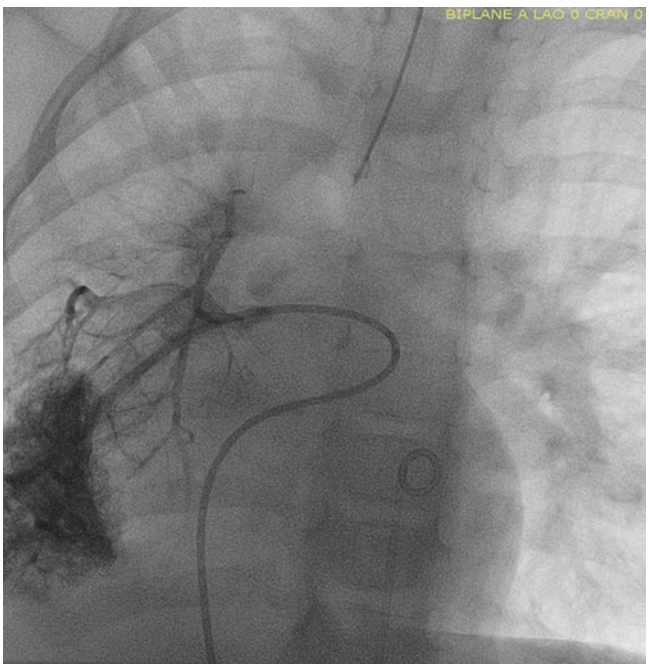



**Fig. 15.18** Patient 4. Ductal origin of a pulmonary artery. Transseptal puncture was performed to facilitate right pulmonary venous wedge angiography, demonstrating a diffusely small but patent right pulmonary artery. This angiogram is also used as a road map for the intervention

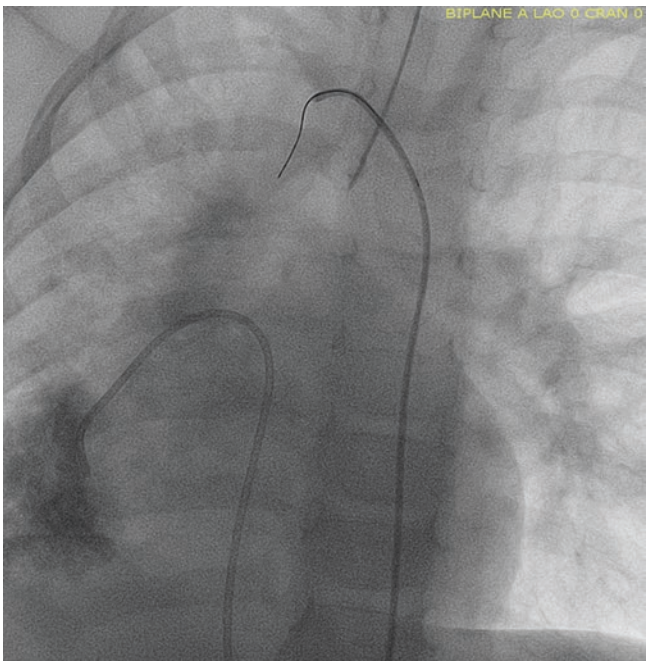



**Fig. 15.19** Patient 4. Ductal origin of a pulmonary artery. A 0.014" Pilot 200 wire (Abbott Vascular, Santa Clara, California, USA) was used to cross the occluded ductal segment

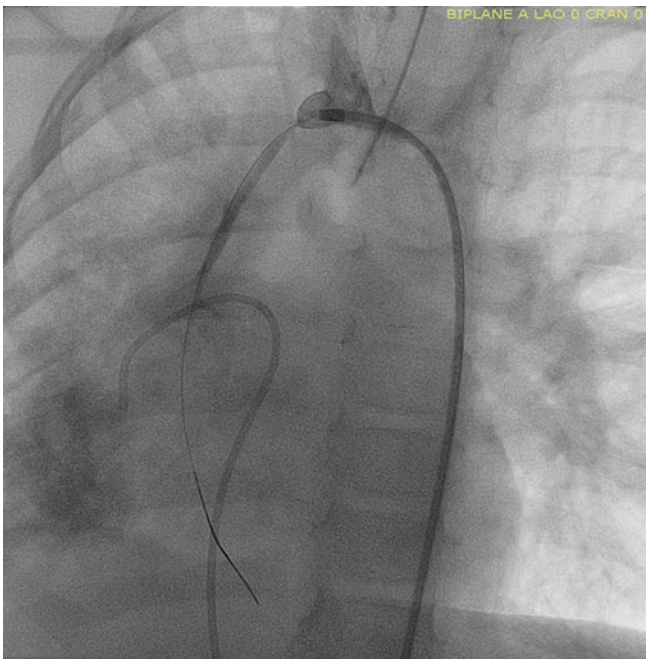



**Fig. 15.21** Patient 4. Ductal origin of a pulmonary artery. Over a 0.014" wire, the duct was angioplastied along its entire length, resulting in a patent ductal tract leading to the RPA

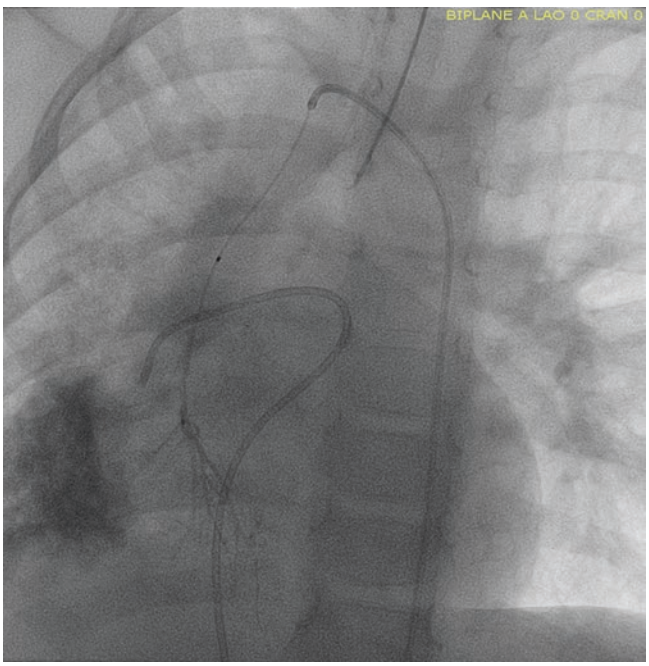



**Fig. 15.20** Patient 4. Ductal origin of a pulmonary artery. After wire removal, an angiogram through a microcatheter confirms the catheter is intravascular in the distal RPA

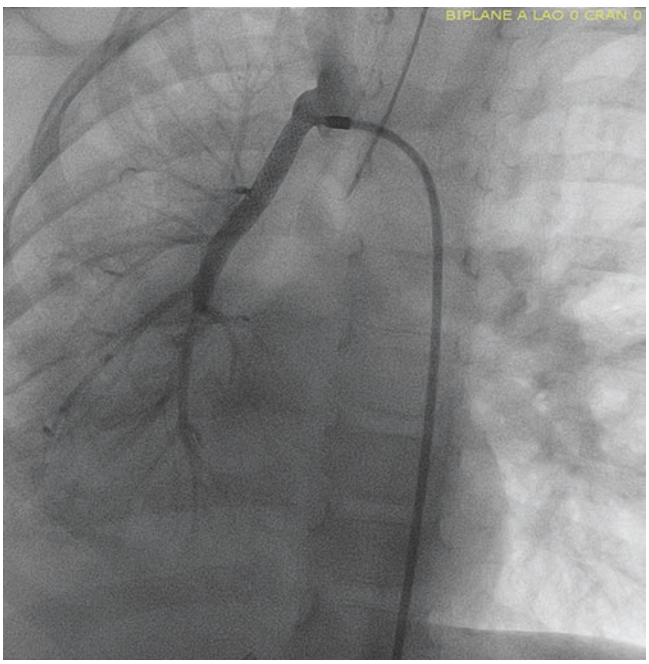



**Fig. 15.22** Patient 4. Ductal origin of a pulmonary artery. The duct was stented with a 4 mm × 24 mm Promus Premier everolimus-eluting platinum chromium stent (Boston Scientific, Marlborough, Massachusetts, USA) with excellent flow seen afterward through the ductal stent to the RPA. Of note, reperfusion injury is seen in the right lung, which is not uncommon after this type of procedure

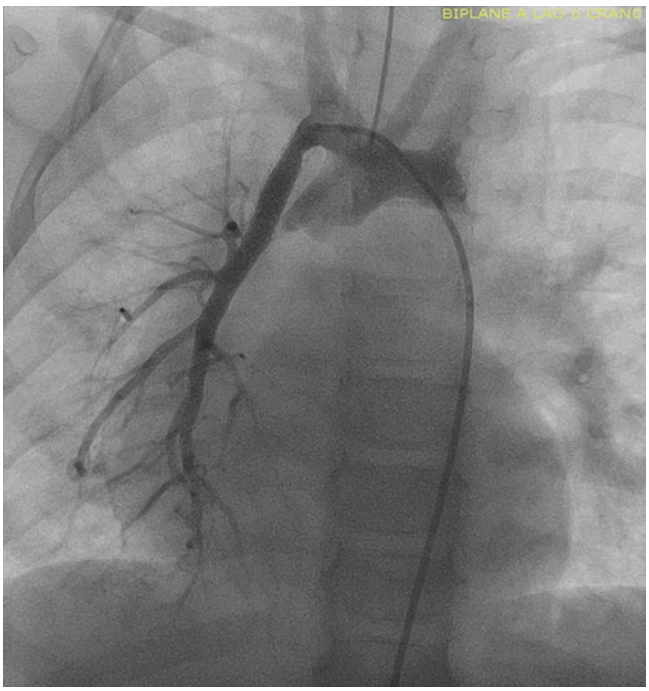


**Fig. 15.23** Patient 4. Ductal origin of a pulmonary artery. A repeat catheterization was performed 5 months later to redilate the ductal stent. After redilation, angiography shows a patent ductal stent with mild proximal in-stent stenosis. She subsequently underwent surgical reconnection of her branch pulmonary arteries

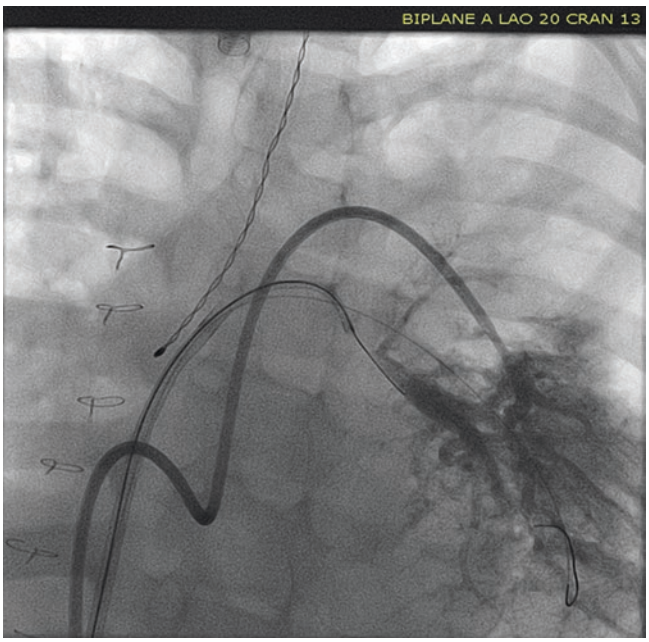



**Fig. 15.25** Patient 5. Pulmonary vein atresia. After transseptal puncture, a 4 Fr JR 2 catheter was positioned within a 7 Fr JR4 guiding catheter (Medtronic, Minneapolis, Minnesota, USA). The occluded left lower pulmonary vein was ultimately crossed with a 0.014" Confianza Pro wire (Abbott Vascular) as seen by left lower lobe pulmonary artery wedge angiography. A wire that has been placed in the patent left lingular pulmonary vein for catheter stability can also be seen

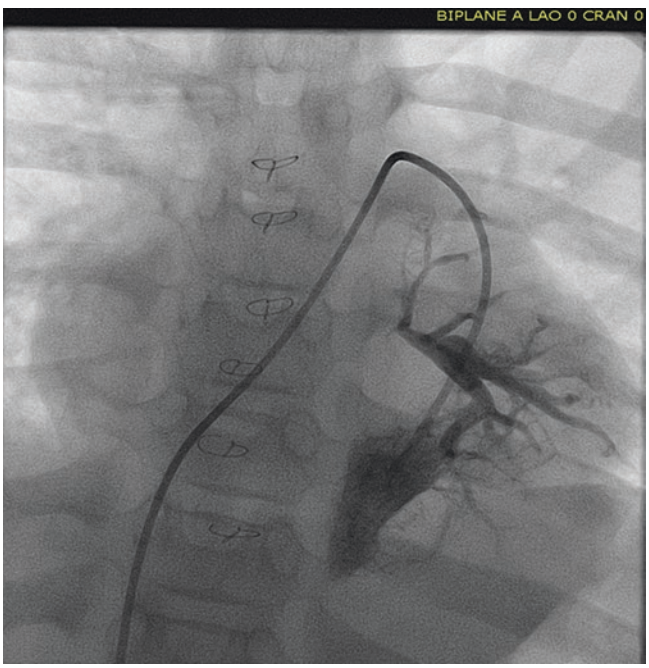



**Fig. 15.24** Patient 5. Pulmonary vein atresia. A 5-year-old girl with cri du chat syndrome had undergone prior surgical repair for pulmonary vein stenosis and an atrial septal defect. She developed restenosis of her pulmonary veins and underwent pulmonary vein balloon angioplasty but subsequently developed left lower pulmonary vein occlusion as seen on this left lower lobe pulmonary artery wedge angiogram

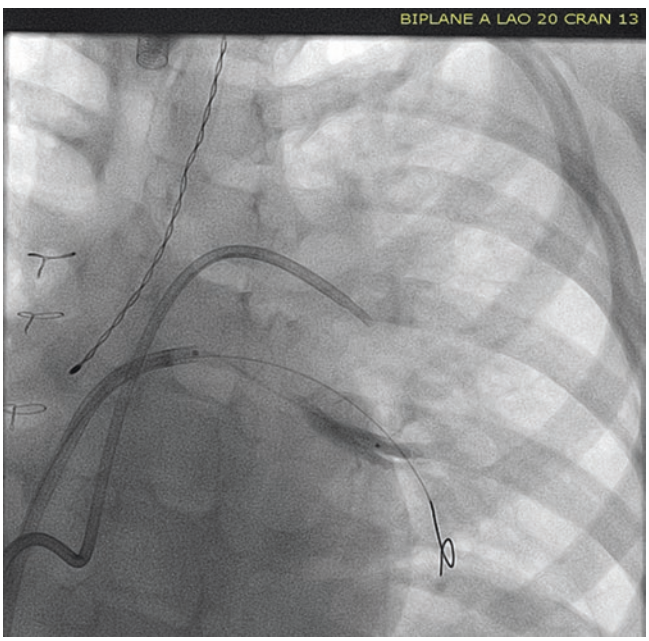



**Fig. 15.26** Patient 5. Pulmonary vein atresia. An angiogram through a microcatheter that was advanced over the guidewire confirms intravascular position in the left lower pulmonary vein before proceeding with angioplasty and stenting

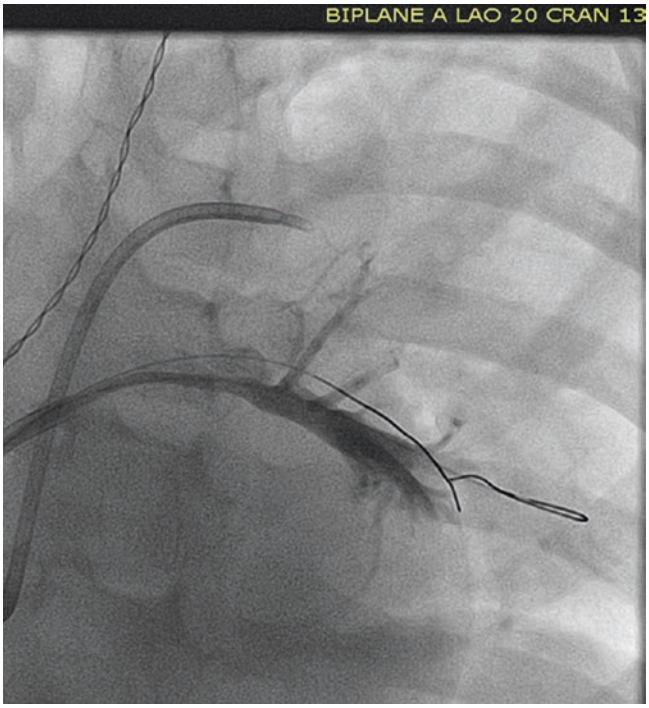



**Fig. 15.27** Patient 5. Pulmonary vein atresia. Angiogram in the left lower pulmonary vein after balloon angioplasty

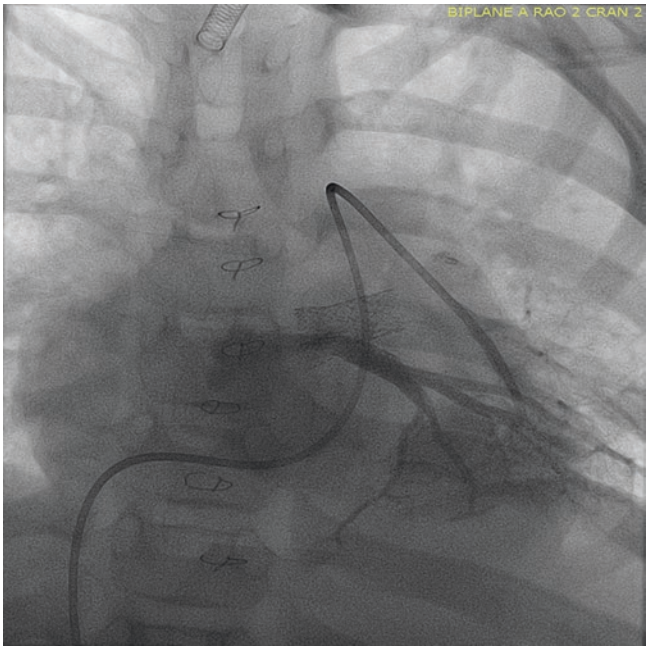



**Fig. 15.28** Patient 5. Pulmonary vein atresia. Two months later, a 4 mm × 16 mm Promus Premier everolimus-eluting platinum chromium stent was implanted in the left lower pulmonary vein. A stent can also be seen in the left upper pulmonary vein

# PDA Stenting in Duct-Dependent Pulmonary Circulation

# 16

Kothandam Sivakumar

## 16.1 Introduction

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 the 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 these 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. Five to twenty percent 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.

## 16.2 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. 16.1).

## 16.3 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:

*Group A*: *Pulmonary atresia in biventricular hearts*
*Group B*: *Pulmonary atresia in univentricular circulation*
*Group C*: *Transient inadequacy of pulmonary circulation*

### 16.3.1 Pre-procedural Imaging

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

1. Duct—morphology, origin and insertion, length, and diameter at aortic and pulmonary end
2. Aortic arch—side of the aortic arch and arch branches for axillary or carotid arterial access
3. Pulmonary arteries—mediastinal and hilar pulmonary artery sizes and stenosis of confluence at duct insertion site
4. Intracardiac anatomy—differentiate groups A,B, and C
5. Ventricular systolic function, atrioventricular valve annulus size and function, aortic root diameter, and aortic valve function
6. Venous anomalies for transvenous approaches
7. Interatrial communication and need for balloon septostomy

K. Sivakumar (✉)
Department of Cardiology, Madras Medical Mission, Chennai, India


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_16

### 16.3.2 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 (3M Bair Hugger), draping sterile warm linen, and warming saline and contrast before use.
- Cross-matched packed red cells are reserved for procedural blood loss.
- Intubation and general anesthesia are preferred if axillary or carotid arterial access is anticipated.
- Intravenous fluids counter PGE1-related hypotension and facilitate quick vascular access.
- 4F or 5F 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.
- 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 (Figs. 16.2 and 16.3). In ducts originating from right arch, RAO 20° Cranial 20° projection is chosen (Fig. 16.4).
- Ductal origin and insertion, morphology, length, and diameter are delineated in lateral view (Fig. 16.1c) in type I–III ducts. In type IV and V ducts, aortogram is done in anteroposterior view.
- 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 extra support 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 **microcatheter** can facilitate further advances of the guidewire.
- The stenting is done in most ducts through long 4F sheaths but sometimes with 5F Judkins right coronary guide catheter.
- Premounted nondrug eluting **coronary stents** are chosen; the 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.

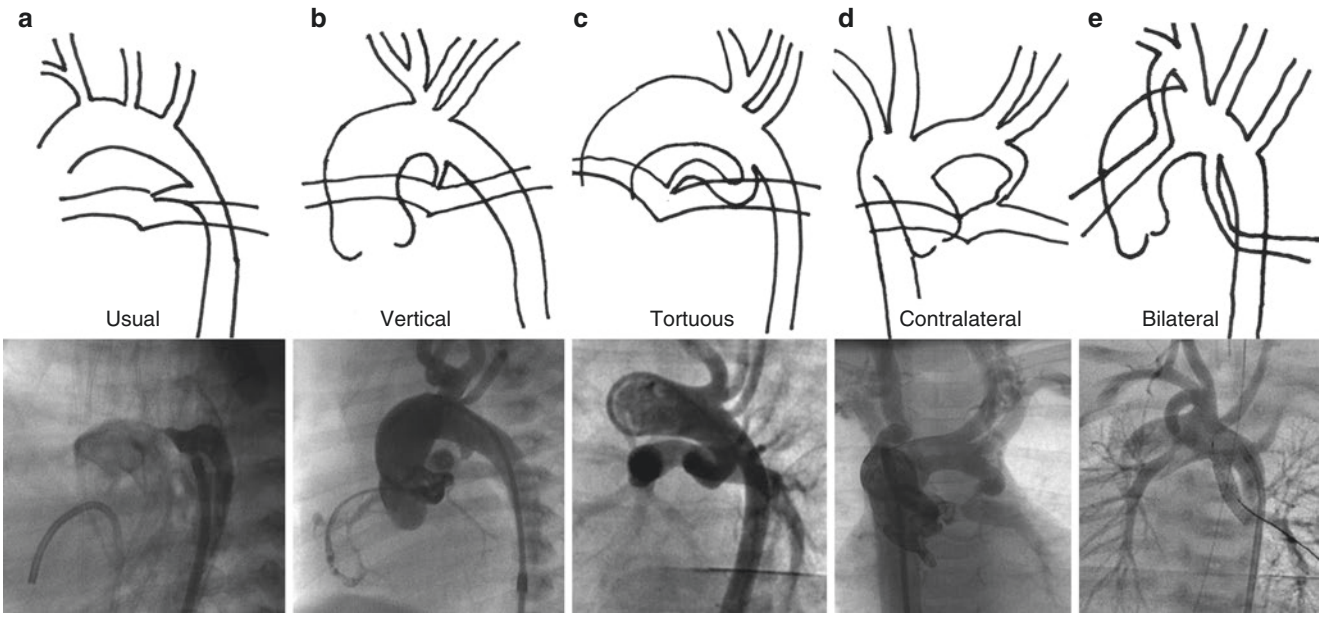



**Fig. 16.1** Different ductal morphology: The ducts in pulmonary atresia can be in various morphologic forms. Type I (usual form): The duct arises from the junction of the arch to descending aorta and courses anteriorly to insert in confluence of pulmonary arteries. Type II (vertical form): The duct arises proximally from the undersurface of the aortic arch and courses vertically down to the confluence. Type III (tortuous form): In this commonest morphological form, the duct takes a C- or S-shaped bend before inserting in the confluence. Type IV (contralateral form): The duct arises opposite to the side of the aortic arch from either the contralateral innominate artery or subclavian artery. Type V (bilateral form): 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

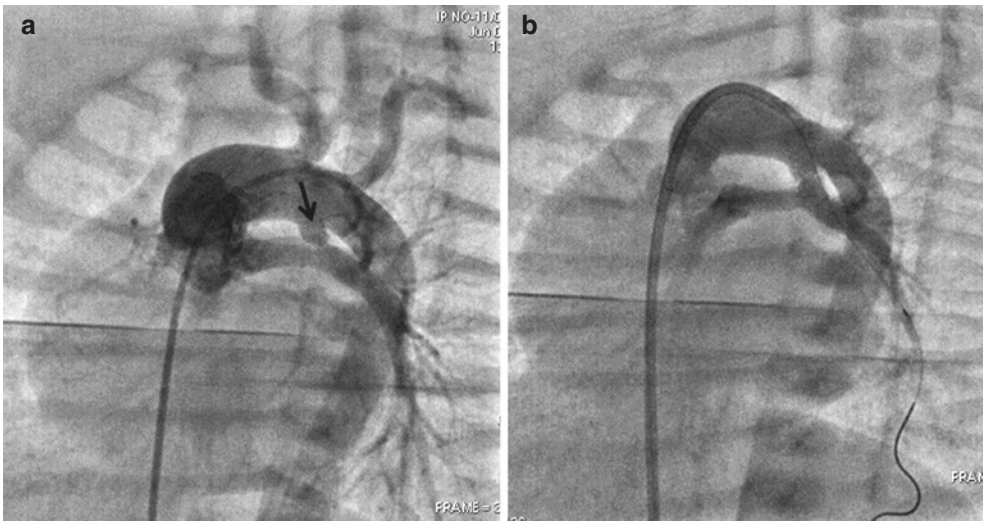



**Fig. 16.2** Aortogram in shallow left anterior oblique view with a pigtail catheter advanced from femoral vein through the right ventricle into the left aortic arch (**a**) shows a vertical duct (arrow) arising from undersurface of the aortic arch opposite to the right innominate artery. In such cases, advancing a guide wire into distal branch of the left pulmonary artery and DS is done more easily (**b**) from favorable angle through the transvenous route

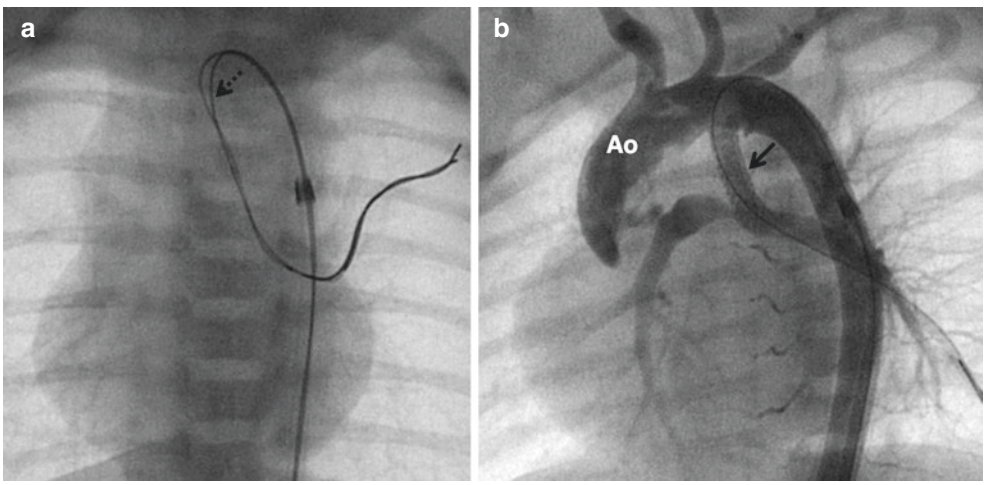



**Fig. 16.3** Through a 4F long sheath, a coronary guidewire was advanced (**a**) through a vertical duct. An additional buddy wire (dotted arrow) was advanced to facilitate the passage of the stent through the acute angulation of vertical duct. After stenting, (**b**) aortogram in shallow left anterior oblique view confirmed the stenting (arrow) of the entire length of the duct

- 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.
- 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. 16.3) 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.

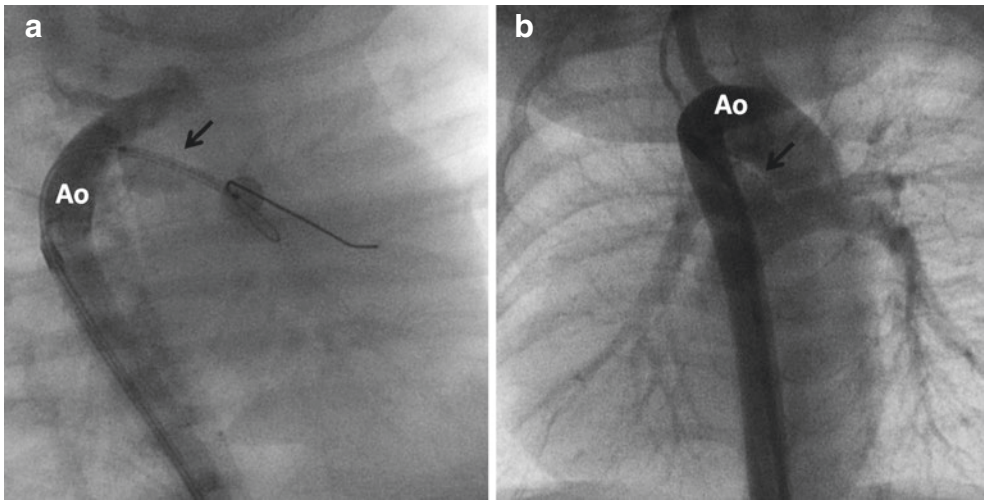



**Fig. 16.4** Angiogram through a 4F long sheath (**a**) in shallow right anterior oblique projection was done to check the position of a 4 mm stent (arrow) placed on a coronary guidewire through the duct advanced into the left pulmonary artery. After stenting, a repeat aortogram (**b**) showed good filling of the pulmonary arteries

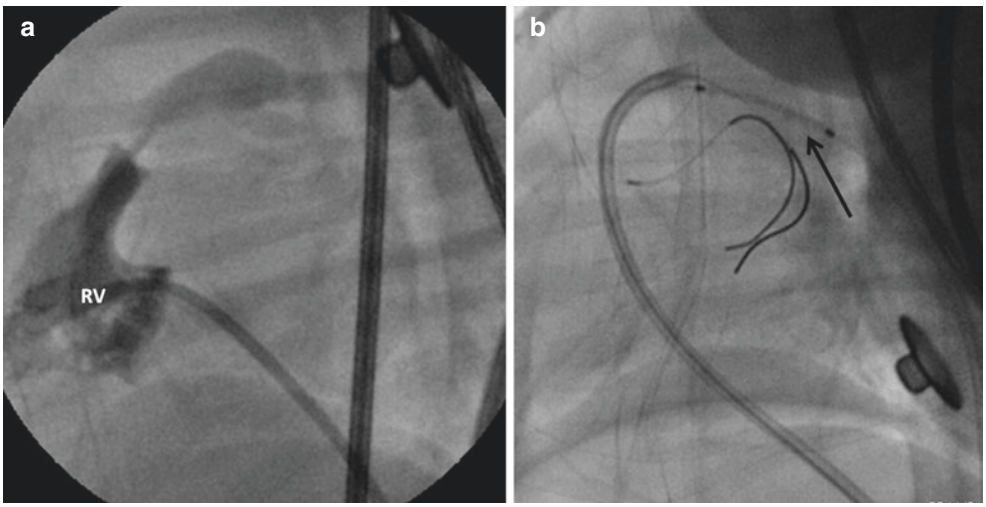



**Fig. 16.5** Right ventricular (RV) angiogram in lateral view (**a**) demonstrates thick pulmonary valve with a narrow contrast jet into the pulmonary artery indicating severe stenosis which was dilated with a balloon. The persistent hypoxia due to inadequate antegrade pulmonary flows was an indication 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

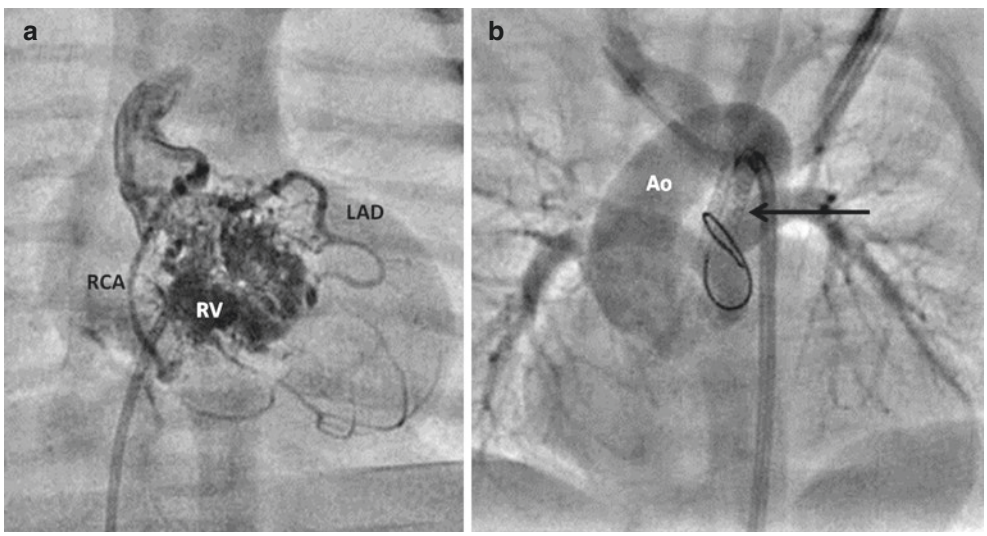



**Fig. 16.6** Right ventricular angiogram (**a**) shows a hypoplastic right ventricle (RV) filling multiple sinusoids which fill the right coronary artery (RCA) and left anterior descending interventricular artery (LAD) indicative of right ventricle-dependent coronary circulation (RVDCC). This precludes 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

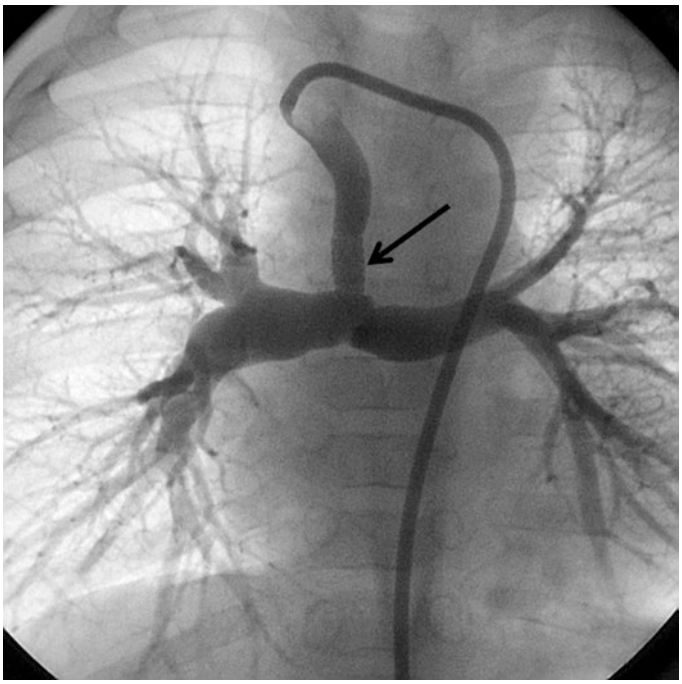


**Fig. 16.7** Injection through a guide catheter passed from the left aortic arch into the right subclavian artery shows the long contralateral duct which inserts into the confluence. The narrowing in the distal insertion of the duct was stented (arrow) with a 3.5 mm stent

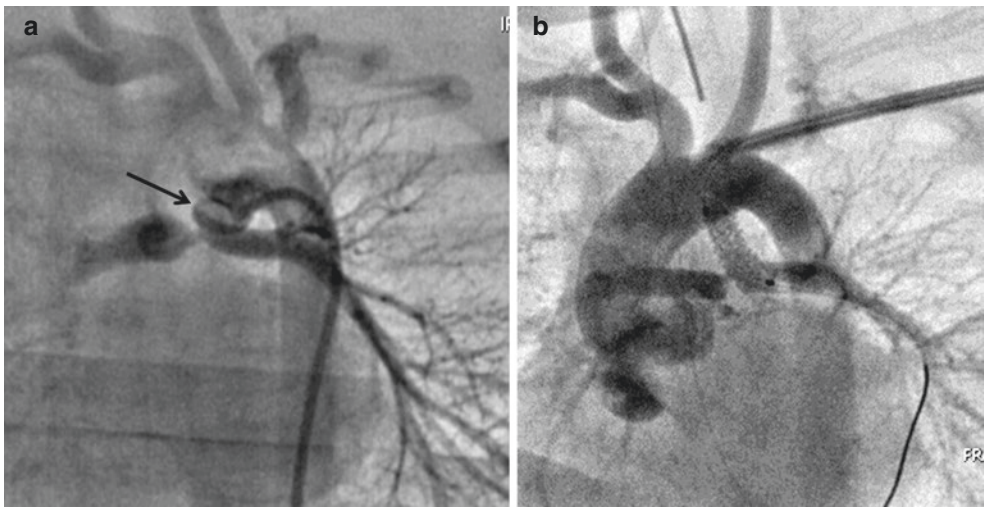



**Fig. 16.8** Femoral access angiogram (**a**) with an end-hole catheter shows a tortuous type III duct (arrow) which needed axillary artery access for successful ductal stenting with a 4 mm coronary stent (**b**). The tip of the guidewire is parked in the lower lobe of the left lung

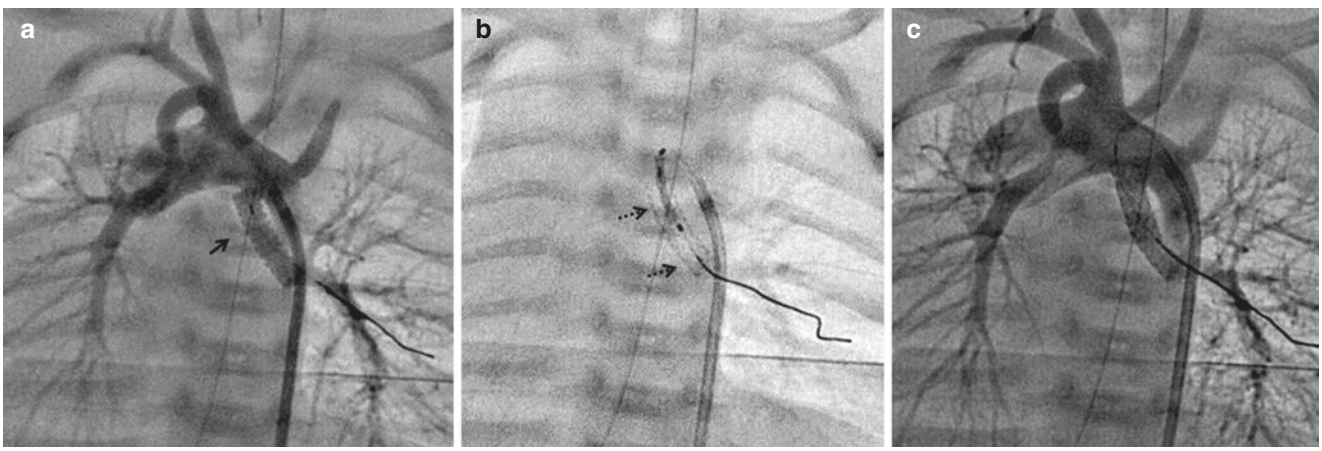



**Fig. 16.9** In a type V bilateral duct in a neonate with single ventricle and pulmonary atresia, aortogram (**a**) after stenting of the left-sided duct from the undersurface of the aortic arch shows that the aortic end of the duct is uncovered by the stent. The guidewire is still in place. A second 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 sitting

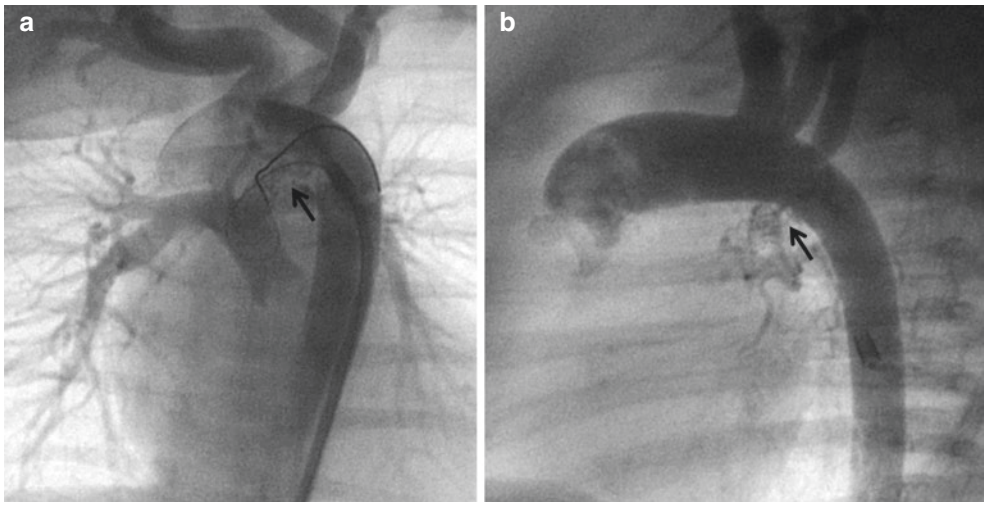



**Fig. 16.10** Unilateral right lung hyperperfusion after ductal stenting in a patient with tetralogy of Fallot with pulmonary atresia

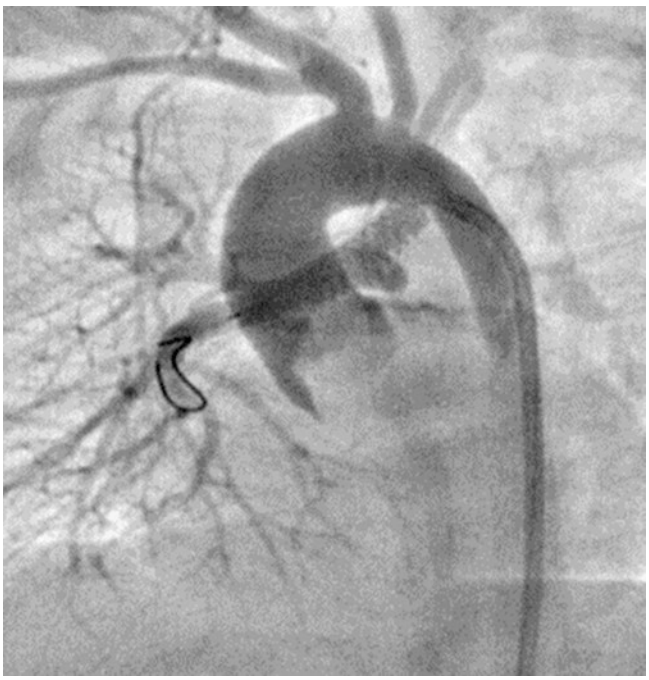


**Fig. 16.11** Aortogram in shallow left anterior oblique view shows successful stenting (arrow) of a type III duct from undersurface of aortic arch in a patient with d-transposition of great arteries, large ventricular septal defect, and pulmonary atresia. After removal of the guidewire, repeat aortogram shows complete thrombotic occlusion of the stent

# Part VI

# Step-by-Step Procedures: Closing or Creating a Defect

# Step-by-Step ASD Closure

17

John D. Thomson

If left untreated, ASDs lead to pulmonary hypertension, atrial dysrhythmia, exercise intolerance and eventual clinical right heart failure. They may be responsible for an excessive burden of respiratory symptoms and occasionally failure to thrive during childhood. Most symptoms and complications are progressive so ideally closure should be performed during the paediatric age range, although it is rarely necessary before the age of 3 years.
Indications for treatment:

- Evidence of right ventricular volume overload (a reliable surrogate for a significant left-to-right shunt) on either transthoracic echocardiography (TTE) or cMRI
- Significant left-to-right shunt (>2:1) as evidenced by oximetry at the time of catheterisation (not strictly necessary in the modern era given the availability of non-invasive imaging techniques)

Patient selection:

- In the main only secundum ASD's can be closed in the catheter laboratory. There are rare reports of closure of certain types of sinus venosus ASD using covered stents by in general these defects and premium ASD's need surgical treatment.
- Currently defects of up to 40 mm in diameter provided the margins are adequate (Figs. 17.1 and 17.2).
- Margins: A 5 mm rim for anchorage of devices in all areas is necessary, save for the retro-aortic rim (Fig. 17.3); although that is somewhat controversial, some operators feel that deficiency of this area is a risk for device related erosion of the myocardium.

Pre-procedure assessment:

- Proper counselling of the patient/family leading to informed consent
- Review of the available imaging, particularly to identify any information that may not have been addressed prior to the procedure, e.g. margins that have not been demonstrated on TTE

Procedure (Figs. 17.4, 17.5 and 17.6):

- Guided by trans-oesophageal echocardiogram (TOE) or (more rarely) intracardiac echocardiography (ICE) in suitable adults.
  - Systematic approach to identify defect size (measured in at least two orthogonal planes (Fig. 17.2), establish individual margins and ensure no anatomic contraindication to closure, e.g. anomalous pulmonary venous drainage.
- Femoral venous access (ideally ultrasound guided).
- Patient heparinisation 100 iu/kg.
- Diagnostic catheter.
  - Systematic, predefined data collection, e.g. pulmonary pressures and shunt by oximetry
- Super-stiff-type wire positioned across the defect into a left-sided pulmonary vein to guide the delivery system (Fig. 17.4).
- Balloon sizing of the defect. The "stop-flow" (abolition of colour flow on TOE) technique should be used to avoid stretching and oversizing the defect (Fig. 17.5) (Note: many operators do not view balloon sizing as a necessary step as defects can be reliably closed on echocardiographic measurements alone).
- Deployment of a device appropriate to the anatomy and size of the defect (Figs. 17.7 and 17.8).
  - Devices available are generally either "self-centring" (with a core), e.g. the Amplatzer septal occluder (ASO) or the Occlutech Figulla, or a "cribriform"-type

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_17) contains supplementary material, which is available to authorized users.

J. D. Thomson (✉)
Department of Congenital Heart Disease,
Yorkshire Heart Centre, Leeds, UK
e-mail: john.thomson8@nhs.net



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_17

occluder (with a central strand rather than a core), e.g. Amplatzer cribriform device or the Gore septal occluder (GSO).

- Device selection depends on laboratory preference, operator expertise and the defect. However by far the most implanted device worldwide is the Amplatzer septal occluder (ASO).

Pitfalls:

- Missing margins: The inferior rim towards the IVC is particularly hard to see on TOE. Ensure this is reliably identified on ultrasound before device insertion (Fig. 17.2).
- Systematic oversizing of defects: This may be a risk factor for erosion which occurs in 0.1–0.3% of cases.
- Device embolisation: This can occur even in experienced hands for many reasons. Devices that migrate to the aorta and pulmonary artery can often be removed by snaring and retrieval into a large (>10F) sheath. Embolised devices that lodge within cardiac chambers (Fig. 17.9) should be approached with great care as permanent damage can be caused when retrieving percutaneously—surgery may be a better option!

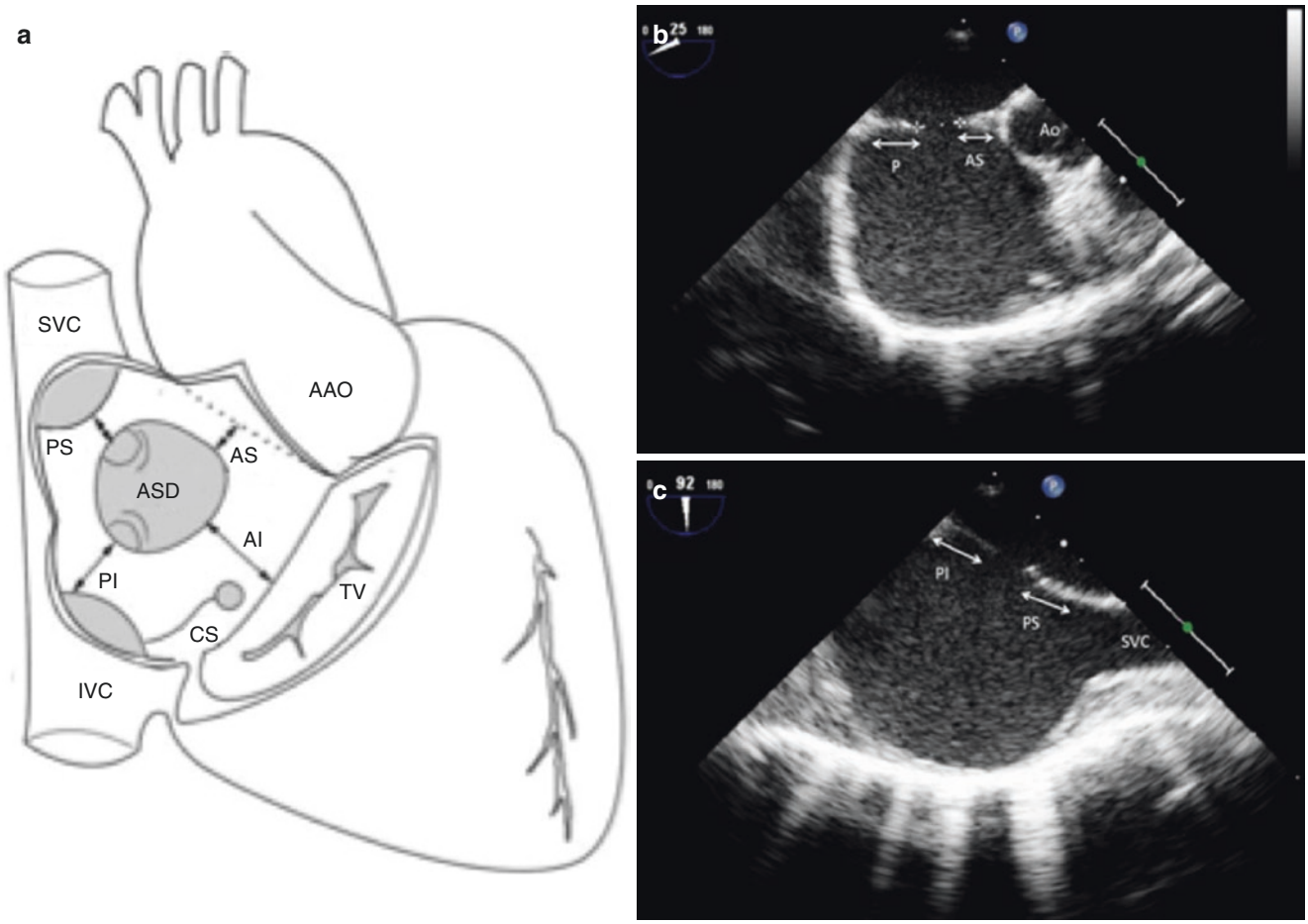



**Fig. 17.1** (**a**) Margins of the atrial septum. PI, postero-inferior atrial rim; PS, postero-superior rim; AI, antero-inferior; AS, antero-superior atrial rim. (**b**) Trans-oesophageal echocardiography (TEE), "30-degree" view. RA, right atrium; LA, left atrium; Ao, aorta; P, posterior atrial rim; AS, antero-superior atrial rim. (**c**) TEE, bi-caval view. PI, postero-inferior atrial rim; PS, postero-superior rim

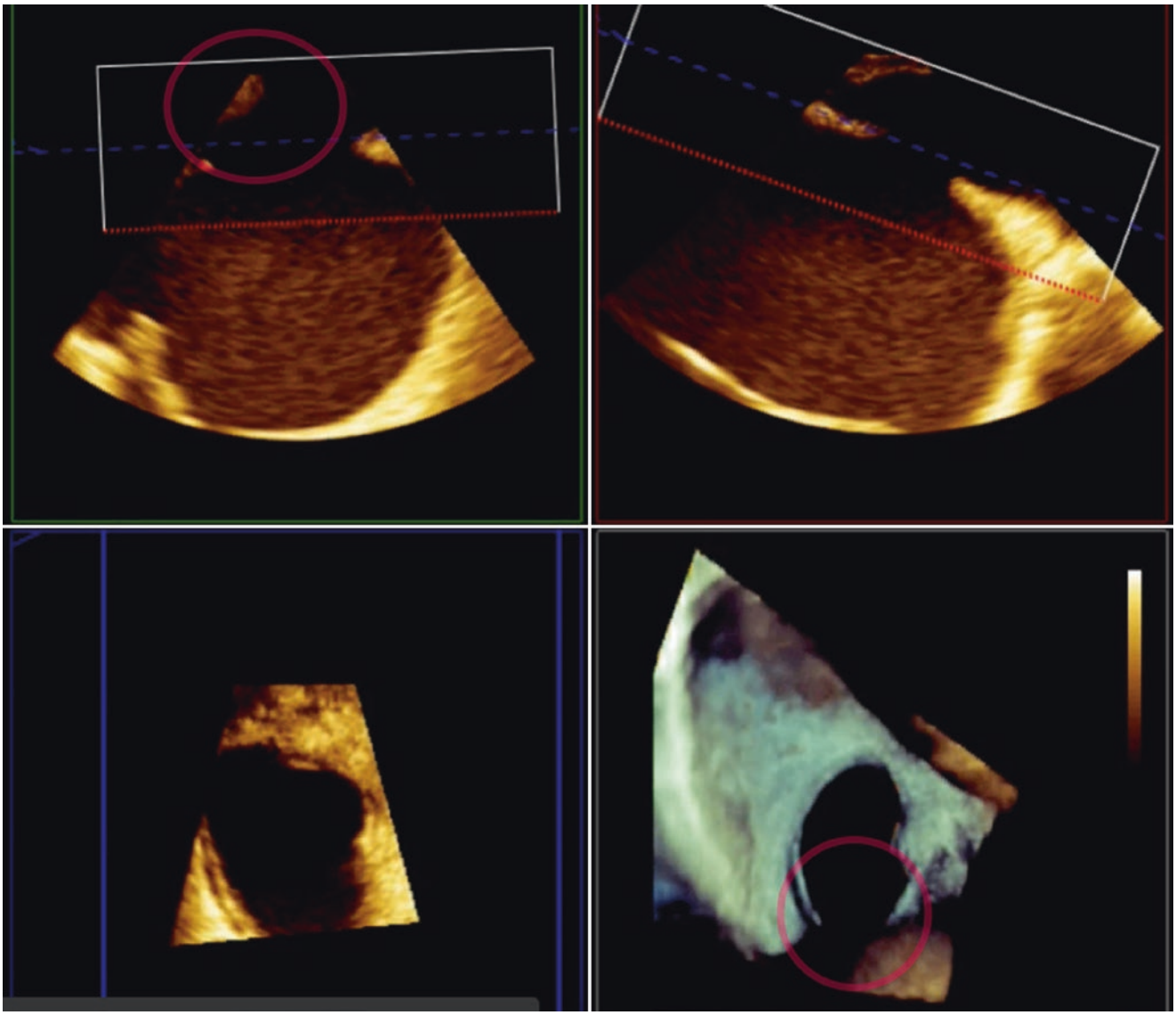


**Fig. 17.2** 3D TTE. Secundum ASD with deficient inferior rim (circled)

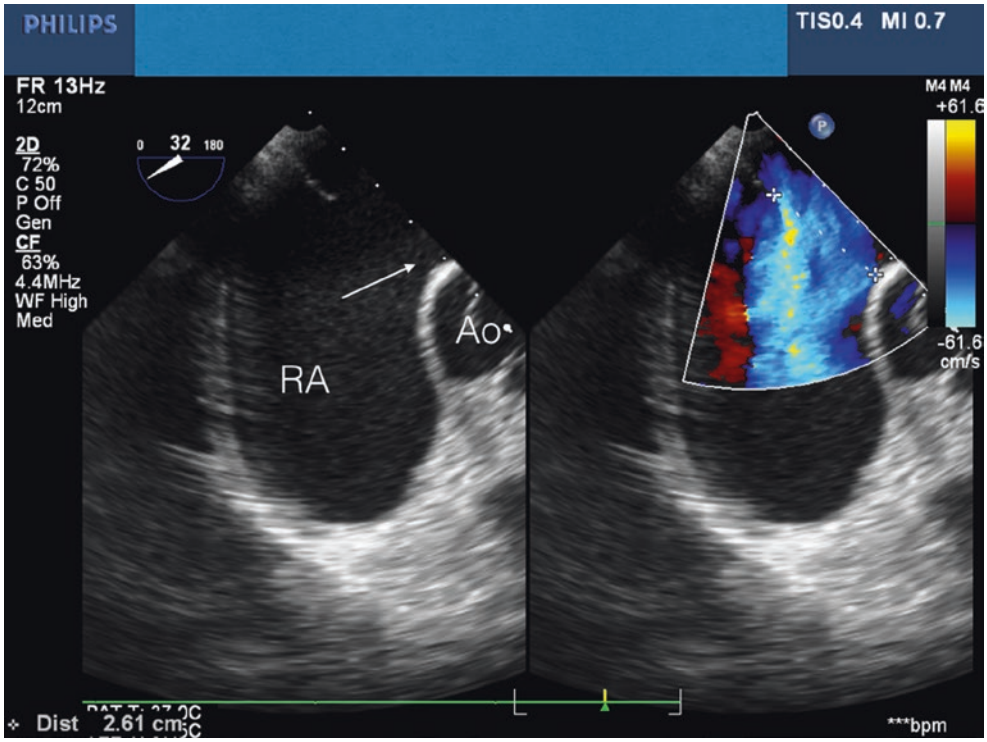



**Fig. 17.3** TEE, "30-degree view"-deficient aortic rim (arrowed). RA, right atrium; Ao, aorta

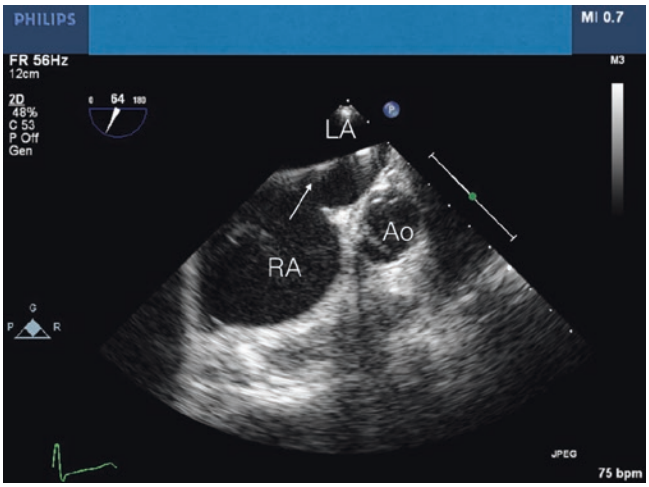



**Fig. 17.4** Stiff wire across atrial septal defect (arrowed)

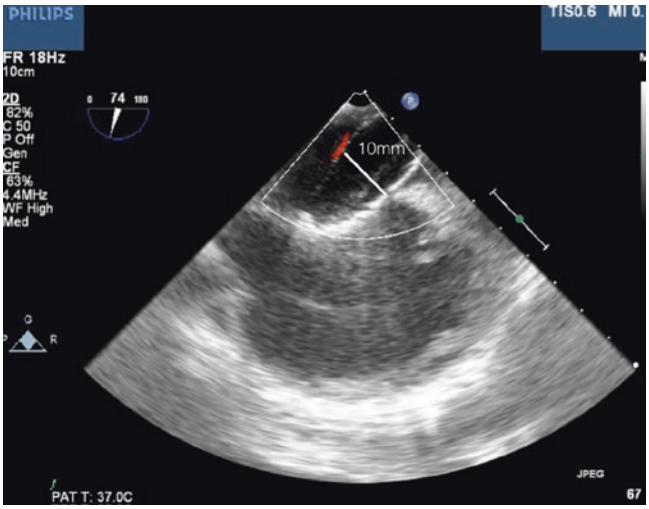



**Fig. 17.5** Stop-flow balloon sizing

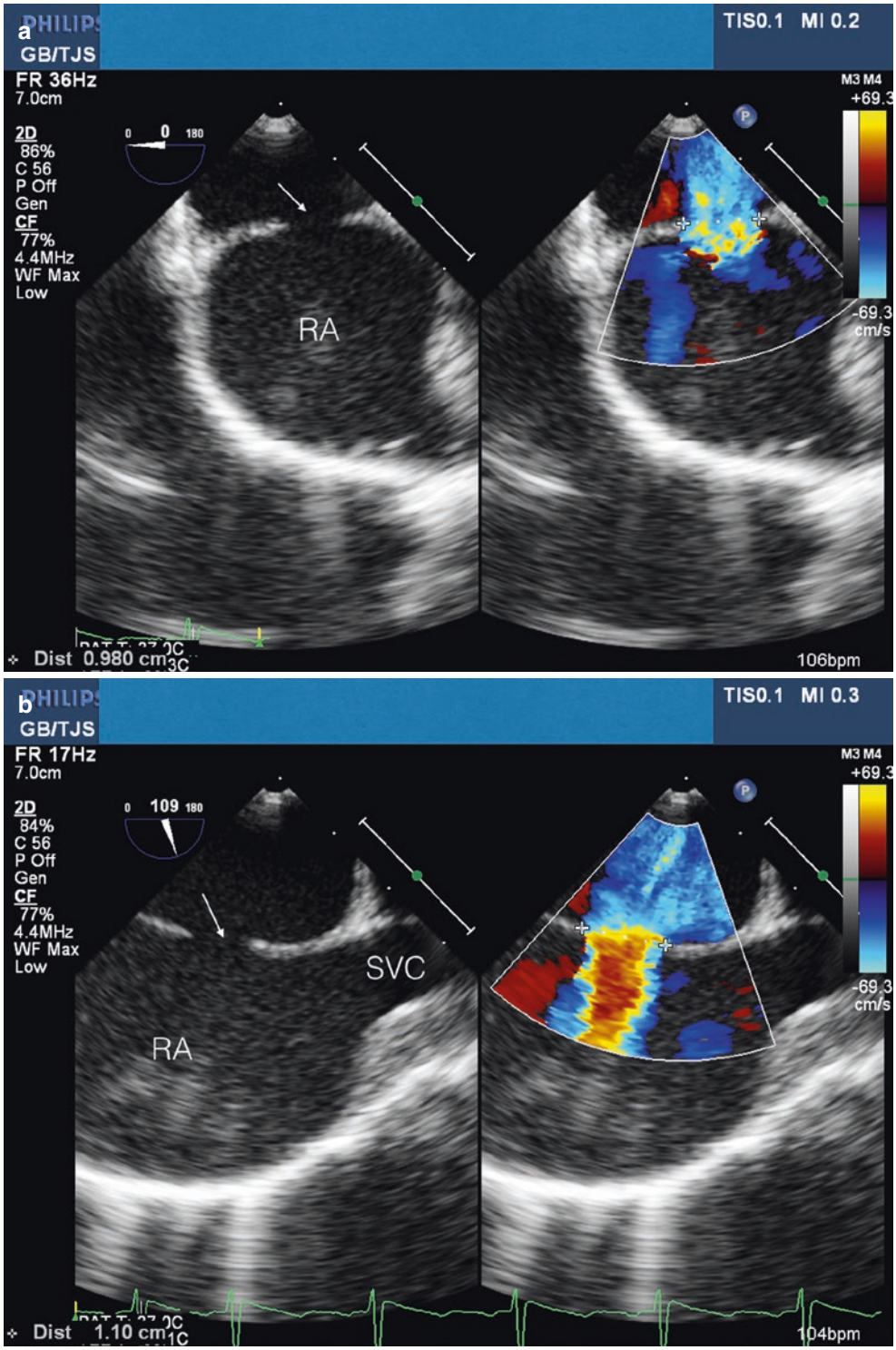



**Fig. 17.6** (a, b) ASD (arrowed) in two orthogonal planes

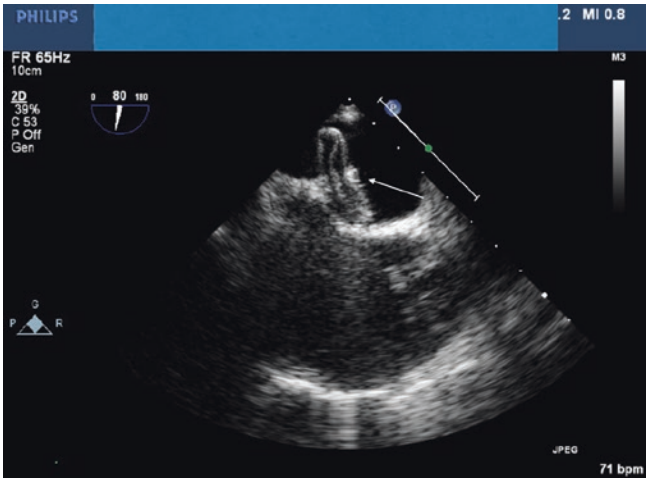



**Fig. 17.7** Deployment of the left atrial disk of a 10 mm Amplatzer septal occluder (arrowed)

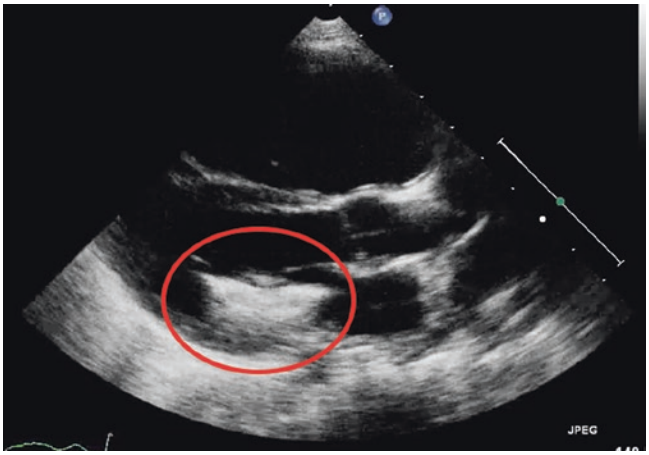



**Fig. 17.9** TTE, parasternal long axis. Embolised Amplatzer device (circled) in the LA/LV

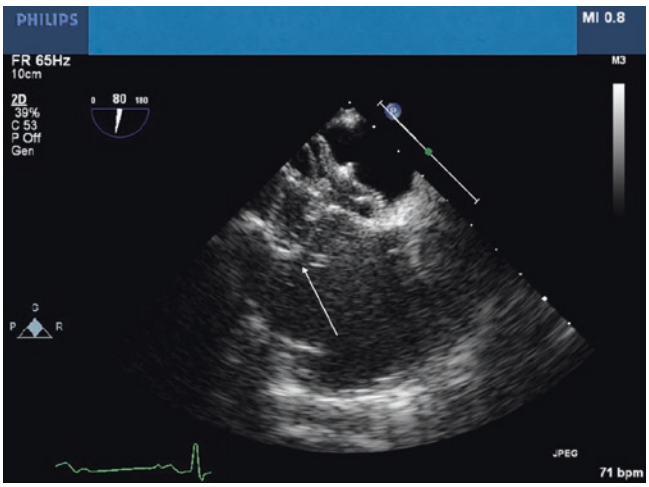



**Fig. 17.8** Deployment of the right atrial risk (arrowed)

**Video 1** Deployment sequence (MP4 6704 kb)

**Video 2** Stability check prior to release (MP4 6666 kb)

# Fontan Fenestration Closure

# 18

Derize E. Boshoff and Marc H. Gewillig

## 18.1 Introduction

Fenestrating the Fontan circuit allows a protective right-to-left shunting at atrial level to overcome the period of low output in the postoperative period, decreasing morbidity and mortality. 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 a few months after completion of the Fontan circulation, when the patient is free of diuretics, if $O_2$ saturations are <90% and test occlusion is tolerated.

## 18.2 Patient Selection

Defining the "ideal" patient for fenestration closure is debatable [2], 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
  - Discontinuation of diuretics within weeks after surgery
- *Clinically:*
  - No evidence of low cardiac output or systemic congestion
  - Normal AV conduction on ECG and absence of significant arrhythmias
- *Imaging (TTE/TEE ± MRI):*
  - Exclusion of a high-velocity shunt through the fenestration on echocardiography
  - Unobstructed Fontan connections and low pulmonary vascular resistance
  - Good ventricular function and absence of significant valve regurgitation
  - Unobstructed systemic outflow and pulmonary venous return
  - Absence of significant venovenous or aortopulmonary collaterals

## 18.3 Catheterization Procedure

Procedure is performed under general anaesthesia. Antibiotic prophylaxis and heparin (100 IU/kg IV, maximum 5000 IU) should be administered routinely.

- *Access:*
  - Femoral (jugular if obstructed) venous and arterial access
- *Hemodynamic evaluation:*
  - Saturations and pressures throughout the Fontan pathway and systemic circulation should be documented on room air.
- *Angiography:*
  - Superior and inferior caval veins to visualize the Fontan connections, surgical fenestration (Figs. 18.1 and 18.2) and additional interatrial leaks.
  - Pulmonary arteries with levo-phase to visualize pulmonary venous return; be aware that external compression of the pulmonary veins (by the conduit or by the

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_18) contains supplementary material, which is available to authorized users.

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


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_18

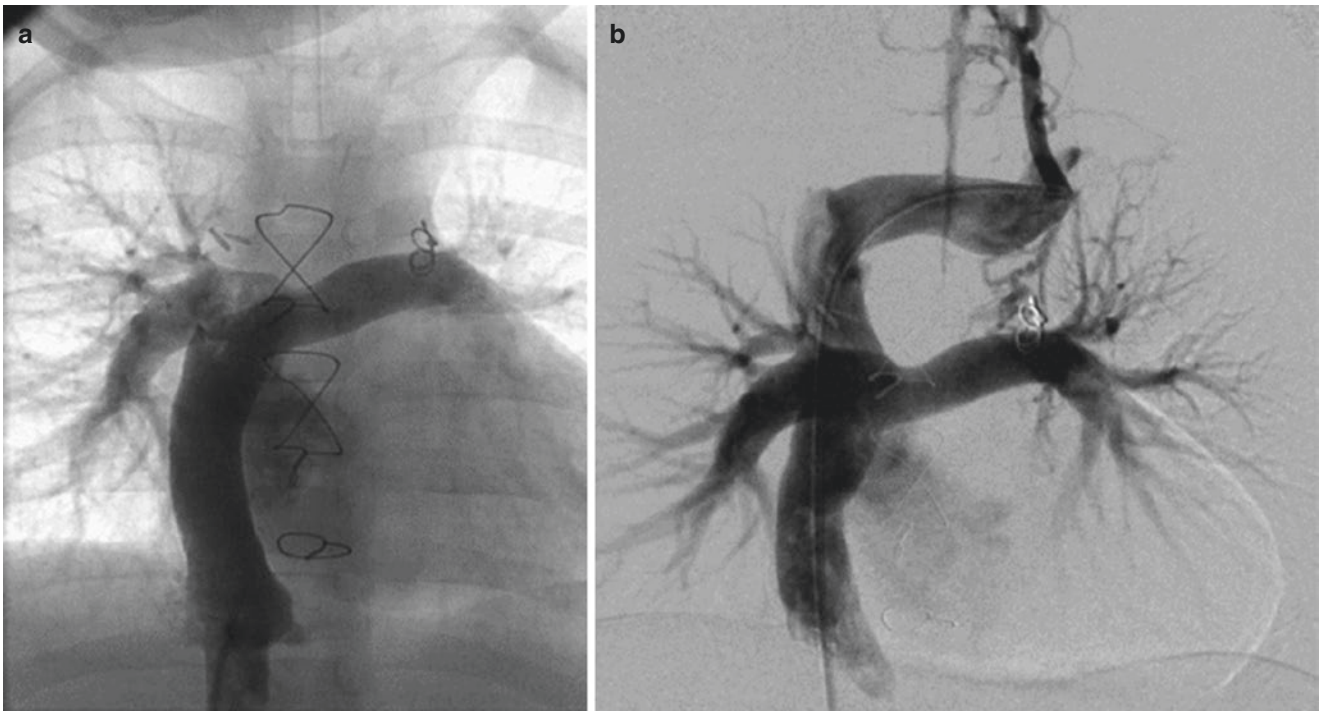



**Fig. 18.1** (**a**) Frontal view of contrast injection in inferior caval vein: a 20-mm conduit is mounted between the inferior caval vein and pulmonary artery; a 4.5-mm fenestration allows right-to-left shunt into the left atrium. (**b**) Selective injection in the innominate vein to exclude significant venovenous connections

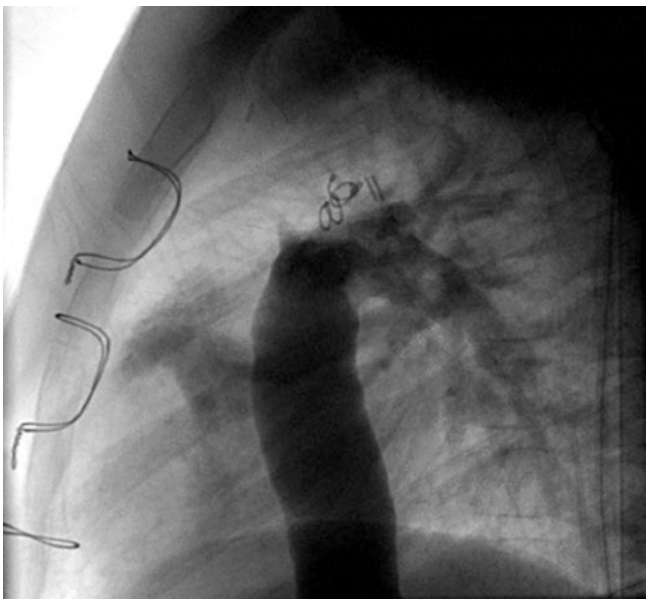


**Fig. 18.2** Lateral projection demonstrating the 4.5-mm fenestration in this 20-mm extracardiac Fontan conduit

left atrium) is easily missed (to exclude: use TEE or 3D angio).
- Selective injection in the innominate vein and right and left hepatic vein to exclude venovenous connections (Fig 18.1a).
- Aortogram to exclude significant aortopulmonary collateral arteries.

- *Anatomical abnormalities amendable to interventional treatment should be treated:*
  - Balloon dilation and/or stenting of obstructed Fontan connections or stenosed/hypoplastic pulmonary arteries
  - Occlusion of significant collaterals if appropriate
  - Treatment of systemic obstruction (i.e. recoarctation) by balloon dilation and/or stenting
  - Pulmonary vein obstruction

### 18.3.1 Test Occlusion of the Fenestration

- *Indications:*
  - It is recommended in case of unfavourable baseline haemodynamics and in high-risk patients by quantifying changes in the systemic or mean venous pressure and systemic saturation.
  - It remains debatable though 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.
- *How to perform:*
  - Pass a 7F balloon-tipped, multi-lumen catheter (Swan-Ganz catheter) over a wire into the systemic atrium; inflate the balloon with 1-$cm^3$ diluted contrast and pull back against the atrial wall/fenestration to allow for temporary occlusion (at least for 15 min).

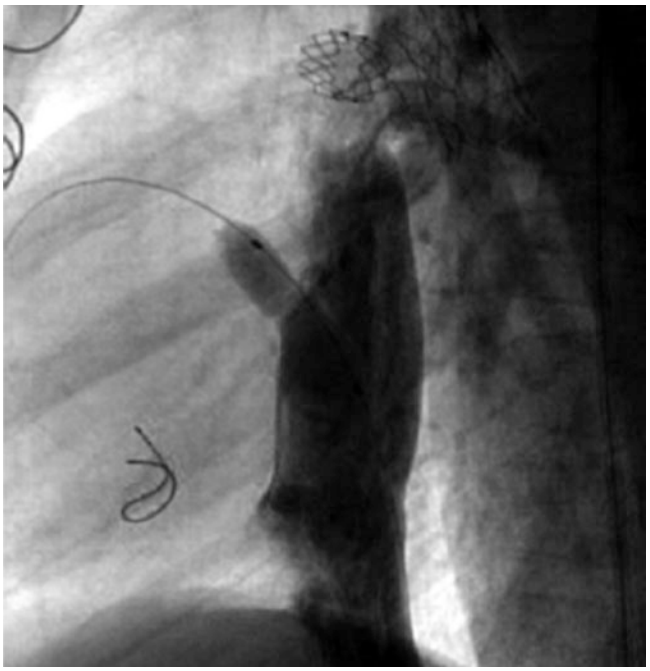


**Fig. 18.3** Low-pressure balloon occlusion of a 4.5-mm fenestration with a 6-mm Tyshak balloon; contrast injection through the venous sheath confirms total occlusion

  - Alternatively, inflate a small compliant balloon (typically a 6–8-mm Tyshak balloon (depending on fenestration size)) within the fenestration, using the femoral sheath for pressure and saturation measurement.
  - Complete occlusion can be confirmed by angiogram (through the proximal port of the balloon-tipped catheter or the femoral sheath) (Fig. 18.3).
- *Closing the fenestration can be considered:*
  - If the systemic venous pressure is <18 mmHg during test occlusion.
  - In the absence of a significant (>4 mmHg) increase in mean systemic venous pressure and reduction in mixed venous saturation of >10% during test occlusion.
  - We would strongly discourage fenestration closure in patients with systemic venous pressure of ≥20 mmHg.

## 18.4 Choice of Device

### 18.4.1 The Ideal Device

It provides complete occlusion with reliable stability and has a low profile without distorting the anatomy or obstructing flow within the Fontan conduit/baffle.

- *Device selection will depend on:*
  - Type of Fontan (extracardiac conduit versus lateral intra-atrial tunnel)
  - Location of the fenestration (intentional fenestration versus baffle leak)
  - Size of the fenestration (usually between 3.5 and 5 mm)
  - Type of fenestration (punch hole versus short PTFE shunt versus baffle leak)
  - Patient characteristics (risk stratification).

### 18.4.2 Devices Used in Our Unit Over the Past Two Decades

- *Rashkind device*, *CardioSEAL*, *Amplatzer ASD occluder* (Fig 18.8), *Amplatzer VSD occluder* and *PFO star type device* (Fig 18.7) have been used for closure of fenestrations or baffle leaks.
- *Amplatzer duct occluder type II (ADO II)* (Fig 18.5):
  - This is currently our device of choice for closing the typical punch-hole-type fenestration performed in our extracardiac Fontan conduits.
  - The device has a high conformability, and its dual articulating discs make placement in the fenestration relatively easy.
  - The fabric-free technology allows for delivery through a low-profile 4F catheter while maintaining a high rate of occlusion without being bulky and potentially obstructive.

## 18.5 Crossing and Outlining the Fenestration/Baffle Leak

### 18.5.1 Imaging to Delineate the Fenestration and/or Baffle Leak

- *Angiography*:
  - Should be done in different views
- *TEE*:
  - May give additional information in patients with baffle leaks, where the size of the atrial chamber is small or when a residual atrial septum may be problematic

**Video 1a** Patient with situs inversus thoracalis, double discordance, large VSD, PA after extracardiac TCPC (20-mm conduit). Frontal and lateral angiography demonstrate the fenestration in the left sided extracardiac conduit. Frontal view angio fenestration situs inversus LTGA (MOV 196054 kb)

**Video 1b** Patient with situs inversus thoracalis, double discordance, large VSD, PA after extracardiac TCPC (20-mm conduit). Frontal and lateral angiography demonstrate the fenestration in the left sided extracardiac conduit. Angio lateral fenestration situs inversus LTGA (MOV 188937 kb)

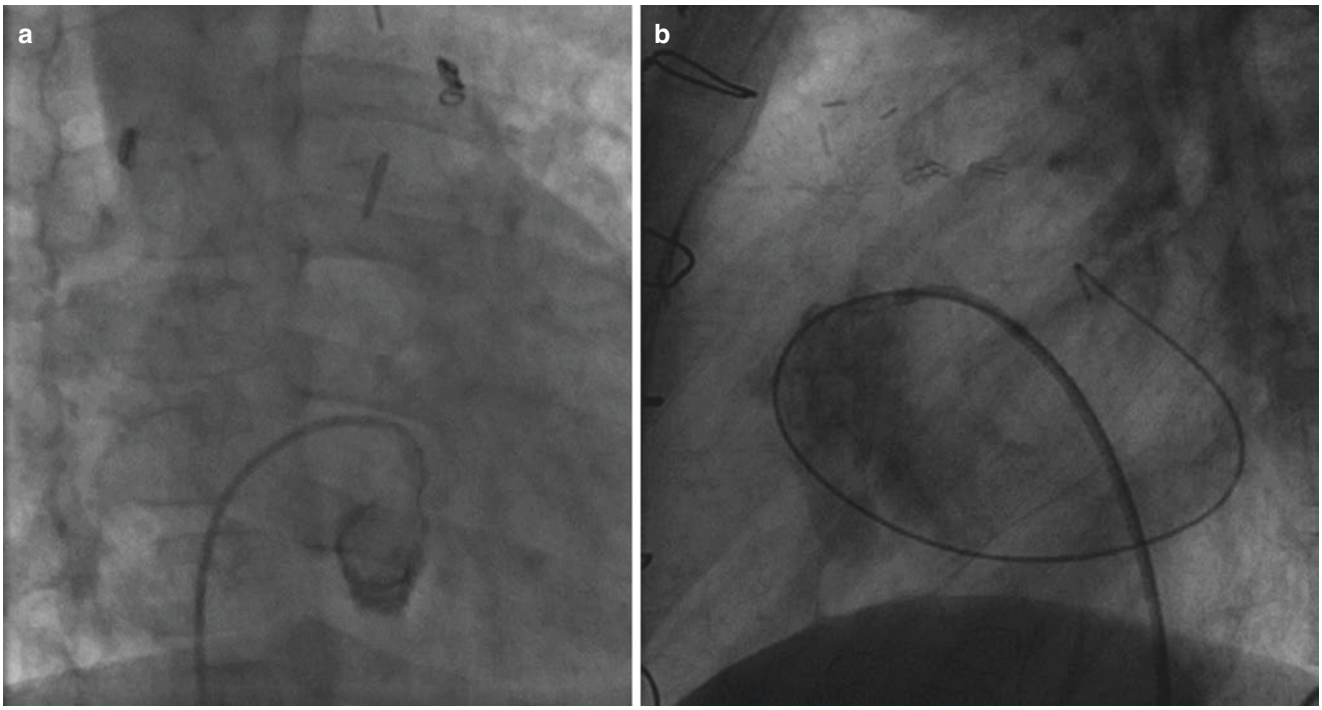



**Fig 18.4** (**a**) Fenestration has been crossed with a 4F right Judkins catheter; a small contrast injection confirms position in the systemic atrium. (**b**) Lateral view of another patient: the fenestration has been crossed with a 6F right Judkins guiding catheter over an Amplatzer 0.035" wire. A 4/4-mm ADO II device (4F delivery catheter) was subsequently implanted via the 6F guiding catheter

### 18.5.2 Crossing and Closing the Fenestration

- The fenestration can be crossed by the aid of various preshaped or custom-shaped catheters such as the right Judkins catheter (Fig 18.4) and a floppy exchange wire [i.e. 0.035" Terumo guide wire or Wholey 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.
- 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.
- 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 (Fig. 18.5a, b) (Fig 18.6a) and on TEE if necessary (Fig 18.6b). Be aware: the distal disc should be free of the AV valve and native atrial septum; the proximal disc should be in the conduit.
- Haemodynamic measurements and saturations should be repeated.

**Video 2** Lateral projection in a patient with an extracardiac Fontan conduit. A 4/4-mm Amplatzer duct occluder type II has been deployed in the fenestration. Angiography is performed via the 4F introducer sheath with the device still attached to delivery cable (MOV 240322 kb)

**Video 3** Lateral projection showing angiography via the 4F introducer sheath after release of the Amplatzer duct occluder type II device. There is still some contrast like "smoke" through the device which will disappear within minutes after release (MOV 216232 kb)

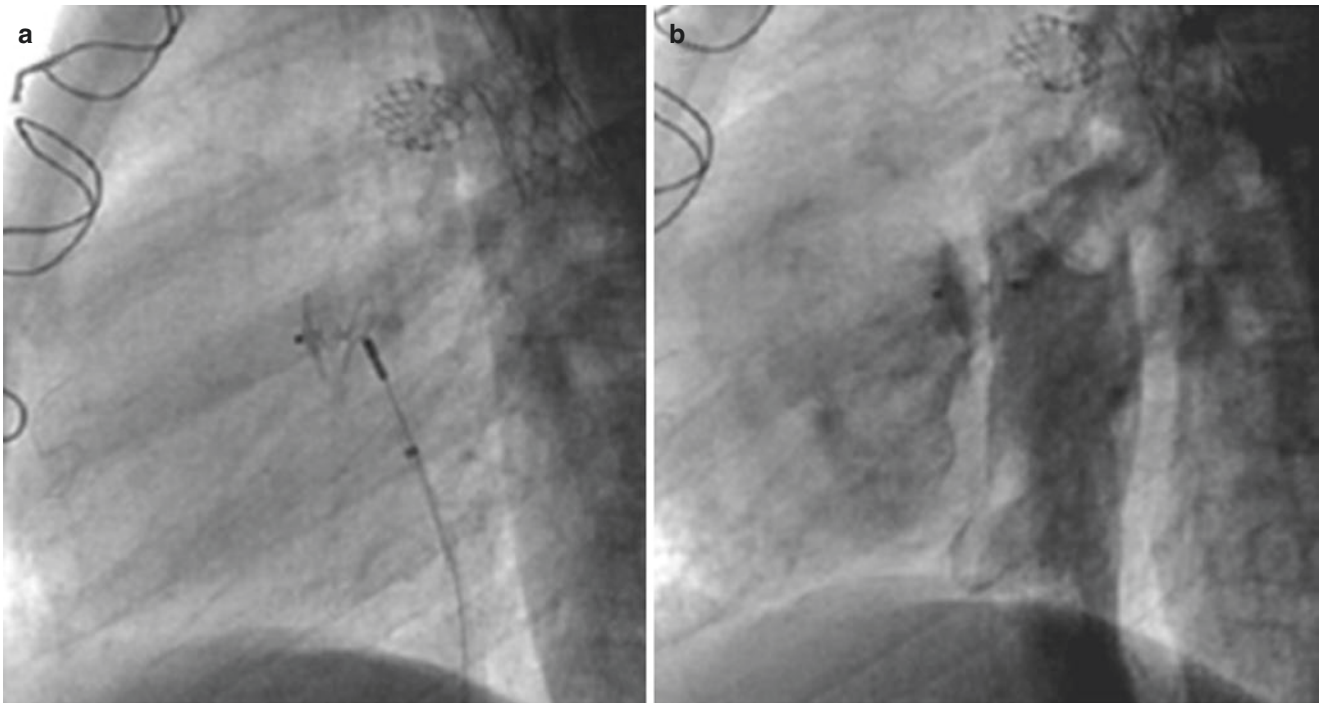



**Fig. 18.5** (**a**) Deployment of Amplatzer 4/4-mm duct occluder type II in fenestration; device is still attached to delivery cable. (**b**) Cavogram after release of device: both discs are clearly at appropriate end of the fenestration; there is still some contrast like "smoke" through the device which will disappear within minutes after release. The device has a lower profile compared to the previously used Amplatzer ASD devices

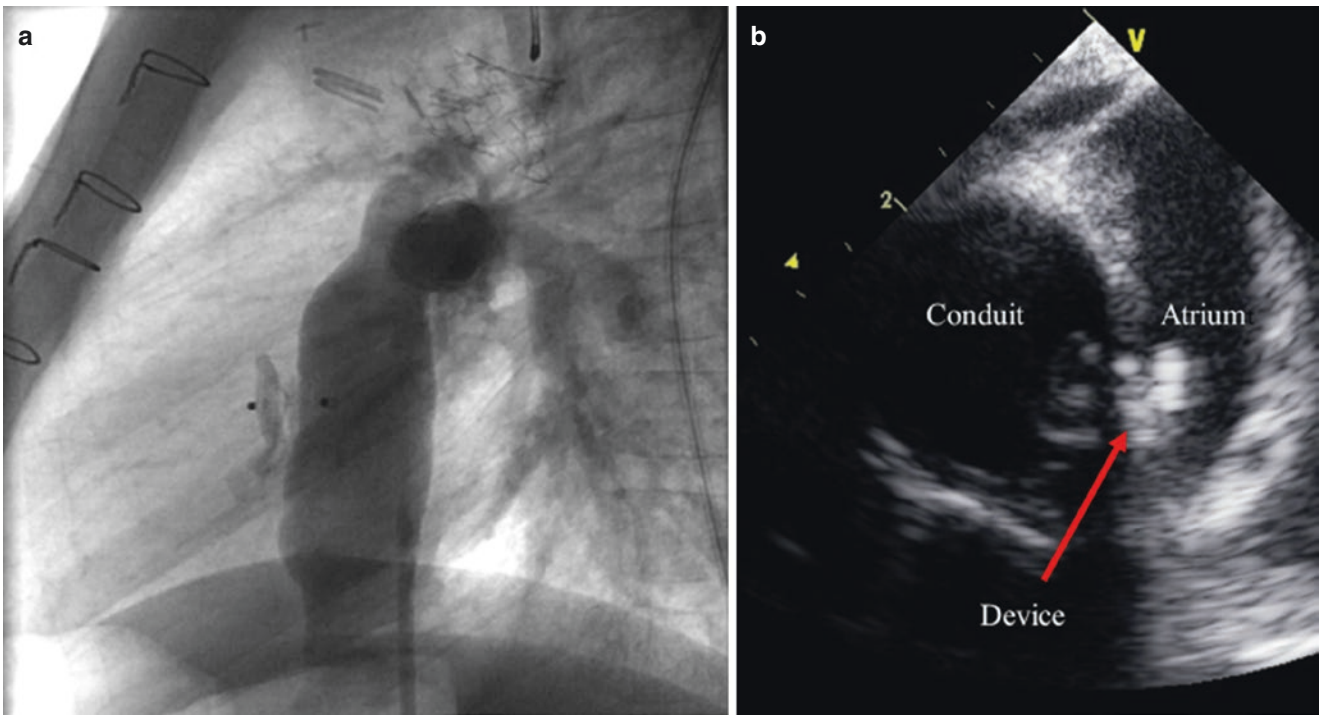



**Fig. 18.6** Closure of fenestration with ADO II device (**a**) Angiography in lateral projection after release of the device. (**b**) TEE demonstrating the position of the device in the fenestration

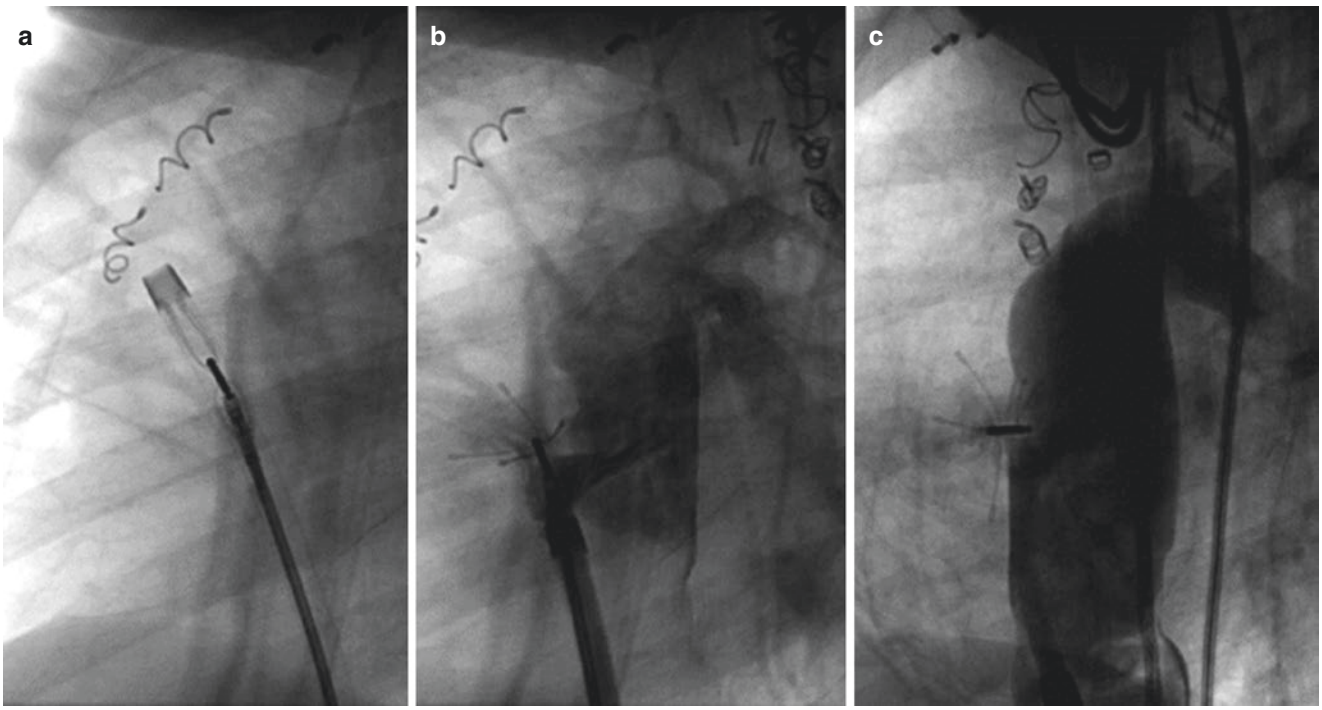



**Fig. 18.7** Lateral projection: Closure of a fenestration with a 15/20-mm PFO star (CARDIA 0115S20) (**a**) 10F-long sheath is positioned through the fenestration in the systemic atrium; device is ready to be deployed. (**b**) Angiography in lateral projection after deployment of distal (atrial) "legs" of the device. (**c**) Angiography after release of the device

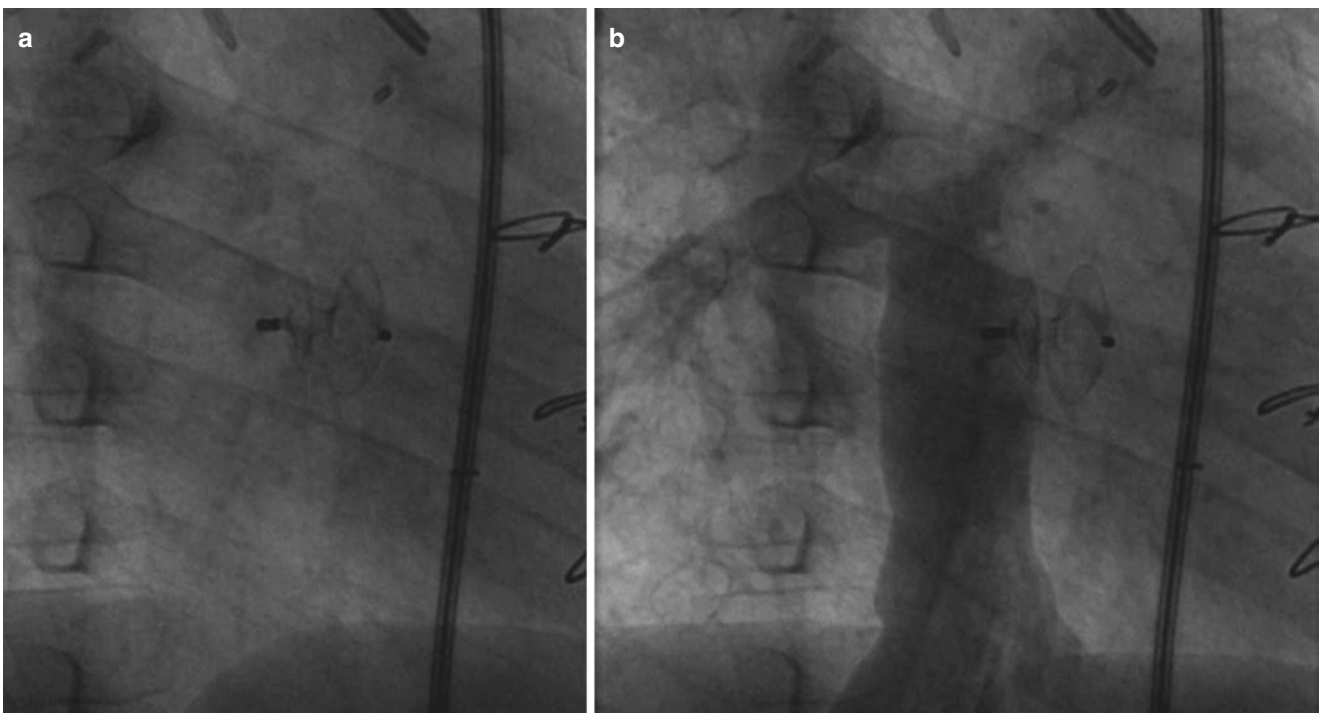



**Fig 18.8** Patient with 22-mm extracardiac Fontan conduit. (**a**) 4-mm Amplatzer ASD device has been implanted through the fenestration. (**b**) The device is clearly more "bulky" than the currently used ADO II device

## 18.6 Alternatives to Device Closure

Technical difficulties in closing fenestrations have been shown in TCPC patients with residual native atrial septum, forming an intermediate chamber on the pulmonary venous side of the fenestration and additionally carrying the risk of systemic embolism in the case of difficult manipulations with wires and sheaths. Implanting a covered stent is also ideal in the setting of a fenestration in combination with conduit stenosis (Fig 18.10).

### 18.6.1 Implantation of a Covered Stent

- *Covered Cheatham Platinum (CCP) stent (NuMED, Hopkinton)*
  - Is an attractive alternative in patients with extracardiac TCPC conduits.
  - May be an ideal solution for occlusion of residual shunting through a previously implanted device (Fig 18.9).
  - Has short procedural/fluoroscopy times and complete immediate fenestration closure.
  - Avoids protrusion of prosthetic material in the pulmonary atrium.
  - Has a lower risk for thrombosis in the systemic atrium and embolism.
  - Should preferably be implanted in patients with a weight >15 kg (due to the large sheath size of ≥12F).
  - Stent is hand crimped onto a BIB balloon catheter (NuMED, Hopkinton).
  - Balloon should be 1–2 mm larger than the angiographic conduit diameter; ideally the final diameter of the conduit in an adult should be 18–20 mm; current Gore-Tex tubes >16 mm can be dilated up to that size.
- *Zenith abdominal aortic aneurysm endograft (Cook Medical)*
  - Device occlusion of fenestrations or baffle leaks in combination with Fontan baffle stenosis may additionally narrow the pathway.
  - Balloon expandable covered stents are less desirable in this setting as there is often a significant size discrepancy between the stenotic area and the largest baffle diameter (risk of incomplete closure or an inadvertent baffle tear).
  - Madan et al. described two patients with the combination of Fontan baffle stenosis and patent fenestration successfully treated with a Zenith abdominal aortic aneurysm endograft.
  - Due to the large delivery sheath size (16Fr), this technique should be reserved for older children or adults.

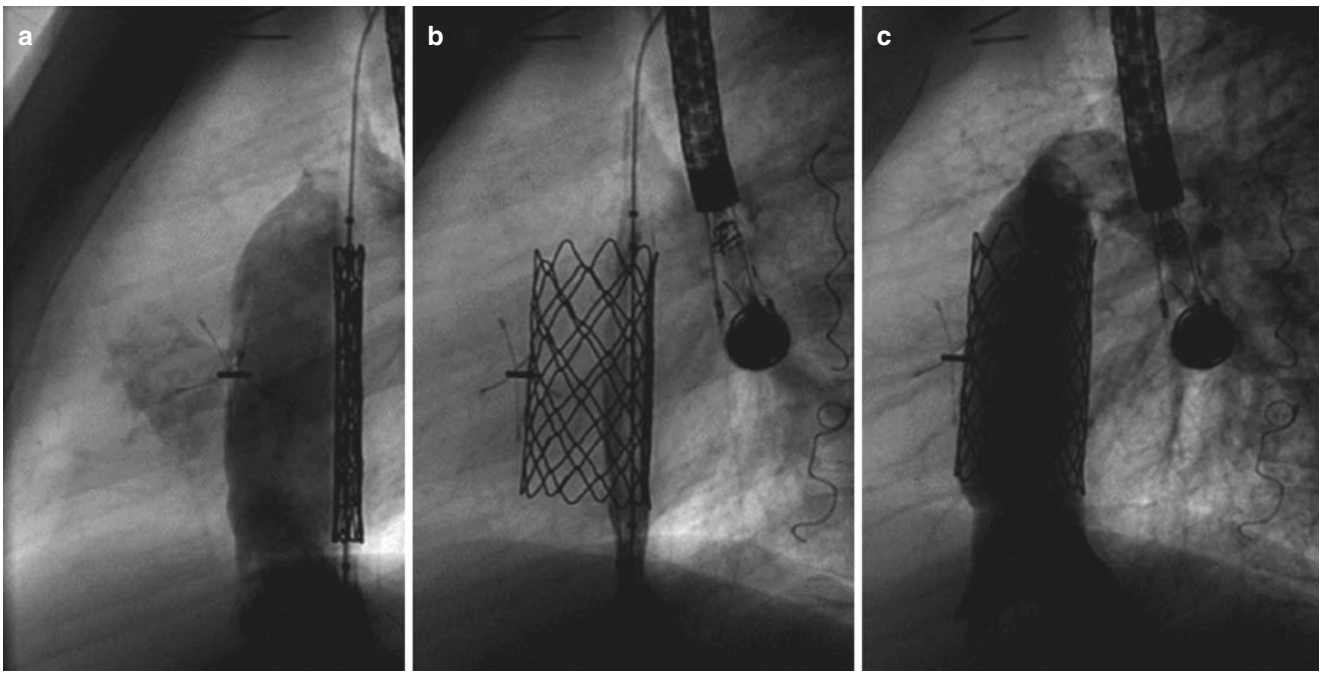



**Fig 18.9** Lateral projections in a patient with a 20-mm extracardiac Fontan conduit. (**a**) Cardia PFO occluder 0115S20 has been implanted in the fenestration, but due to residual right-to-left shunting, a 45-mm CCP stent was implanted. (**b**) An 0.035" stiff wire (Amplatzer) has been positioned in the VCS. The stent was hand crimped onto a 22-mm BIB balloon catheter and implanted via a 14F-long Mullins sheath. (**c**) Angiography after stent implantation demonstrates a non-obstructed conduit and complete occlusion of the fenestration

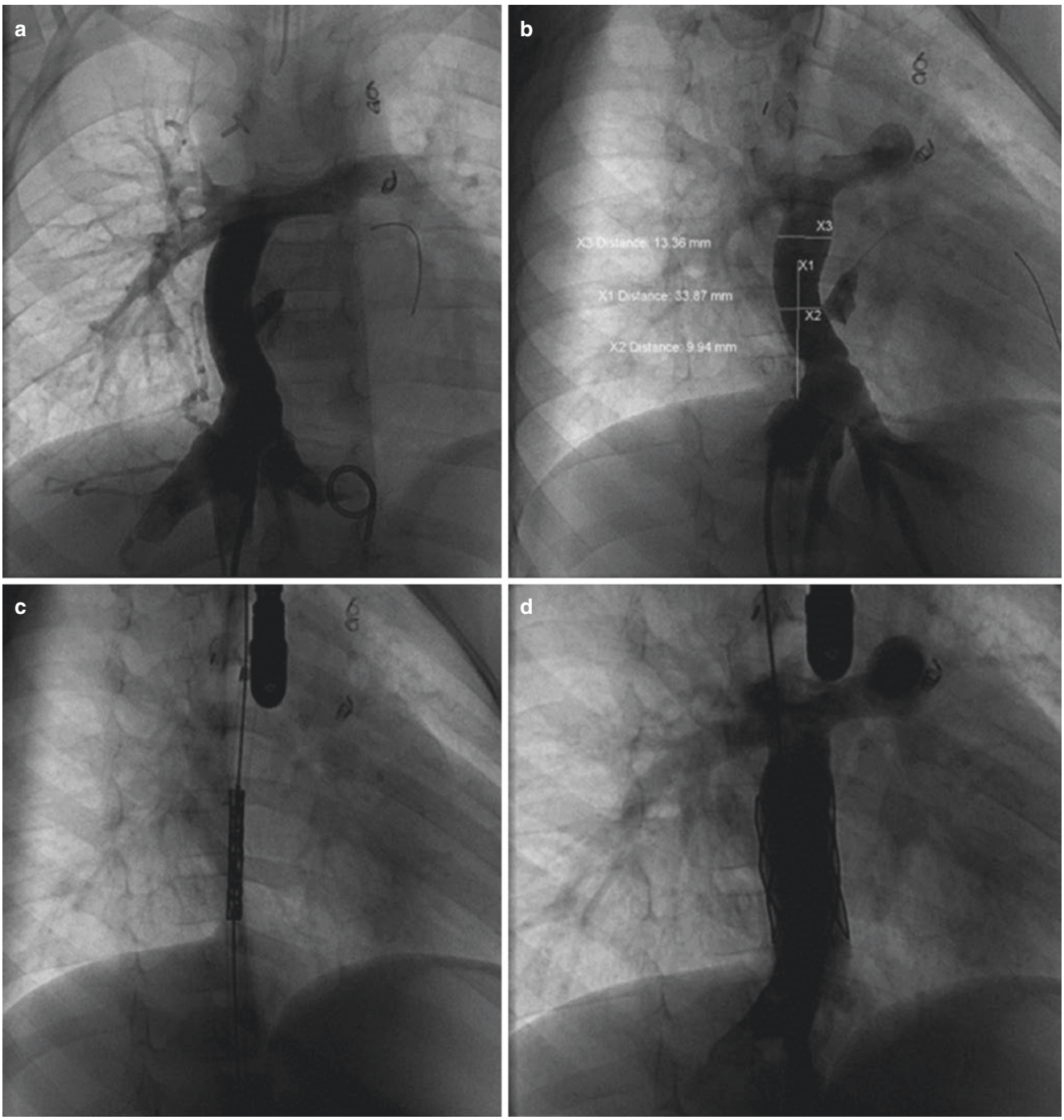



**Fig 18.10** AP projections in a patient with a 18-mm extracardiac Fontan conduit. (**a**) Angiography during test occlusion of the fenestration with a 5-mm Tyshak balloon over a 0.014 ironman wire. The conduit is relatively small compared to the VCI. (**b**) Measurements have been made to decide stent length and balloon diameter. (**c**) An 0.035" stiff wire (Amplatzer) has been positioned in the VCS. A 34-mm CCP stent was hand crimped onto a 18-mm BIB balloon catheter and implanted via a 13F-long Mullins sheath. (**d**) Angiography after stent implantation demonstrates a non-obstructed conduit, with larger diameter and complete occlusion of the fenestration

## 18.7 Closing the Stented Fenestration

- Future reclosing of stented fenestrations after patients have improved haemodynamically might pose some challenges.
- Figure 18.11 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. An Amplatzer duct occluder II was implanted within the stent after 10 months. Three years later,

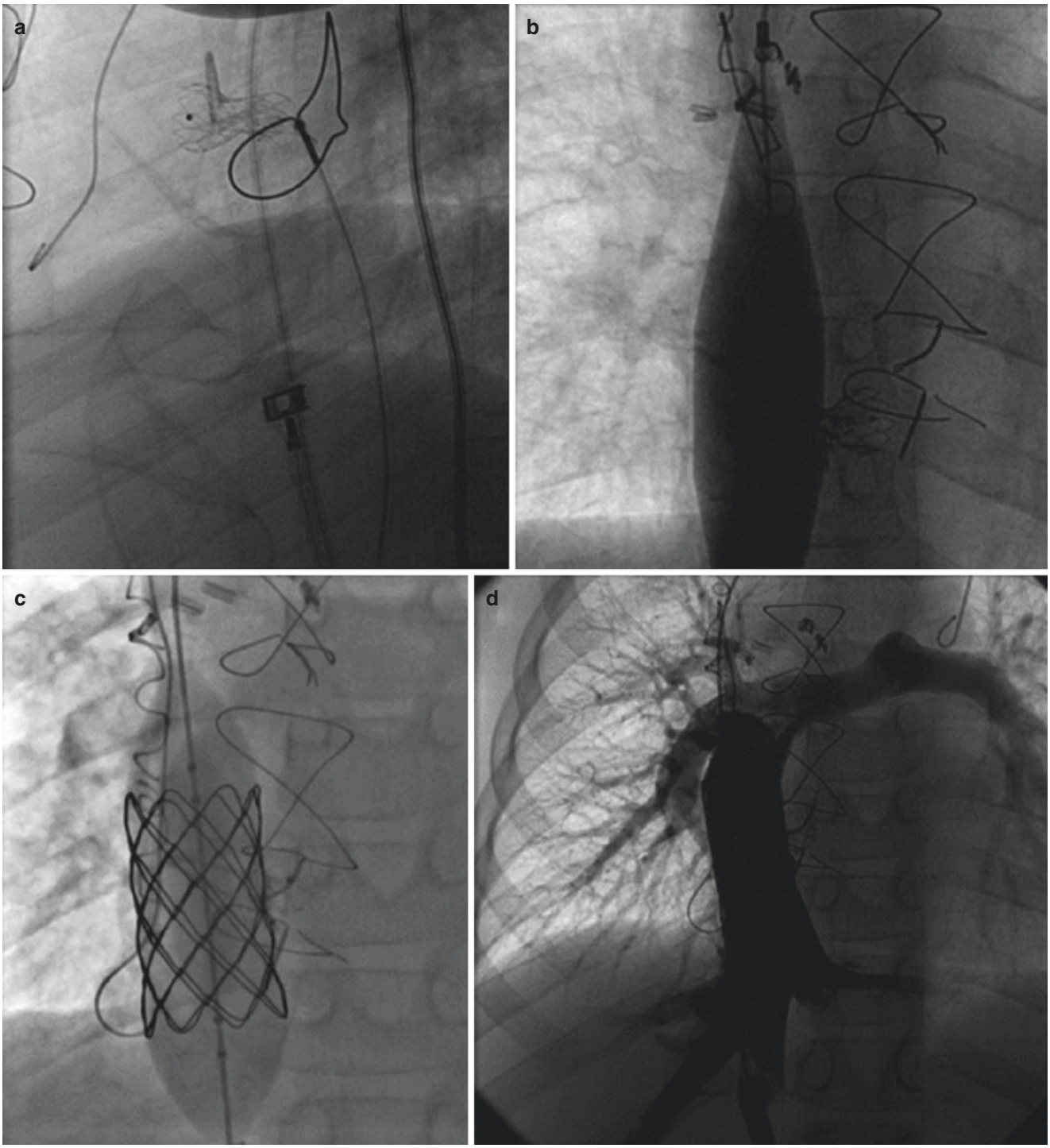



**Fig. 18.11** Stented fenestration with previous attempt of closure with an Amplatzer duct occluder type II. (**a**) The distal (conduit) part of the stent was snared from the femoral side and gradually pulled caudally against the conduit wall, to prevent sharp edges sticking into the conduit. (**b**) The stent was 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. (**c**) A 45-mm covered CP stent was implanted using a 22-mm BIB balloon. (**d**) Stent implantation resulted in complete closure of the fenestration and a non-obstructive conduit

**Video 4** Stented fenestration with previous attempt of closure with an Amplatzer duct occluder type II. A 45-mm covered CP stent is deployed using a 22-mm BIB balloon (MOV 240322 kb)

saturations persisted below 88% (residual right-to-left shunting). Clinical and haemodynamic evaluations were favourable for complete fenestration closure, but technically the procedure proved to be challenging.

- The technique used in this patient is outlined in Fig. 18.11a–d.

## 18.8 Devices for Partial Occlusion

Some Fontan patients with a large fenestration (created due to suboptimal physiology) may be severely cyanotic but not yet stable enough for complete closure of the fenestration. Partially closing such a fenestration may be an attractive option in this setting.

### 18.8.1 115S PFO Star (CARDIA™)

- *Device:*
  - Customized fenestrated atrial septal occlude.
  - Designed by removing two opposite quadrants from the right atrial disc.
  - 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).
- Results:
  - Sixteen of eighteen patients had mild to moderate shunting 1 month after implantation.
  - Six months after device implantation, residual shunting was still documented by echocardiography in 12 of these patients (saturations 90 ± 3%).

## 18.9 Follow-Up After Fenestration Closure

### 18.9.1 Evaluation

Clinical and echocardiographical evaluation should be performed at 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. TTE may fail to detect some thrombi, and although routine TEE is more invasive, it should probably be considered in certain high-risk patients.

#### 18.9.1.1 Anticoagulation Regimen

The optimal anticoagulation regimen after Fontan completion is unknown. Treatment protocol before and after fenestration closure differs depending on institutional protocol and risk stratification in individual patients.

*Treatment protocol in our institution:*

- Acetylsalicylic acid 1–2 mg/kg/day and clopidogrel 0.2 mg/kg/day.
- Clopidogrel is usually stopped after 6 months, 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 2.0–2.5.

# Ventricular Septal Defects

# 19

Massimo Chessa

Muscular ventricular septal defects (MVSD) are suitable for percutaneous closure in most cases (Fig. 19.1). Perimembranous ventricular septal defects (PMVSD) closure may be performed percutaneously, although the procedure is more challenging in this anatomic form given the proximity of aortic valve, tricuspid valve, and conduction tissue (Fig. 19.1). Surgical repair is indicated for double committed or supracristal defects and for perimembranous defects associated with aortic regurgitation.

## 19.1 Indications for VSD Closure

1. Symptoms of heart failure attributed to left-right shunt through the VSD
2. Asymptomatic patients with left ventricle volume overload evidence on echocardiography
3. Defects with neither symptoms of heart failure nor overload but with history of infective endocarditis

## 19.2 Before the Procedure

1. Eradication of any active infection before catheterization
2. Elaboration of a strategy for the eventually associated anomalies (pulmonary valve or branch stenosis, atrial septal defect)
3. Informed consent for all the planned procedures

## 19.3 The Procedure (Figs. 19.2–19.10)

1. General anesthesia and orotracheal intubation.
2. Monitoring with arterial line, two peripheral venous lines or a central venous line, vesical catheter.
3. A transesophageal echocardiography (TEE) guidance is mandatory. 3D TEE is preferred over 2D TEE.
4. Administration of heparin (100 IU/kg IV). Hourly check of the activated clotting time.
5. Antibiotic prophylaxis.
6. Caution for strict asepsis.
7. Bilateral femoral access preparation.
8. A femoral vein access is used for PMVSD closure, and an internal jugular vein (if possible) should be used for MVSD closure.
9. Arteriovenous circuit must be created.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_19) contains supplementary material, which is available to authorized users.

M. Chessa (✉)
Children and Adults Congenital Heart Disease, Policlinico San Donato, San Donato Milanese, Italy
e-mail: MASSIMO.CHESSA@GRUPPOSANDONATO.IT


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_19

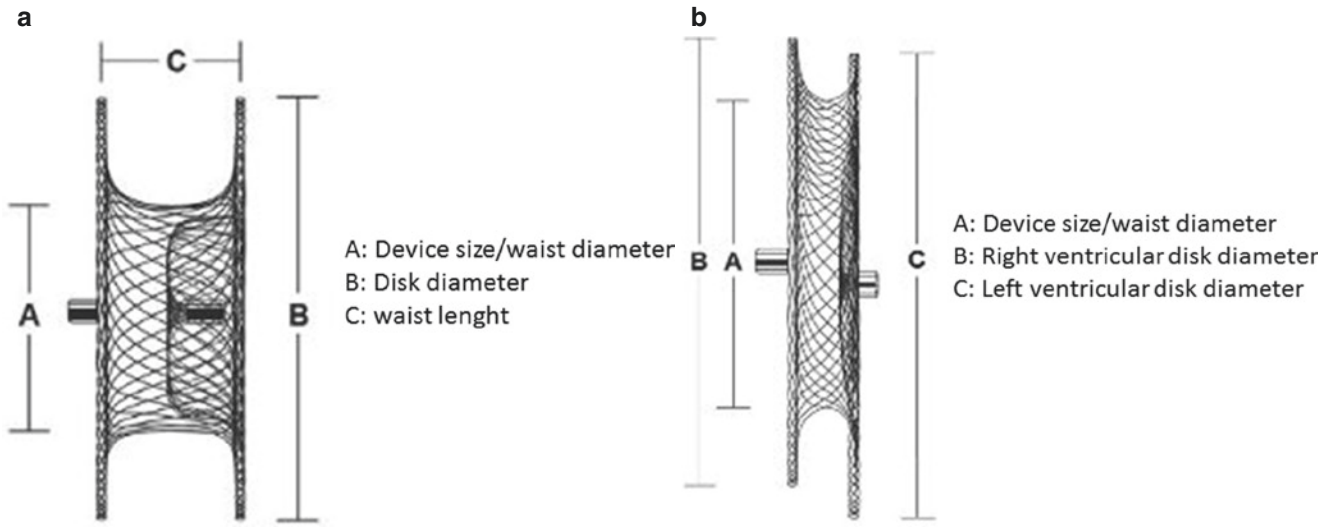



**Fig. 19.1** The Amplatzer VSD occluders are self-expandable devices made of nitinol wires and are composed of three parts: a left ventricular disk, a central waist, and a right ventricular disk. The device is mounted on a delivery cable and inserted into a delivery sheath. The MVSD occluder (**a**) is symmetric; the two disks are 8 mm larger than the waist. The size of the device is determined by the diameter of the waist that ranges from 4 to 18 mm. The PMVSD occluder (**b**) is asymmetric. The aortic rim of the left ventricular disk exceeds the dimensions of the connecting waist by only 0.5 mm reducing the risk of aortic valve compromise during implantation. The apical part of the left disk is 5.5 mm larger than the waist and is indicated by a platinum marker. The right disk is 2 mm larger than the waist

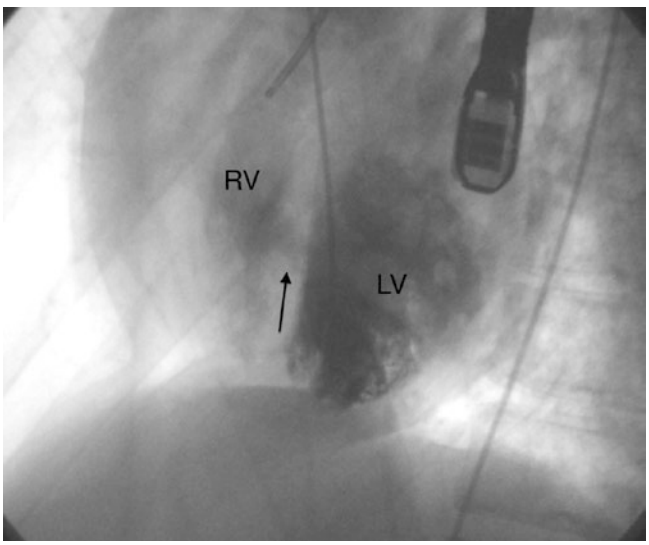



**Fig. 19.2** Left ventricular angiography. Left ventriculography in the hepatoclavicular projection (35° left anterior oblique/35° cranial) showing a mid-ventricular MVSD (arrow). 60° left anterior oblique/20° cranial view left ventriculography is preferred to visualize anterior defects. Left ventricular angiographies should determine anatomic characteristics of the VSD: size, number, and location. *RV* right ventricle, *LV* left ventricle

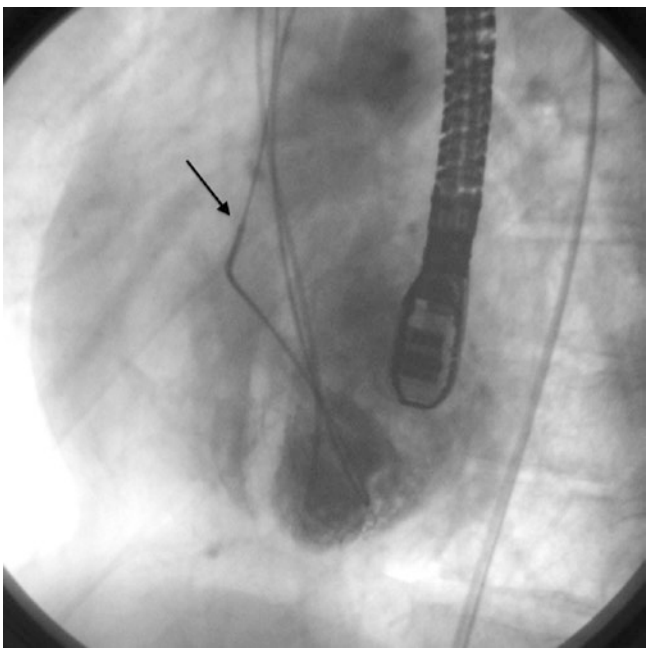


**Fig. 19.3** MVSD crossing and arteriovenous circuit creation. A 5F Judkins right catheter is positioned across the aortic valve into the left ventricle and is used to cross the VSD from left to right side. Crossing is assisted with a 0.035 in. hydrophilic wire. The wire is advanced to the SVC (or IVC or pulmonary artery) and then snared with a Goose Neck snare (20–25 mm in adults; 10–15 mm in children). The wire is exteriorized from the right internal jugular vein creating an arteriovenous loop (arrow)

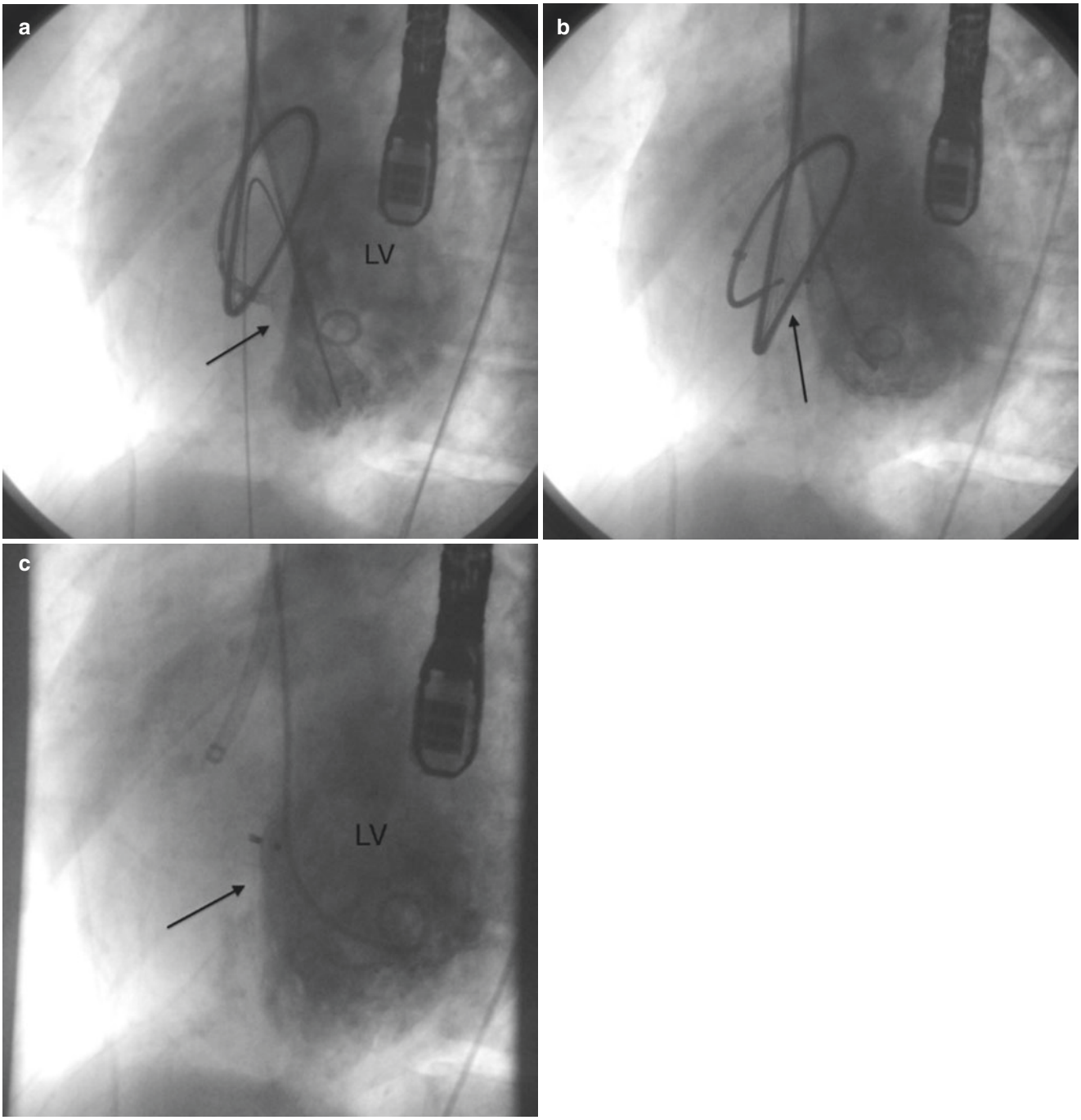



**Fig. 19.4** The left disk is deployed into the left ventricle (**a**). TEE evaluation makes sure that the disk is not impinged in the mitral valve apparatus; then the entire system is withdrawn toward the septum. The central waist and the right disk are deployed (**b**). The device is released by counterclockwise rotation of the cable (**c**). According to angiography and echocardiography, a muscular VSD occluder 1–2 mm larger the maximum size of the defect is chosen (black arrow)

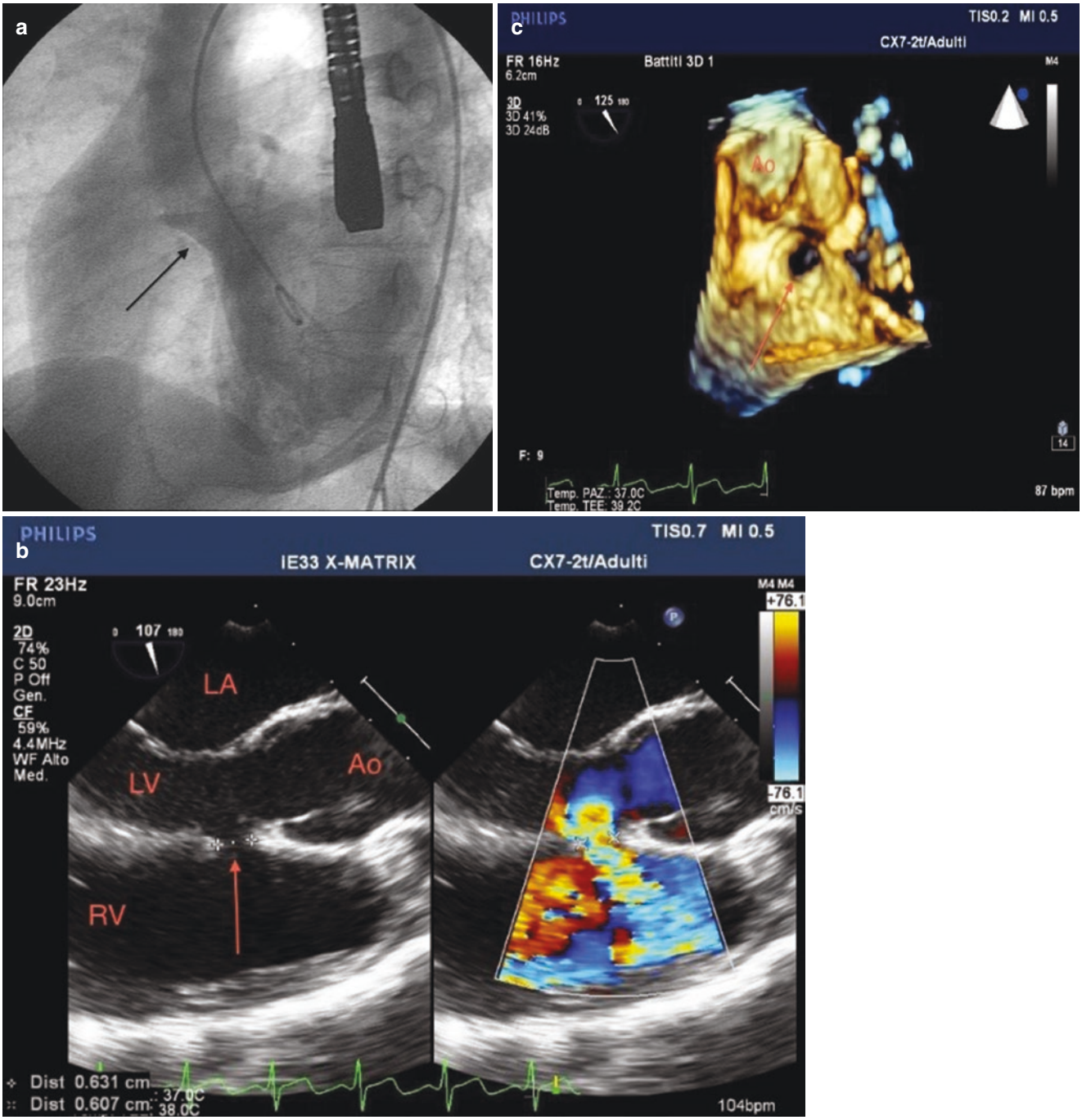



**Fig. 19.5** PMVSD evaluation. Angiographic evaluation of PMVSD is performed in 60° LAO with 20° cranial angulation to define localization and size of the defect (**a**). Aortic angiography is also performed to check for aortic regurgitation. 2D TEE guidance allows sizing of the defect (red arrow) and evaluation of the aortic, tricuspid, and mitral valve before device implantation (**b**). 3D TEE can be helpful for a better anatomic evaluation of the defect (**c**). *LA* left atrium, *Ao* aorta, *LV* left ventricle, *RV* right ventricle

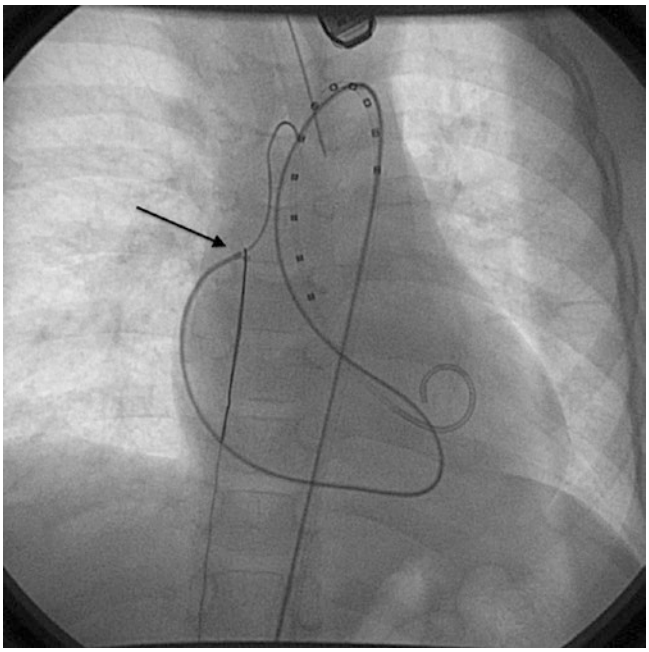


**Fig. 19.6** A Judkins right 5fr catheter and a 0.035 hydrophilic wire are used to cross the PMVD from left to right side. The catheter is advanced over the wire until it reaches the SVC. The wire is then replaced by the soft exchange dedicated noodle wire. The noodle wire is snared (arrow) and exteriorized from the femoral vein creating an arteriovenous circuit

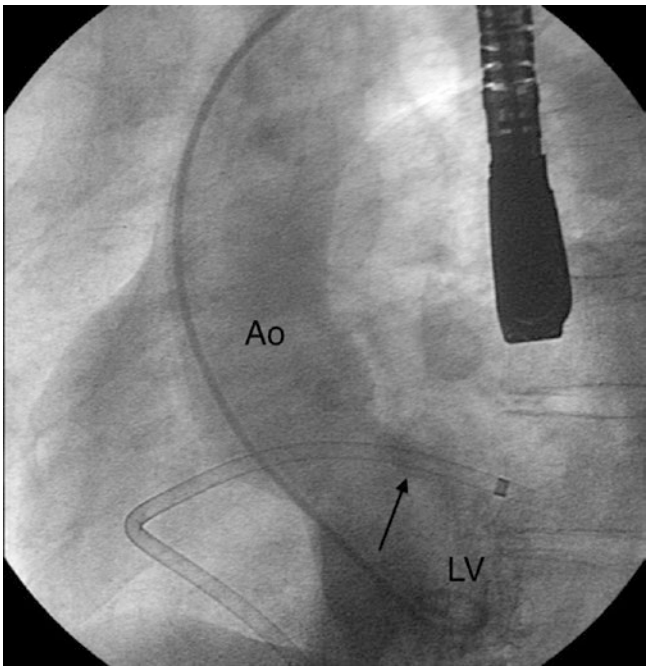
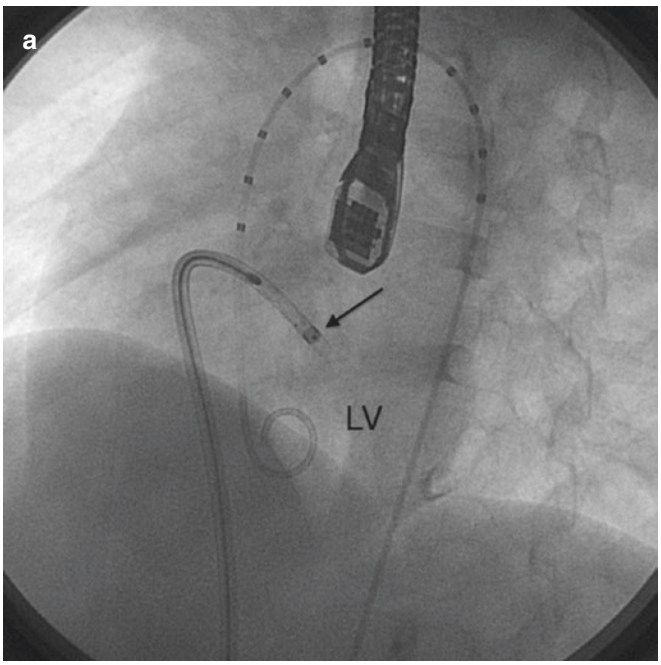



**Fig. 19.7** The dilator is then withdrawn, and the sheath is slowly pulled back in the left ventricle. The arterial catheter is advanced making a loop of the wire in the left ventricular apex. The sheath is finally advanced over the wire until it reaches the right position into the left ventricle (black arrow)



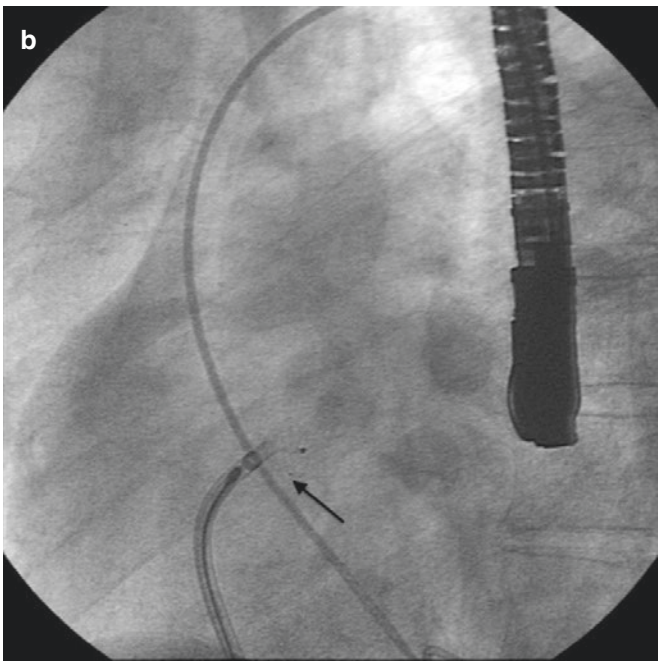




**Fig. 19.8** Device delivery for PMVSD closure. The chosen device attached to the delivery cable is advanced until the tip of the sheath (**a**). The left disk is deployed (**b**) under echographic monitoring to confirm normal function of aortic, mitral, and tricuspid valve. The platinum marker of the left disk should be oriented downward (black arrow). The entire system is then withdrawn back to the septum and the right disk is deployed (**c**). Final angiography is performed after device releasing (**d**)

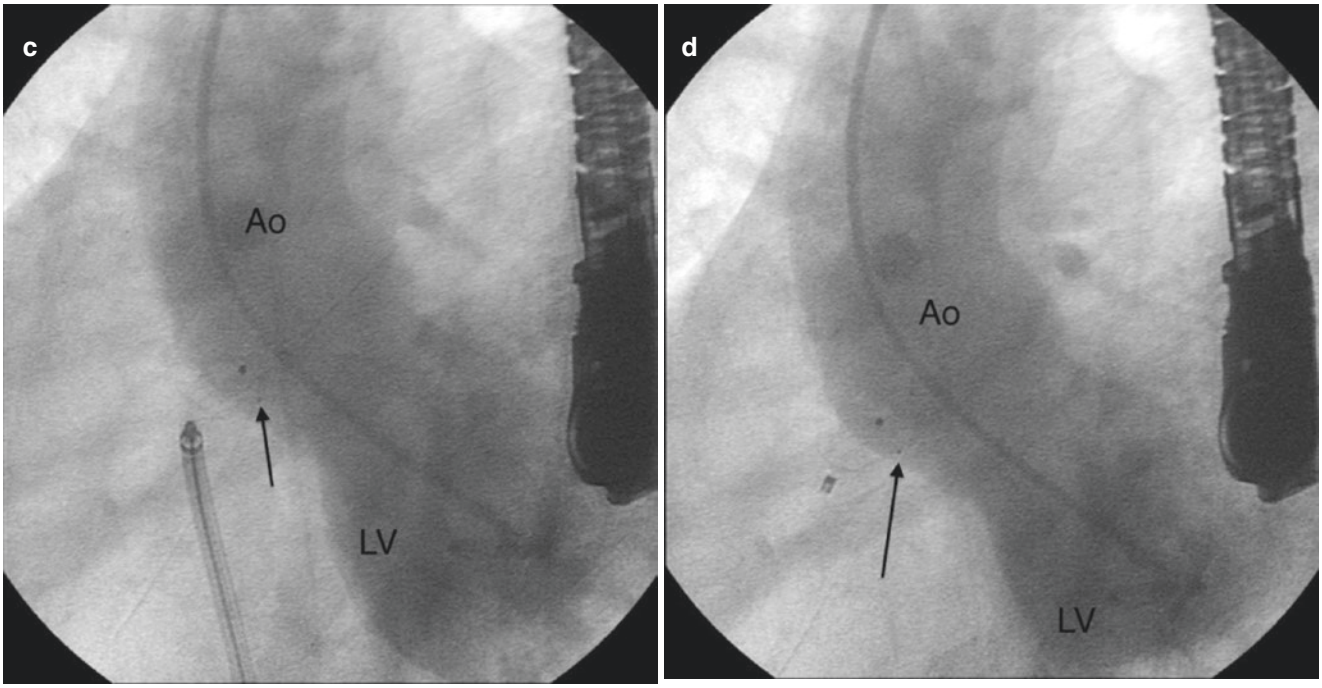



**Fig. 19.8** (continued)

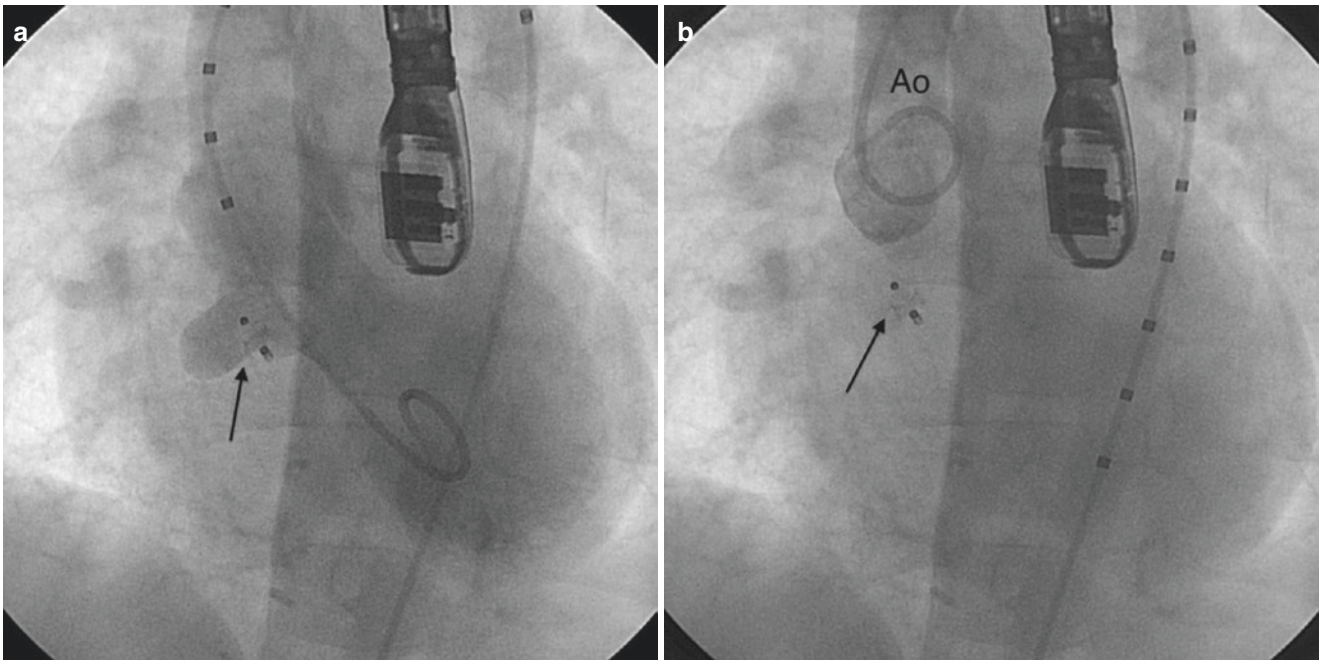



**Fig. 19.9** PMVSD with septal aneurysm. Septal aneurysm is the most common anatomic variation of PMVSD. When the aneurysm is relatively small, a closure of the anatomic defect is recommended (**a**). In this case, the device (black arrow) could cover both the hole and the aneurysm (**b**)

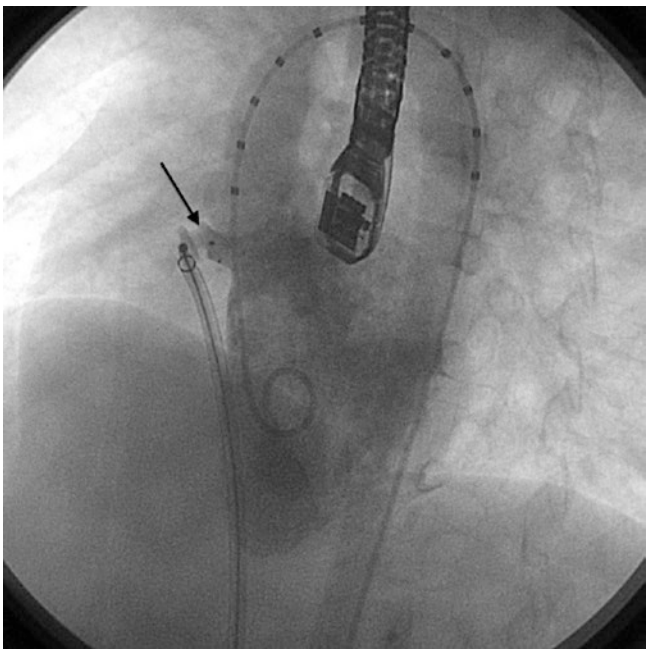


**Fig. 19.10** Large septal aneurysm. In case of large aneurysm, the device may be implanted within the aneurysm itself (black arrow) and might close the anatomical hole. The aim of this technique is to avoid implantation of an oversized device

**Video 1** A complete procedure of a MVSD closure from the RIJV (MOV 68222 kb)

**Video 2** A complete procedure of a MVSD closure from the RFV because of occlusion of the RIJV (MOV 16306 kb)

**Video 3** A complete procedure of a pmVSD closure from the RFV (MOV 44050 kb)

**Video 4** A complete procedure of a pmVSD + aneurism closure from the RFV opening the device from the aorta (MOV 37888 kb)

**Video 5** VSD closure in a retrograde way from the aortic side (MOV 38988 kb)

# Patent Ductus Arteriosus Closure

20

Ahmed Mohammed Al Kamali

A patent ductus arteriosus (PDA) is a cone-shaped tube connecting the descending thoracic aorta and the origin of the left pulmonary artery. Clinically, PDA may be present in patients with congestive heart failure, but it can also be present in asymptomatic subjects. It is necessary to close a PDA if there are symptoms of heart failure or if there is a continual heart murmur without symptoms. Transcatheter closure of a PDA can be done with either a mushroom-shaped device or a coil. For small babies weighing below 5 kg, surgical closure is advisable.

The selection of either a device or a coil depends on the shape and morphology, and the narrowest diameter, of the PDA. For diameters of less than 3 mm with a long duct, a coil can be used. For cone-shaped or elongated ducts with the narrowest diameter greater than 3 mm, the PDA can be closed with a ductal occluder device. Some window-shaped ducts can be closed with larger devices such as an atrial septal defect occluder device.

For the placement of a device or coil, puncture should be done from both the femoral vein and the femoral artery to obtain clear angiographic anatomy of the duct. The PDA device should be delivered from the venous side, and before release, aortic angiography can delineate the correct placement of the device. A Flipper coil can be delivered from the venous or arterial side; the coil has a higher risk of dislodgment and residual leak than a device.

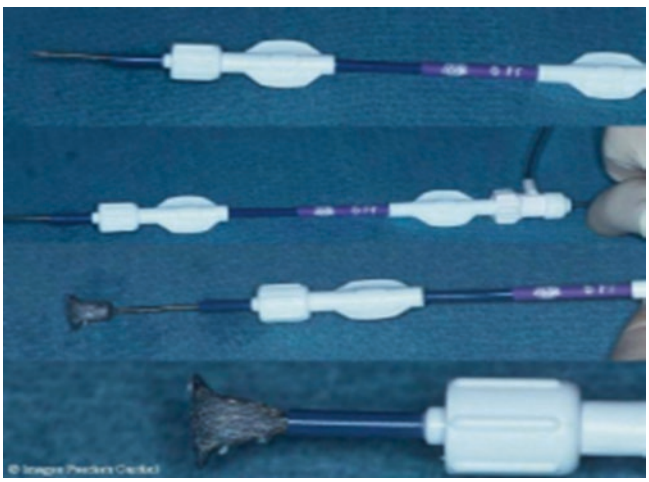


**Fig. 20.1** Assembling the delivery system of a ductal occluder. The delivery cable is introduced through the loader and, via a pin vise, to the ductal occluder device. Then the loader is screwed into a long Mullins-type sheath that is delivered over a stiff guidewire, crossing the PDA from the venous to the arterial side

The original version of this chapter was revised. A correction to this chapter can be found at https://doi.org/10.1007/978-3-319-72443-0_46

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_20) contains supplementary material, which is available to authorized users.

A. M. Al Kamali (✉)
AL Qassimi Children Hospital, Sharjah, UAE



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_20

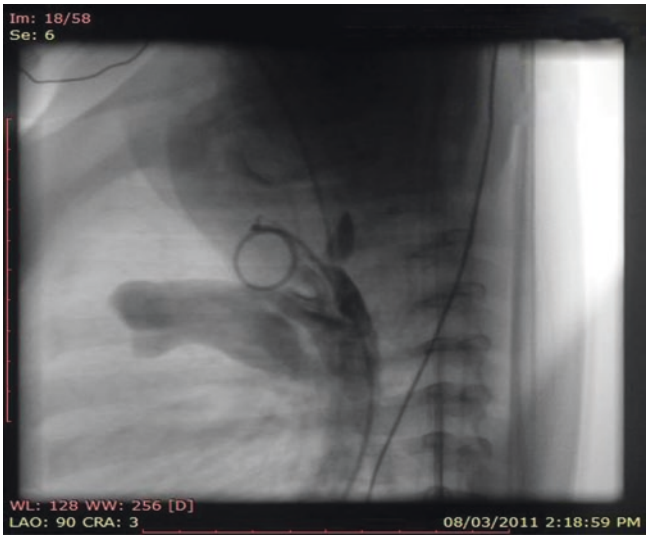



**Fig. 20.2** Lateral aortic angiogram showing type A elongated patent ductus arteriosus (PDA) with left-to-right shunt filling the main pulmonary artery and descending aorta with the narrowest diameter around 3 mm. This type of PDA can be closed with a ductal occluder device

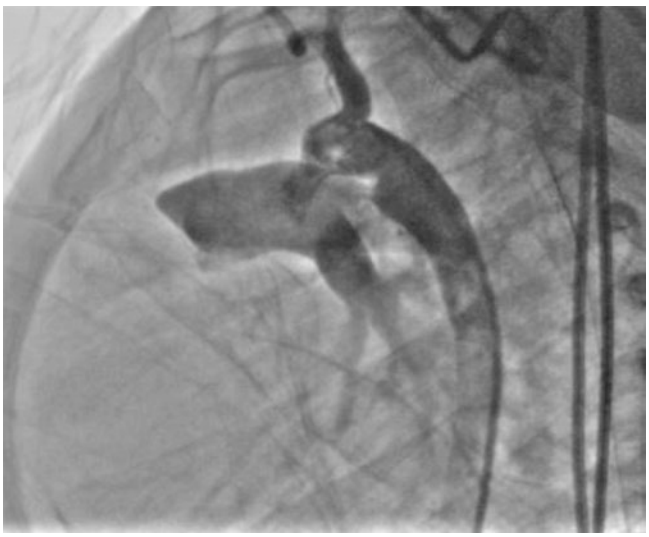


**Fig. 20.3** Lateral aortic angiogram showing type A PDA with left-to-right shunt filling the main pulmonary artery and descending aorta and left subclavian artery with the narrowest diameter around 3 mm

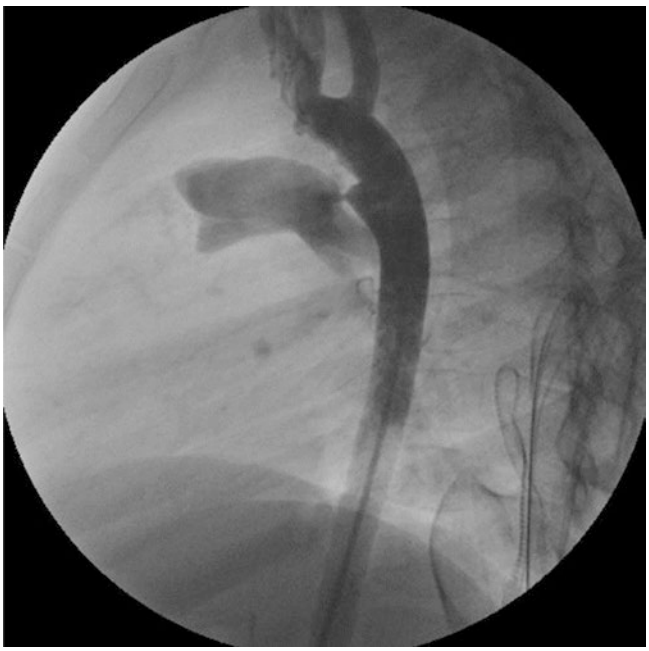


**Fig. 20.4** Lateral aortogram showing small type A PDA with the narrowest diameter of 2 mm with a left-to- right shunt. This type of PDA can be closed with a Flipper coil or small ductal occluder device

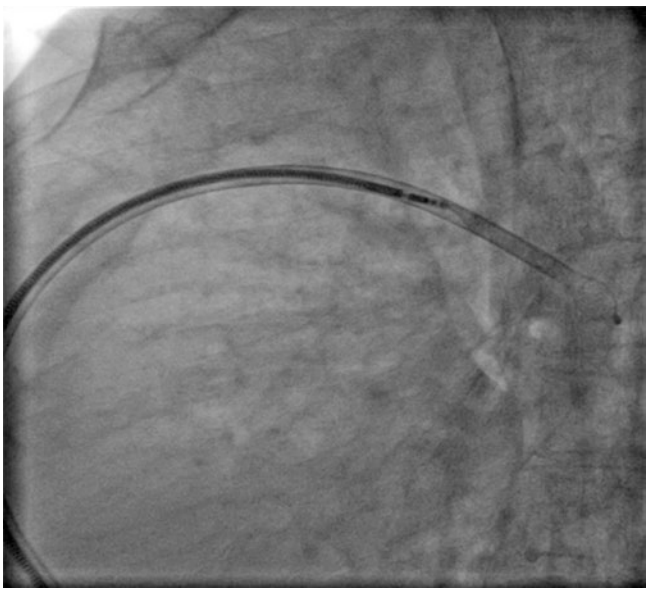


**Fig. 20.5** A 7-French PDA device delivery catheter introduced from the venous right side up to the PDA to the descending aorta, with the PDA occluder device screwed into the delivery cable. The tip of the device's aortic disc protrudes from the delivery catheter

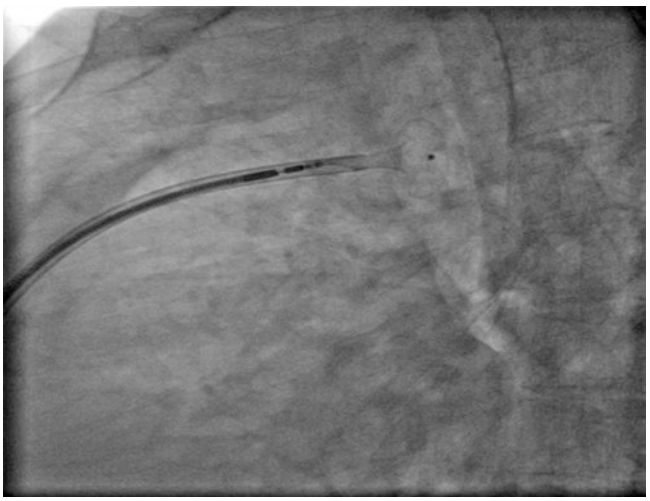


**Fig. 20.6** PDA occluder device that has been been introduced into a 7-French delivery system with the entire aortic disc delivered into the aorta

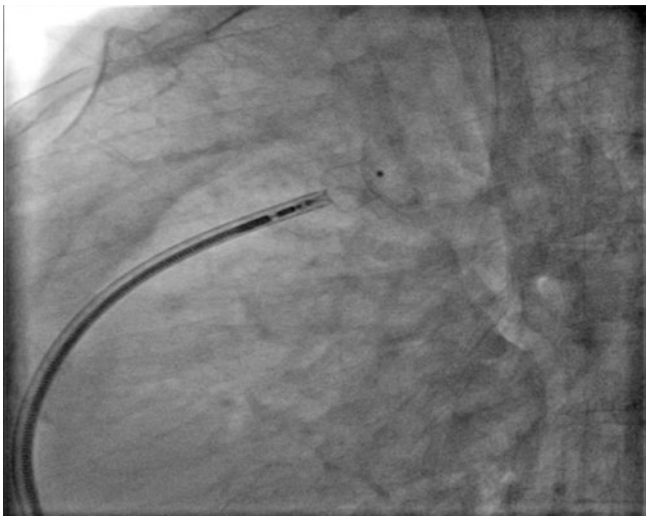


**Fig. 20.7** Completely delivered PDA device still attached to the delivery cable

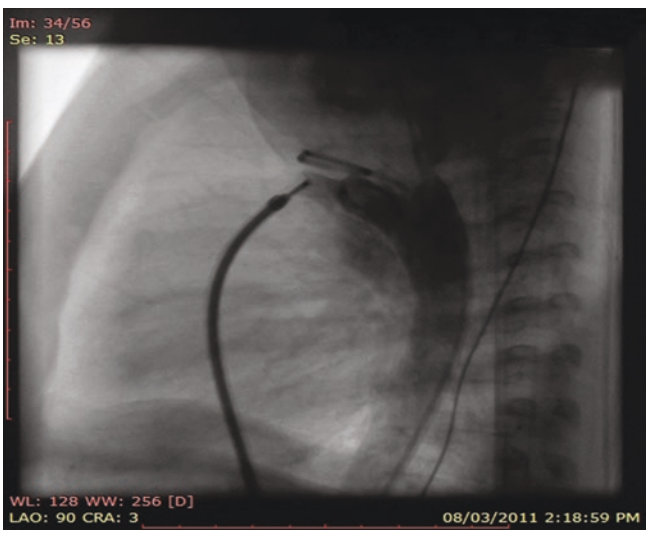



**Fig. 20.8** Lateral aortic angiogram showing the ductal occluder in a good position, with no residual PDA leak. The delivery cable is still attached to the device

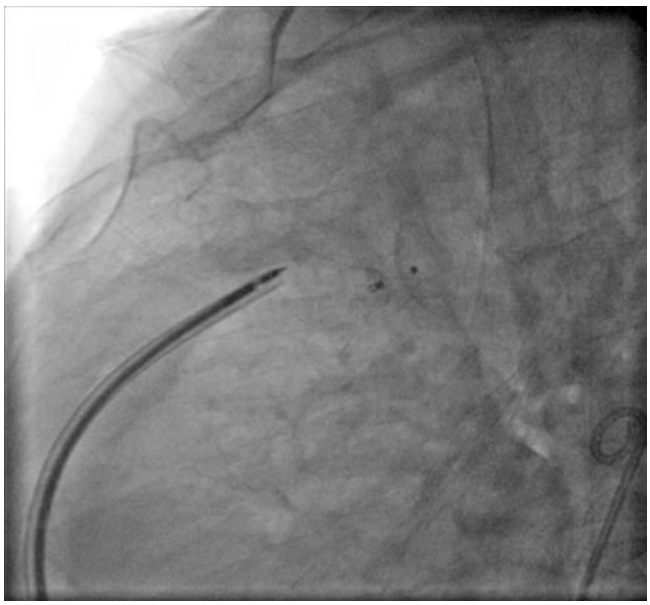


**Fig. 20.9** Ductal occluder released from the delivery system. Good position of the device; to be confirmed by lateral aortic angiogram

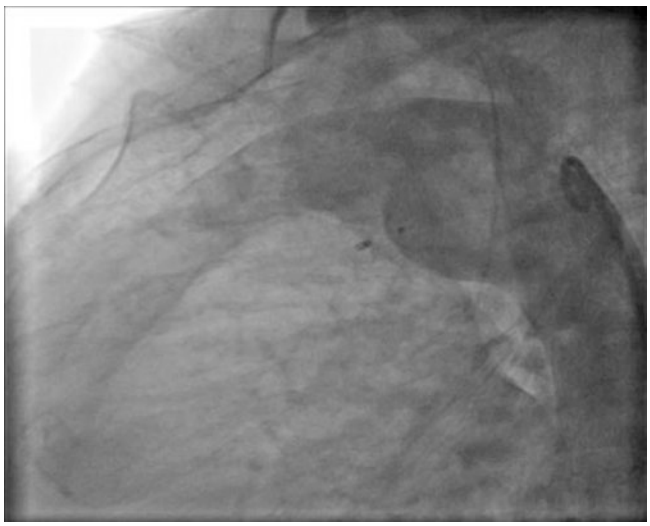


**Fig. 20.10** Lateral aortic angiogram showing good position of the ductal occluder, with no residual PDA leak and good flow in the patent aortic arch

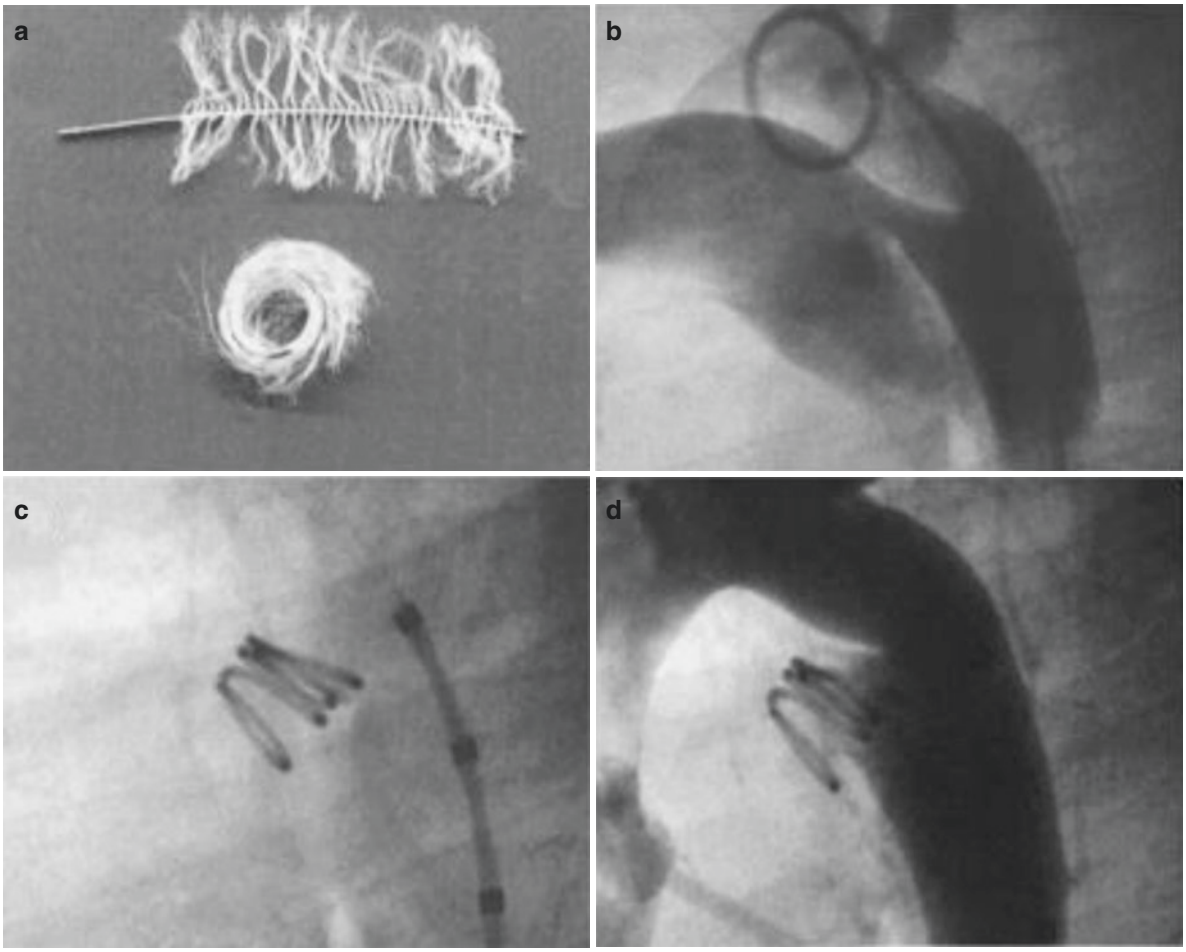



**Fig. 20.11** Flipper coil used for a small PDA with diameter less than 3 mm. This appliance has a detachable coil that makes it easier for the embolization coils to be controlled and delivered. The detachable coil is Covered with synthetic fibers that make it work faster and close the PPA immediatly. The coil can be deliver with a 4.0-Fr or 5.0-Fr catheter with a 0.038-in end hole and can work with the Flipper delivery system (**a**). Example of coil occlusion of a PDA (**b**, **c**, **d**)

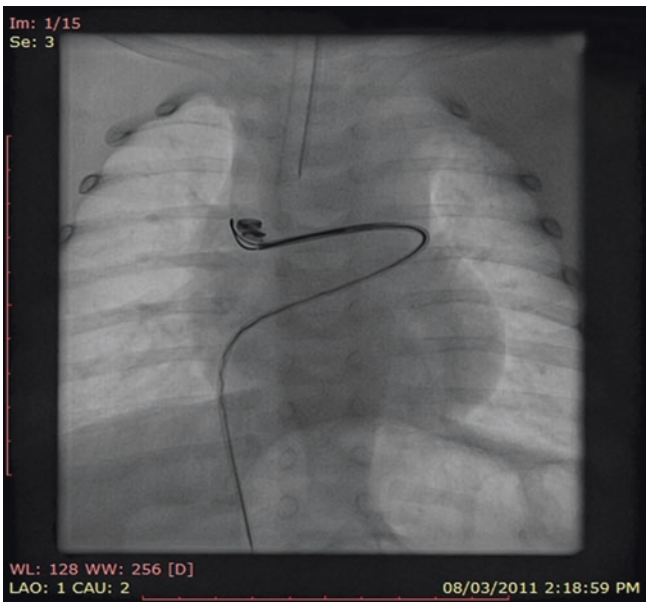



**Fig. 20.12** Flipper coil emboli in the right pulmonary artery have been retrieved with a Gooseneck snare. Ideally, such emboli should be retrieved through the delivery sheath

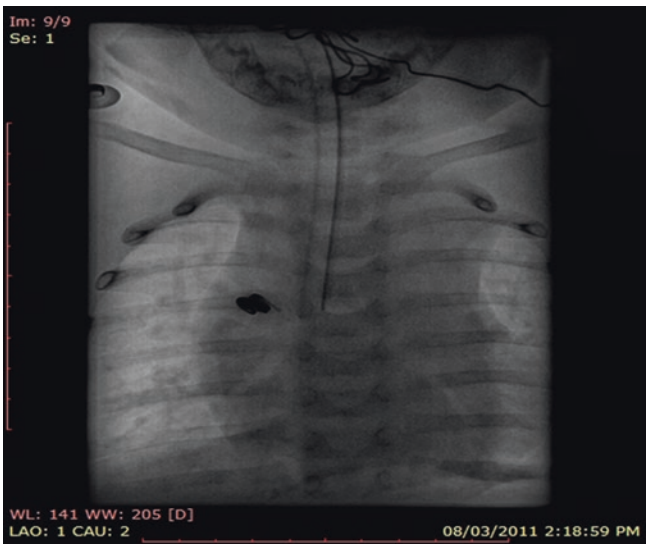



**Fig. 20.13** Flipper coil emboli in the right pulmonary artery should be retrieved in the same setting. The emboli are usually easy to retrieve from this position with any type of snare

**Video 1** Aortography in lateral view showing a large PDA (WMV 1653 kb)

**Video 2** Aortic disc is opened on the aortic side. A check aortography is performed (WMV 1614 kb)

**Video 3** Device is implanted across the PDA. A Check angiography is performed (WMV 1629 kb)

**Video 4** Device release (WMV 2309 kb)

**Video 5** Aortography in lateral video showing device position and PDA closure (WMV 2075 kb)

**Video 6** Coil implantation in lateral view (WMV 4614 kb)

**Video 7** Device release (WMV 2567 kb)

**Video 8** Aortography in lateral video showing device position and PDA closure (WMV 2317 kb)

# Catheter Closure of Coronary Artery Fistula

# 21

Kothandam Sivakumar, Ajit Mullasari, and Bharat Dalvi

## 21.1 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 (LCx) arteries in that order. Rarely, fistulae may arise from more than one coronary artery. Over 90% of the CAFs drain in the right heart chambers.

## 21.2 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 has been uncommonly reported. Very rarely, spontaneous thrombosis of a slow-flowing fistula may result in its natural closure.

## 21.3 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. 21.1).

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_21) contains supplementary material, which is available to authorized users.

K. Sivakumar · A. Mullasari
Department of Cardiology, Madras Medical Mission, Chennai, India

B. Dalvi (✉)
Glenmark Cardiac Centre, Mumbai, India



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_21

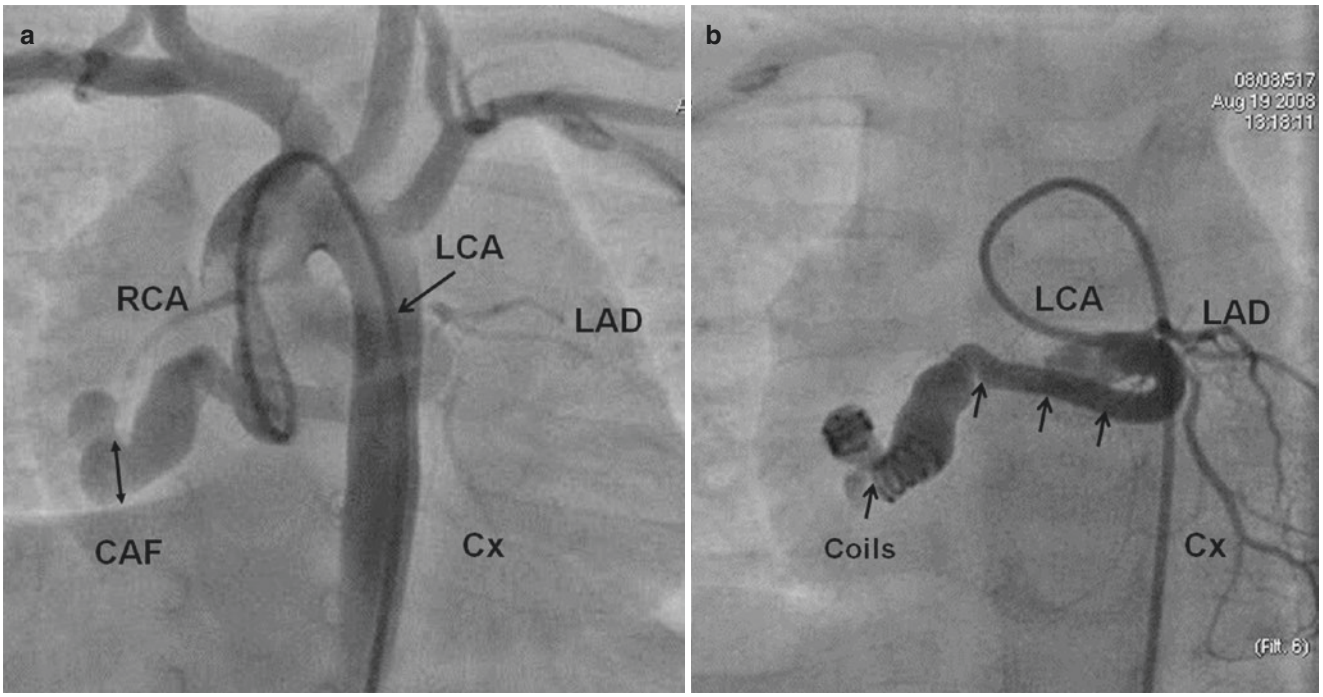



**Fig. 21.1** (**a**) Aortic root angiogram in anteroposterior view shows tortuous coronary artery fistula (CAF) (arrow) arising from the left coronary artery (LCA) entering the right atrium (RA). Normal branching of the LCA into the left anterior descending (LAD) and left circumflex (Cx) arteries is well seen. Proximal portion of the right coronary artery (RCA) is also opacified. (**b**) It was closed with two Gianturco coils (arrow)

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. 21.2).

Case study 3: Angiogram of a 15-day-old presenting with heart failure showed large fistula from LCA to the RA coursing posterior to the aortic root. Surgery was performed through midline sternotomy, and a large coronary fistula from the left coronary sinus behind the aortic root was identified in the transverse sinus of the heart. The fistula was clipped in its course without the use of cardiopulmonary bypass. Disappearance of the thrill was taken as confirmation of closure of the fistula, and sternotomy was closed. The neonate remained ventilator dependent due to significant residual flow through the fistula. Lung infections complicated the course of the child further. Transcatheter closure of the fistula was prompted by persistent heart failure, growth failure, and recurrent pneumonia warranting ventilatory support (Fig. 21.3).

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. 21.4).

Case study 5: A 21-year-old asymptomatic man was identified to have 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. 21.5). Magnitude of the shunt and elevation of pulmonary artery pressure were the indications for its closure.

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. 21.6).

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. 21.7). Symptoms and documentation of reversible myocardial ischemia dictated its closure.

Case study 8: A 3-year-old asymptomatic young boy was diagnosed to have a fistula from a single left coronary artery

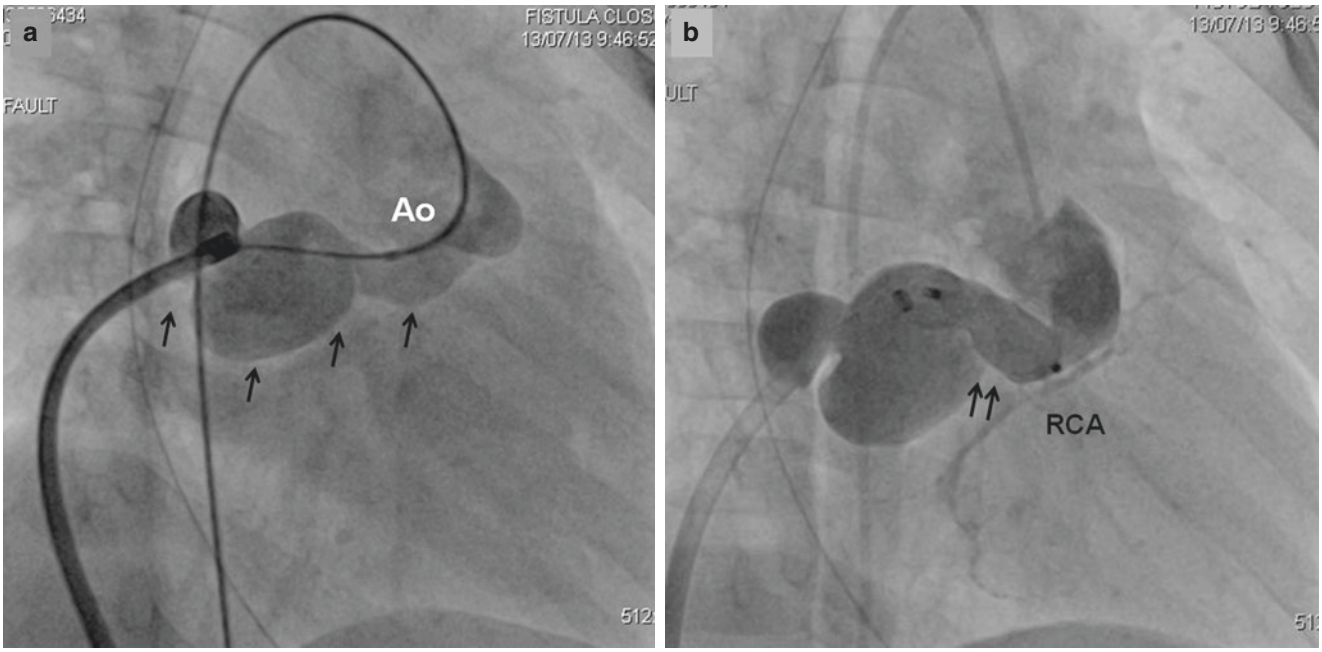



**Fig. 21.2** (**a**) Angiogram through venous sheath after AV loop formation in right anterior oblique projection shows a large fistula (arrows) from the proximal part of right coronary artery (RCA) coursing posteriorly to enter the right atrium. Faint opacification of the aortic root (Ao) is also seen.The 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

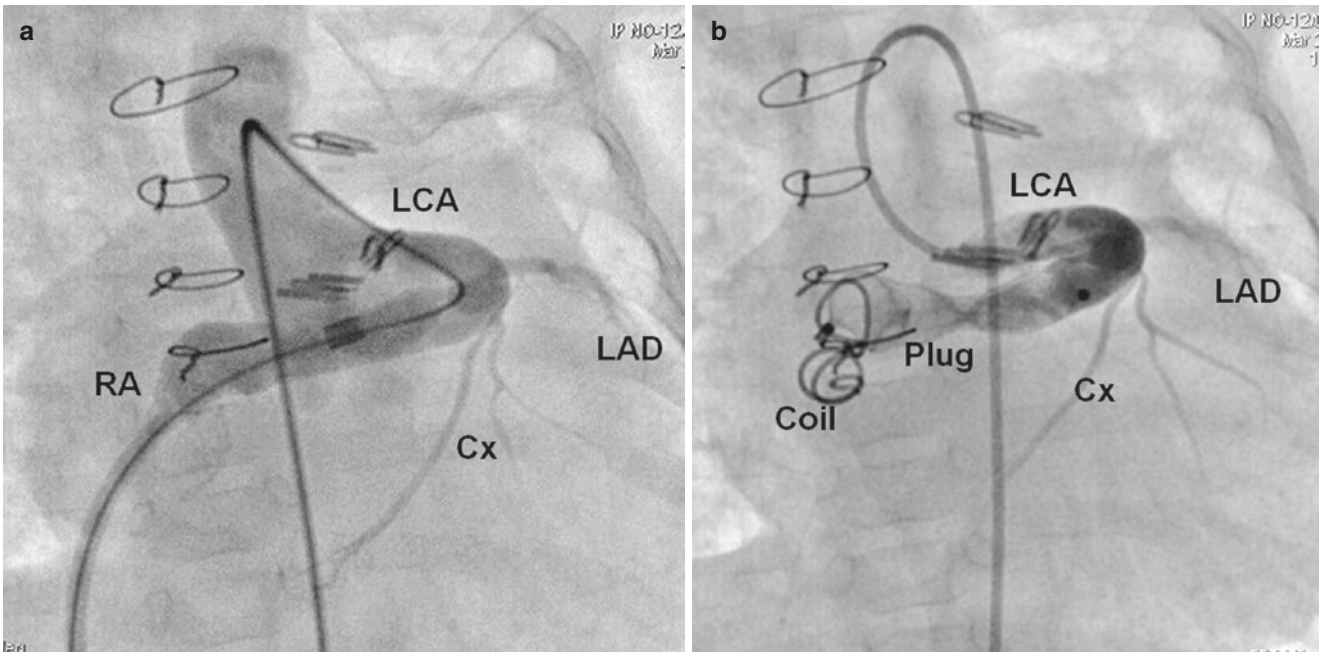



**Fig. 21.3** (**a**) Angiogram through venous sheath after arteriovenous (A-V) loop formation in a patient with a significant residual flow from the left coronary artery (LCA) to the right atrial (RA) fistula after surgical ligation shows the long fistulous tract and normally branching LCA into the left anterior descending (LAD) and the left circumflex (Cx) arteries. Sternal wires and multiple clips placed to close the fistula surgically are seen. (**b**) Repeat angiogram after placement of Amplatzer vascular plug II through the venous sheath and an additional coil at the most distal end shows a complete closure of the fistula with better visualization of the branches of the LCA

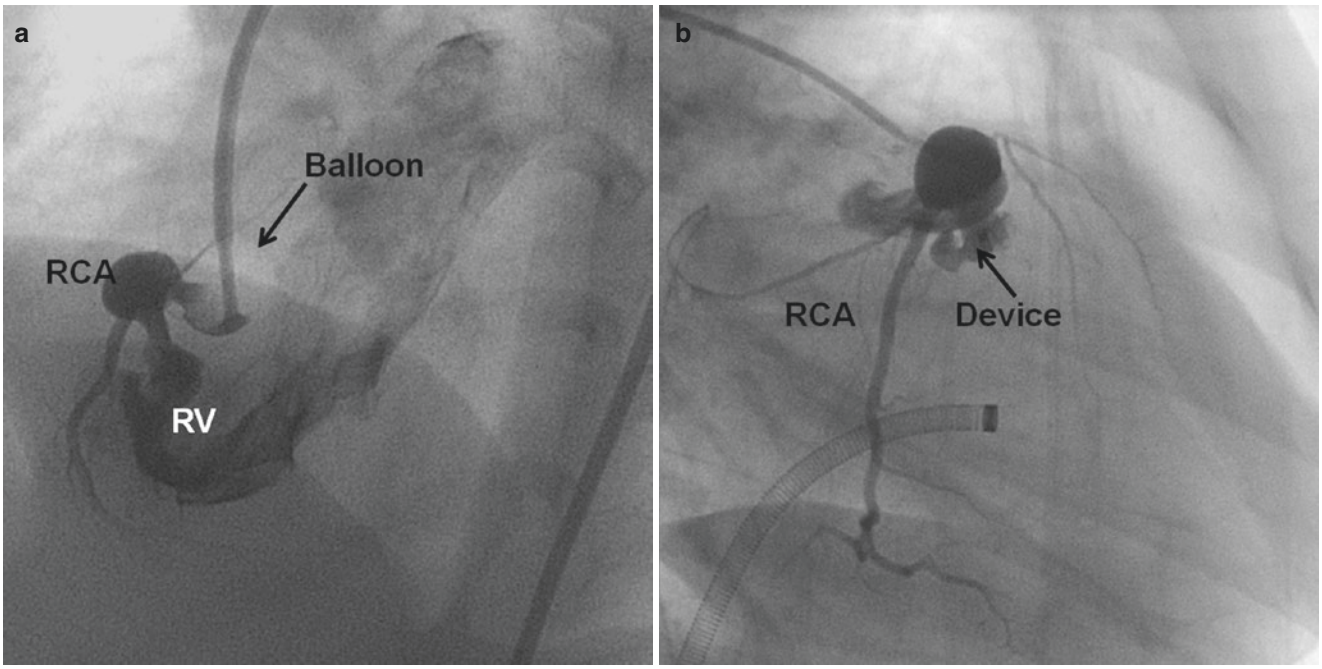



**Fig. 21.4** (**a**) Aortogram in left anterior oblique view with cranial angulation using a balloon-tipped catheter shows large fistula from proximal right coronary artery (RCA) to the right ventricle (RV). The branches of the RCA distal to the fistula are seen due to proximal balloon occlusion. (**b**) After closure with a duct occluder I, angiogram in right anterior oblique view shows the RCA with branches

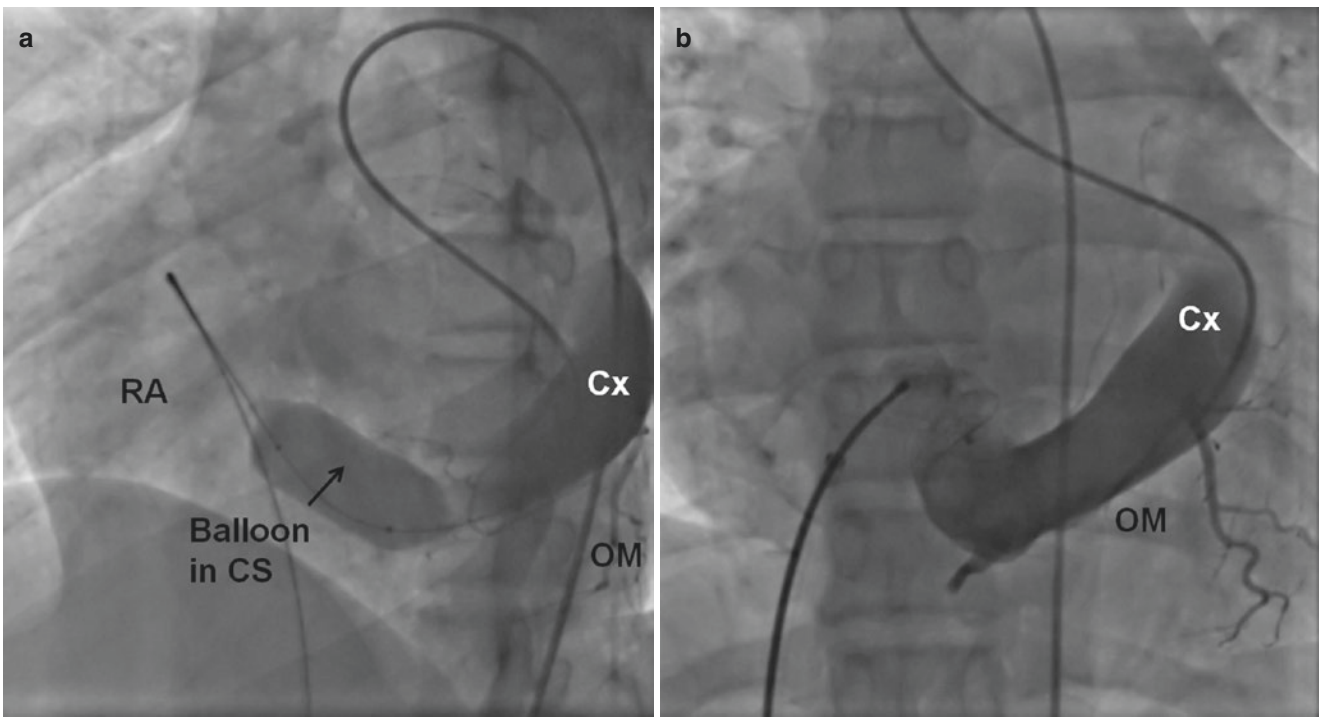



**Fig. 21.5** (**a**) Selective angiogram in the left circumflex artery (Cx) after occluding the fistula opening into the right atrium (RA) by a 25 mm Tyshak II valvuloplasty balloon shows the obtuse marginal (OM) branches of the Cx. (**b**) After the distal end of fistula was closed with a large duct occluder I, the contrast is seen to opacify the Cx and OM branches more intensely

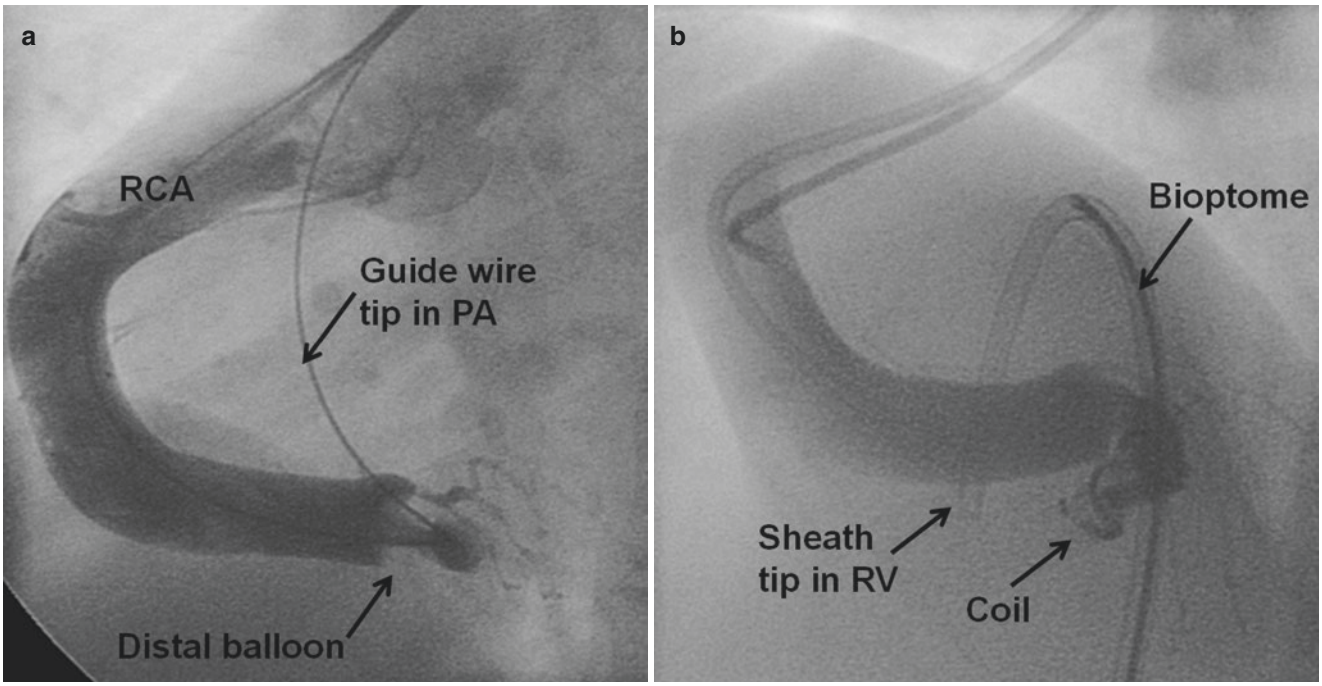



**Fig. 21.6** (**a**) Selective right coronary artery (RCA) angiogram in left anterior oblique view shows a large RCA to right ventricle (RV) fistula. Through a second arterial access, a guidewire was advanced through the fistula into the pulmonary artery, and a distal balloon occlusion was done. The 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

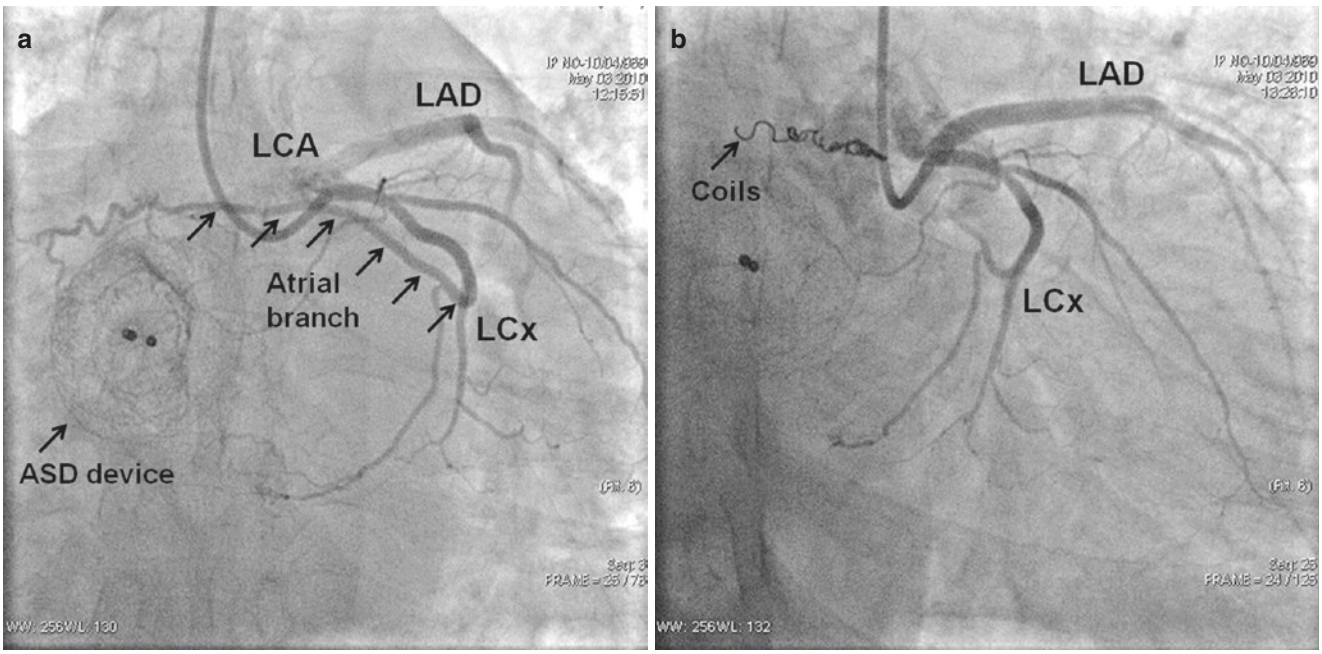



**Fig. 21.7** (**a**) Left coronary artery (LCA) injection through right radial arterial access in right anterior oblique view shows a small fistula (multiple arrows) from the left circumflex artery (LCx) coursing posteriorly and terminating in the right atrium in a patient who had closure of atrial 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 microcoils (0.018″ Hilal embolization coils, Cook Medical) using a microcatheter through a left coronary artery guide catheter

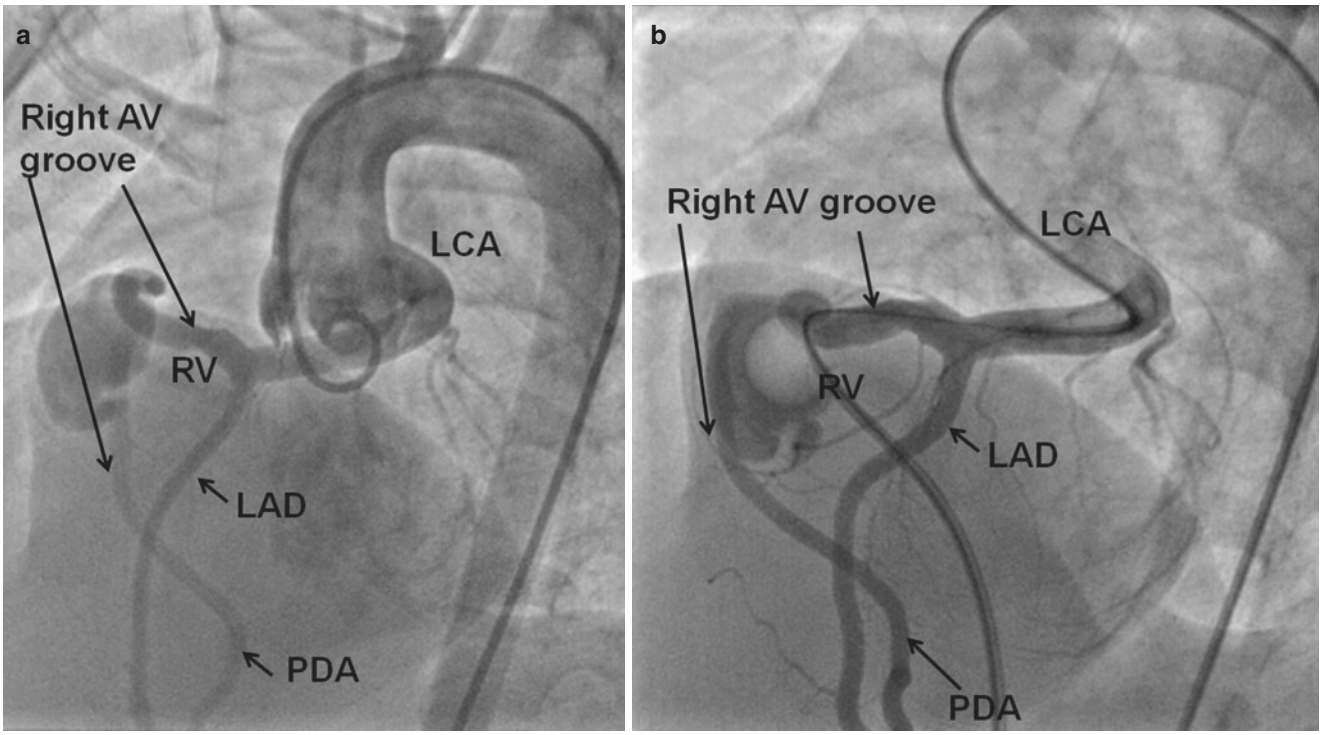



**Fig. 21.8** (**a**) Aortic root angiogram in left anterior oblique projection shows absence of the right coronary artery with faint opacification of the left coronary artery (LCA). A dilated left anterior descending (LAD) artery continues beyond the apex in the posterior interventricular groove as posterior descending artery (PDA) and then subsequently courses in the posterior right atrioventricular groove in the 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) and 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)

to the right ventricle. The LAD continued beyond the apex in the posterior interventricular groove as the posterior descending artery (PDA) and subsequently coursed in the posterior right atrioventricular groove and terminated in a sac before entering the right ventricle. There was a large right-sided branch from the LAD that coursed in the right anterior interventricular groove which also terminated in the same sac 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. 21.8).

*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 echo 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

*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*: 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 features (presence of aneurysmal sac, progressive enlargement of the feeding vessel, arrhythmias either at rest or exercise induced) which can result in life-threatening complication.

*Anatomy of the fistula*: Proximal side branch fistulas 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 arteries, 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.

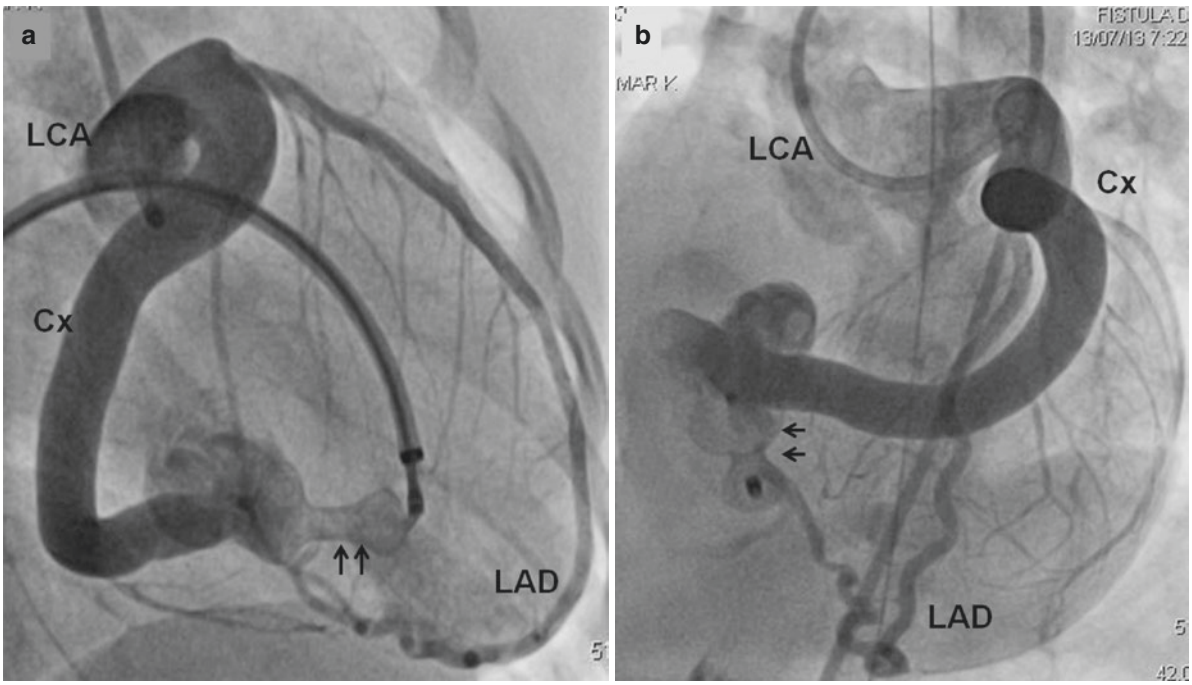



**Fig. 21.9** Left coronary artery (LCA) injections in right anterior oblique projection (**a**) and left anterior oblique view (**b**) show a large feeder from the dilated left circumflex (Cx) artery and smaller additional feeder from the terminal part of the left anterior descending (LAD) artery entering 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)

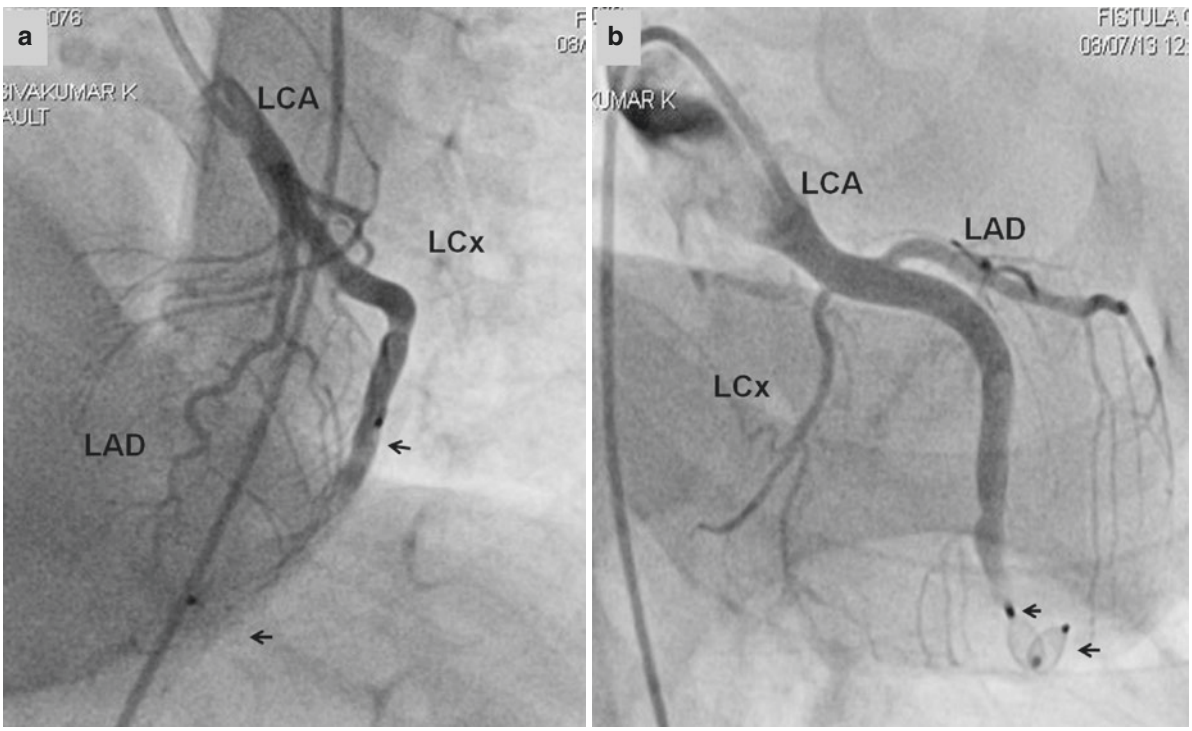



**Fig. 21.10** (**a**) Left coronary artery (LCA) injection in left anterior oblique and (**b**) right anterior oblique views shows a fistula from the ramus intermedius branch of the LCA arising between the left anterior descending (LAD) and the left circumflex (LCx) branches. This fistula was closed 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

## 21.4 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 especially in the absence of a continuous murmur, 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.

## 21.5 Pre-procedural 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, the presence of side branches proximal and distal to its drainage, and the size of the fistula at various sites (Fig. 21.11). 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.

## 21.6 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. 21.7). In cases where AV loop formation and transvenous delivery of the device are planned, it is necessary to have an

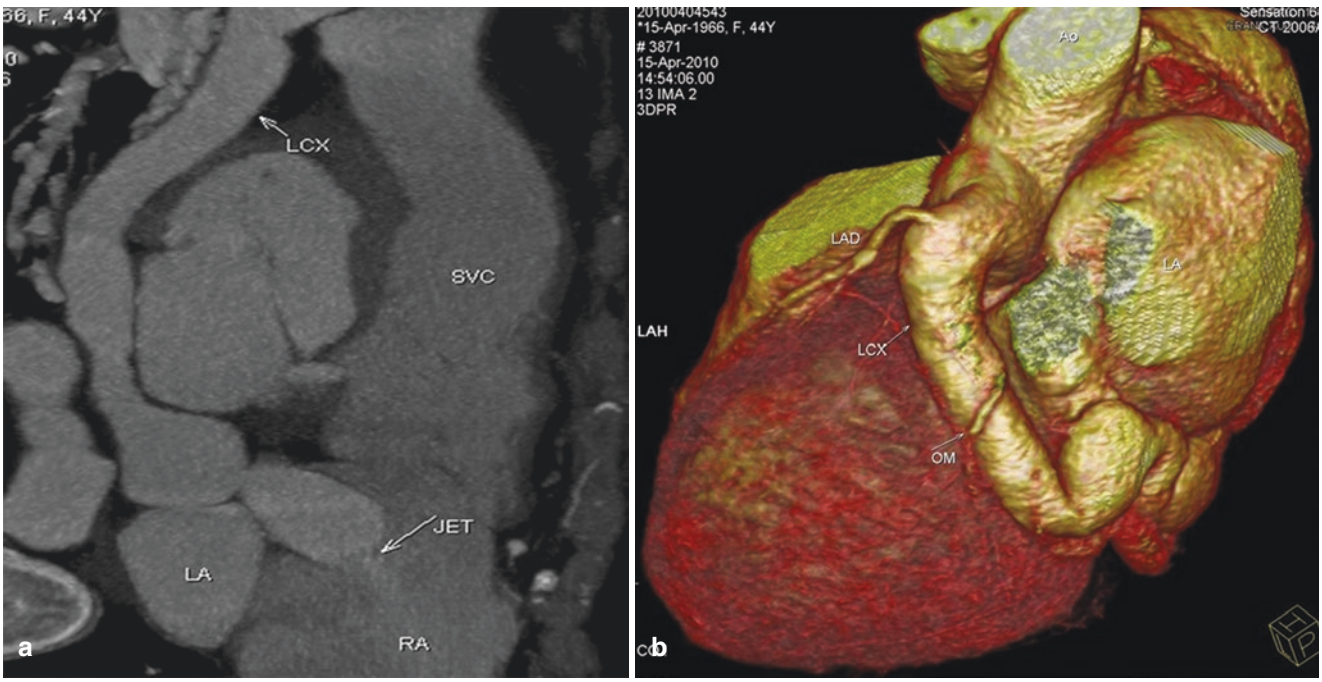



**Fig. 21.11** (**a**) CT coronary angiogram showing a fistula arising from the left circumflex artery (LCx) coursing in front of the left atrium (LA) and draining into the RA just below the entry of superior vena cava (SVC). Severe restriction at the point of exit into the RA is producing a jet effect. 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 (LAD) artery 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

additional venous access. In some cases where balloon occlusion angiography is planned, an additional arterial access may be necessary.

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. Precise anatomical delineation of the fistula is of utmost importance in planning the intervention, and hence every effort needs to be made in defining the origin, course, and site of drainage very precisely. In a large flow fistula with dilated and tortuous fistulous tract, routine angiographic techniques may not work. Selective coronary angiogram in patients 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 (Video 1). In some cases, a delivery sheath can be used to do a diagnostic angiogram prior to making a final decision about the size and type of the device and the site of device delivery (Video 2). Better delineation of the fistulous tract and the coronary branches can also be achieved by making a pump angiogram under pressure using either a cut or a non-cut pigtail catheter (Video 3a, 3b).
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. 21.8).
8. In high-flow fistula, the coronary branches are delineated better by proximal or distal balloon occlusion angiogram (Figs. 21.4, 21.5, and 21.8) (Video 4a, 4b).
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 balloon with care to avoid balloon rupture (Fig. 21.4).
10. Distal occlusion with balloon floatation wedge catheters (Figs. 21.5 and 21.8) or compliant occlusion balloons (Fig. 21.6) is carried out after forming an arteriovenous (A-V) loop. The waist on the balloon gives additional information regarding the size of the distal exit (Video 5). 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. 21.2). 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. 21.5 and 21.9).
12. Forming A-V 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" glide wire (Terumo) is

**Video 1** Selective right coronary angiogram using a 6F guide catheter to delineate a large flow fistula opening into the right ventricle (AVI 6371 kb)

**Video 2** Selective left coronary angiogram done antegradely using a delivery sheath prior to making a final decision regarding the type and size of the device as well as the site of placement. The angiogram clearly demarcates the origin, course, and the drainage site of the fistula which was arising from the left coronary sinus and was draining into the right atrium (AVI 7451 kb)

**Video 3a and b** Selective left circumflex angiogram in RAO (**a**) and LAO (**b**) projections using a cut pigtail catheter and pump injection in a patient with a significantly tortuous and dilated fistulous tract. 12 mL contrast was injected at 8 mL/s with a PSI of 600 (WMV 614 kb)

**Video 4a and b** Retrograde, distal occlusion balloon angiography done with a Berman catheter. It helps in delineating multiple side branches (**a**) which were not obviously seen on a routine angiogram (**b**) (WMV 15208 kb)

**Video 5** Balloon occlusion angiography using a Tyshak II balloon passed over the A-V loop from the venous side. Test occlusion gives a good idea about whether ischemia is likely to occur following device closure. It also helps in defining the narrowest portion of the fistula which is useful in deciding the type and size of the device to be used for closure (WMV 15387 kb)

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 glide/coronary wire with a Noodle wire (St Jude Medical) for forming the A-V loop.

13. For a more proximal occlusion, a retrograde closure from the 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 a 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 for 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. 21.5 and 21.6).

## 21.7 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″ glide wires (Terumo Corporation), 0.014″ floppy coronary guidewires, Noodle wire.

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, Amplatzer vascular plugs I–IV (St Jude Medical).

Long sheaths: Flexor sheaths and Mullins sheaths (Cook Medical), Torque view 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: Goose neck snare (eV3 medical), bioptome (Cook)

## 21.8 Tips and Tricks

1. A-V 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 0.014″, 0.018″, or 0.025″ guidewires may be more easily advanced to form the A-V loop. Microcatheter helps in manipulating the guidewire along the tortuosities (Videos 6, 7, and 8). Hypotension occurs during passage of rigid braided sheaths after A-V loop formation; however, the blood pressure quickly recovers once the A-V loop is broken. This is more commonly seen in small neonates or infants.
2. Braided hydrophilic sheath (Flexor sheath, Cook Medical) is preferred to avoid kinks, but it tends to be more rigid.
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 retrograde closure from the aortic end, a diagnostic catheter is carefully advanced deep into the fistula to the selected occlusion site. Embolization coils and Amplatzer vascular plug IV can be delivered through 0.038″ lumen diagnostic catheters (Fig. 21.10). 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. 21.6). Bioptome helps in controlling the coil delivery.

**Video 6** Diagnostic angiogram showing a very tortuous fistula arising from the left circumflex artery and draining into the coronary sinus (AVI 18438 kb)

**Video 7** Microcatheter and 0.014" soft-tipped PTCA wire being manipulated in the proximal part of the fistula (AVI 3332 kb)

**Video 8** The microcatheter and PTCA wire seen going all the way into the coronary sinus taking all the twists and turns. This probably would not have been possible without the support of the microcatheter (AVI 5637 kb)

## 21.9 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.

## 21.10 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. Occasionally, distal, end artery fistula having a long, dilated proximal fistulous tract can develop acute thrombosis immediately after fistula closure due to "slow flow" (Videos 9, 10, 11, 12, and 13). If 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 a ST segment elevation and evidence of acute myocardial ischemia/infarction, thrombolysis using lytic drugs can be useful (Video 14). 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 atropine to combat bradycardia may rarely be required if there is hemodynamic compromise.
3. Coronary artery dissection: The 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.

## 21.11 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.

## 21.12 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, computerized stress test is recommended every year to confirm 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.

**Acknowledgments** The authors wish to thank Prof. Prafulla Kerkar for allowing us to share Videos 4a, 4b, 9, 10, 11, 12, 13, and 14. These have been reproduced with his permission.

**Video 9** Balloon occlusion angiogram showing a distal, end artery fistula arising from the left anterior descending (LAD) artery and opening into the right ventricle. There are a large number of septal and diagonal branches arising from the fistulous LAD (WMV 15208 kb)

**Video 10** The fistula is occluded using AVP II (WMV 2426 kb)

**Video 11** Angiogram showing a "slow flow" in the LAD after 5 min of closure (WMV 6286 kb)

**Video 12** Angiogram showing further slowing of flow after 10 min of occlusion (WMV 8786 kb)

**Video 13** Angiogram showing "no flow" in the LAD with ECG showing ST elevation with drop of blood pressure (WMV 4325 kb)

**Video 14** Post-urokinase angiogram showing recanalization of the fistula with resolution of the ECG changes and normalization of hemodynamics (WMV 5997 kb)

# 22 Vessel Embolization: Transcatheter Embolization of Pulmonary Arteriovenous Malformations and Aortopulmonary Collateral Arteries

Liang Tang, Zhen-Fei Fang, and Sheng-Hua Zhou

## 22.1 Transcatheter Embolization of Pulmonary Arteriovenous Malformations

Pulmonary arteriovenous malformations (PAVMs) are direct communications between the pulmonary arteries and veins, bypassing the normal pulmonary capillary bed and resulting in an intrapulmonary right-to-left shunt. Large or diffuse PAVMs can cause systemic hypoxemia and cerebrovascular complications secondary to paradoxical embolism as well as pulmonary hemorrhage due to sac rupture. Transcatheter embolization has emerged as the preferred treatment for PAVMs with favorable long-term follow-up results.

### 22.1.1 Indications and Patient Selection

All symptomatic patients

– Evidence of significant systemic hypoxemia

Asymptomatic patients

– Risk for paradoxical embolism or pulmonary hemorrhage
– Having a documented history of a paradoxical embolic event
– Discrete lesions with feeding arteries greater than 3 mm in diameter

Partial or staged closure may also be indicated in some patients with diffuse PAVMs in order to alleviate symptoms.

Although traditionally coils have been used to occlude PAVM, the size of the feeding vessels involved is usually more effectively occluded using the Amplatzer vascular plug (AVP) series (AGA Medical Corp., MN, USA) (Fig. 22.1). Transcatheter occlusion of a large PAVM in a 11-year-old female patient using the AVP I is shown in Figs. 22.2–22.6 and videos 1–3.

## 22.2 Transcatheter Embolization of Aortopulmonary Collateral Arteries

Aortopulmonary collateral arteries (APCs) can be detected frequently in patients with 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.

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.

### 22.2.1 Indications and Patient Selection

#### 22.2.1.1 Indications

Transcatheter occlusion of APCs is indicated for the treatment of aortopulmonary collateral vessels with documented large left-to-right shunting that results in congestive heart failure, pulmonary overcirculation, and respiratory compromise or development of pleural effusion or protein-losing enteropathy.

**Relative Indications**

1. Asymptomatic single ventricle patients with moderate-sized collaterals undergoing routine pre-Glenn or pre-Fontan cardiac catheterization
2. Patients with pulmonary atresia and APCs that have adequate dual supply from native pulmonary arteries

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_22) contains supplementary material, which is available to authorized users.

L. Tang · Z.-F. Fang · S.-H. Zhou (✉)
Department of Cardiology, The Second Xiangya Hospital of Central South University, Changsha, Hunan, China



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_22

**Contraindications**

1. Transcatheter occlusion is not recommended for the presence of APCs of any size in 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.

Transcatheter occlusion of multiple APCs in three patients using coils or the combination of coils with AVP are shown in Figs. 22.7–22.24.

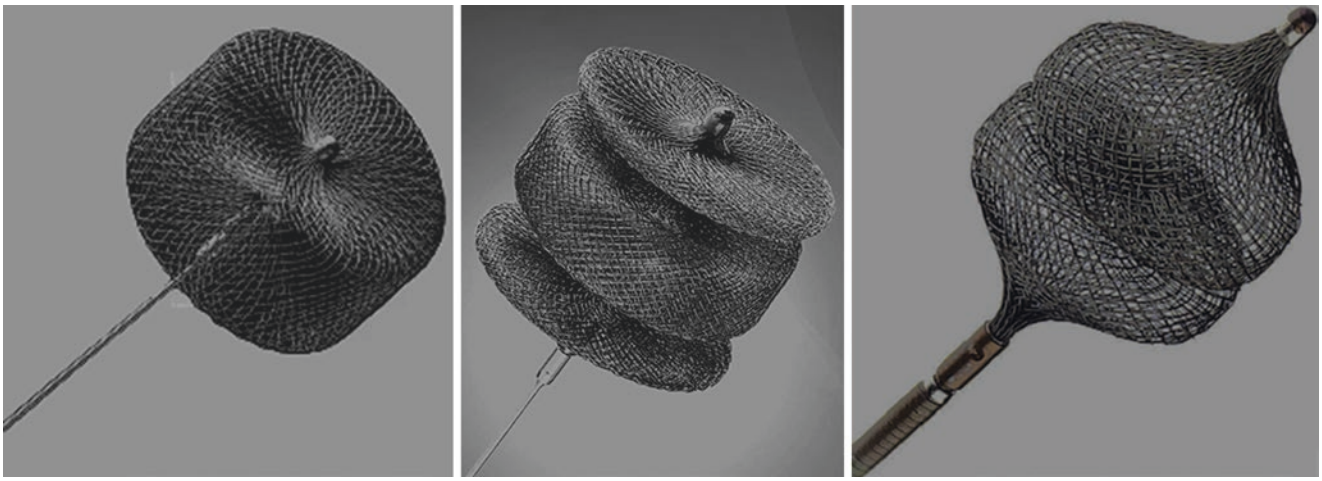


**Fig. 22.1** Amplatzer plug I, Amplatzer plug II, and Amplatzer plug IV. The AVPs are particularly suitable for embolization of large high-flow feeding vessels. They are a woven nitinol wire cylinder that can be delivered via small catheters such as standard 5–8-Fr coronary guiding catheters. The size of the AVP selected for embolization was approximately 1.5–2 times the caliber of the feeding vessel

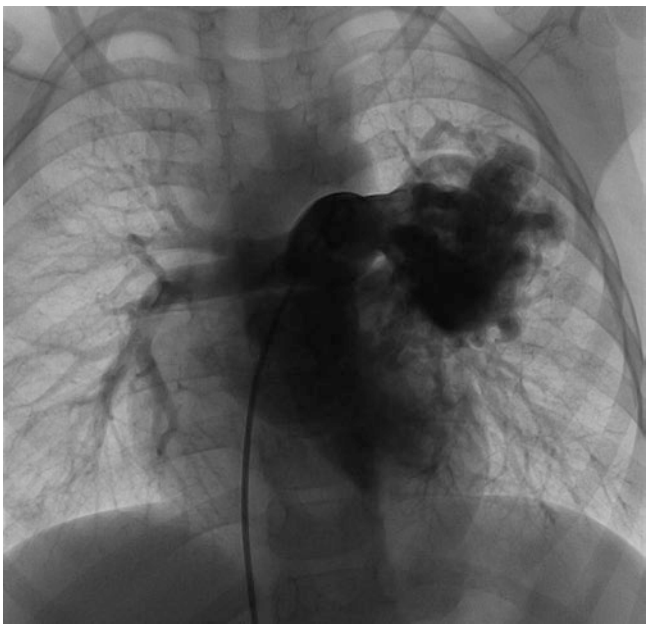


**Fig. 22.2** Patient 1. A 11-year-old female patient with a diagnosis of hereditary hemorrhagic telangiectasia was referred for transcatheter treatment of multiple PAVMs. Pulmonary angiogram revealed diffuse PAVMs of the left lung, with major PAVFs identified in the upper lobes

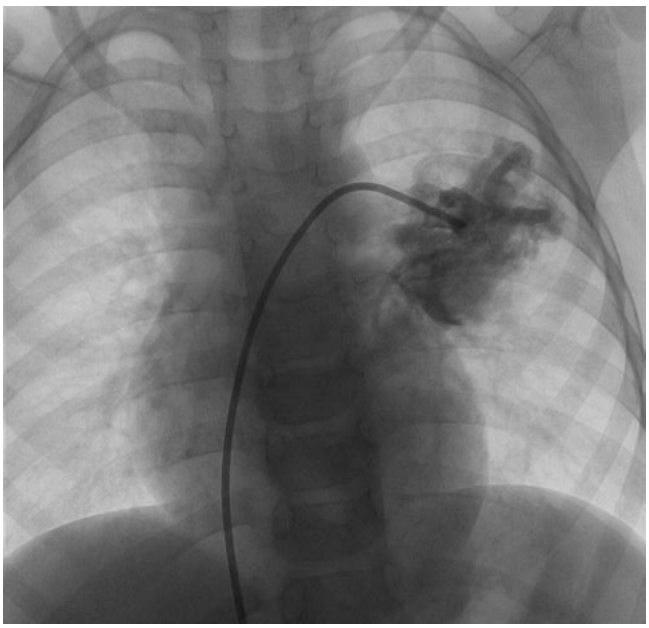


**Fig. 22.3** Patient 1. The feeding vessel of the major PAVM located in the left upper was selectively engaged with the 5F VER135° angiographic catheter(Cordis, USA) and the feeding vessel measuring approximately 8 mm in diameter

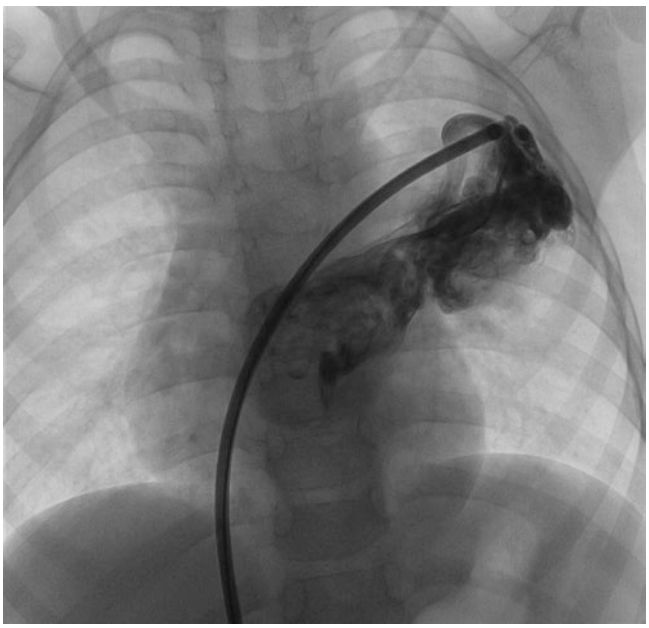


**Fig. 22.4** Patient 1. The VER135° angiographic catheter was then exchanged for an 8 Fr, 80-cm long delivery sheath through which a 100 cm, 7 Fr Judkins Right 3.5 guide catheter was introduced. The guide catheter was then advanced as distally as possible within the feeding vessel beyond any branches to normal lung

**Video 1** Patient 1. The VER135° angiographic catheter was then exchanged for an 8 Fr, 80-cm long delivery sheath through which a 100 cm, 7 Fr Judkins Right 3.5 guide catheter was introduced. The guide catheter was then advanced as distally as possible within the feeding vessel beyond any branches to the normal lung (AVI 15365 kb)

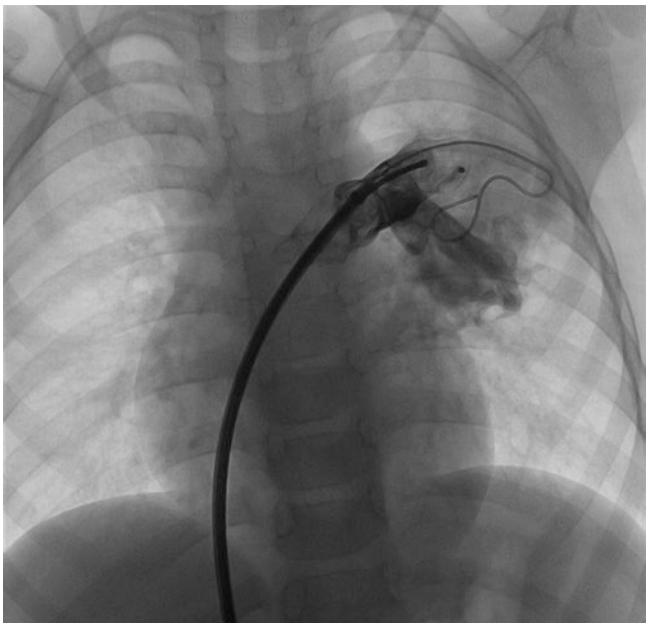


**Fig. 22.5** Patient 1. A 12 mm AVP I (AGA Medical Corporation, Minnesota) was selected so as to be 50% larger than the target vessel and was delivered through the guide catheter to the feeding vessel. Selective arteriogram after deployment of the AVP in the feeding vessel showed near-complete occlusion of the left upper PAVM

**Video 2** Patient 1. A 12 mm AVP I (AGA Medical Corporation, Minnesota) was selected so as to be 50% larger than the target vessel and was delivered through the guide catheter to the feeding vessel. Selective arteriogram after deployment of the AVP in the feeding vessel showed near-complete occlusion of the left upper PAVM (AVI 23814 kb)

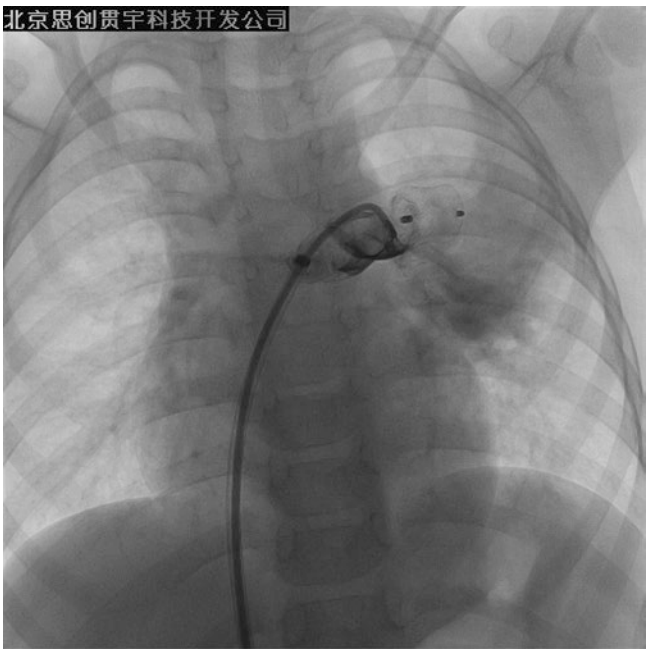



**Fig. 22.6** Patient 1. Repeated selective arteriogram a 5 min later after the device release documented the AVP I in good position with no residual flow into the left upper PAVM. Upon deployment of the AVP, her peripheral arterial oxygen saturation improved from 86 to 93% at rest. Due to the length of the procedure and the amount of contrast given, no further embolization attempts to another PAVM were performed

**Video 3** Patient 1. Repeated selective arteriogram a 5 min later after the device release documented the AVP I in good position with no residual flow into the left upper PAVM. Upon deployment of the AVP, her peripheral arterial oxygen saturation improved from 86 to 93% at rest. Due to the length of the procedure and the amount of contrast given, no further embolization attempts to another PAVM were performed (AVI 24070 kb)

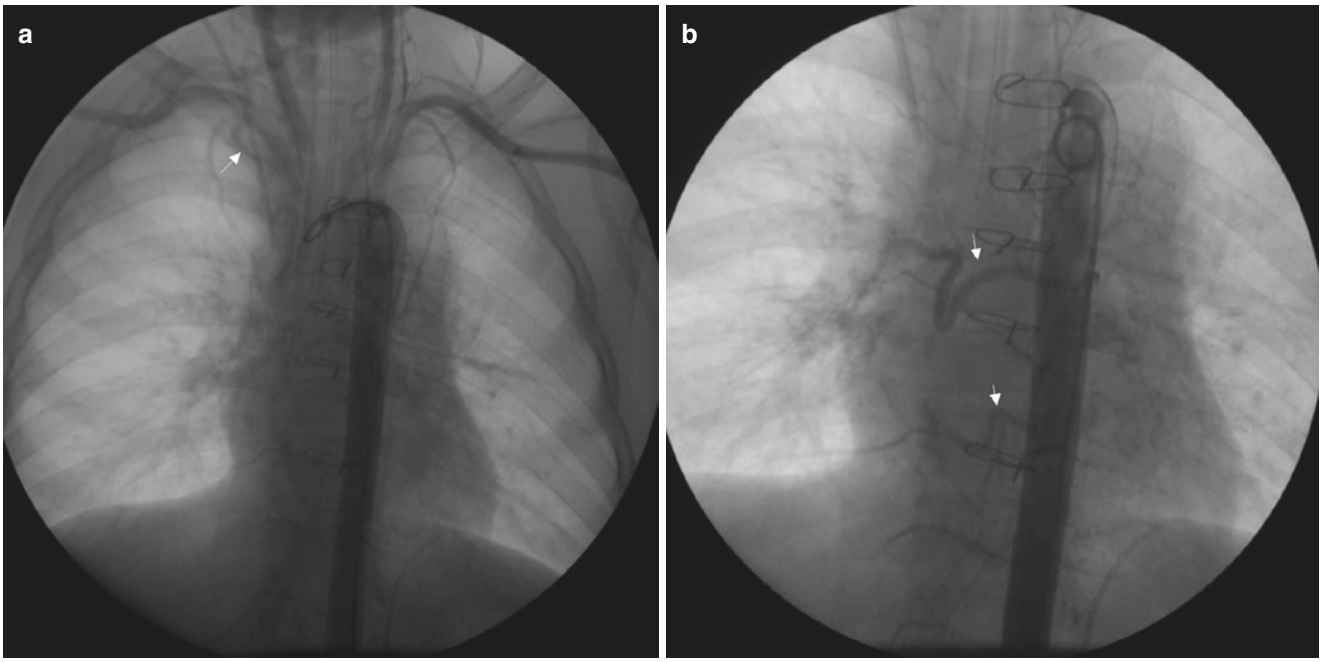



**Fig. 22.7** (**a**, **b**) Patient 1.An 11-year-old boy (24 kg) who had undergone surgical correction for TOF was referred for transcatheter occlusion of multiple APCs due to failure to wean from ventilation. Ascending and descending aortogram showing multiple APCs arising from the right subclavian artery and descending aorta (arrows)

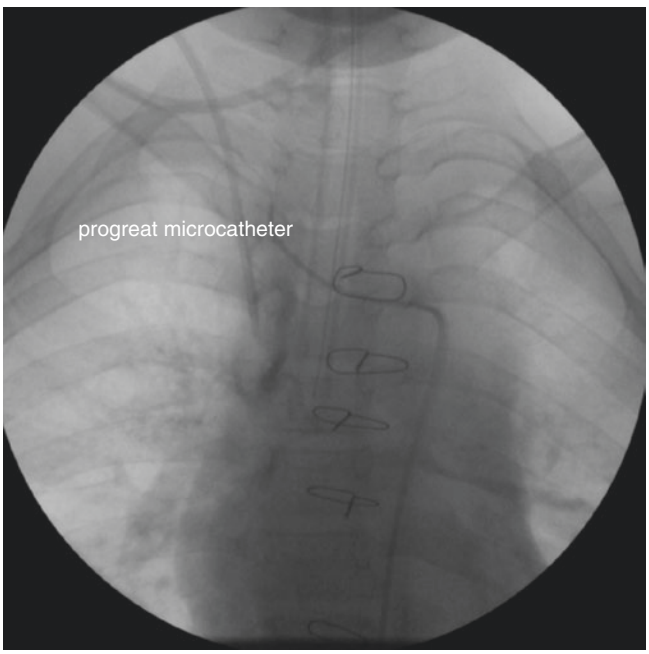



**Fig. 22.8** Patient 1. The aortopulmonary collateral artery arising from right subclavian artery was selectively engaged by a 100 cm, 5-Fr Cobra catheter (Terumo, Japan) through which a 0.014 in. guidewire was advanced into the target vessel distally. Over the guidewire, a Progreat microcatheter (Terumo, Japan) was introduced into the target vessel as deep as possible

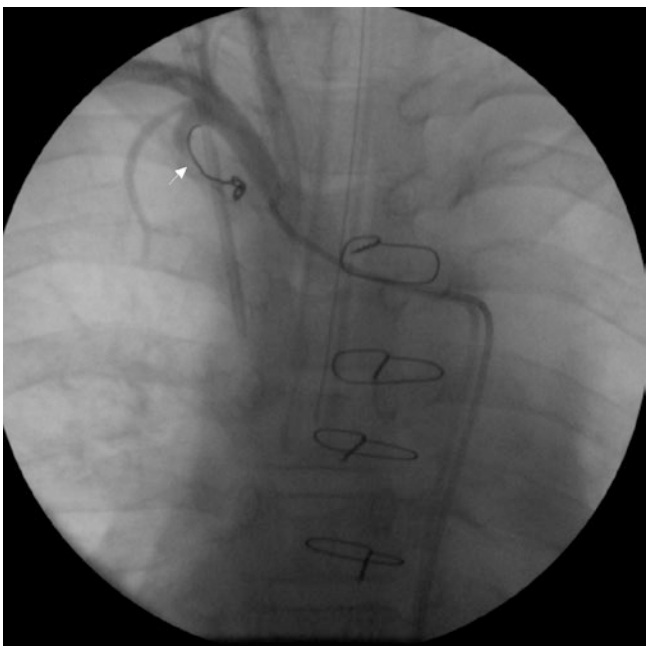


**Fig. 22.9** Patient 1. Then, two MWCE-18S-3/2 coils (COOK, USA) were delivered to the target vessel through the Progreat microcatheter (arrow). After the coils were positioned, an angiogram performed through the Cobra catheter confirming complete occlusion of the collateral artery

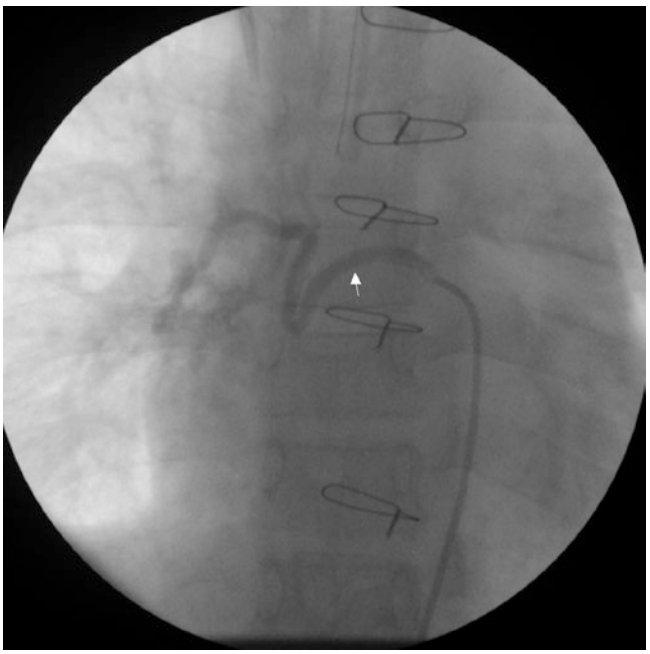


**Fig. 22.10** Patient 1. The aortopulmonary collateral artery arising from the descending aorta was cannulated selectively with the Cobra catheter. Selective angiogram of the target vessel performed via the Cobra catheter showing its tortuous course to the right middle pulmonary artery branch (arrow)

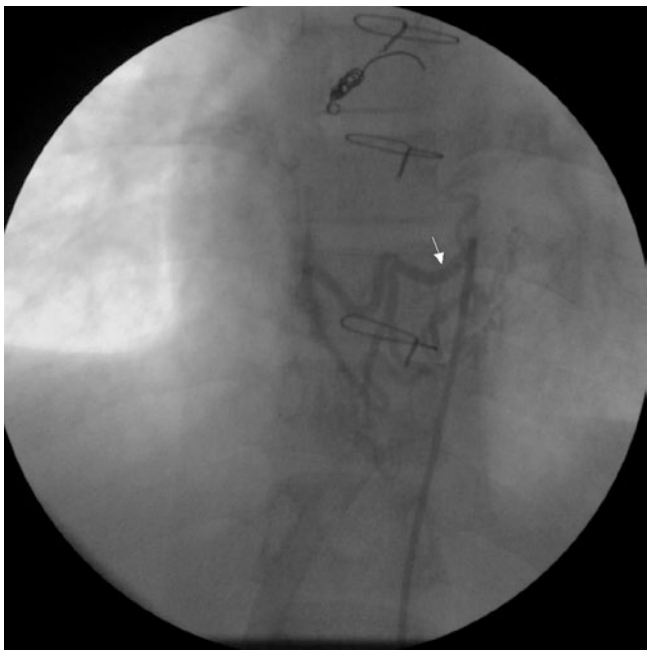


**Fig. 22.12** Patient 1. Next, the Cobra catheter was placed into another aortopulmonary artery that arises from the descending aorta and a selective angiogram showing the tortuous features of the target vessel (arrow)

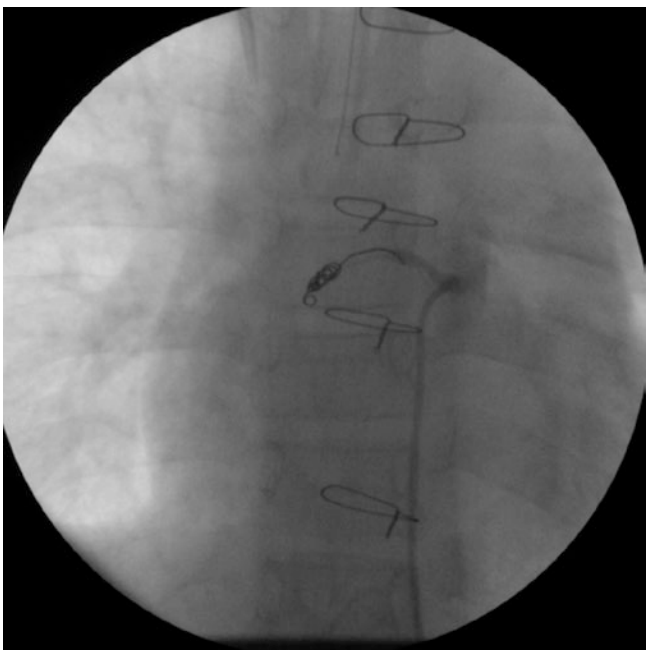


**Fig. 22.11** Patient 1. A MWCE-18S-4/2 coil (COOK, USA) was deployed at the narrowest segment of the collateral artery. Repeated descending aortogram confirming complete occlusion of the collateral artery

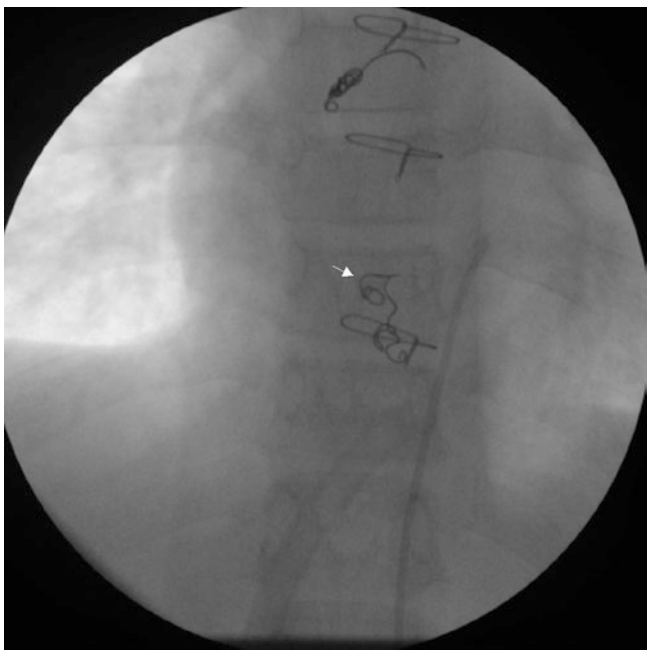


**Fig. 22.13** Patient 1. Three Coils (MWCE-18S-5/2, 4/2, 3/2 coils, COOK, USA) were positioned at the appropriate position of the collateral artery in a similar fashion as previously described (arrow)

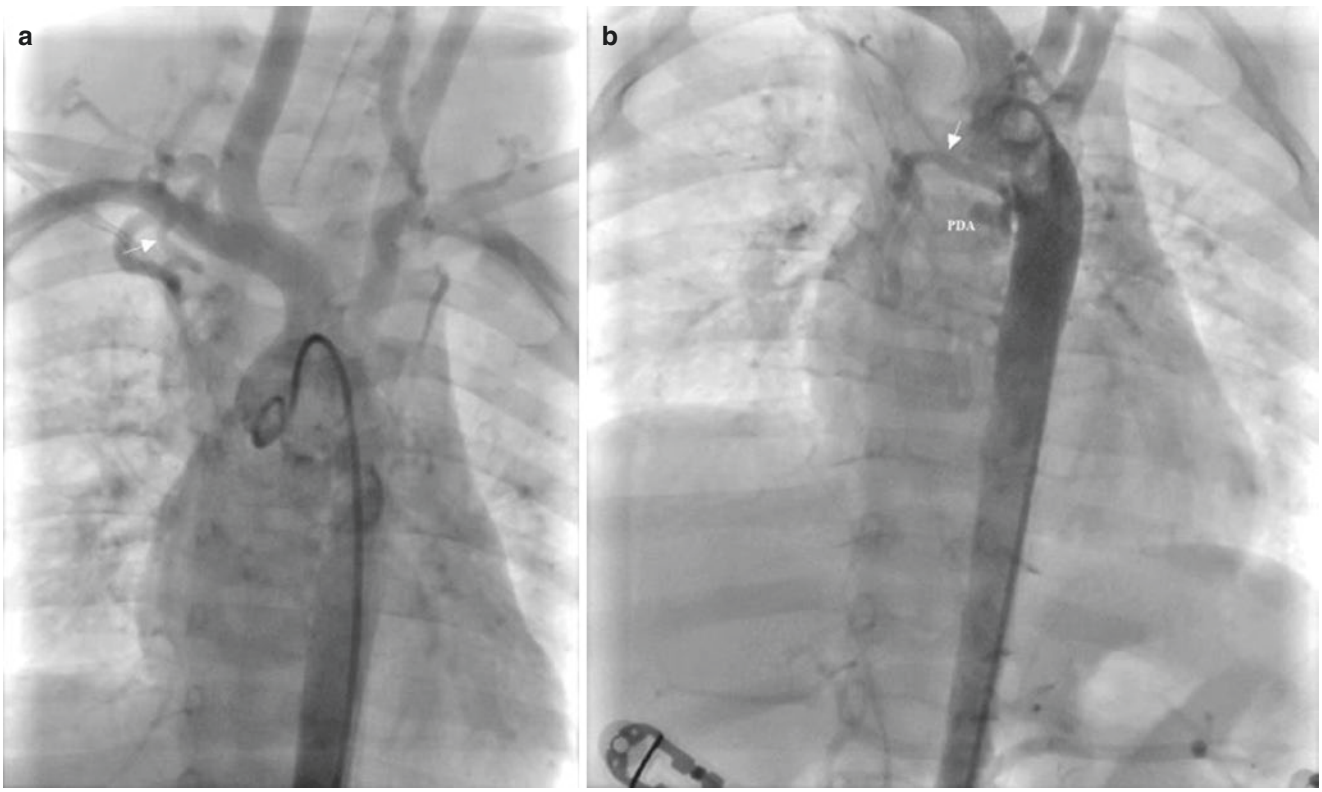



**Fig. 22.14** (**a, b**) Patient 2. A three-year-old boy (15 kg) was diagnosed to have transposition of great arteries (TGA) with ventricular septal defects (VSD) and pulmonary valve stenosis (PS) was referred for transcatheter occlusion of APCs due to recurrent symptomatic heart failure and frequent lower airway infection. Angiogram in the ascending and descending thoracic aorta demonstrating multiple APCs arising from the right subclavian artery and descending aorta (arrows) with a PDA measuring approximately 4 mm in diameter at its narrowest (white arrow)

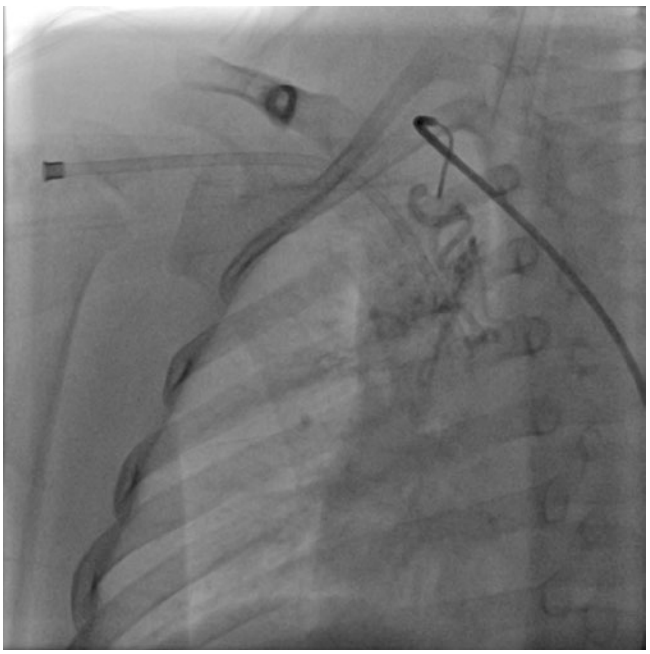


**Fig. 22.15** Patient 2. The target collateral artery arising from right subclavian artery was selectively engaged using a coaxial guide system with an outer 5-Fr Cobra catheter (Terumo, Japan) and inner Progreat microcatheter (Terumo, Japan)

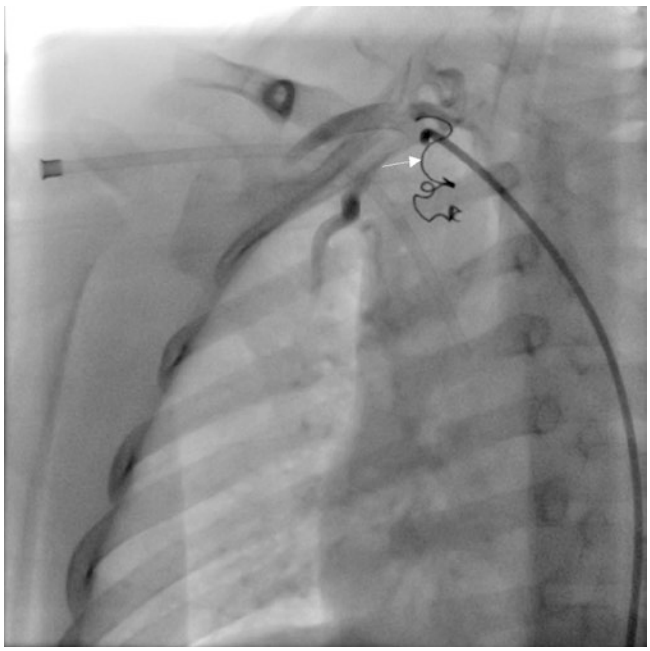


**Fig. 22.16** Patient 2. Then, three appropriate sized coils (arrow) were delivered to the target vessel through the Progreat microcatheter as distal as possible. After the coils were deployed, an angiogram was performed through the Cobra catheter confirming complete occlusion of the collateral artery

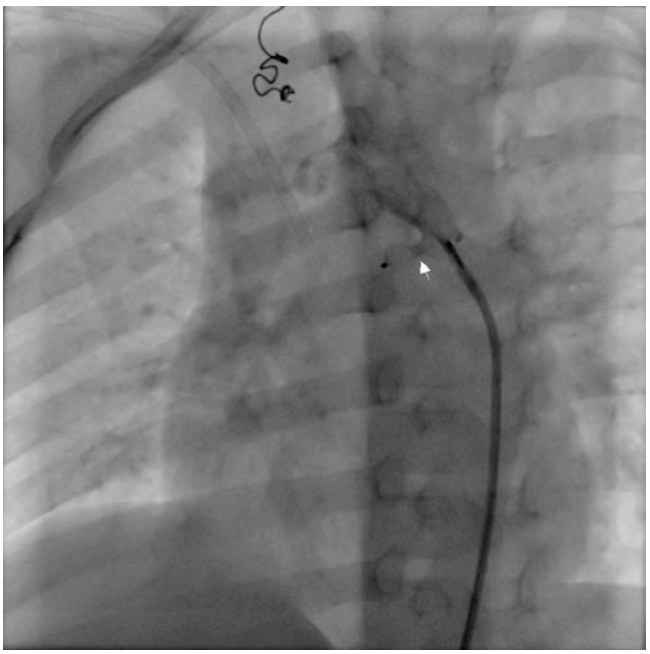


**Fig. 22.17** Patient 2. A 5-Fr Judkins right guide catheter was advanced over a hydrophilic-coated guidewire into the PDA as deeply as possible. Then an 8 mm AVP was advanced via the guide catheter into the PDA and deployed at the suitable position (arrow)

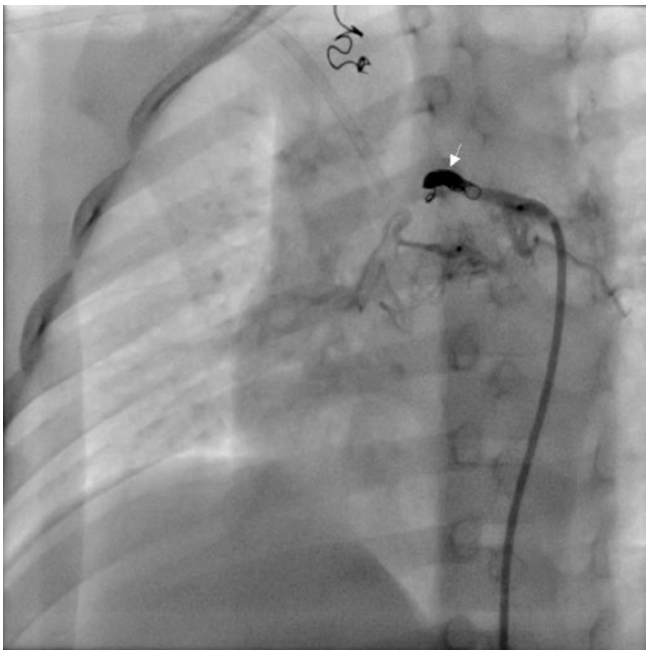


**Fig. 22.19** Patient 2. Following implant of two MWCE-18S-6/2 Tornado coils (COOK, USA), this aortopulmonary collateral artery was completely occluded

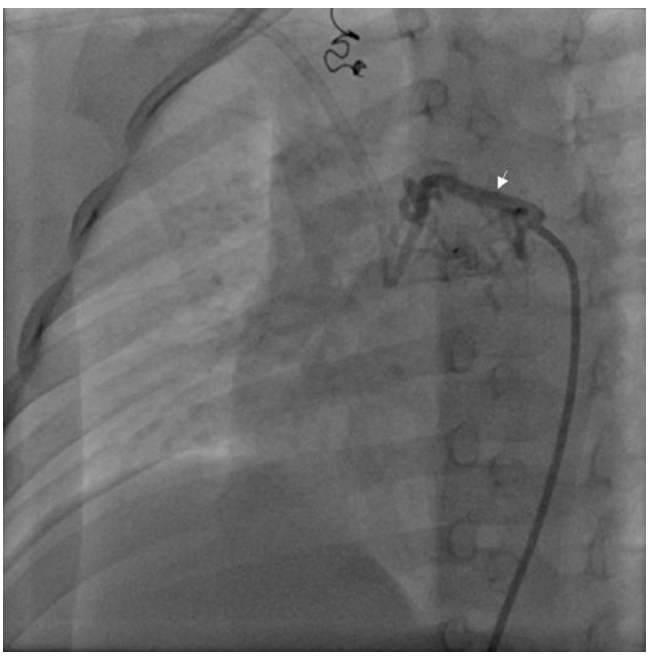


**Fig. 22.18** Patient 2. An angiogram performed after the deployment of the AVP, indicating complete occlusion of the PDA, but there remains a large collateral artery above the AVP device (arrow)

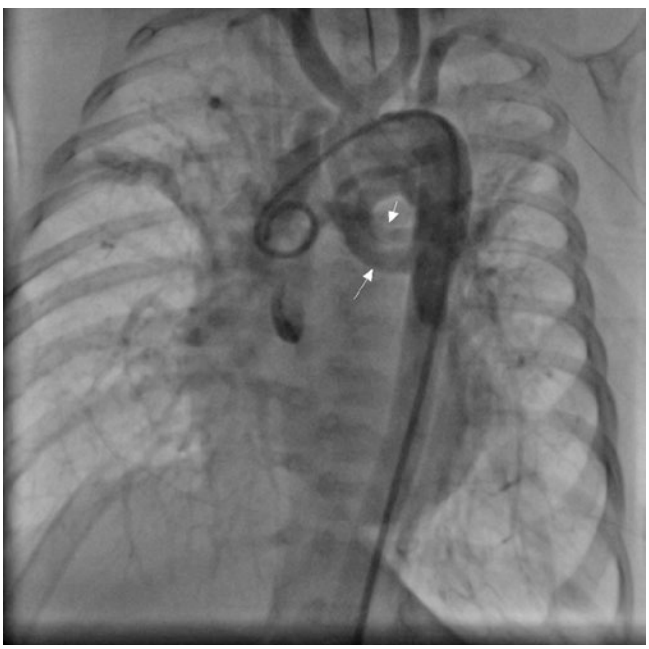


**Fig. 22.20** Patient 3. A two-month-old boy (3.9 kg) diagnosed with ventricular septal defect (VSD) and pulmonary atresia (PA) was referred for transcatheter occlusion of APCs prior to surgical correction. An angiogram in the ascending aorta demonstrating the presence of multiple APCs (arrows) arising from the descending aorta, which supplying the left and right lungs

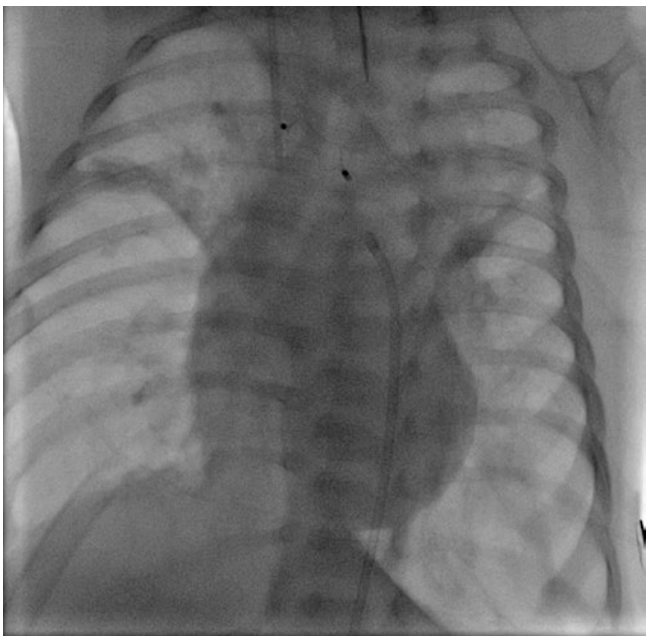


**Fig. 22.21** Patient 3. The larger collateral vessel was entered using a 5-Fr Judkins right 4.0 guide catheter (Cordis, USA), and an appropriate sized AVP was deployed at the narrowest position of the collateral vessel

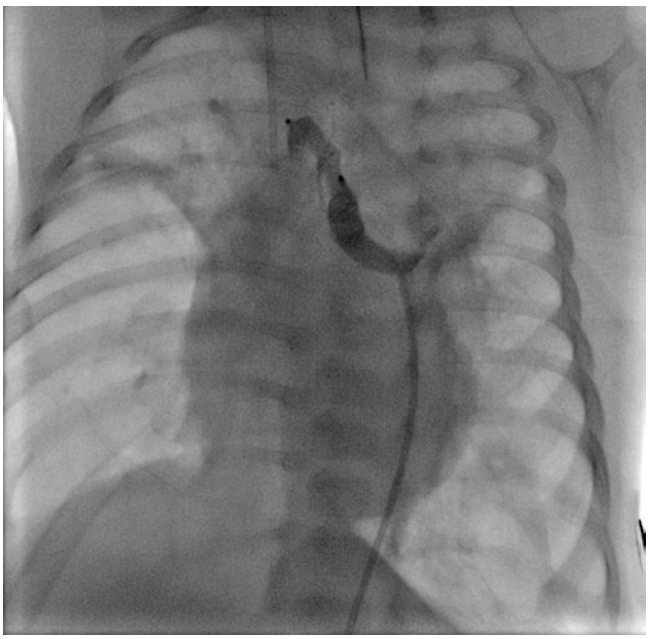


**Fig. 22.23** Patient 3. Therefore, an addition MWCE-18S-8/5mm Tornado coil (COOK, USA) was implanted, resulting in immediate, complete occlusion of this vessel

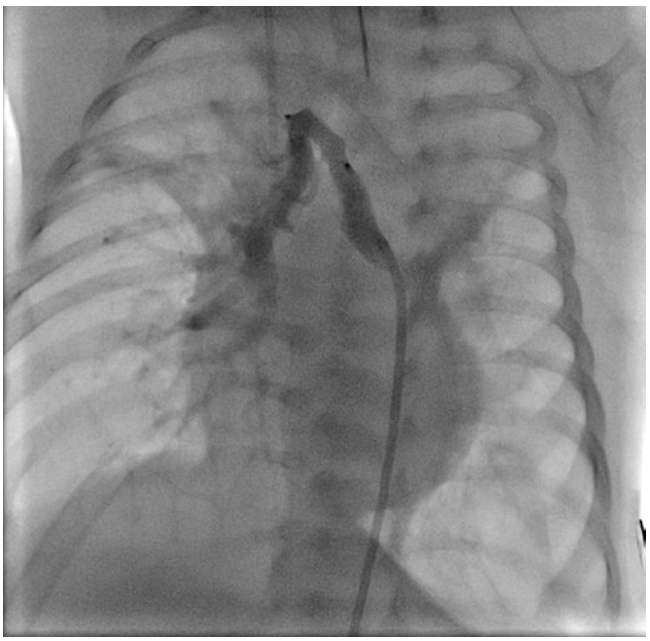


**Fig. 22.22** Patient 3. An angiogram was performed 5 min later after the AVP released, indicating a considerable residual shunt remained through the device

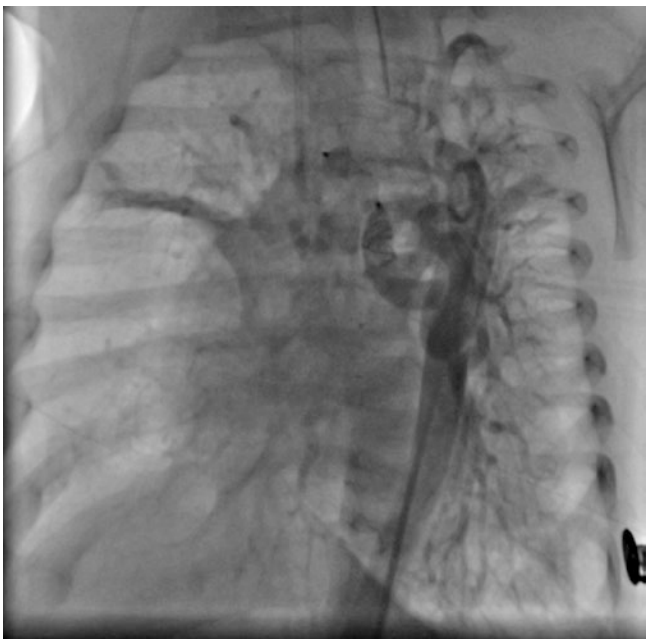


**Fig. 22.24** Patient 3. Repeat descending aortogram performed 10 min later, demonstrating no residual shunting across this collateral vessel

# Closure of Residual Postsurgical Defects

# 23

G. Kaleschke and H. Baumgartner

## 23.1 Anatomic Description and Physiopathology

Residual leaks occur in a wide anatomic variety after repair of heart defects and may not rarely be underdiagnosed especially after complex surgery.

*Residual defects* after *surgical ASD closure* (direct suture or patch) are rare and have been reported in 2–7% of patients. Defects may occur at any site but are most likely posterior-inferior where surgical closure is more demanding. Compared to native ASD, residual defects are more rigid. Again, inferior leaks are frequently complex and complicated by deficient rims making them not suitable for interventional closure. "Simple" residual ASDs result—when large enough (greater than 5–10 mm)—in significant left-to-right shunt and right ventricular volume overload. Inferior residual defects may cause right-to-left shunt in the 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 then 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 for a long enough time untreated. 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, 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. 23.1).

*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. Shunt direction und shunt portion depend on defect size and hemodynamics determined by ventricular filling characteristics and associated pathologies. Additional obstructions have particular impact and may cause right-to-left shunt. These patients are at risk for *paradoxical embolic events* or may be cyanotic and present with secondary *erythrocytosis*, but the latter findings are rare. Given the increased risk of thromboembolism after permanent pacemaker implantation, every patient after atrial switch operation should meticulously be examined with echocardiography and catheterization because smaller leakages may easily be missed. Intracardiac echo might facilitate leak detection especially in inferior defects where the accuracy of transoesophageal echo is limited. Defects can virtually occur at all sites of central venous return but are more often located at superior or inferior caval connection. 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.

G. Kaleschke · H. Baumgartner (✉)
Department of Cardiology III – Adult Congenital and Valvular Heart Disease, University Hospital Muenster, Cardiol, Muenster, Germany
e-mail: Helmut.Baumgartner@ukmuenster.de



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_23

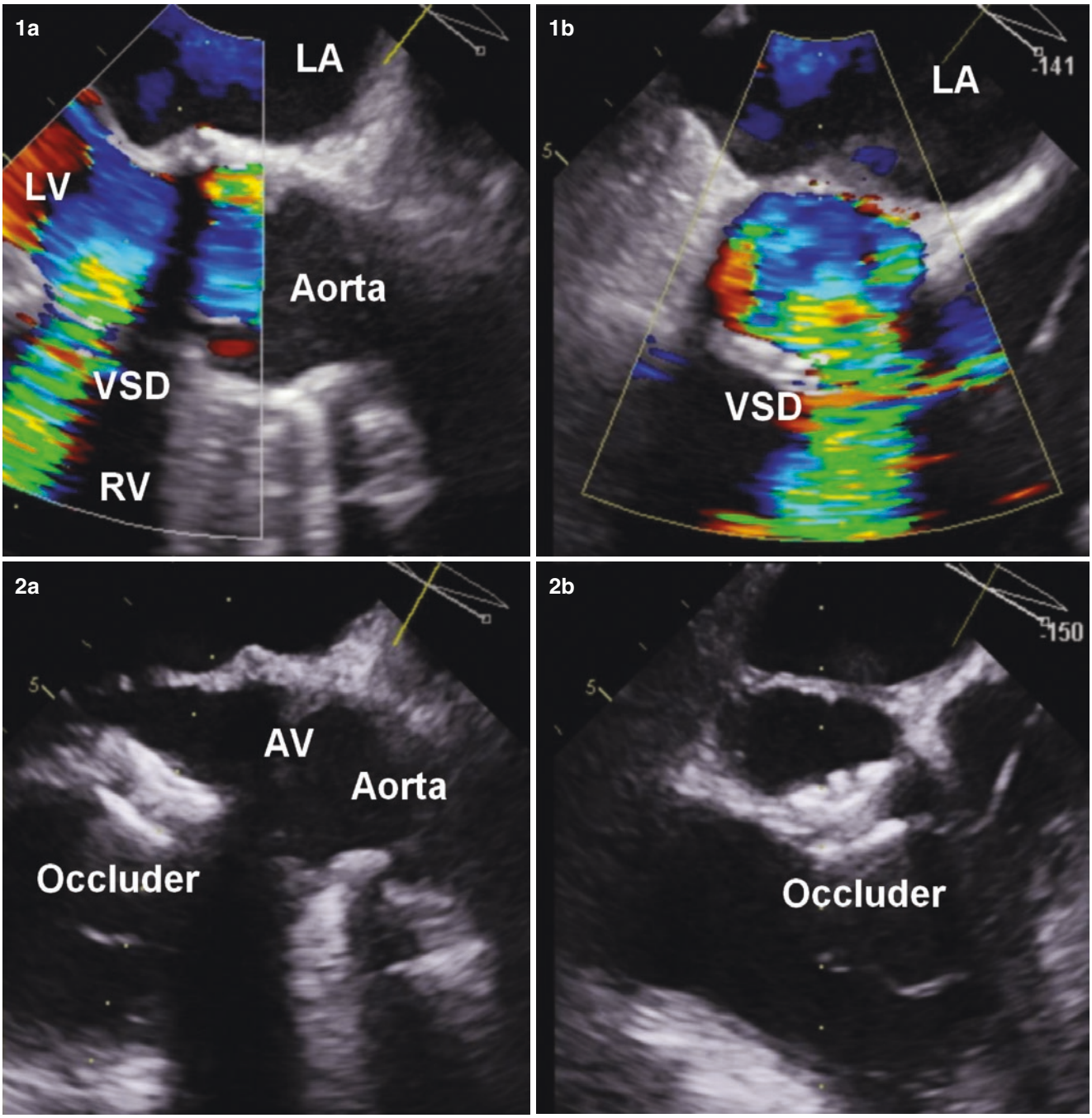



**Fig. 23.1** Example of postsurgical muscular VSD closure. Transesophageal echocardiography of a postsurgical muscular VSD in a 73-year-old male after recurrent aortic valve surgery and myectomy (Qp/Qs ratio 2, evidence of pulmonary hypertension, (**1a**) color Doppler long axis and (**1b**) short axis view visualizing the defect). A maximum defect diameter of 7 mm was measured by echo and angiography; direct retrograde closure could be achieved with an Amplatzer™ Muscular VSD Occluder 10 mm without residual shunting (**2a**: long axis view, **2b**: short axis view with implanted occluder)

## 23.2 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.

## 23.3 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*

## 23.4 Treatment Options

For most residual leaks after surgical ASD or VSD closure, self-centering double-disc devices or their derivatives (e.g., Amplatzer™ Abbott Santa Clara, CA. 95054 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).

## 23.5 Pre-procedural Imaging

Most appropriate information can be gained from *transesophageal echocardiography*. In residual ASDs native size of the defects, the 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 the right to the 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. In cases of insufficient visibility of the defect (inferior position, shadowing by mechanical valves etc) intracardiac echo provides attional information and might facilitate intervention. 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.

## 23.6 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., cephazoline) are generally recommended. Furthermore heparin is administered (70–100 U/kg, ACT 200-250s) 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 guidewire facilitates probing the leaks. In order to achieve a stable wire position that can be maintained during balloon testing of the defect and advancing the delivery sheath, the exchange to a stiff guidewire (e.g., Amplatz Extra Stiff, Cook Medical, IN, USA) is recommended.

*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

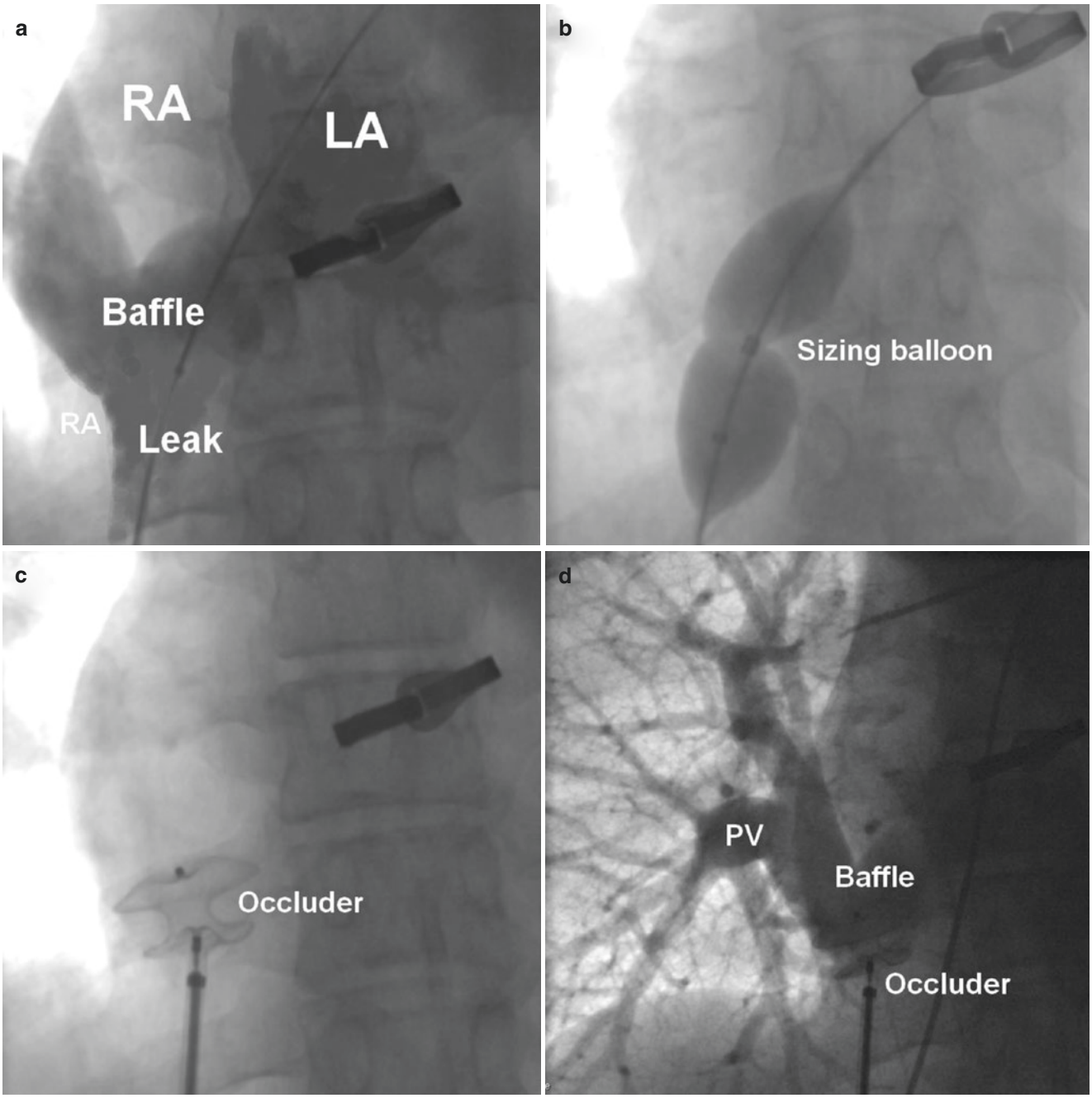



**Fig. 23.2** Example of baffle leak closure. 45-year-old female, correction of scimitar syndrome at the age of 10 years, recurrent embolic events (TIA, myocardial infarction without evidence of coronary artery disease, meningitis). Diagnosis of baffle leak based on contrast echo was confirmed during 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

meticulously before device release by echocardiography and angiography. Increase in wedge pressure (compared to the contralateral side) should be ruled out (Fig. 23.2). When covered stents are used for the treatment of leaks (especially in combination with baffle stenosis) balloon interrogation of the area with special regard to potential obstruction of the pulmonary venous return should be considered. Interference of the device with surrounding structures, mainly valves, must be excluded.

*Multiple devices*. If there are two or more adjacent leaks that necessitate the implantation of two devices, the smaller occluder should be placed first and initially being screwed upon the delivery cable, until the second occluder is in place. By doing so, correction of position remains possible, and optimal overlap of the discs can be achieved.

The same principles also apply for the intervention of postsurgical ASD and VSD. In the latter device type and size selection are 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; 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. Besides the Amplatzer™ VSD devices, the Duct Occluder 2 may be an option and fit within the aneurysm with its left disc.

## 23.7 Expected Results

Data about results and closure rates of interventional therapy in postsurgical ASD or VSD defects are sparse, 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.

## 23.8 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 discs 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, 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.

## 23.9 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. Laboratory tests of hemolysis have to be performed and repeated in cases of residual shunts especially when high pressure cavities are involved.

# ASD Closure in Special Situations: Elderly, PA-IVS

# 24

Giuseppe Santoro, Mario Giordano,
Maria Teresa Palladino, Carola Iacono, Gianpiero Gaio,
Marco Di Maio, Berardo Sarubbi,
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.

## 24.1 ASD Closure in the Elderly

Over time, ASD tends to progressively increase its hemodynamic burden due to pathophysiologic changes ensuing in the elderly. Indeed, physiologic decrease of left ventricular (LV) compliance and/or associated chronic diseases, such as aortic valve sclerosis, essential arterial hypertension, and coronary artery disease, tend to increase left-to-right atrial pressure gradient and hence atrial shunt. These pathophysiologic changes may cause progressive increase of pulmonary overcirculation as well as relative LV pre-load decrease and deconditioning, resulting in high risk of atrial arrhythmias and drop of systemic cardiac output.

Transcatheter approach allows to "test" the pathophysiologic consequences of defect closure by temporary balloon test occlusion (Fig. 24.1) which abolishes atrial shunt and suddenly decreases pulmonary blood flow in a high-resistant circulation. These changes may decrease pulmonary venous drainage and, hence, systemic cardiac output. At the same time, sudden increase of left chamber pre-load may be poorly tolerated by an under-trained LV (Fig. 24.2) and potentially result in pulmonary edema.

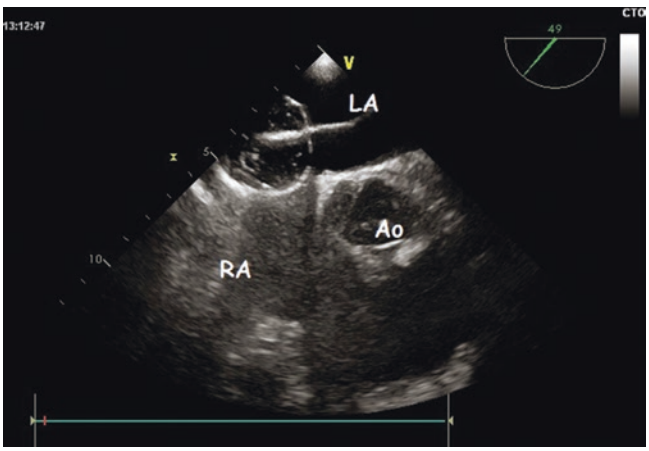



**Fig. 24.1** Temporary ASD closure in elderly patient to test the hemodynamic changes resulting from sudden left-to-right shunt disappearance. The occluding balloon is inflated into the right atrium and carefully advanced to the atrial septum. ASD dynamic balloon occlusion from the right atrial aspect (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 it is less interfering with the volume and compliance of the left heart chambers. *Ao* aorta, *LA* left atrium, *RA* right atrium

During balloon testing, relative contraindications to shunt closure are:

- Persistent increase of LV end-diastolic pressure (>20 mmHg and/or increase >50% compared to baseline)
- Decrease of systemic arterial pressure as high 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)

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_24) contains supplementary material, which is available to authorized users.

G. Santoro (✉)
Paediatric Cardiology and GUCH Unit, Fondazione Toscana-CNR "G. Monasterio", Massa, Italy

M. Giordano · M. T. Palladino · C. Iacono
G. Gaio · M. Di Maio · B. Sarubbi · M. G. Russo
Paediatric Cardiology and GUCH Unit, A.O.R.N. "Ospedali dei Colli", II University of Naples, Naples, Italy


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_24

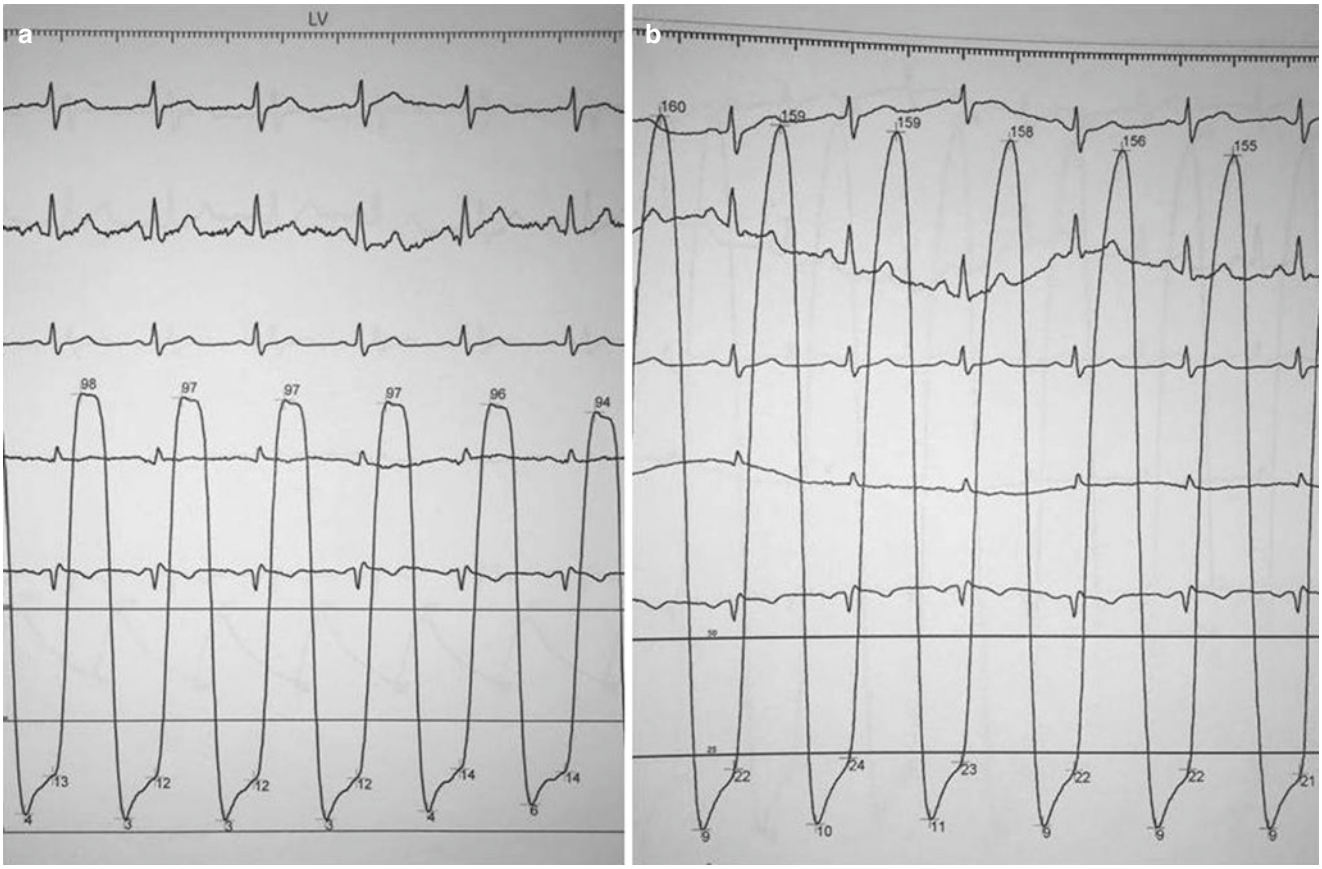



**Fig. 24.2** Left ventricular (LV) pressure tracing before (**a**) and during (**b**) temporary dynamic ASD balloon occlusion performed from the right atrium showing a significant increase of both proto- and end-diastolic pressure. These changes are almost universally recorded in elderly patients during test occlusion. However, in the vast majority of cases, they tend to normalize in a few minutes of balloon occlusion. If LV diastolic pressures continue to increase during balloon occlusion or do not show any trend toward normalization, a further test of ASD closure should be performed under diuretic and ino-lusitropic drugs

This subset of patients may benefit of:

- Partial ASD closure with a fenestrated device (Fig. 24.3). Fenestration may be obtained by perforating a self-centered occluding device using a Seldinger technique with 10–12 Fr femoral sheath, 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. The fate of fenestration is usually spontaneous closure within a few months.
- Pharmacologic trial with intravenous anti-congestive drugs (diuretics and ACE inhibitors) for 3–5 days or with oral drugs (diuretics and ACE inhibitors) for 3 months followed by further re-evaluation of the hemodynamic data during ASD balloon occlusion. Also in this setting, an improved but persistent borderline hemodynamic profile should advise for partial ASD closure with fenestrated devices.

Technical steps of transcatheter ASD closure in the elderly are not significantly different from what are widely described in younger patients. However, based on the previous pathophysiologic considerations, the procedure demands some additional tips:

- Adequacy of the respiratory pattern, obtained either with a comfortable position in the awake patient or optimizing the mechanical ventilation in anesthetized ones.
- Systemic artery and left ventricle pressure recording by small-size catheters both at baseline and during challenging tests.
- Right atrial, pulmonary artery, and pulmonary capillary wedge pressures as well as systemic pressure recording at the end of a quiet respiratory cycle. Superior vena cava, inferior vena cava, pulmonary artery, and femoral artery spO2 measurements in triplicate.
- Evaluation of pulmonary venous saturation through the ASD.

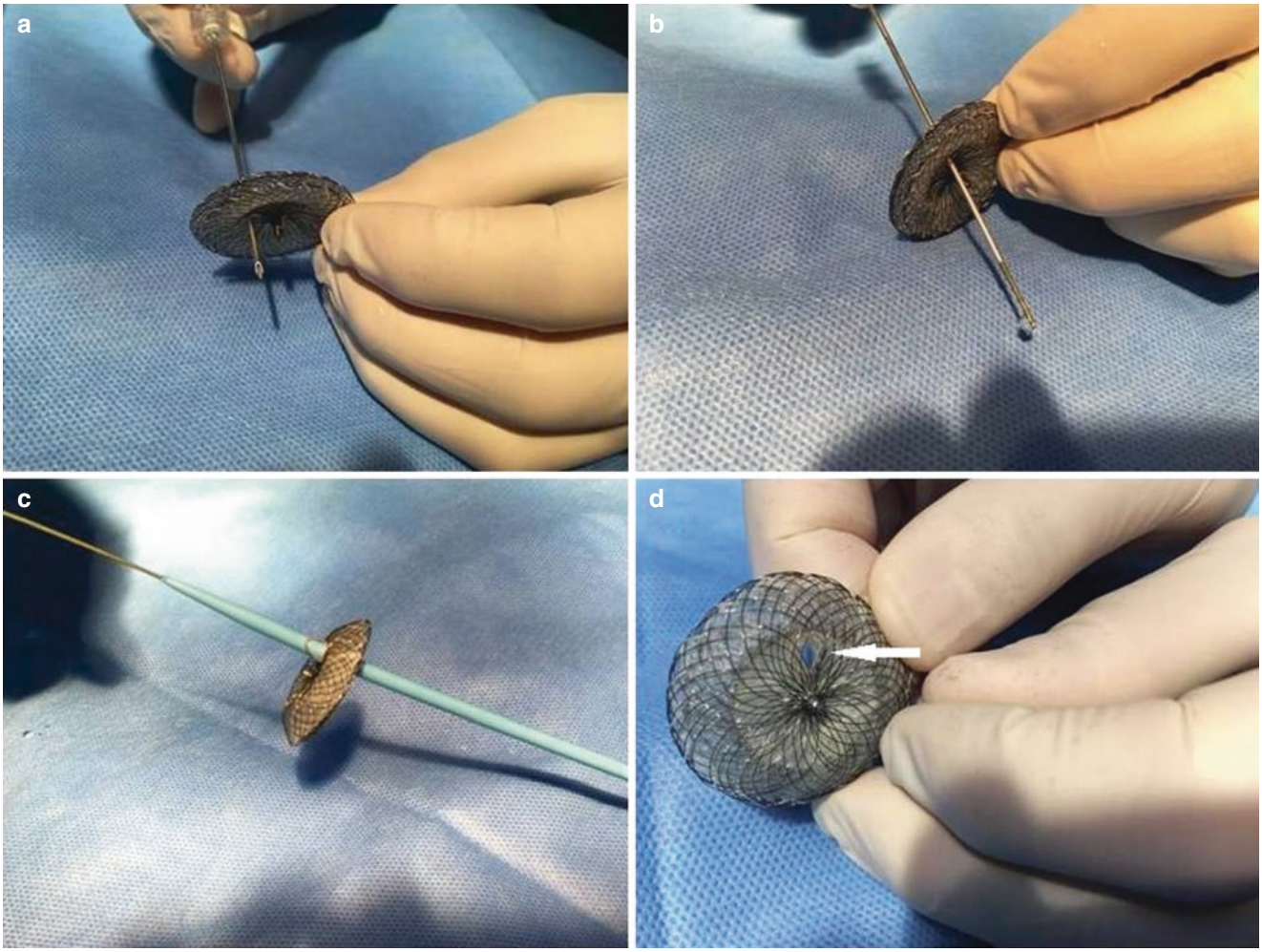



**Fig. 24.3** Handmade fenestrated ASD-occluding device. (**a**) The device is perforated close to the central hub using an 18 G Seldinger needle. (**b**) Then, the stiff end of a 0.035″ standard guidewire is passed through the needle. (**c**) Finally, the dilator of a standard 10 Fr introducer is firmly advanced over the wire. (**d**) As a final result, a 3–4-mm-large hole (arrow) is created

- Evaluation of potential coronary artery pathophysiologic abnormalities by coronary angiography and/or intracoronary pressure and/or flow recordings since any significant LV pre-load increase resulting from atrial shunt closure might potentially unmask subclinical, borderline coronary artery stenoses.
- Temporary balloon occlusion test for 15 min that mimes device deployment. In patients with borderline coronary artery stenoses, balloon occlusion is maintained for a longer time, looking for ischemic EKG changes or regional systolic/diastolic LV abnormalities.
- Careful evaluation of LV end-diastolic pressure and systemic arterial pressure before, during, and after balloon testing.
- Pulmonary artery pressure evaluation during and after balloon testing in subjects with high baseline values.
- ASD closure in elderly patients is performed following a routinely, well-described technique.

## 24.2 ASD Closure in Pulmonary Atresia with Intact Ventricular Septum

ASD or patent foramen ovale (PFO) is almost invariably present in the setting of pulmonary atresia with intact ventricular septum (PA-IVS) submitted to right chamber decompression by percutaneous or surgical valvotomy. Right ventricular (RV) hypoplasia and/or abnormal compliance almost always burdens on long-term pathophysiology of this malformation, resulting in right-to-left atrial (Fig. 24.4) shunt which may act as a safety valve either to unload right chambers or to increase 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.

ASD closure should be always performed in patients with PA-IVS submitted to RV decompression, avoiding the right-

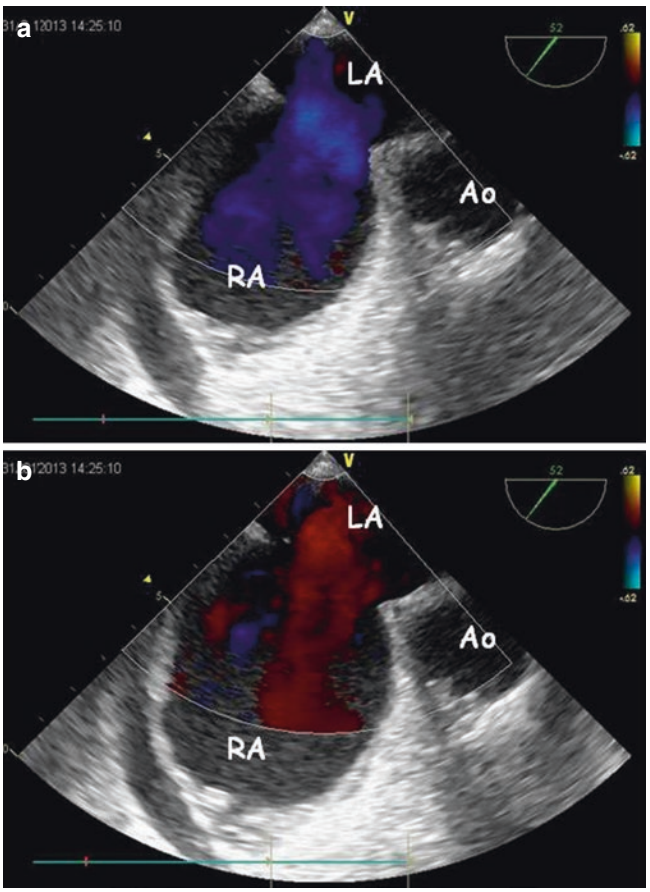



**Fig. 24.4** ASD closure in a patient with pulmonary atresia and intact ventricular septum (PA-IVS) submitted to right ventricular (RV) decompression. Bidirectional left-to-right (**a**) and right-to-left (**b**) ASD shunt as imaged at TEE color Doppler analysis. *Ao* aorta, *LA* left atrium, *RA* right atrium

to-left shunt caused by a borderline right chamber size/compliance that causes 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.

Technical steps of transcatheter ASD closure in these patients demand some tips:

- Complete right and left heart catheterization.
- Transesophageal echocardiographic and fluoroscopic monitoring.
- Balloon occlusion testing maintained for 15 min. Balloon occlusion test should be performed from the left atrium (Fig. 24.5) to avoid any interference with the right chamber volume and compliance. Therefore, dynamic balloon testing is preferable to static ASD occlusion.
- During test occlusion, the following parameters should be monitored: right atrial pressure (Fig. 24.6), systemic arterial pressure, and systemic oxygen saturation. Ideally, right atrial pressure should not increase >20%, systemic arterial pressure should not decrease >20%, and oxygen saturation should increase to >94% as compared to baseline values. In the 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. As an alternative, device fenestration could be considered.
- ASD/PFO closure in PA-IVS is performed following a routinely, well-described technique (Fig. 24.7).

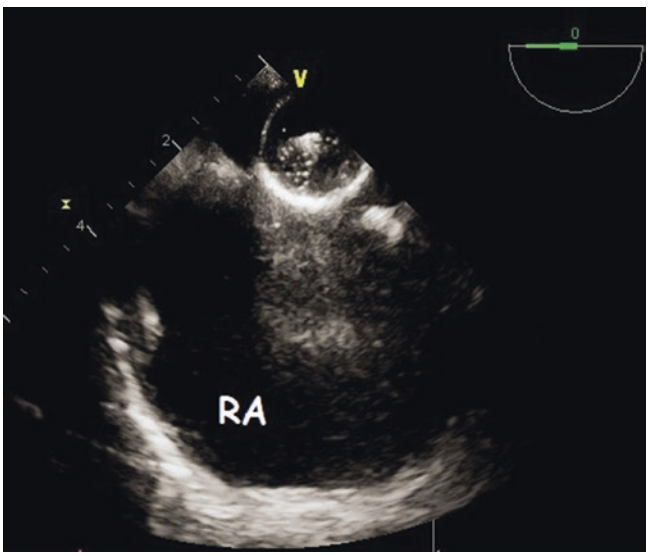



**Fig. 24.5** Temporary ASD closure in a patient with PA-IVS submitted to RV decompression to test the hemodynamic changes resulting from sudden right-to-left shunt disappearance. The occluding balloon is inflated into the left atrium and carefully pulled back toward the atrial septum in order to reduce the volumetric impact into the right atrium. *PA-IVS* pulmonary atresia with intact ventricular septum, *RA* right atrium, *RV* right ventricle

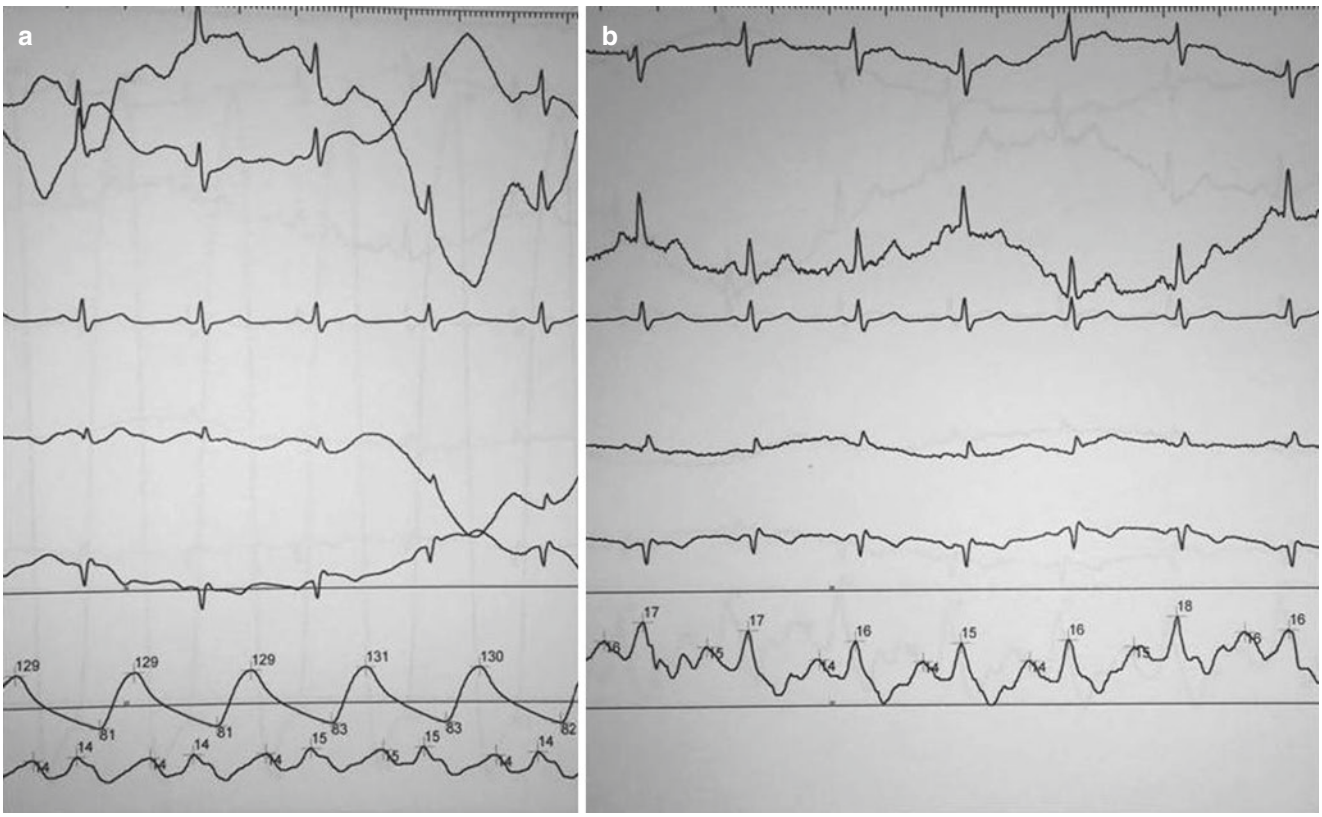



**Fig. 24.6** Right atrium (RA) pressure tracing before (**a**) and during (**b**) temporary dynamic ASD occlusion. Noteworthy, during ASD closure, RA pressure does not significantly change as a result of decrease of the right-to-left shunt and hence LV pre-load. Conversely, significant and persistent increase of RA pressure and/or drop of aortic pressure would preclude ASD closure

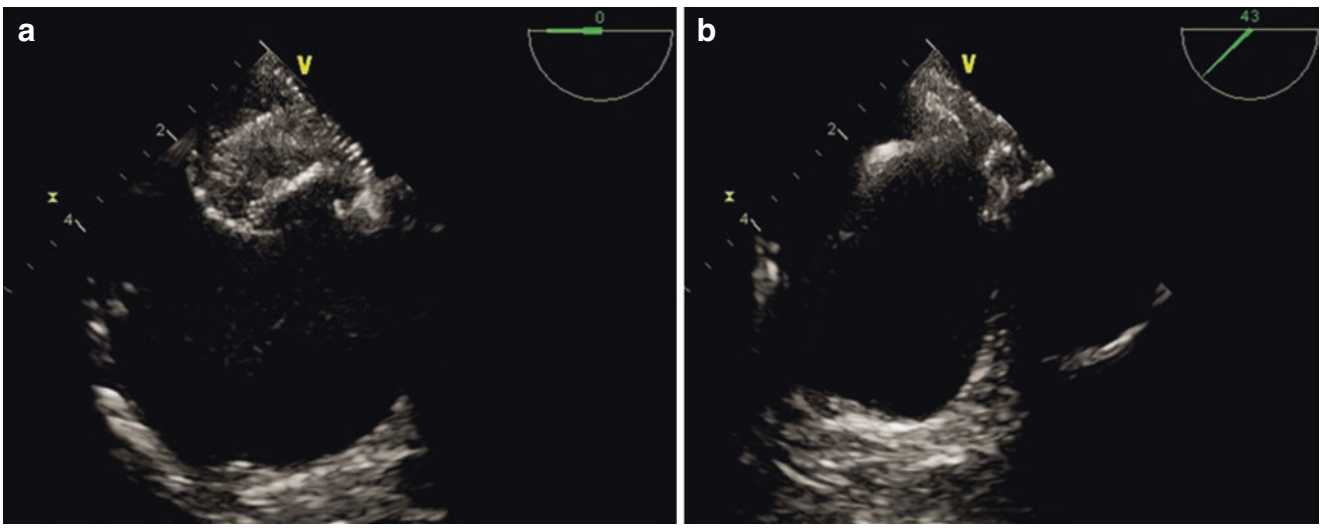



**Fig. 24.7** Steps of ASD occluding device implantation as imaged at TEE. (**a**) The left disk and the central waist of the device are sequentially opened in the left atrium and carefully approached to the atrial septum. As soon as the central waist fills the atrial septal defect, the right disk of the device is deployed in order to stabilize the device. (**b**, **c**) Then, a "push and pull" maneuver is performed to test device stability. (**d**) Finally, the device is released and any residual shunt is looked for at color Doppler analysis

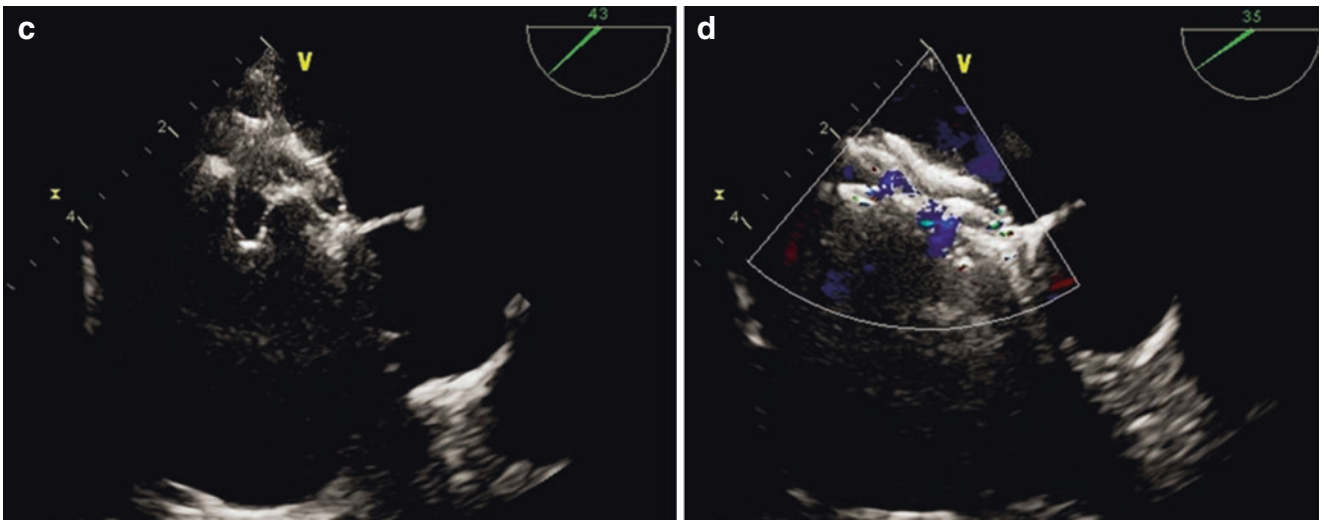



**Fig. 24.7** (continued)

**Video 1** Temporary ASD balloon occlusion in elderly patient to size the defect as well as to evaluate any hemodynamic change resulting from the sudden left-to-right shunt disappearance. The occluding balloon is inflated into the right atrium and carefully advanced to the atrial septum until the shunt disappears, as evaluated at color Doppler analysis (MP4 4507 kb)

**Video 2** Step-by-step description of percutaneous ASD closure as imaged at fluoroscopy. After ASD sizing, the left disk and the central waist of the device are sequentially opened in the left atrium and carefully approached to the atrial septum. As soon as the central waist fills the atrial septal defect, the right disk of the device is deployed in order to stabilize the device. Then, a "push and pull" maneuver is performed to test the device stability. Finally, the device is released (MP4 14243 kb)

**Video 3** Bidirectional shunt at atrial level in a patient with PA-IVS previously submitted to RV decompression by pulmonary valve perforation and angioplasty, as imaged at TEE color Doppler analysis (MP4 4664 kb)

**Video 4** Step-by-step description of percutaneous ASD closure, as imaged at TEE. After ASD sizing, the left disk and the central waist of the device are sequentially opened in the left atrium and carefully approached to the atrial septum. As soon as the central waist fills the atrial septal defect, the right disk of the device is deployed in order to stabilize the device. Then, a "push and pull" maneuver is performed to test the device stability. Finally, the device is released and any residual shunt is imaged by color Doppler analysis (MP4 6051 kb)

# Creating an Interatrial Communication

# 25

Derize E. Boshoff and Marc H. Gewillig

## 25.1 Indications for Creating an Interatrial Communication

*Creation or enlargement of interatrial communications may be important to:*

- Enhance mixing in patients with simple/complex transposition of the great vessels
- Augment cardiac output in case of obstructive lesions of the right side of the heart
- Off-load the right side of the heart in pulmonary vascular obstructive physiology
- Decompress the right atrium in postoperative right ventricular failure
- Relieve left atrial hypertension in left-sided obstructive lesions
- Relieve left atrial hypertension from the non-ejecting left heart during extracorporeal membrane oxygenation for circulatory support
- Lessen systemic venous hypertension, improve systemic perfusion, and perhaps relieve protein-losing enteropathy in patients with a failing Fontan circulation

## 25.2 Catheterization: General Principles

### 25.2.1 Vascular Access

- *Femoral vein*:
  - Access from right femoral vein allows straight passage to the fossa ovalis and into the LA. Left femoral venous access gives a sharp angulation between the left iliac vein and VCI, making transseptal crossing more difficult. Occlusion of the femoral veins may be considered as an anatomical limitation for transseptal access.
- *Jugular vein*:
  - Atrial septal interventions need an access that will be perpendicular to the plane of the atrial septum [1]. Jugular access will only achieve a perpendicular approach to the atrial septum with special (steerable) sheaths and special puncture techniques (perforating wire, radio frequency).
- *Umbilical vein*:
  - Is a good alternative in the newborn baby to perform balloon atrial septostomy but obliterates due to fibrosis a few days after birth. The tortuosity from the umbilicus to atrial septum through the venous duct and left hepatic veins may pose difficulty for septal crossing. It cannot be used for transseptal puncture or stenting [1].
- *Transhepatic access*:
  - Can be used safely even in small infants
- *Hybrid surgical access*:
  - If no vascular access is available, hybrid surgical access (via thoracotomy or sternotomy) through a double-layered purse string suture around the right atrial wall may be required [1].

### 25.2.2 Procedure

- Should be performed under *general anesthesia* (except routine atrial septostomy in newborns in the neonatal intensive care unit under echocardiographic guidance).
- *Prophylactic antibiotics* and *heparin sulfate* (100 units/kg) should be given intravenously. If radio-frequency (RF) transseptal perforation is performed, heparin should be given after entering the left atrium.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_25) contains supplementary material, which is available to authorized users.

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



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_25

## 25.3 Imaging Techniques

### 25.3.1 Fluoroscopy

*Biplane fluoroscopy is preferred in*:

- A very large or very small atrium
- A large dilated aortic root
- No vena cava inferior access to the atrial septum
- Any abnormal cardiac chamber or great vessel positional abnormalities

*Angiographic projections*:

- Different projections have been described in chapter # to best visualize the interatrial septum during transseptal procedure.

### 25.3.2 Echocardiography

*Transthoracic echocardiography* (*TTE*):

- Permits visualization of the interatrial septum and the adjacent structures
- Has a limited role in guiding complex transseptal catheterization (i.e., stent implantation) due to poor image quality, difficulty to identify the fossa ovalis correctly, and disruption of the sterile field

*Intracardiac echocardiography* (*ICE*):

- The fossa ovalis can be accurately located using ICE.
- Limitations are sheath size, additional puncture in the femoral vein, possible longer procedural time, and significantly higher costs.

*Transesophageal echocardiography* (*TEE*):
TEE is the modality of choice in addition to fluoroscopy, particularly to visualize

- A specific area of the fossa ovalis to be punctured
- The thickness of the septum at that point
- The degree of anterior-posterior direction of the intended puncture
- In case of complex anatomy when stent implantation is performed

*TEE with 8-French AcuNav* (*ACUSON AcuNav*, *Siemens Medical Solutions*, *USA*) *probe*

- Can be used transesophageally in small infants (limited reports).
- It is a monoplane probe and does not have an attached thermistor, but the quality of the pictures seems to be sufficient, and thermal damage in the esophagus does not seem to be an issue.

## 25.4 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.

### 25.4.1 Balloon Catheters

BAS catheters are available from various manufacturers and in different designs. Currently, there are four different catheters that can be used for this purpose:

- *The Miller-Edwards catheter* (*Edwards Lifesciences*)
  - Single-lumen catheter, 5-Fr shaft, requires a 7-Fr sheath.
  - 35° hockey stick angle 2 cm from the tip (allows easy entry into the LA).
  - Compliant latex balloon, accepts 4–5 mL of fluid (balloon diameter 17–18 mm).
  - Large balloon inflations are required to perform a septostomy (disadvantage, especially in small infants <3 kg, or a small LA).
- *The Rashkind balloon catheter* (*USCI-CR Bard*)
  - Recessed, low-profile balloon requires 6-Fr sheath.
  - Accepts 1.5 mL of contrast (balloon diameter 12–13 mm).
  - Larger volumes will elongate the balloon without increasing the diameter.
- *The Fogarty* (*Paul*) *balloon catheter* (*Edwards Lifesciences*)
  - Introduced via a 6-Fr sheath.
- *The NuMED Z-5 Atrioseptostomy catheter* (*NuMED*)
  - End hole can be advanced over a 0.014/0.018-in. wire (5-Fr/6-Fr sheaths).
  - Can confirm position by injecting contrast into LA.
  - Balloon sizes of 1 mL (9 mm diameter) and 2 mL (13.5 mm diameter).
  - Noncompliant nature and relatively small catheter size offer advantages when performing BAS in patients with a small LA size (i.e., HLHS).
  - Radiopaque marker is located in the midportion of the balloon; wrapped balloon extends beyond the end of the catheter shaft.
  - Operator should avoid pushing stiff tip against the LA wall or appendage when advancing the catheter (can induce atrial tachycardia) [2].

## 25.4.2 Procedure

### 25.4.2.1 Tips When Using the Umbilical Venous Approach

*Fluoroscopy*:

- AP projection: catheter passes from the right of the midline superiorly toward the RA.
- Lateral projection: catheter passes from front to back.

*In case of stenosis or closure of the ductus venosus*:

- A 4-French end-hole catheter with 0.018″ guidewire can be introduced into the umbilical vein and manipulated into the RA.
- Then exchange for appropriate-sized sheath to introduce the septostomy catheter.

*Using a sheath in the umbilical vein*:

- The tip of the sheath is often inside the RA.
- May impede withdrawal of the inflated balloon across the septum.
- Must be withdrawn into the ductus venosus before performing the septostomy.

### 25.4.2.2 Performing the Septostomy

- Balloon is positioned in the LA (fluoroscopy and/or echocardiography).
- 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).
- 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.
- The stopcock is closed and the balloon advanced 1–2 mm of the atrial septum.
- Balloon is jerked/pulled briskly to the RA/VCI junction.
- Balloon is subsequently advanced promptly to the mid RA and deflated as quickly as possible.
- Care must be exercised as to how vigorously the balloon is pulled into the VCI.

**Video 1** The septum is easily crossed with a Fogarty catheter (AVI 1797 kb)

**Video 2** Balloon septostomy is performed: balloon volume is 1.5 mL (AVI 596 kb)

- The process is repeated at least once until there is no resistance to passage of the full balloon across the defect.
- Gradient across the septum may be measured; if still significant, BAS may be repeated.
- Echocardiography with Doppler assessment of the residual gradient may be used to determine the adequacy of the septostomy.

### 25.4.2.3 Tips for Crossing the "Difficult" Septum

- Direct advancement of the pre-shaped catheter is successful in most cases.
- Advancing a sheath across the interatrial septum may facilitate passage of the septostomy catheter [Cordis 6-FR BRITE TIP sheath (Cordis Corp., Miami, FL) has a smooth transition to pass over a 0.018-in. guidewire into the left atrium]. The sheath should be pulled back into the VCI prior to performing the BAS.
- Advancing a low-profile balloon (i.e., NuMED Tyshak Mini (NuMED, Hopkinton, NY) across the septum may allow predilation of the interatrial communication to subsequently allow passage of the septostomy catheter.

## 25.4.3 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).

*Brockenbrough needle*:

- May be cumbersome for transseptal puncture in patients with complex anatomy or a small LA (HLHS and variants)
- Has the potential risk of atrial perforation

*Nykanen radio-frequency (RF) perforation wire and coaxial injectable catheter (180-cm 0.035-in. outer diameter) (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.

**Video 3** Needle septostomy in a patient with complex DORV; 0.014″ guidewire placed in the LA (AVI 2594 kb)

## 25.5 Blade Atrial Septostomy

### 25.5.1 Indications

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 BAS or for surgical atrial septostomy that otherwise would be needed in the older infant.

### 25.5.2 Blade Septostomy Catheters (Cook, Bloomington, IN)

- Available in three blade lengths; sheath is one size larger than the catheter size for smooth introduction:
  - *0.94 cm* (*PBS 100*, *6F catheter*, *7F sheath*)
  - *1.34 cm* (*PBS 200*, *6F catheter*, *7F sheath*)
  - *2.0 cm* (*PBS 300*, *8F catheter*, *9F sheath*)
- 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 (Fig. 25.1).

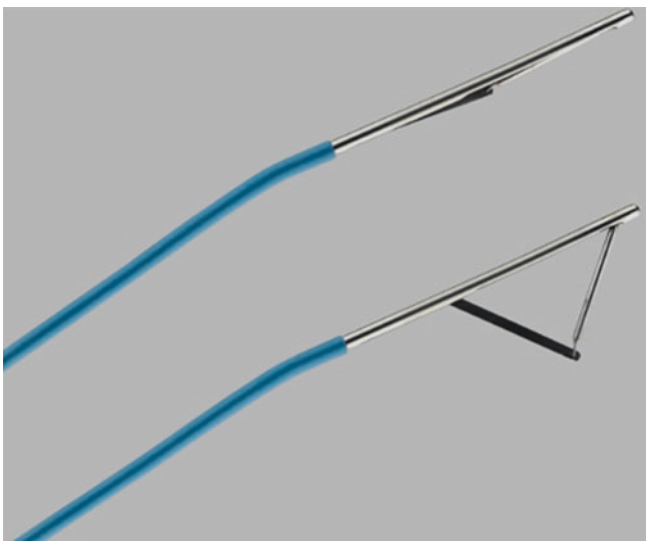


**Fig. 25.1** The blade should be tested outside the patient to be sure it opens and closes fully without resistance

### 25.5.3 Procedure

- The blade catheter is advanced through a long Mullins sheath into the LA; the sheath is withdrawn well into the VCI.
- The blade is opened in the LA (continuously observed on fluoroscopy and TEE).
- The tip is directed anteriorly and either to the patient's right or left side.
- The blade catheter is withdrawn slowly in a controlled maneuver.
- Resistance may be considerable, so bracing one's hands against the patient's leg (and pulling with the fingers) may prevent sudden retraction of the open blade down the VCI.
- The introducer sheath must be withdrawn simultaneously with the septostomy catheter to prevent possible cutting of the distal tip of the sheath.
- If the septum is 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 and changing the blade direction 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 be avoided in patients with complex anatomy and small LA size (combination of cutting balloon septostomy and static balloon septostomy is a safer alternative; stenting is safer with a more predictable and longer lasting result).

## 25.6 Cutting Balloon Septostomy

### 25.6.1 Indications

In patients with a thickened interatrial septum, the combination of static cutting balloon septoplasty (Boston Scientific, Boston, MA), followed by the use of larger diameter static balloons or standard balloon atrial septostomy, is a valuable alternative to blade atrial septostomy. 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.

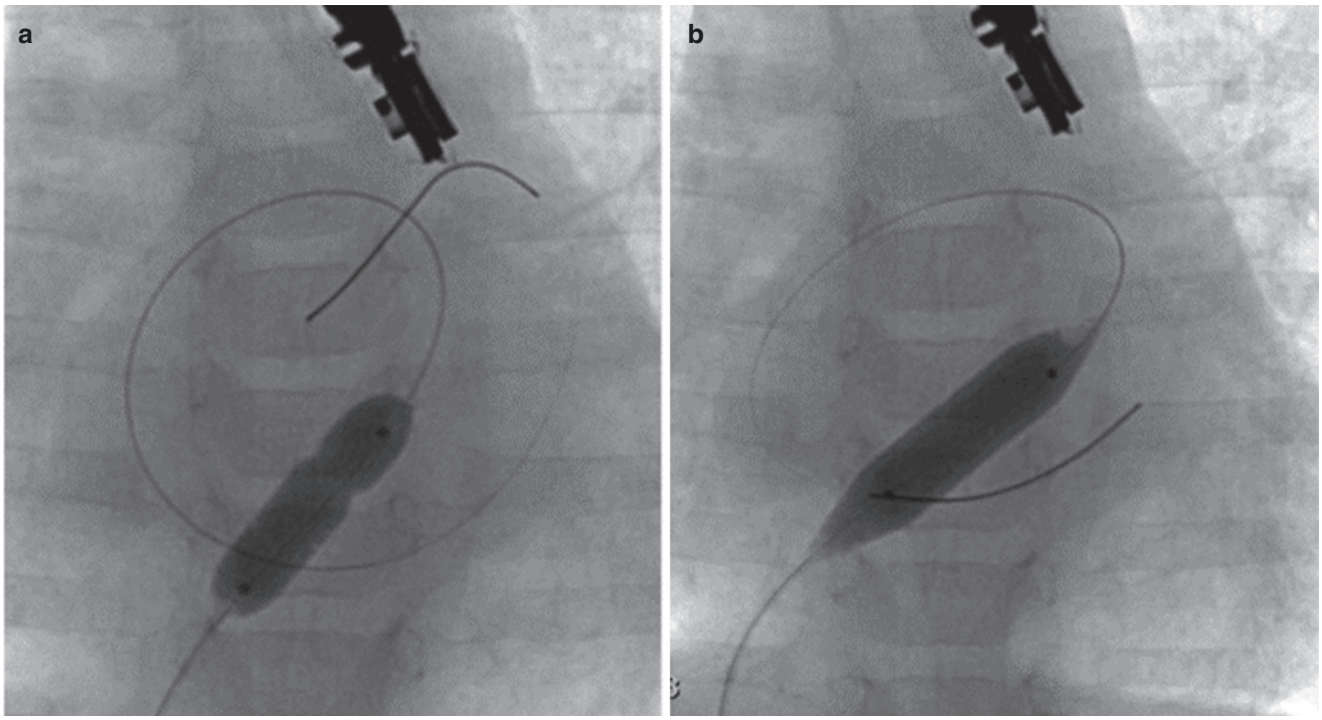



**Fig. 25.2** (**a**) Static balloon dilation with 7 mm Tyshak balloon shows recoil at deflation. (**b**) Balloon dilation with 8 mm cutting balloon: full expansion at six atmospheres. The wire has been curled in the body of the LA

### 25.6.2 Procedure

- 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).
- The wire is positioned in the left upper pulmonary vein or alternatively curled in the body of the LA (Fig. 25.2).
- Inflate and deflate slowly (use screw of indeflator) in order to prevent the blades to cut the balloon itself; rotation of the cutting balloon followed by repeat inflations may tear the interatrial septum in different locations and improve the response to static balloon septoplasty.
- The smaller the preexisting septal defect, the higher the likelihood that the use of a cutting balloon will achieve an adequate result.
- If the existing interatrial communication is "stretchable" (i.e., floppy valve), cutting balloon dilation will be inefficient. In this situation, it may be better to perform a transseptal puncture and start with a new diminutive opening to obtain a better result with cutting balloon septoplasty.

## 25.7 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. Balloon diameter will depend on patient/atrial size and underlying cardiac anomaly (Fig. 25.3). In some patients, predilation of the septum after needle puncture is necessary before advancing of the long sheath needed for subsequent stent implantation (Fig. 25.4).

## 25.8 Stent Implantation in Congenital Heart Defects: Nonrestrictive Technique

### 25.8.1 Indications

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. Atrial stent septostomy can provide a reliable long-lasting restrictive or nonrestrictive interatrial communication (Video 4).

### 25.8.2 Procedure

The 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.

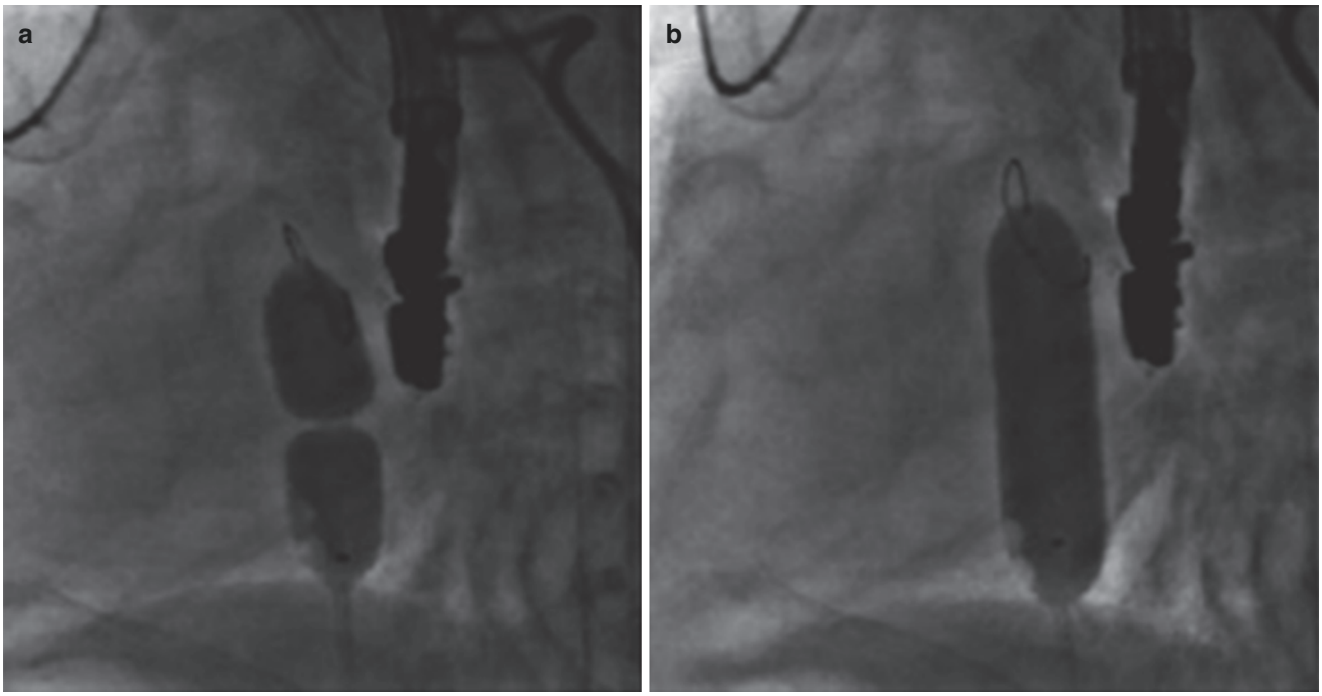



**Fig. 25.3** Static balloon dilation of restrictive atrial communication (**a**) 12 mm balloon inflated at mild pressure delineating the small atrial communication (**b**) full inflation stretches-tears the septum

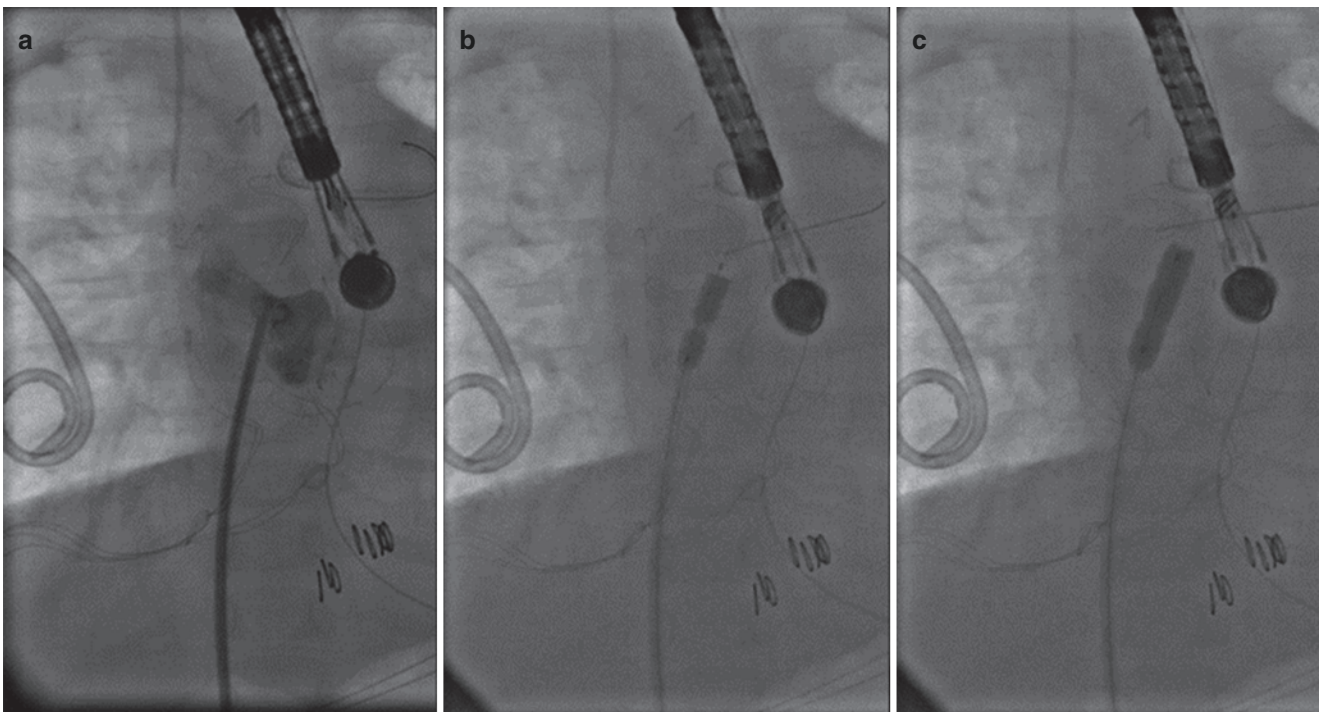



**Fig. 25.4** Patient with complex DORV, sub-PS/PS, intact IAS. (**a**) Long sheath cannot be advanced after needle puncture (**b**) predilation with a 6/20 mm Maverick balloon. (**c**) Balloon fully inflated. Long sheath could subsequently be advanced, followed by stent implantation

**Video 4** Static balloon dilation shows recoil: stent implantation can provide a reliable long-lasting nonrestrictive interatrial communication (AVI 1765 kb)

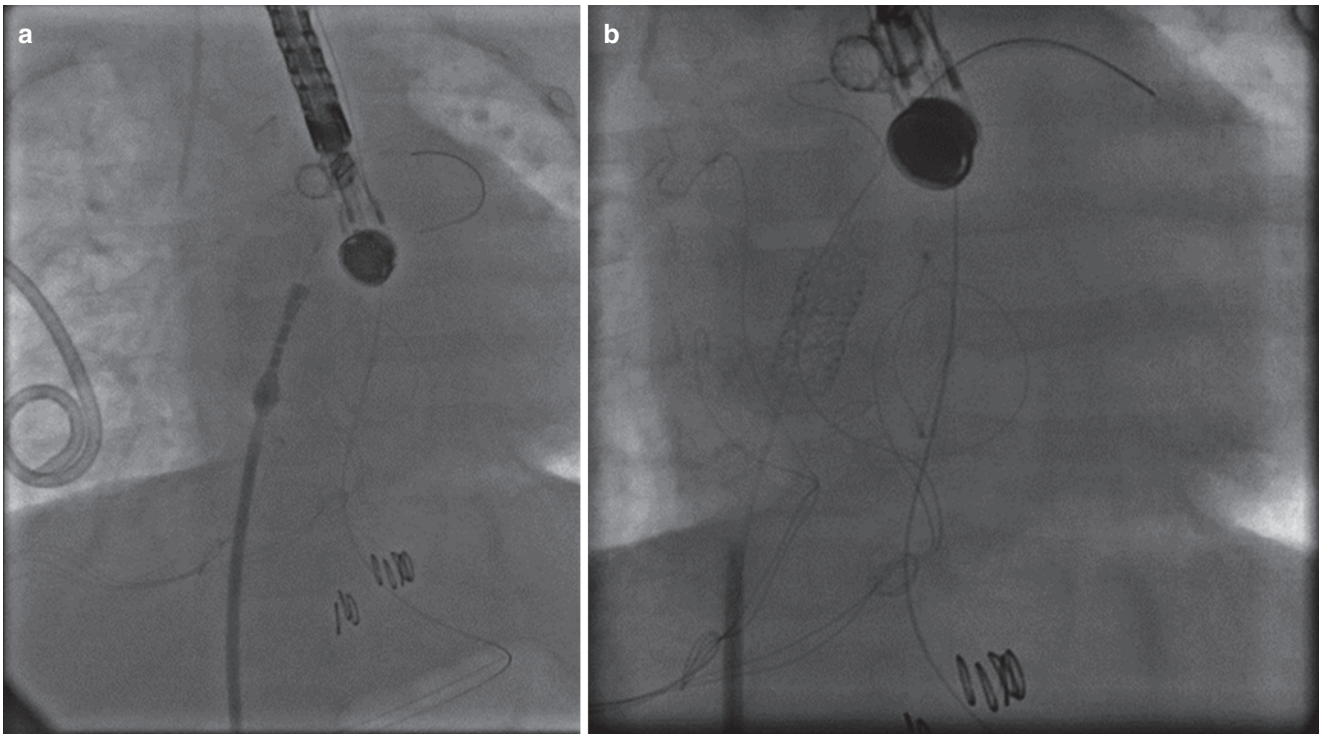



**Fig. 25.5** Patient with complex DORV, sub-PS/PS (**a**) implantation of a 7/16 mm Cook Formula premounted balloon-expandable stent. (**b**) Stent fully expanded

**Video 5** Implantation of a 10/30 mm Sinus-Superflex self-expandable stent in the IAS. Shorter stent would have been better (10/20 mm) (AVI 2189 kb)

*Type of stent*:

- Premounted balloon-expandable stents are preferable in this setting (Fig. 25.5).
- Self-expandable stents have been used with success in some patients (Video 5; Fig. 25.6).

*Stent diameter depends on*:

- The age and size of the patient
- The type of congenital anomaly (especially atrial size)

*Stent length*:

- The stent should be long enough to allow adequate stabilization within the interatrial septum and minimize the risk of embolization due to movement during inflation or foreshortening after expansion.
- The stent should not be too long nor big due to the risk of atrial erosion, thrombus formation, and obstruction of the pulmonary veins (Video 5).

*Technique of sequential stent flaring is outlined stepwise in Fig. 25.7*:

- Adequacy of stenting should be assessed using TTE or TEE (Fig. 25.8).
- Anti-aggregation should be given after stent implantation to prevent thrombus formation (acetylsalicylic acid 2–5 mg/kg/day)

## 25.9 Stenting of the Interatrial Septum: Restrictive Technique

### 25.9.1 Indications

#### 25.9.1.1 Severe Pulmonary Arterial Hypertension

Atrial septostomy improves cardiac index and functional class by the creation of a right-to-left atrial shunt, delays dystrophy, and cardiac cachexia while on the transplant waiting list and may even improve the survival in some patients. The

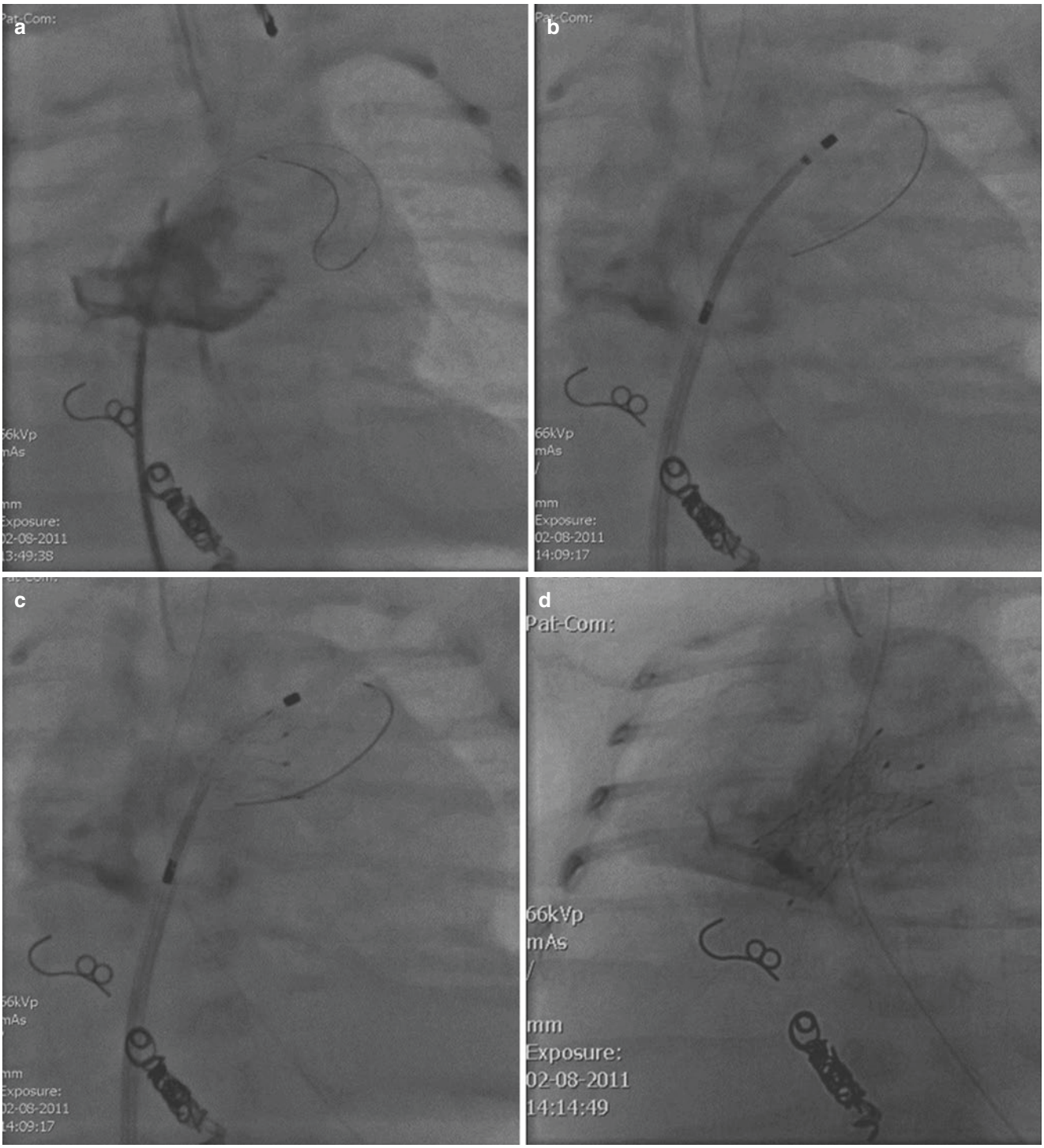



**Fig. 25.6** Patient with UVH, DORV, MS, small LA, and sub-PS/PS. (**a**) Position of the sheath is confirmed with a small contrast injection in the LA. Residual staining of the septum and pericardiac space can be seen. Needle puncture was technically difficult due to the small LA size. (**b**) 10/20 mm Optimed sinus-SuperFlex Visual self-expandable stent is in position and ready to be deployed. (**c**) Distal end of stent has been deployed; repositioning is still possible. (**d**) Stent fully deployed

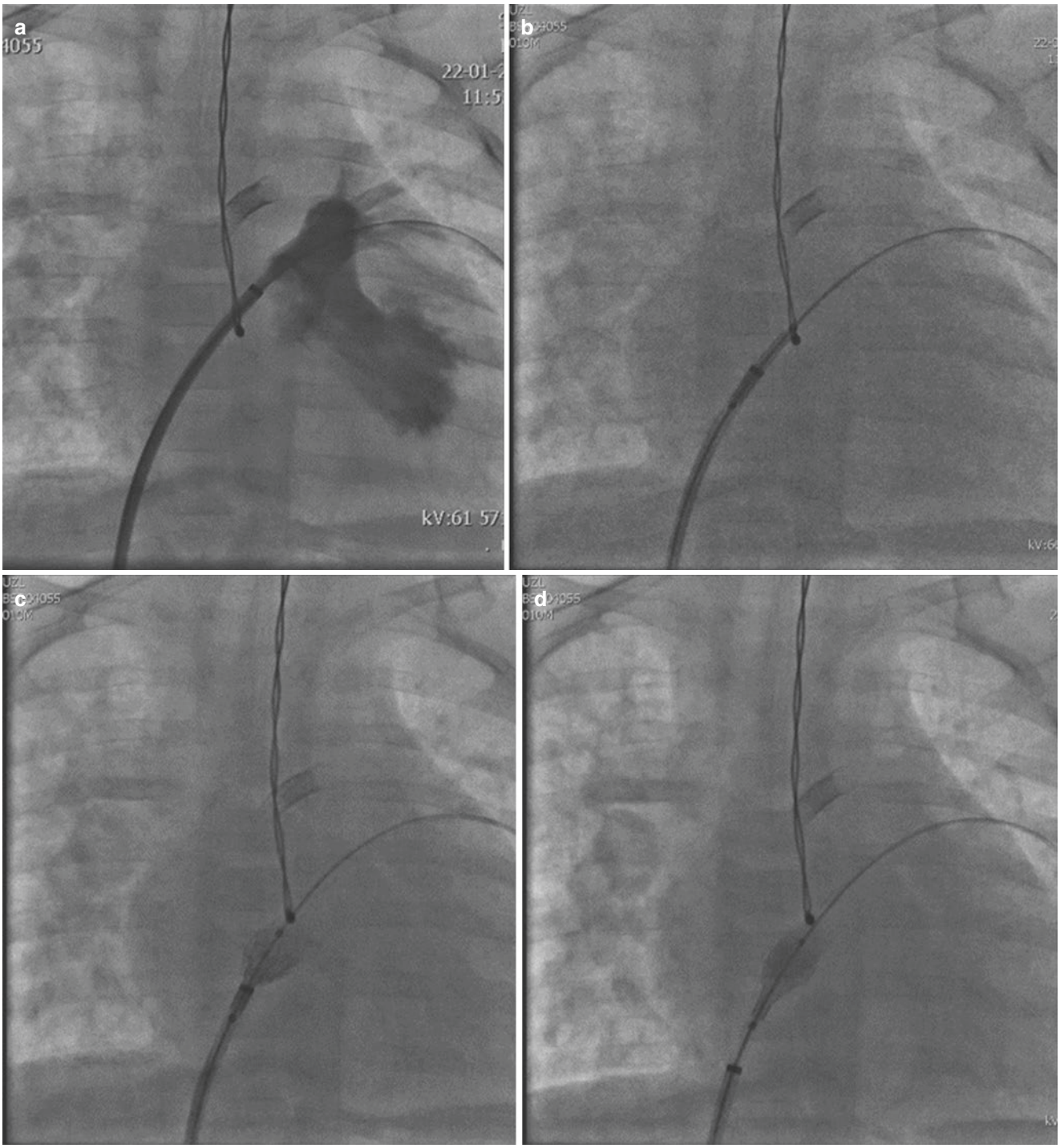



**Fig. 25.7** Patient with DORV, TGA, and PS. Technique of sequential stent flaring is used to implant a 10/17 mm Valeo stent in the IAS. (**a**) 0.035″ Amplatz wire has been positioned in the left upper pulmonary vein. Small contrast injection is given via the 7F Cook sheath to confirm position in LA. (**b**) Half of the stent is exposed by pulling back the sheath. (**c**) The balloon is inflated in the LA, expanding the distal half of the stent. The pressure in the balloon is maintained using a stopcock. The entire system is firmly pulled back against the atrial septum. (**d**) The pressure in the balloon is slightly released, allowing the RA portion of the stent to be unsheathed. (**e**) The balloon is then fully inflated, opening the proximal portion of the stent. (**f**) The deflated balloon should be removed carefully out of the stent into the long sheath, avoiding stent dislodgment. The newly implanted stent should not be crossed unless certainty of adequate fixation

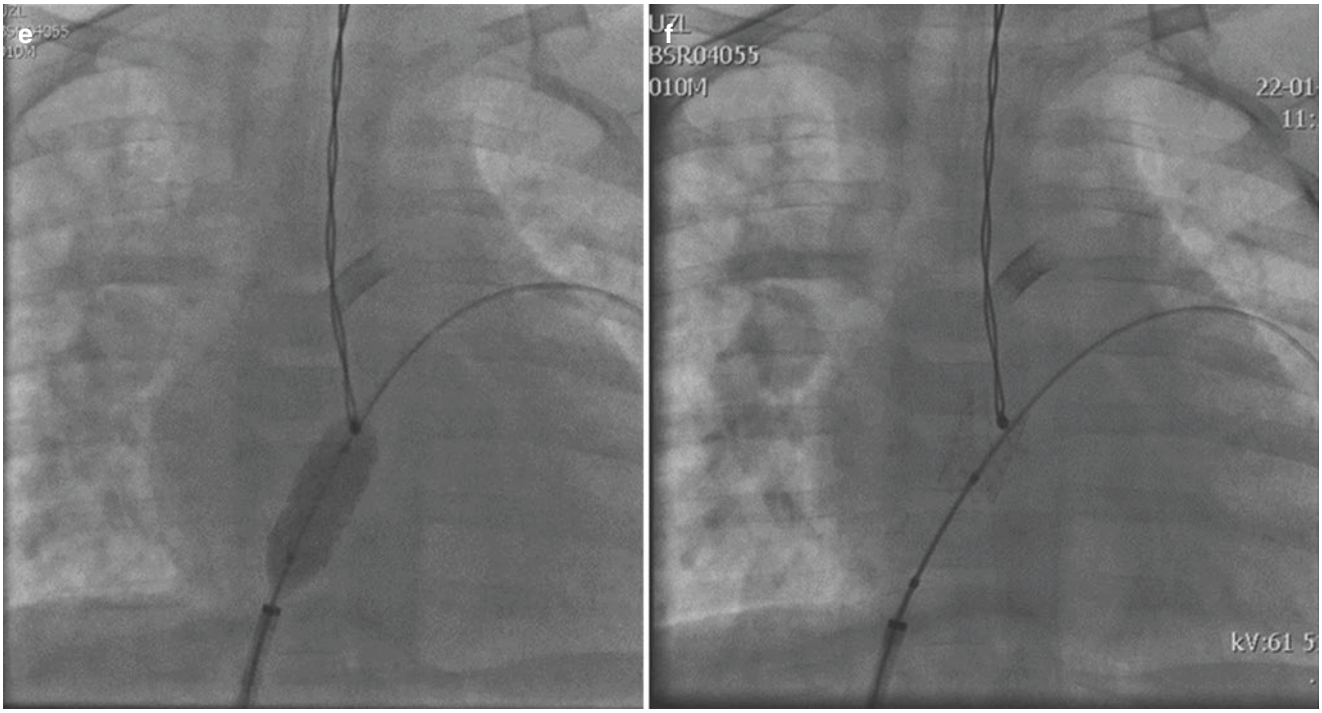



**Fig. 25.7** (continued)

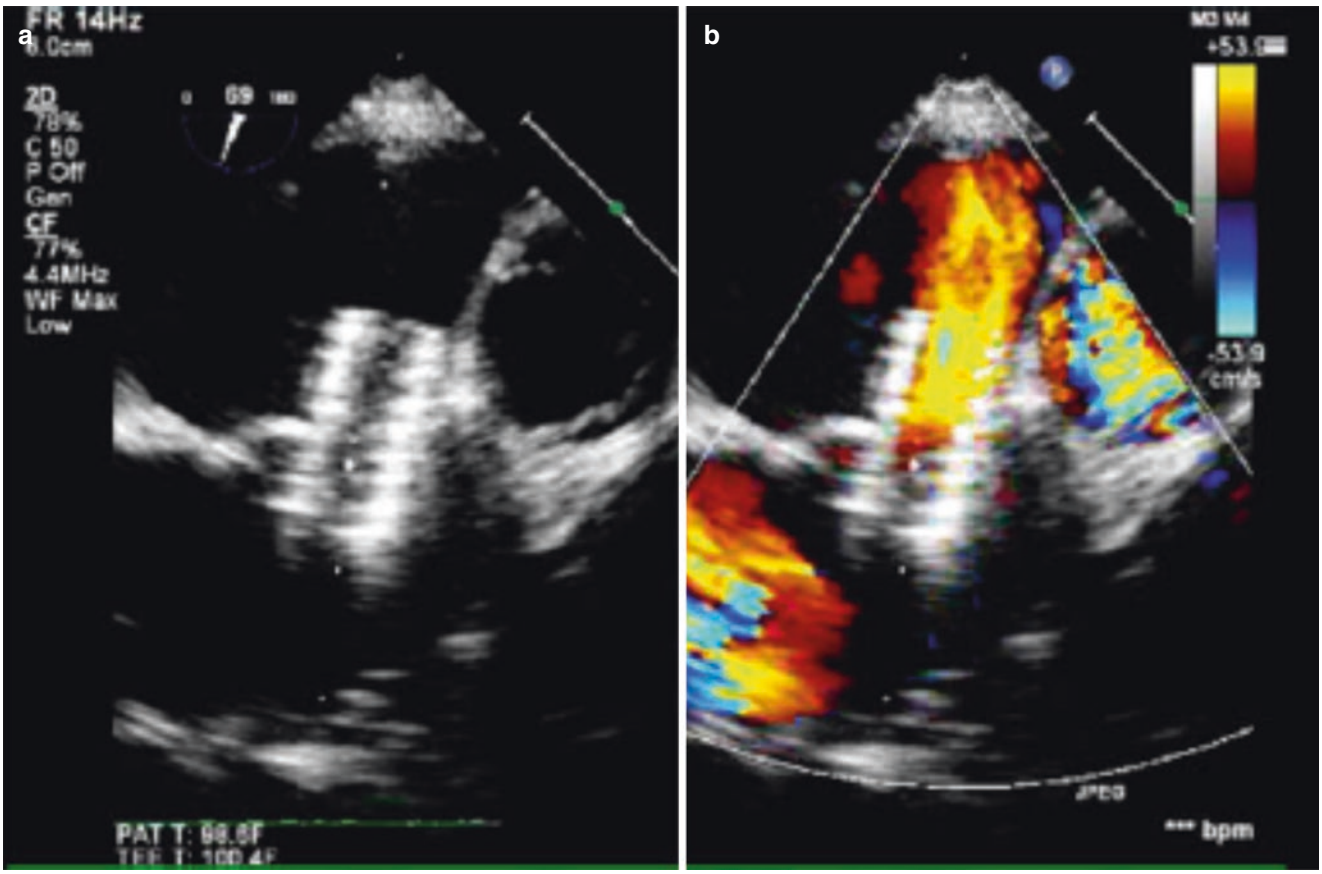



**Fig. 25.8** TEE of stent across atrial septum. (**a**) Stent is nicely positioned across the atrial septum. (**b**) Color flow mapping shows right-to-left shunt across the stent

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.

#### 25.9.1.2 Failing Fontan Circulation

Secondary fenestration is a valuable technique to improve the hemodynamic condition of these patients. 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 [3].

### 25.9.2 Fenestration Technique

The fenestration technique currently used in our unit has been adapted from [4], who described a small mixed series of primary PAH patients and patients with a failing Fontan circulation.

#### 25.9.2.1 Septal/Conduit Puncture

*Pulmonary arterial hypertension*:

- A venous sheath up to 12 Fr is placed into the right femoral vein.
- Puncture of the interatrial septum with a Brockenbrough needle.

*Extracardiac Fontan circuit*:

- Fenestration may be more challenging, due to separation of the different wall layers during needle puncture and sheath placement [5].
- The optimal perforation site is the point that has the most acute angle coming from the VCI (exceptionally the VCS) and which is in contact with the atrium. We prefer puncture of the VCI just below the conduit (Fig. 25.9).
- Puncturing the conduit with a Brockenbrough needle may require considerable force and consequently adequate immobilization of the patient.
- 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) (Video 6 and Fig. 25.10).

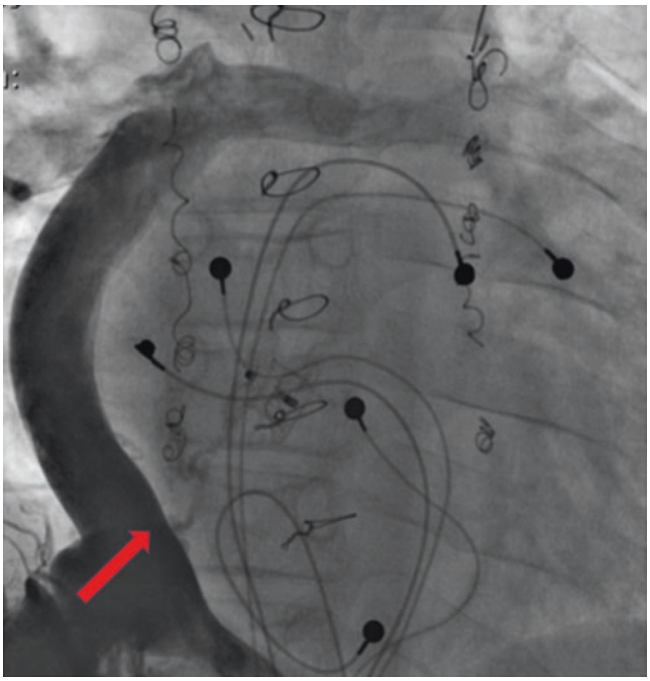


**Fig. 25.9** The optimal perforation site is the point that has the most acute angle coming from the VCI and which is in contact with the atrium. We prefer puncture of the VCI just below the conduit (red arrow)

**Video 6** Patient with a failing Fontan circulation. Restrictive stenting of the conduit is intended. The Gore-Tex wall has been crossed, and the atrial wall has been tagged with some contrast overflow in the pericardial space. The needle is advanced until it pops through the atrial wall (AVI 129796 kb)

- A 0.014-in. coronary wire is advanced preferably into the left upper pulmonary vein.
- Advancing the dilator and sheath over the Brockenbrough needle may 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.

#### 25.9.2.2 Stent Preparation (Fig. 25.11)

- A loop of 3–5 mm diameter is created using stiff wire (a set of temporary epicardial pacing wires or surgical wire).
- Epicardial pacing wire: the needle ends and distal 5 cm length of isolative coating are removed, allowing making

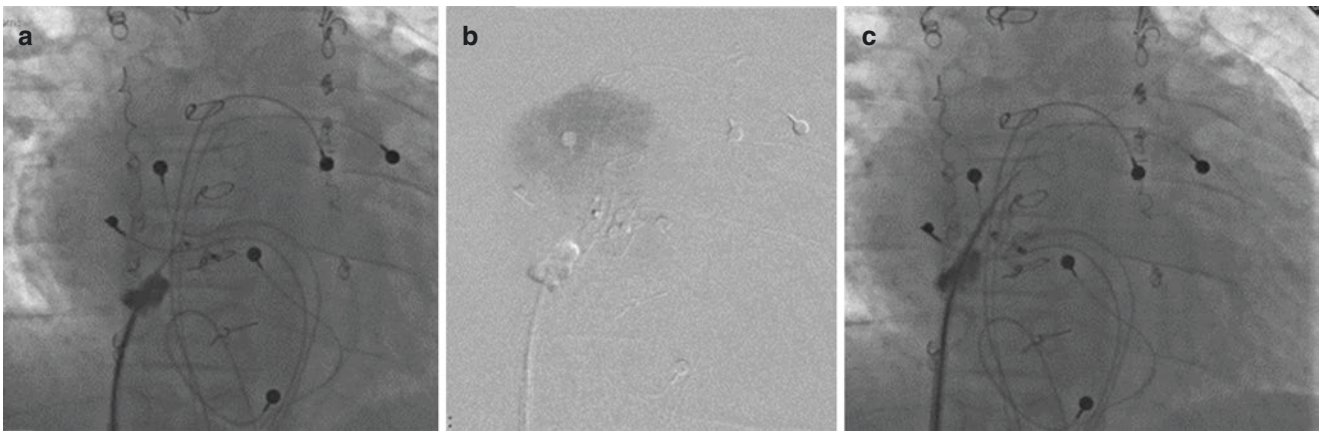



**Fig. 25.10** Patient with failing Fontan circulation. (**a**) The VCI just below the conduit has been punctured with a Brockenbrough needle; a small contrast injection has tagged the atrial wall with some overflow in the pericardial space. The needle has been advanced through the atrial wall. (**b**) The position of the needle is confirmed by a small contrast flush. (**c**) A 0.014-in. coronary wire has been advanced into the left upper pulmonary vein. Advancing the dilator and sheath over the Brockenbrough needle may require considerable force and wringing of the sheath

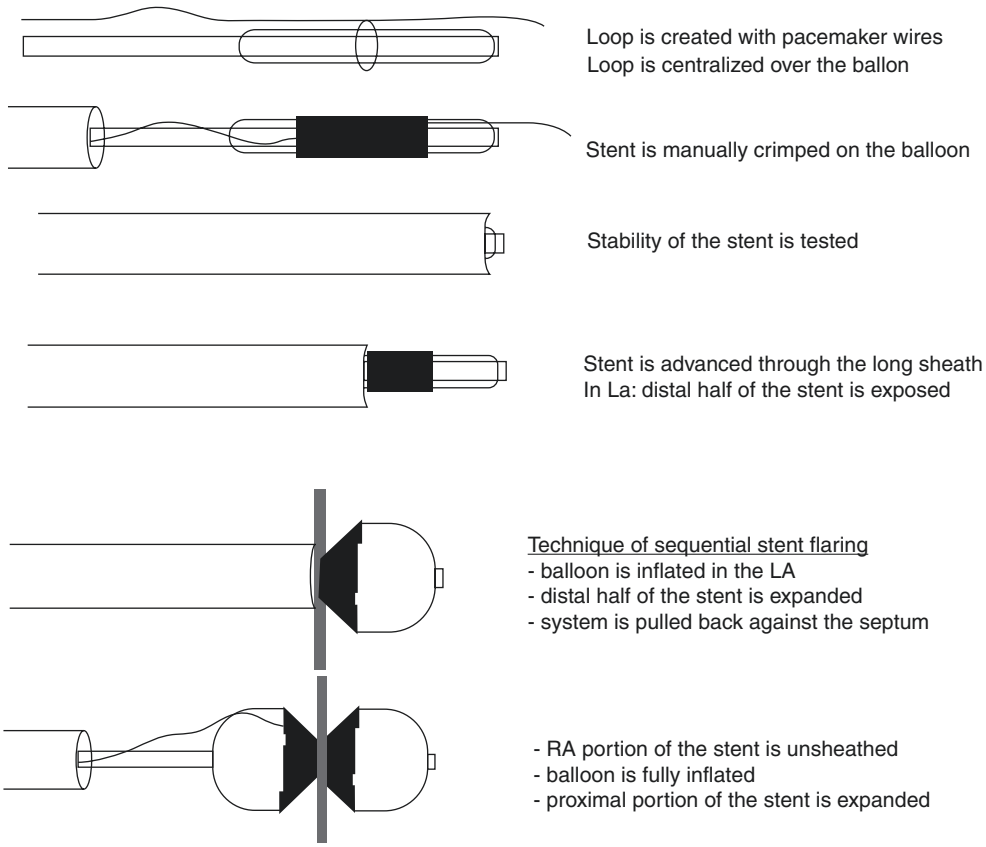



**Fig. 25.11** Cartoon with various steps for a diabolo stent

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 loop is then placed over the midportion of a standard 12–15 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 mounted on the valvuloplasty balloon over the loop, taking care that the loop is placed accurately in the center of the balloon and the stent.
– The stent is then manually crimped, and its stability tested.

#### 25.9.2.3 Stent Deployment (Sequential Stent Flaring) (Figs. 25.12 and 25.13)

- 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 [4].
- After the stent has been deployed in diabolo across the septum, the balloon (with the metal-knot wire) is removed.
- The size of the fenestration can be gradually increased until arterial saturation has decreased down to 80–85% (Fig. 25.14).

### 25.9.3 New Devices

Several manufacturers are currently evaluating ASD occluder like devices with a pre-made perforation. After puncturing the atrial septum, the self-expanding device can be opened across the atrial septum. The technique appears easier, but the fenestration is more difficult to modulate after deployment.

## 25.10 Complications and How to Treat/Avoid

### 25.10.1 Complications of Balloon Atrial Septostomy

- May cause tears to the LA, pulmonary vein, and RA and atrial dysrhythmias (usually transient).
- Procedure-related mortality in patient with HLHS 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.

### 25.10.2 Complications Inherent to Atrial Septal Puncture

- Cardiac perforation and puncture of an inappropriate atrial septal site.
- Pushing an extremely floppy valve with the tip of the catheter, may extend the catheter to the lateral wall of the

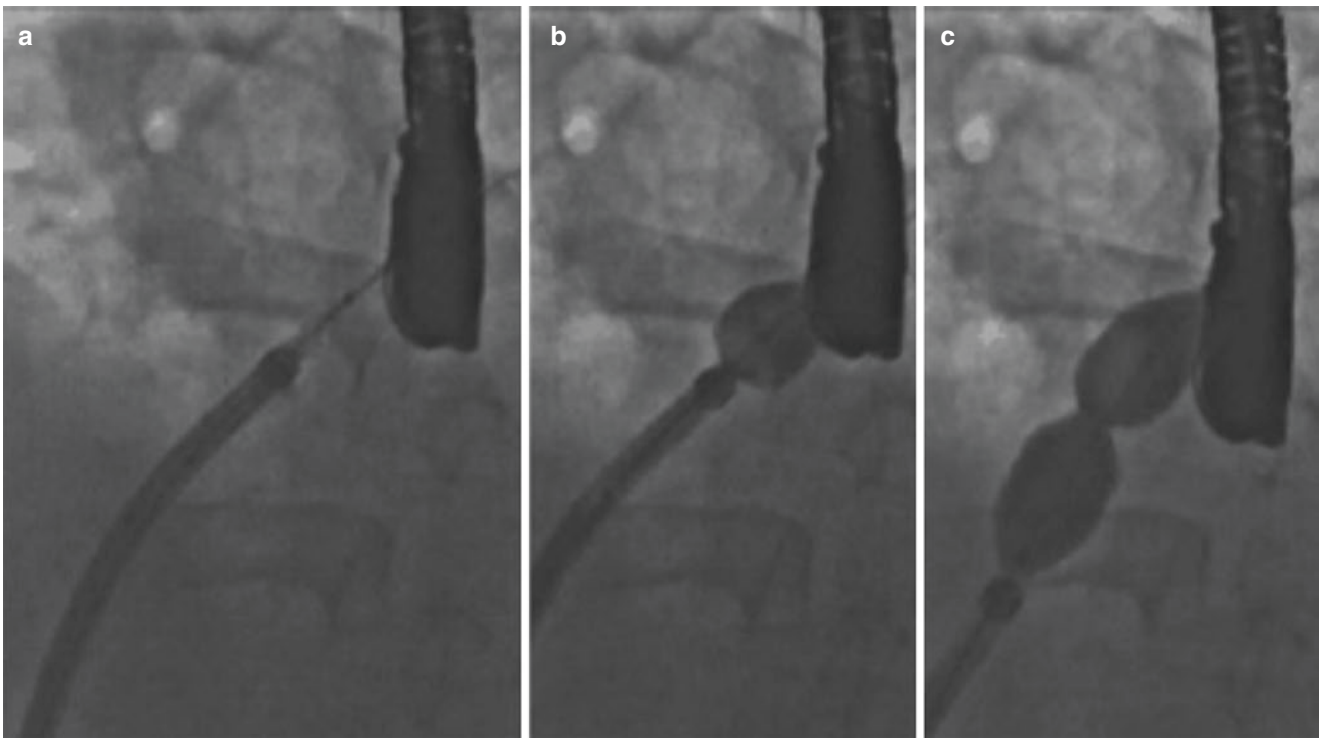



**Fig. 25.12** Deployment of a 1910 Genesis stent in a patient with PAH (**a**) 10F sheath in LA, stent is partially uncovered (**b**) inflation of 15 mm balloon results in flaring of distal end of the stent (**c**) after pulling whole system against the septum; the sheath is pulled back to uncover the RA end and further inflation of the balloon results in diabolo shape of the stent across the atrial septum

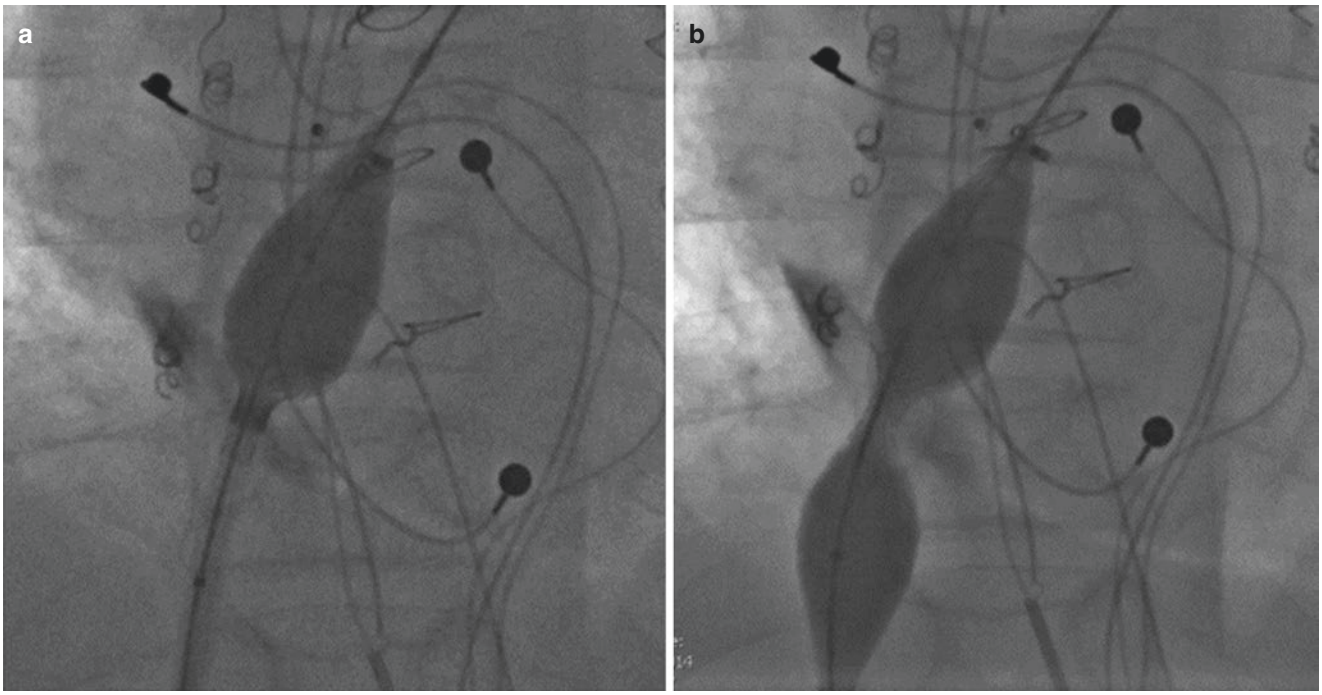



**Fig. 25.13** Deployment of a 1910 Genesis stent in a patient with a failing Fontan circulation (extracardiac conduit). (**a**) The 10F has been withdrawn to partially uncover the stent; inflation of 16 mm Atlas Gold balloon results in flaring of distal end of the stent. (**b**) After pulling the whole system against the atrial wall, the sheath is pulled back to uncover the conduit end and further inflation of the balloon results in a diabolo shape of the stent across the Fontan conduit. Sequential stent flaring allows for reapproximation of the different layers during stent deployment

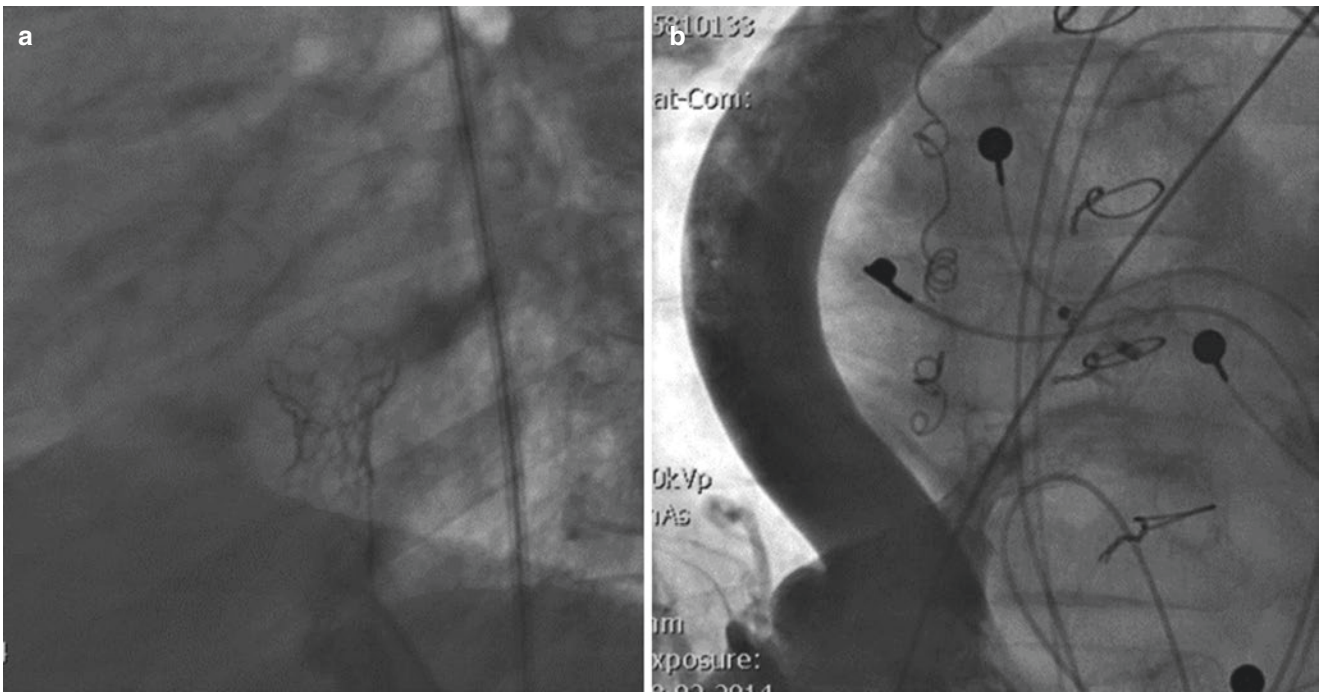



**Fig. 25.14** (**a**) Lateral protection: stent was gradually balloon dilated up to 8 mm, resulting in a diabolo shape of the stent across conduit. (**b**) AP projection: angiography shows adequate but restrictive right-to-left shunting across the stented Fontan conduit

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.

### 25.10.3 Complications of IA Stenting

– Stent embolization: to be avoided by adequate imaging, sequential stent flaring, and diabolo-shaped (bow tie or dog bone stent).
– Avoid stenting a tunnel-shaped fossa ovalis (insufficient grip), consider new septal puncture.
– Embolized stent should be brought down into the VCI and expanded below the hepatic veins (above renal veins).

## 25.11 Conclusions

Creation or enlargement of interatrial communications can be achieved using a variety of transcatheter techniques including transseptal needle puncture or RF perforation, balloon septostomy, blade septostomy, and stent implantation. The procedure can improve hemodynamics acutely in a variety of compromised circulations or provide effective palliation until definitive surgery can be attempted.

# Part VII

# Step-by-Step Procedures: Valve Implantation

# Melody Valve Implantation in Pulmonary Position

# 26

Gianfranco Butera, Alessia Lunardini, and Massimo Chessa

## 26.1 Introduction: Clinical Indications and Patients Selection

Percutaneous pulmonary valve implantation (PPVI) can replace surgery for treatment of right ventricular outflow tract (RVOT) dysfunction after repair of congenital heart disease. Commonly accepted indications are:

- Severe pulmonary stenosis: RV systolic pressure >2/3 systemic in presence of symptoms of heart failure (HF), or >¾ systemic in asymptomatic patients.
- Severe pulmonary regurgitation and one of more of the following:
  - Symptoms of HF.
  - RV dysfunction and/or significant dilatation (on echocardiography and/or cardiovascular magnetic resonance imaging, cMRI).
  - Reduced exercise capacity (peak VO2 < 65–70% of predicted).
  - Arrhythmias: sustained atrial or ventricular arrhythmias.

## 26.2 Procedure

### 26.2.1 Vascular Access

- Femoral vein: more common; both sides should be prepared.
- Internal jugular vein: if the femoral pathway is not patent, or if the access in the RV from the inferior caval vein is difficult.

### 26.2.2 Catheterization and Hemodynamic Evaluation

- Complete right and left heart catheterization are performed by the standard techniques.

### 26.2.3 Complications

A rate of 5–7% has been reported:

- Device instability and migration/embolization.
- RVOT rupture.
- Compression of the coronary arteries.
- Injury to a distal branch pulmonary artery or tricuspid valve.
- Melody Stent fractures.
- Endocarditis.
- Vascular access complications.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_26) contains supplementary material, which is available to authorized users.

G. Butera (✉)
Department of Pediatric Cardiology and Adult with Congenital Heart Disease, Evelina London Children's Hospital, St Thomas' Hospital, London, UK
e-mail: gianfranco.butera@gstt.nhs.uk

A. Lunardini
Congenital Cardiology—Policlinico San Donato IRCCS, San donato Milanese, Italy

M. Chessa
Children and Adults Congenital Heart Disease, Policlinico San Donato, San Donato Milanese, Italy


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_26

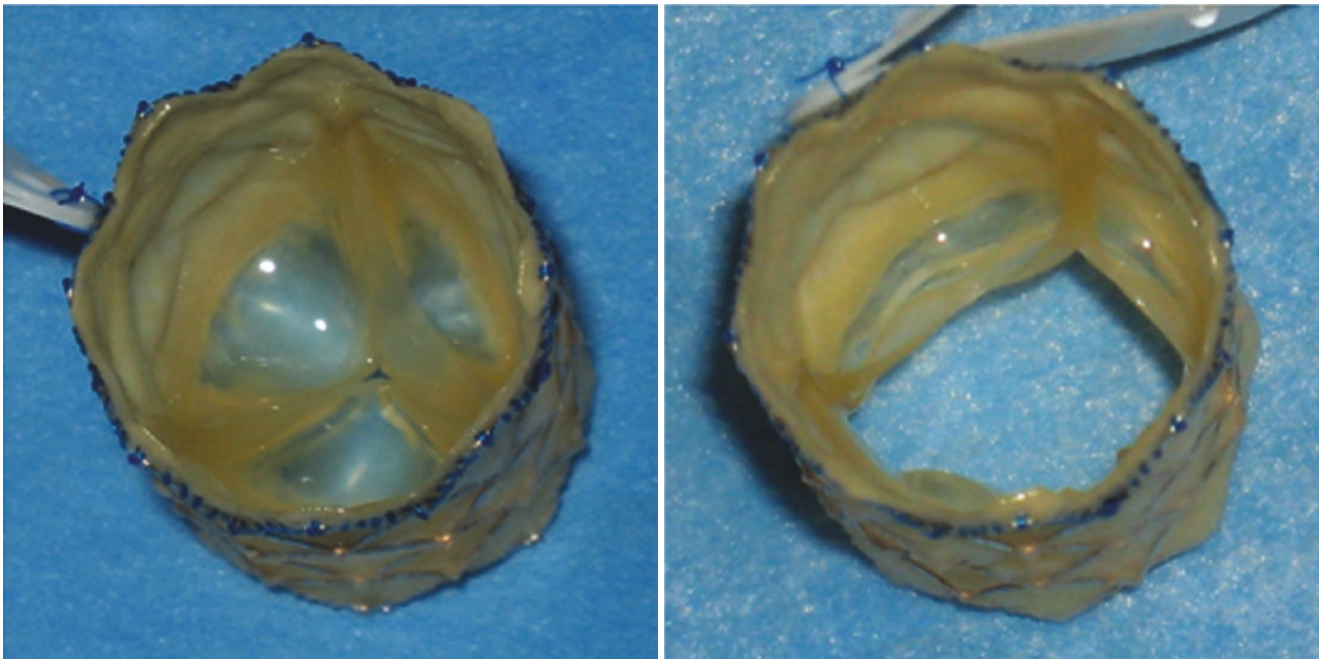


**Fig. 26.1** The device. The Melody Transcatheter Pulmonary Valve (TPV) (Medtronic Inc., MN, USA) is composed of a segment of bovine jugular vein (BJV) with a central valve, and a 28-mm long platinum-iridium stent, reinforced at each strut insertion with gold weld. The venous segment is sutured within the stent by continuous 5-0 polypropylene sutures around the entire circumference at the inflow and outflow portion, as well as discretely at each strut insertion. The suture is white except for the outflow line, which is blue. The combined device is sterilized and then packaged in a solution containing glutaraldehyde and isopropyl alcohol. The valved stent is available in two sizes: Melody TPV 20 (16 mm BJV tissue), which can be dilated up to 20 mm, and Melody TPV 22 (18 mm BJV tissue), which can be dilated up to 22 mm. Before use, it has to be rinsed for 2 min each in 500-mL saline baths to remove the glutaraldehyde

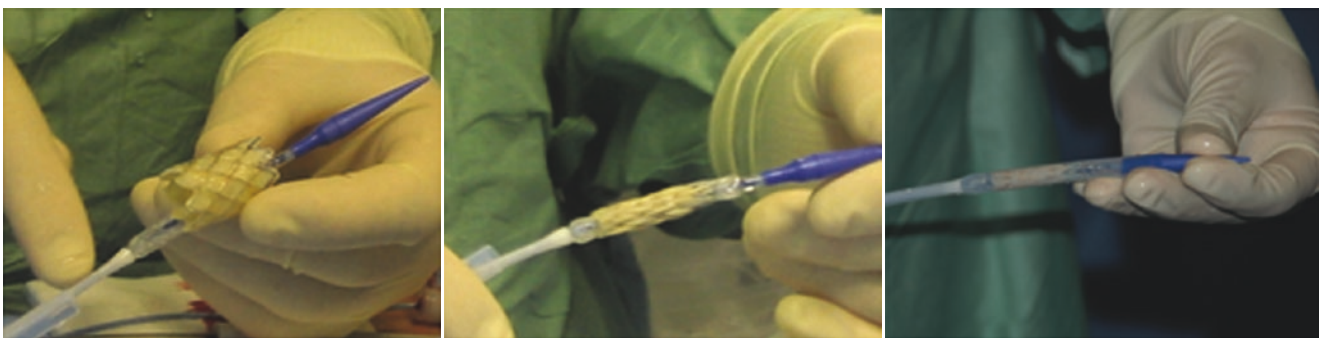


**Fig. 26.2** The delivery system. The Ensemble delivery system (Medtronic Inc., MN, USA) includes: a balloon-in-balloon (BiB), available with three outer balloon diameters (18, 20 and 22 mm) onto which the valved stent is front-loaded and crimped; a tapered tip, blue-colored to correspond with the blue outflow suture on the device; a body consisting of a one-piece Teflon sheath containing a braided-wire reinforced elastomer lumen, to minimize the risk of kinking whilst optimizing flexibility and pushability; a retractable sheath covering the valve during delivery, to be withdrawn just prior to deployment. Proximally there are three ports: a green one for the guidewire, an indigo one for the inner balloon, and an orange one for the outer balloon

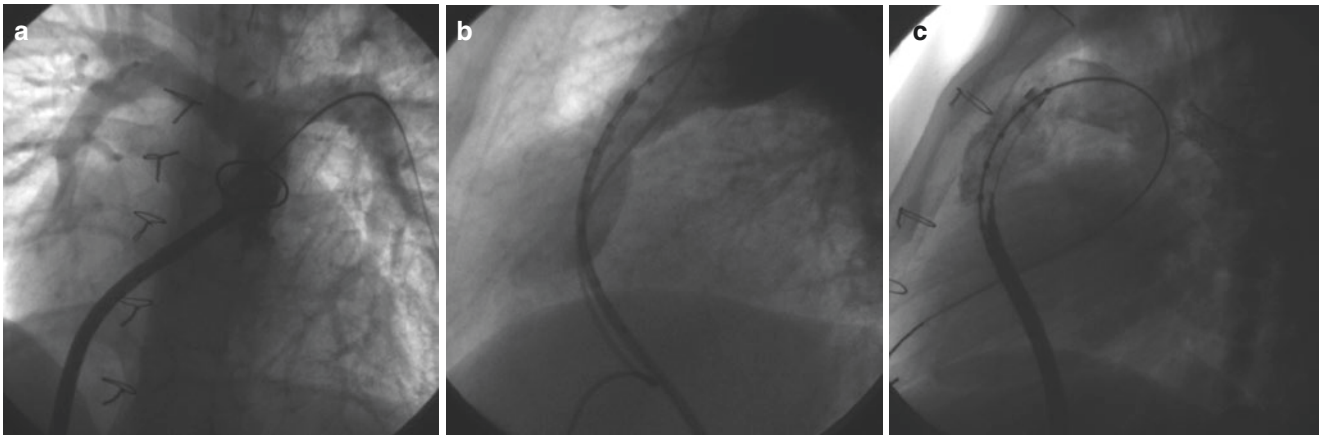



**Fig. 26.3** Angiographies are performed in RVOT and/or RV and ascending aorta. Anteroposterior (AP) view with cranial angulation and left anterior oblique (LAO) view (**a**): Relationship between the bifurcation and distal end of the stent; Landing zone, final target diameter to be achieved with Melody valve. Lateral (LL) view (**b**, **c**): Anterior chest, landing zone and proximal end of the stent; Coronary arteries during balloon interrogation of RVOT

**Video 1** (**a**, **b**, **c**) Angiographies are performed in RVOT and/or RV and ascending aorta. Anteroposterior (AP) view with cranial angulation and left anterior oblique (LAO) view (**a**): Relationship between the bifurcation and distal end of the stent; Landing zone, final target diameter to be achieved with Melody valve. Lateral (LL) view (**b** and **c**): Anterior chest, landing zone and proximal end of the stent; Coronary arteries during balloon interrogation of RVOT (MOV 11905 kb)

**Video 2** (**a**, **b**, **c**) Angiographies are performed in RVOT and/or RV and ascending aorta. Anteroposterior (AP) view with cranial angulation and left anterior oblique (LAO) view (**a**): Relationship between the bifurcation and distal end of the stent; Landing zone, final target diameter to be achieved with Melody valve. Lateral (LL) view (**b** and **c**): Anterior chest, landing zone and proximal end of the stent; Coronary arteries during balloon interrogation of RVOT (MOV 9922 kb)

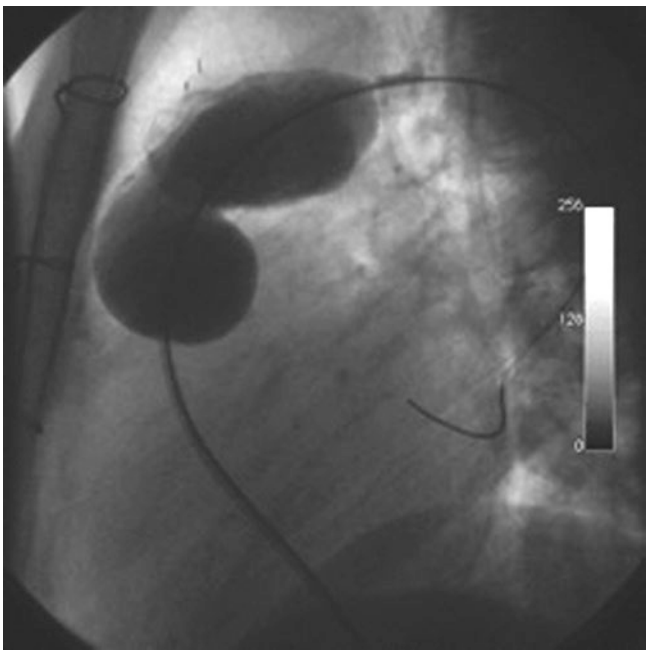


**Fig. 26.4** Balloon interrogation is used to evaluate the characteristics of RVOT: A sizing balloon, such as an Amplatzer sizing balloon II (St. Jude Medical Inc., USA), gives a negative image of the RVOT and landing zone; Low-pressure angioplasty balloons, such as Z-Med X (NuMED Inc., NY, USA) or Crystal balloons (Balt, Montmorency, France), evaluate tissue distensibility; High-pressure balloons, such as Mullins X (NuMED Inc., Hopkinton, NY) or Atlas Gold PTA balloon dilatation catheter (Bard, Tempe, AZ, USA) are used if RVOT is poorly distensible with low-pressure balloons. Balloon testing with angioplasty balloons may cause tear or rupture of RVOT

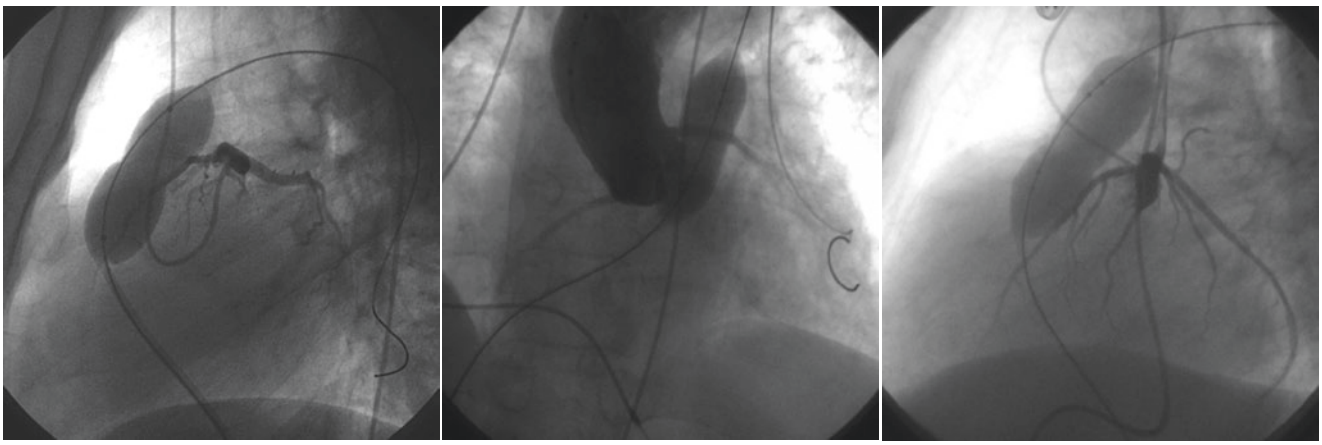


**Fig. 26.5** Coronary artery compression/distortion has to be ruled out before PPVI. Pre-interventional evaluation with cMRI or angio-CT scan may identify patients at high risk of coronary involvement. However, aortography or selective coronary angiography in multiple projections and during balloon interrogation of RVOT are required in all patients in whom PPVI is attempted: Low-pressure dilatation if both coronary arteries are distant from RVOT; Full balloon inflation (with either low or high-pressure balloons) if coronary arteries are near. If coronary compression occurs or appears highly probable, PPVI is contraindicated

**Video 3** Coronary artery compression/distortion has to be ruled out before PPVI. Pre-interventional evaluation with cMRI or angio-CT scan may identify patients at high risk of coronary involvement. However, aortography or selective coronary angiography in multiple projections and during balloon interrogation of RVOT are required in all patients in whom PPVI is attempted: Low-pressure dilatation if both coronary arteries are distant from RVOT; Full balloon inflation (with either low or high-pressure balloons) if coronary arteries are near. If coronary compression occurs or appears highly probable, PPVI is contraindicated (MOV 10816 kb)

**Video 4** Coronary artery compression/distortion has to be ruled out before PPVI. Pre-interventional evaluation with cMRI or angio-CT scan may identify patients at high risk of coronary involvement. However, aortography or selective coronary angiography in multiple projections and during balloon interrogation of RVOT are required in all patients in whom PPVI is attempted: Low-pressure dilatation if both coronary arteries are distant from RVOT; Full balloon inflation (with either low or high-pressure balloons) if coronary arteries are near. If coronary compression occurs or appears highly probable, PPVI is contraindicated (MOV 6829 kb)

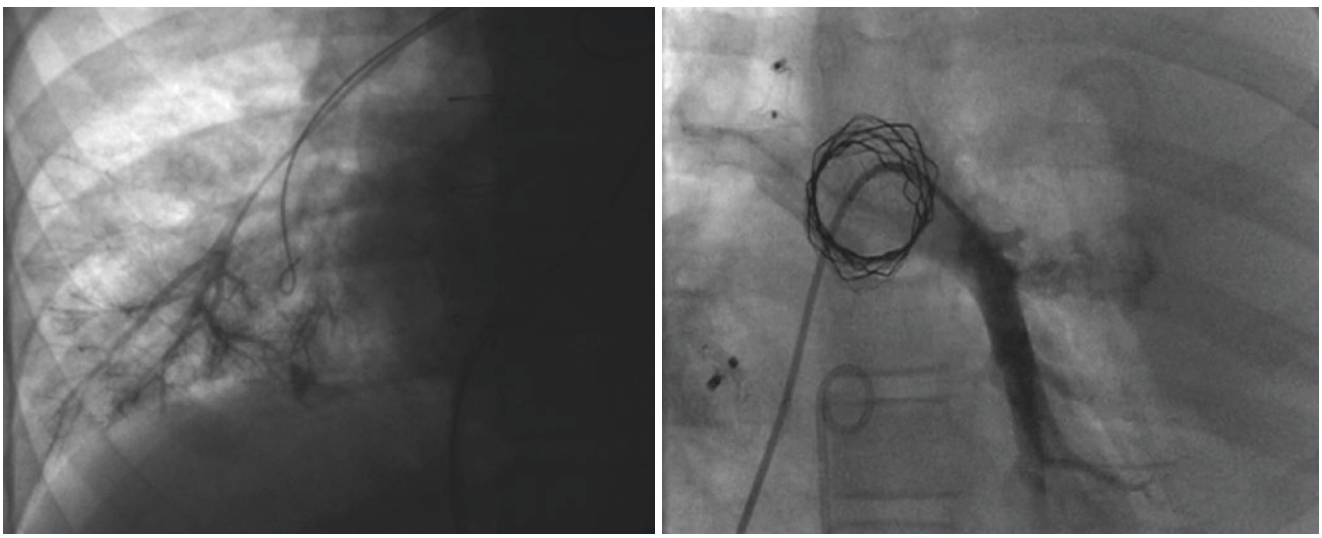


**Fig. 26.6** Angiographies in RVOT and pulmonary branches are performed to exclude extravasation, after each inflation of angioplasty balloons in RVOT, or whenever hemodynamic instability occurs. If a covered stent (CP covered stent, NuMED Inc., Hopkinton, NY) is implanted to treat a tear: Its length should exceed RVOT length by 1–2 zigs proximally and distally; Full expansion is not needed, especially if coronary compression is possible: appose the stent to RVOT walls and flare the ends against wall to seal the tear

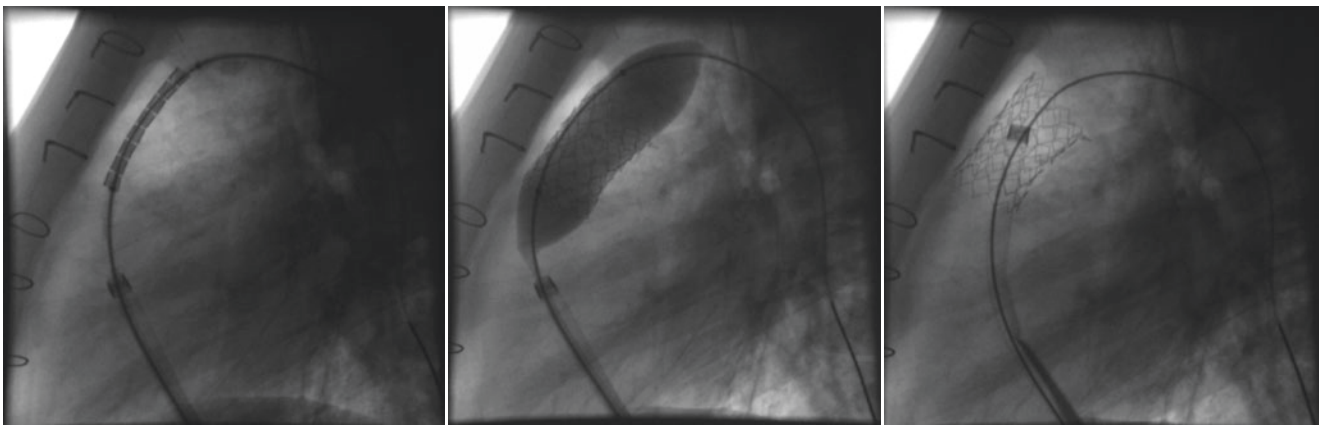


**Fig. 26.7** Pre-stenting. A 14Fr Mullins long sheath is used. Length of the stent should be enough to cover the stenotic area and the entire length of the Melody valve. Implant a second or third stent in case of significant recoil at fluoroscopy. Repeat coronary angiogram before further dilation in case of any doubt, Covered stent(s) are used in case of extravasation or in cases with significant calcifications

**Video 5** Pre-stenting. A 14Fr Mullins long sheath is used. Length of the stent should be enough to cover the stenotic area and the entire length of the Melody valve. Implant a second or third stent in case of significant recoil at fluoroscopy. Repeat coronary angiogram before further dilation in case of any doubt, Covered stent(s) are used in case of extravasation or in cases with significant calcifications (MOV 6157 kb)

**Video 6** Valve implantation into the landing zone. Inner balloon is already inflated. It's important to have a simultaneous view of right atrium, target zone, and distal tip of wire (MOV 16391 kb)

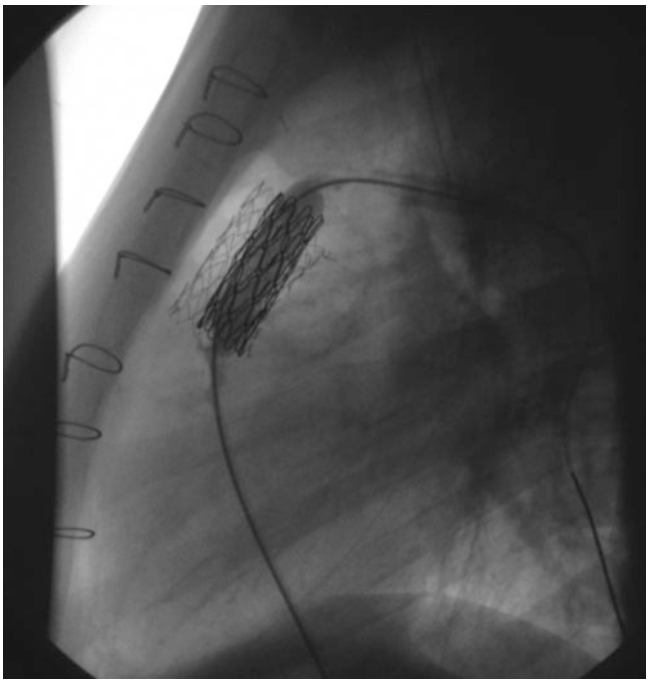


**Fig. 26.8** Valve implantation into the landing zone. Inner balloon is already inflated. It's important to have a simultaneous view of right atrium, target zone, and distal tip of wire

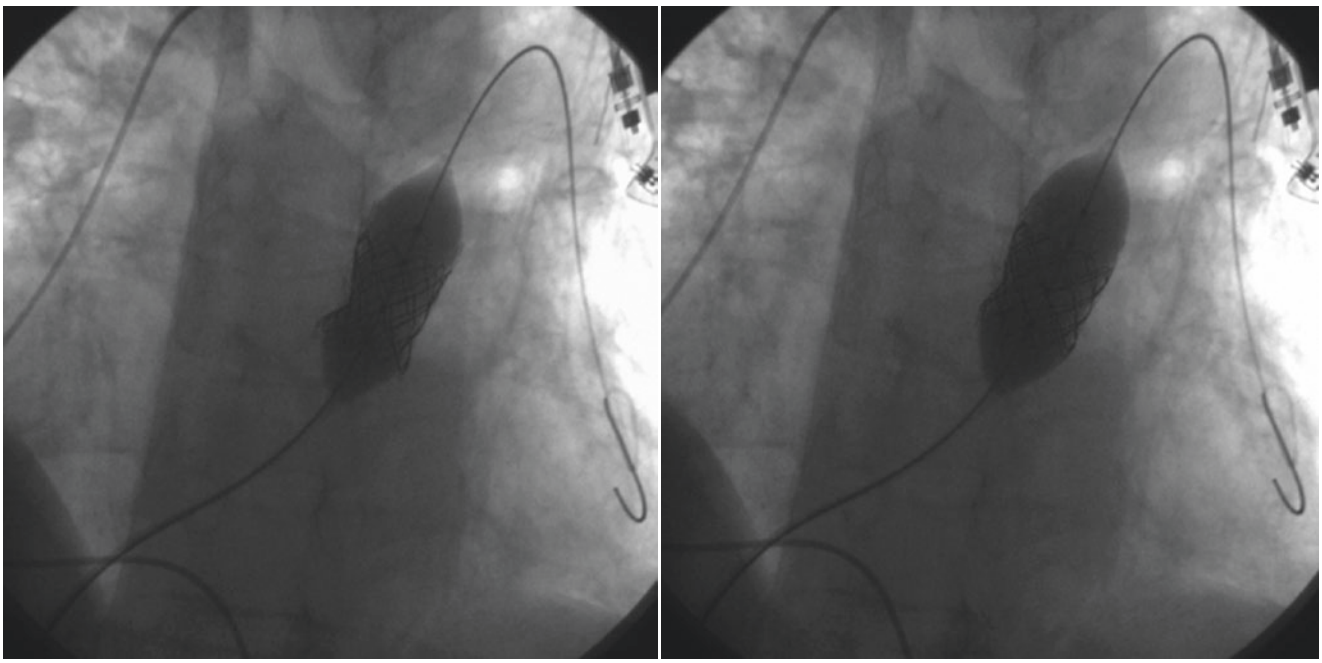


**Fig. 26.9** Post-dilatation of the valve may be needed in the presence of a residual gradient (>20 mmHg) and incomplete expansion of the valved stent. Use appropriately sized ultra-high-pressure balloon with a maximum balloon size of 24 mm

**Video 7** Post-dilatation of the valve may be needed in the presence of a residual gradient (>20 mmHg) and incomplete expansion of the valved stent. Use appropriately sized ultra-high-pressure balloon with a maximum balloon size of 24 mm (MOV 8979 kb)

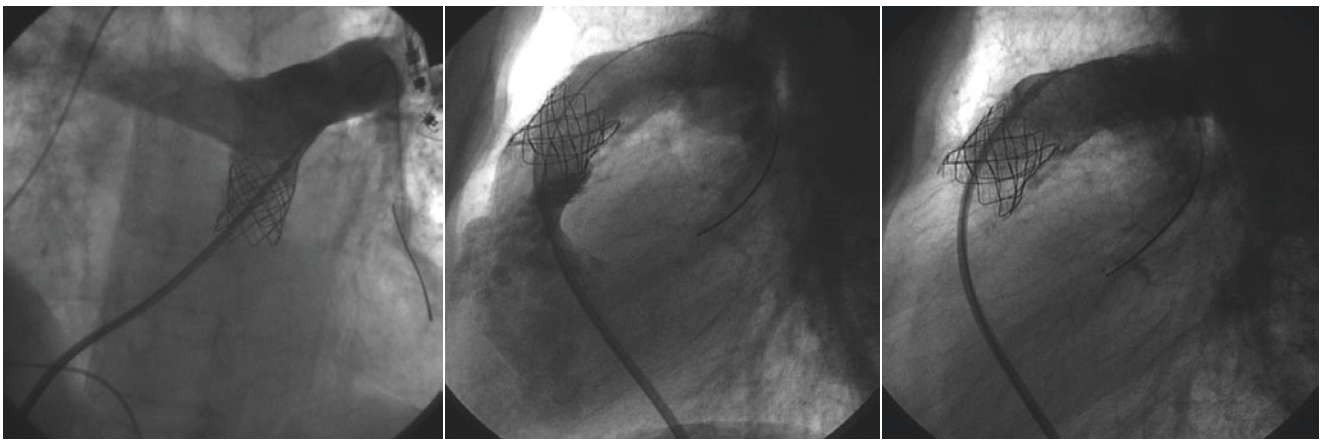


**Fig. 26.10** Final angiographies in the infundibulum (LL, AP with cranial angulation) and the pulmonary trunk/stent (AP with cranial angulation ± LAO) are performed to: rule out extravasations of contrast; evaluate stent apposition to the walls of RVOT, valve competence and flow into the pulmonary branches

**Videos 8** Final angiographies in the infundibulum (LL, AP with cranial angulation) and the pulmonary trunk/stent (AP with cranial angulation ± LAO) are performed to: rule out extravasations of contrast; evaluate stent apposition to the walls of RVOT, valve competence and flow into the pulmonary branches (MOV 8133 kb)

**Video 9** Final angiographies in the infundibulum (LL, AP with cranial angulation) and the pulmonary trunk/stent (AP with cranial angulation ± LAO) are performed to: rule out extravasations of contrast; evaluate stent apposition to the walls of RVOT, valve competence and flow into the pulmonary branches (MOV 10175 kb)

**Video 10** Final angiographies in the infundibulum (LL, AP with cranial angulation) and the pulmonary trunk/stent (AP with cranial angulation ± LAO) are performed to: rule out extravasations of contrast; evaluate stent apposition to the walls of RVOT, valve competence and flow into the pulmonary branches (MOV 10371 kb)

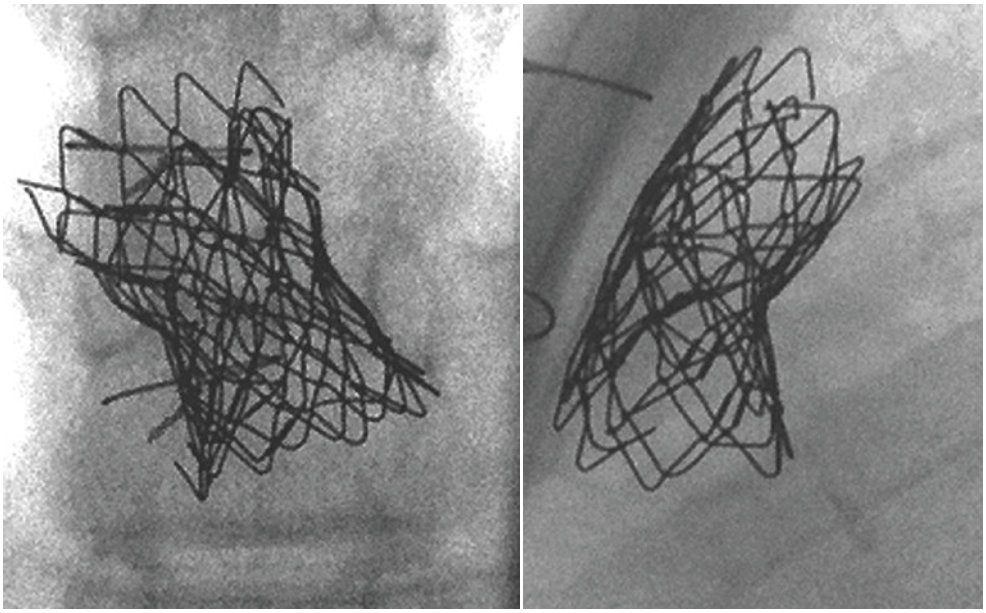


**Fig. 26.11** Melody stent fractures. Repeat Melody implantation can be performed for stent fracture and residual stenosis. The procedure is feasible and has excellent and sustained hemodynamic results

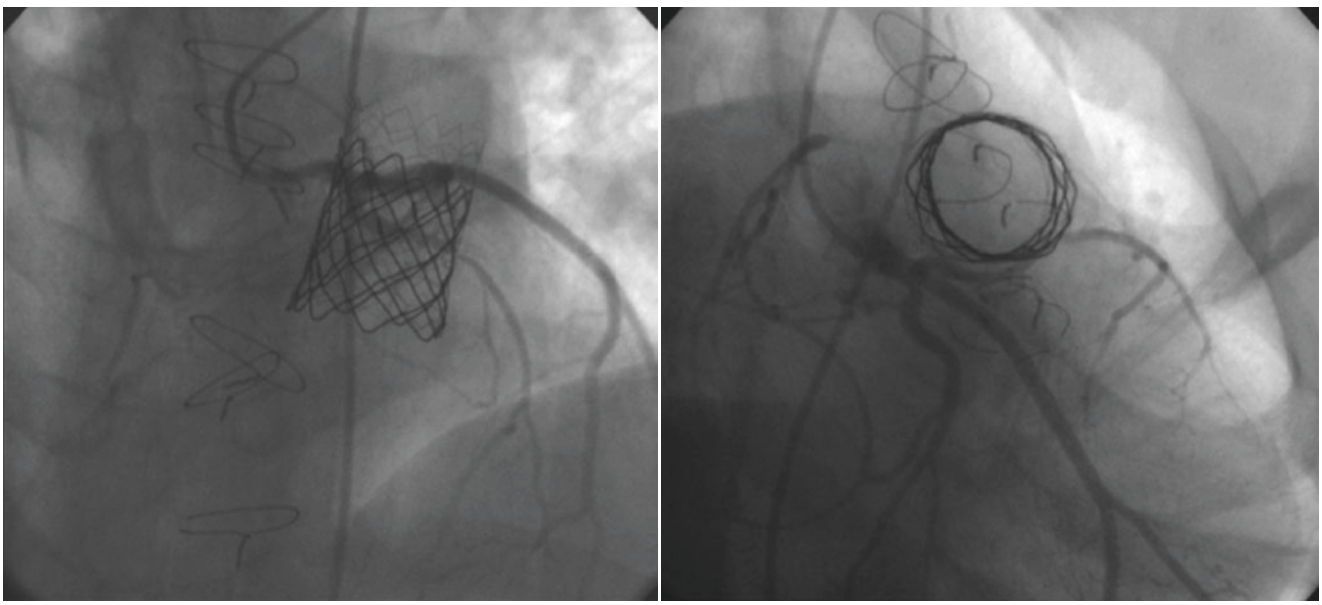


**Fig. 26.12** Sometimes coronary compression occurs after deployment of the valve, despite a careful pre- and intra-procedural evaluation. In these cases, urgent surgery is needed

# Edwards SAPIEN XT Valve Implantation in the Pulmonary Position

27

Noa Holoshitz, Gurdeep Mann, and Ziyad M. Hijazi

## 27.1 Introduction

There are currently two commercially available valves in the United States for use in the pulmonic position, the Melody valve (Medtronic Inc., Minneapolis, MN) and the Edwards SAPIEN XT valve (Edwards Lifesciences LLC, Irvine, CA). The initial use of the SAPIEN was as a percutaneous substitute to surgical valve replacement in the *aortic* position; however, it has emerged as an alternative to the Melody valve in the pulmonic position and has been used successfully in the right ventricular (RV) to pulmonary artery (PA) conduits since 2005. In March 2016, the SAPIEN XT (second generation of the Edwards SAPIEN transcatheter heart valve) (Figs. 27.1, 27.2, and 27.3) gained the approval of the US Food and Drug Administration (FDA) for use in dysfunctional RV to PA conduits.

## 27.2 Patient Selection and Pathophysiology

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. 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. The procedure 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.

## 27.3 Pre-procedural Imaging

Echocardiography (echo) is usually the first imaging test performed in patients who may be candidates for transcatheter pulmonary valve replacement (tPVR). From the initial echo, the patient's right and left ventricular function (Fig. 27.4d) can be evaluated as well as the amount of pulmonic insufficiency using color (Fig. 27.4b) and continuous wave (Fig. 27.4c) Doppler. The RVOT can also be measured for initial measurements (Fig. 27.4a). If there is concern that intervention may be indicated, cardiac magnetic resonance imaging (MRI) is the next imaging test that 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 whom have been trained at performing and interpreting congenital cardiac MRI. The MRI can help evaluate the degree of pulmonic valve dysfunction by calculating the pulmonary regurgitant fraction, the RV ejection fraction, and the RV end-diastolic volumes. Moreover, valuable information about the patient's anatomy can be obtained by MRI such as native RVOT dimensions (Fig. 27.5a, b), degree of conduit stenosis, geometry of the RVOT (Fig. 27.5c), and the distance of the coronary arteries from the outflow tract or conduit, which is a critical step in the evaluation.

N. Holoshitz
Ascension Columbia-St. Mary's Cardiovascular Specialists, Milwaukee, WI, USA

G. Mann
Department of Radiology, Sidra Medical & Research Center, Doha, Qatar

Z. M. Hijazi (✉)
Department of Pediatrics, Sidra Medical & Research Center, Doha, Qatar
e-mail: zhijazi@sidra.org



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_27

**Fig. 27.1** The SAPIEN XT valve is made of three bovine pericardial leaflets, hand-sewn into a cobalt-chromium balloon expandable stent. A fabric cuff covers the lower end of the stent to achieve a seal with the calcified conduit that will prevent para-valvular leak. It is available in 23 mm, 26 mm, and 29 mm diameters with heights of 14.3 mm, 17.2 mm, and 19.1 mm, respectively. It can therefore be used in calcified conduits up to 29 mm at the time of transcatheter valve replacement

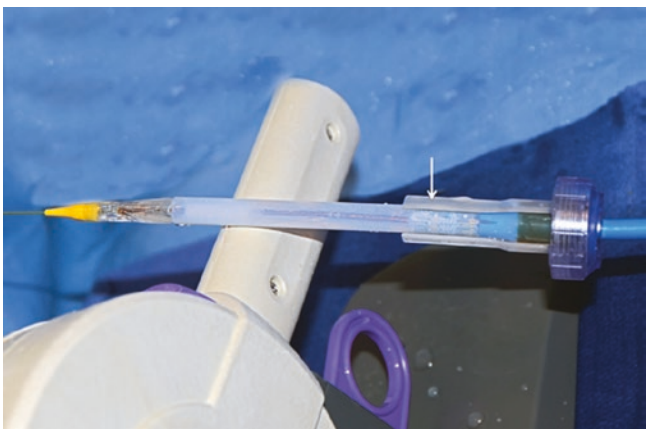


**Fig. 27.2** The Edwards SAPIEN XT heart valve (arrow) is already crimped on the shaft and covered with the loader. The delivery system used is the NovaFlex+ (Edwards Lifesciences). It has a tapered nose cone-shaped balloon catheter with a deflectable tip. It requires either a 14 Fr for the 23 and 26 mm valve or 16 Fr for the 29 mm valve expandable sheaths. With this delivery system, the valve is crimped proximally to the balloon tip (white arrow), which allows for a smaller delivery profile. Once the delivery system is past the sheath, in a straight part of the vena cava, the balloon is pulled back onto the crimped valve

**Fig. 27.3** Close-up image of the NovaFlex+ delivery handle showing the two wheels: the flex wheel (short white arrow) and the fine alignment wheel (long white arrow). The flex wheel (short arrow) of the NovaFlex+ delivery system is used to maneuver the valve across the right ventricular outflow track (RVOT). The valve alignment wheel (long arrow) is used to position the valve more precisely between the valve alignment markers after it is retracted onto the balloon

## 27.4 Technique

The femoral vein is the preferred route of delivery for the valve, but it is also possible to deliver it through the internal jugular vein. 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 to keep the activated clotting time of >250 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 has not been uniformly enforced in our practice. All patients should be given antibiotic prophylaxis (first dose at the start of the procedure and then two subsequent doses 8 h apart).

- Standard right and left heart catheterization is carried out to evaluate the baseline hemodynamics and the pressure gradient across the dysfunctional conduit. Further, calculating the pulmonary vascular resistance is important in such patients.
- Angiographic evaluation of the RV-PA conduit is performed (preferably in biplane) through a side hole catheter (Fig. 27.6a, b) to assess the degree of pulmonary regurgitation and the shape of the conduit (arrow demonstrating narrowing in the conduit) and the presence of calcifications and to rule in/out any distal branch pulmonary artery narrowings.

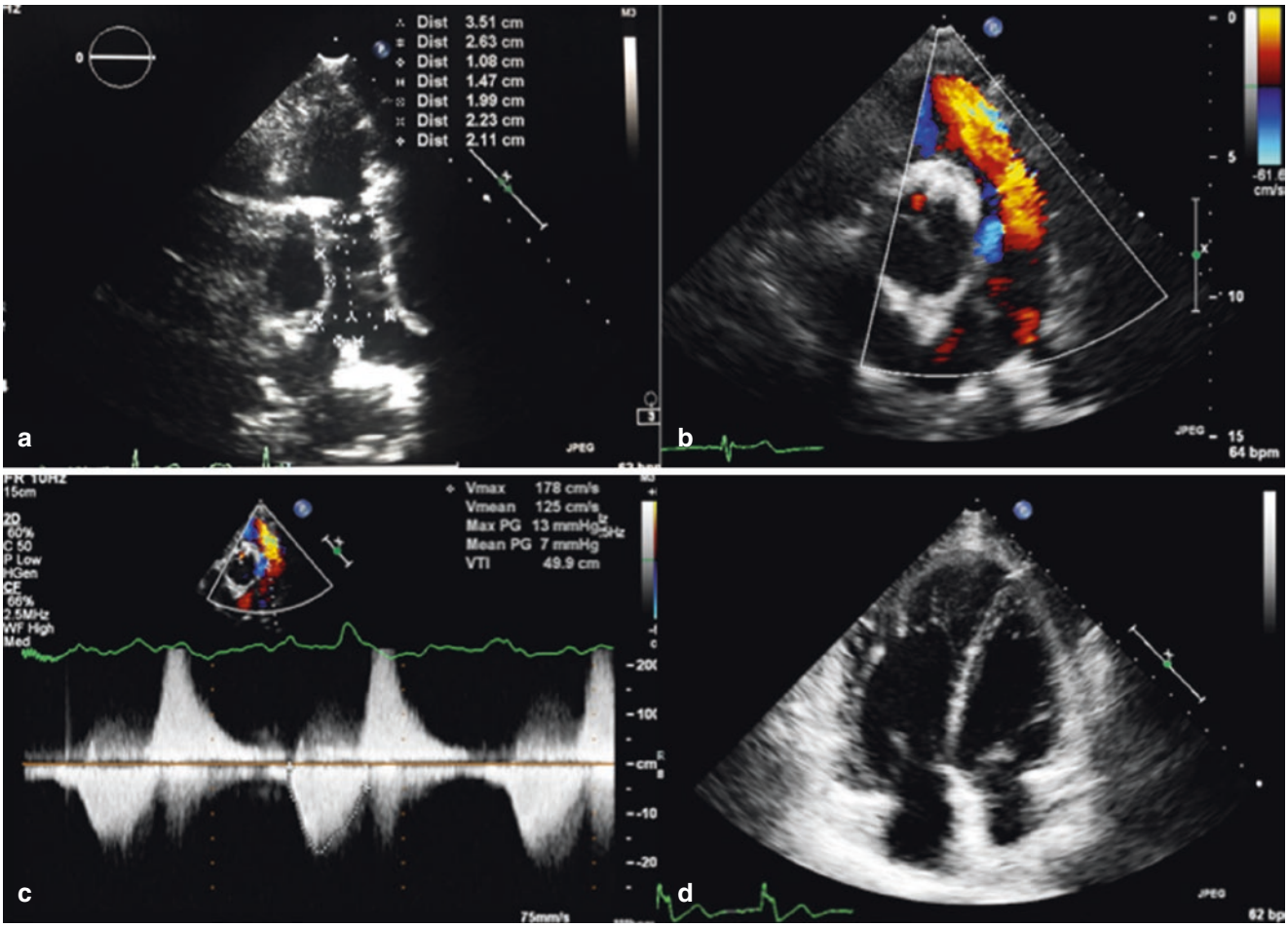



**Fig. 27.4** Transthoracic echocardiography images in a patient with a dysfunctional homograft between the right ventricle and pulmonary artery. (**a**) Short axis view demonstrating various measurements obtained. (**b**) Color Doppler in short axis demonstrating severe regurgitation. (**c**) Continuous wave Doppler across the right ventricular outflow tract demonstrating severe regurgitation. (**d**) Four-chamber view demonstrating large right ventricle

- The minimum diameter of the conduit is measured by inflating a sizing balloon across the pulmonic valve. The choice of the sizing balloon is important. If from angiography the distance between the RVOT and aortic root is far, then we have been using a compliant balloon (such as an ASD sizing balloon); however, if the distance is within 10 mm, it is important to use a high-pressure balloon to dilate the conduit gradually to its final diameter prior to implantation of a stent in the conduit.
- Aortic root angiography or selective coronary angiography is carried out with simultaneous balloon inflation in the RVOT (Fig. 27.7a, b) to evaluate for coronary artery compression. 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.
- Bare-metal stent implantation (pre-stenting) as a landing zone is performed. The stent (white arrow) is deployed on a BiB (balloon-in-balloon) catheter (NuMED Inc., Hopkinton, NY, USA) (preferable) or a single balloon over a stiff guide wire placed in one of the pulmonary arteries, preferably in the left pulmonary artery (Figs. 27.8a, b, 27.9a, b, 27.10a, b, and 27.11a, b).
- 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 heavily calcified conduits, which are at a higher risk for rupture/dissection, a covered stent may be used in place of a bare-metal stent with gradual inflation of larger balloons. With the recent approval of NuMED covered CP stent in the United States, many operators now prefer to implant covered stents as landing zone in heavily calcified conduits.

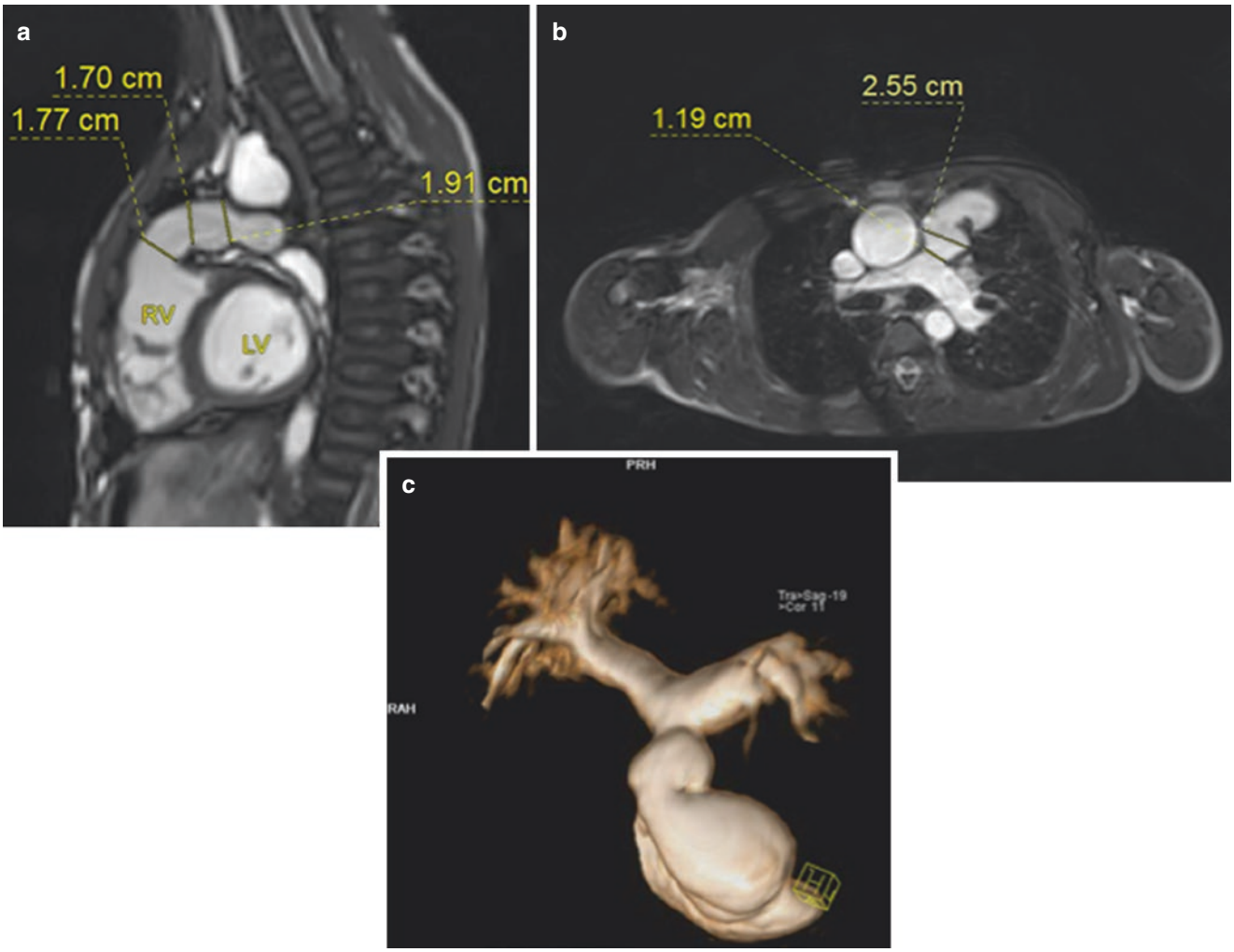



**Fig. 27.5** Cardiac MR images (SSFP) in a patient with dysfunctional homograft between the right ventricle and pulmonary artery. The regurgitant fraction in this patient was 51%. (**a**) Cine SSFP sagittal-oblique image through the right ventricular outflow tract demonstrating various measurements obtained. (**b**) Axial oblique 3D SSFP demonstrating main and branch pulmonary arteries and measurements. (**c**) Three-dimensional time-resolved contrast-enhanced MRA of the right ventricular outflow tract and branch pulmonary arteries. SSFP, steady-state free precession

- We always repeat angiography after stent deployment (Fig. 27.11a, b) to look for any potential complication (such as dissection or extravasation of dye from the conduit).
- 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 that does not recoil. After stent deployment, it is important to assess pressure gradient across the stent. We usually do not accept any gradient higher than 15 mmHg; if it is higher than 15 mmHg, the stent is further expanded with a higher-pressure balloon, such as Atlas until the residual gradient is less than 15 mmHg.
- The valve stent is crimped symmetrically using a specialized crimping tool proximally to the balloon on the shaft of the balloon (Fig. 27.2).
- The venous sheath is upsized to the appropriately sized Edwards Expandable Introducer sheath (14 Fr for 23 and 26 mm valves and 16 Fr for 29 mm valve). Ensure that the sheath tip is in the mid-inferior vena cava for appropriate positioning of the valve over the balloon. The NovaFlex+ delivery system is inserted through the sheath via the loader until the valve exits the sheath. The loader is retracted to the proximal end of the delivery system. In a straight section of the inferior vena cava, valve alignment is then initiated by pressing the release button and pulling back on the balloon catheter. Continue pulling back on the balloon catheter until the delivery system locks into the valve alignment position, and then use the valve alignment wheel (Fig. 27.3) to position the valve between the markers.

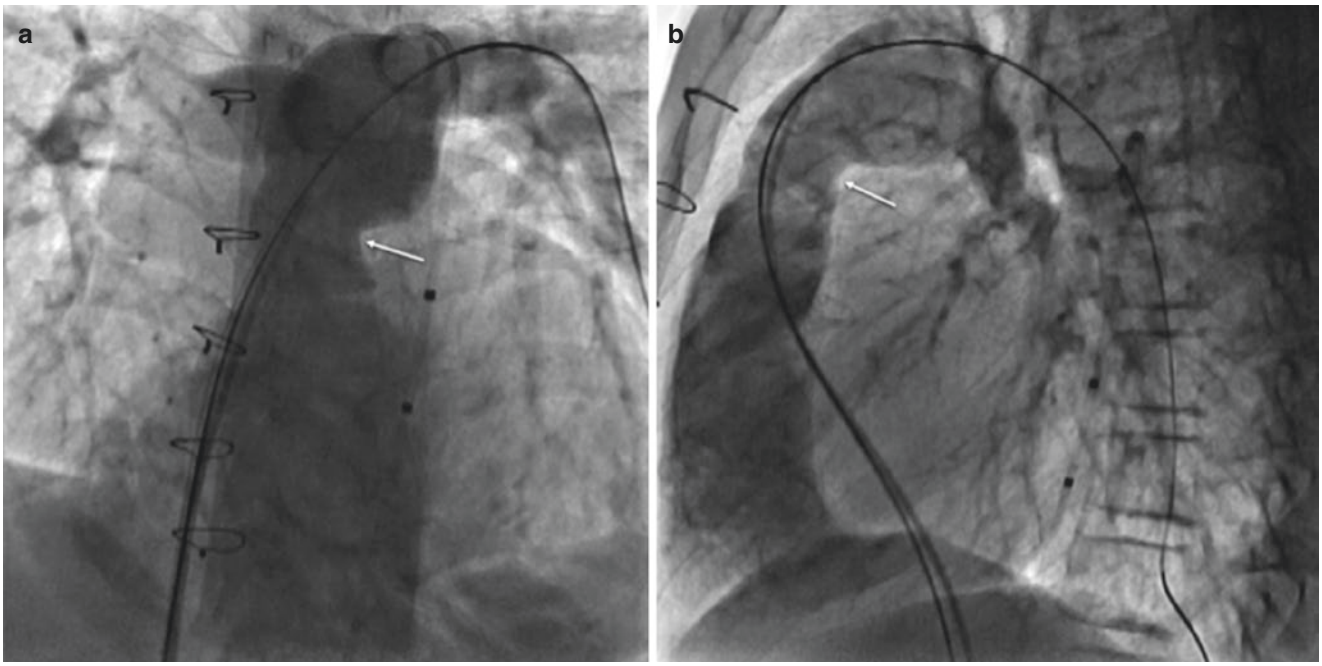



**Fig. 27.6** Cine angiographic/fluoroscopic images in a 13-year-old male patient with dysfunctional 22 mm homograft in two views (frontal: left anterior oblique with cranial angulation and straight lateral). (**a**, **b**) Angiogram in the conduit showing free regurgitation and narrowing in the conduit (arrows)

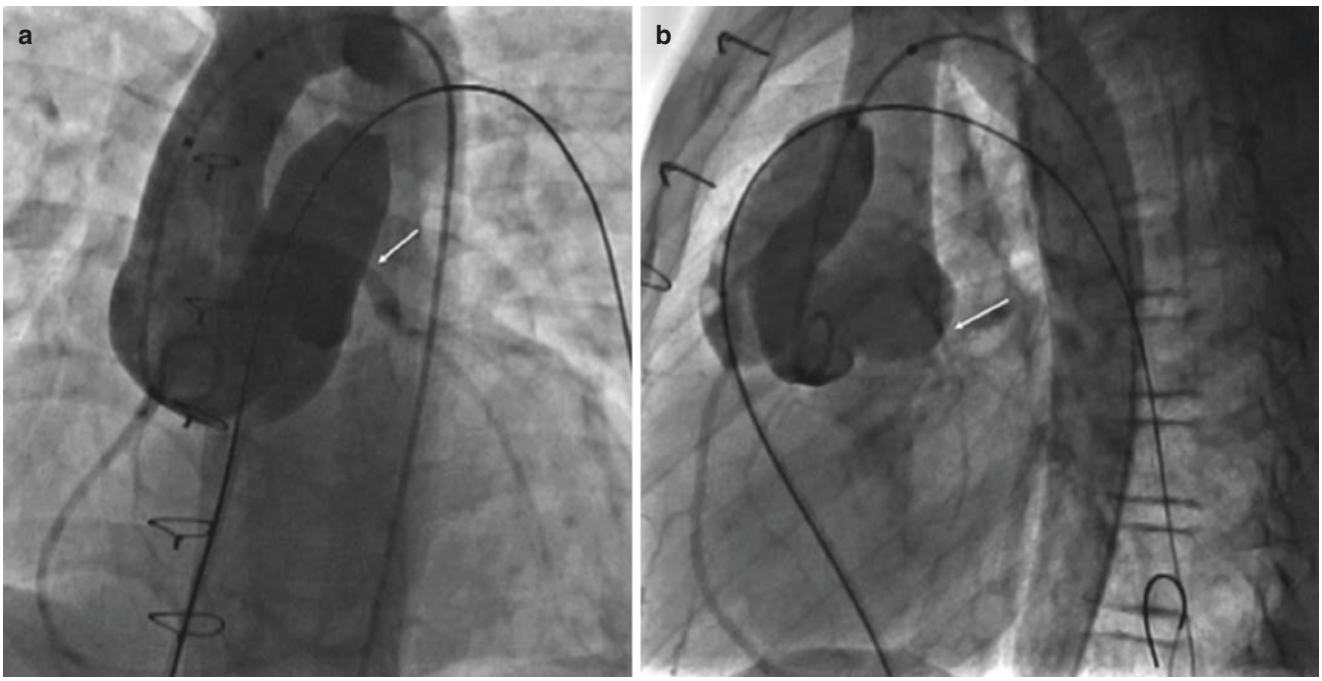



**Fig. 27.7** Cine angiographic/fluoroscopic images in a 13-year-old male patient with dysfunctional 22 mm homograft in two views (frontal: left anterior oblique with cranial angulation and straight lateral). (**a**, **b**) Ascending aortic root angiogram during balloon inflation in the homograft showing normal filling of the coronary arteries (arrows)

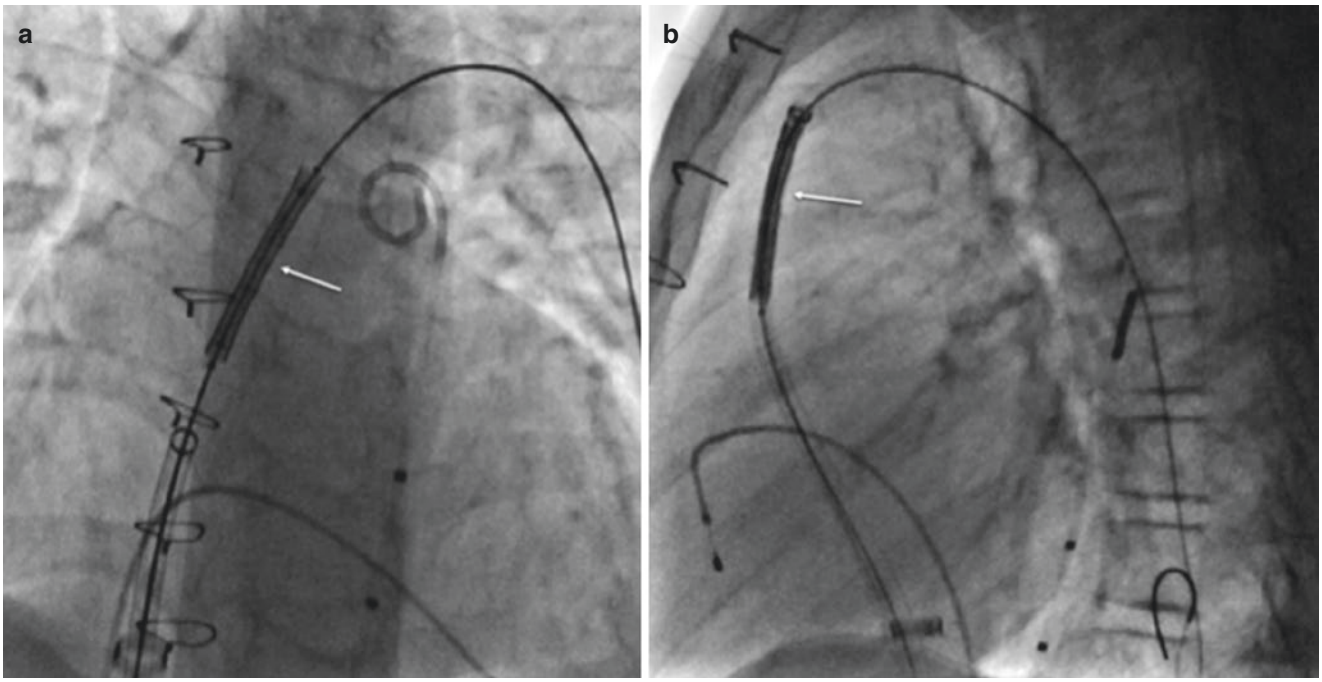



**Fig. 27.8** Cine angiographic/fluoroscopic images in a 13-year-old male patient with dysfunctional 22 mm homograft in two views (frontal: left anterior oblique with cranial angulation and straight lateral). (**a**, **b**) Cine fluoroscopy during deployment of a P3110 bare-metal stent (arrows) as a landing zone, mounted on a 22 mm BiB catheter

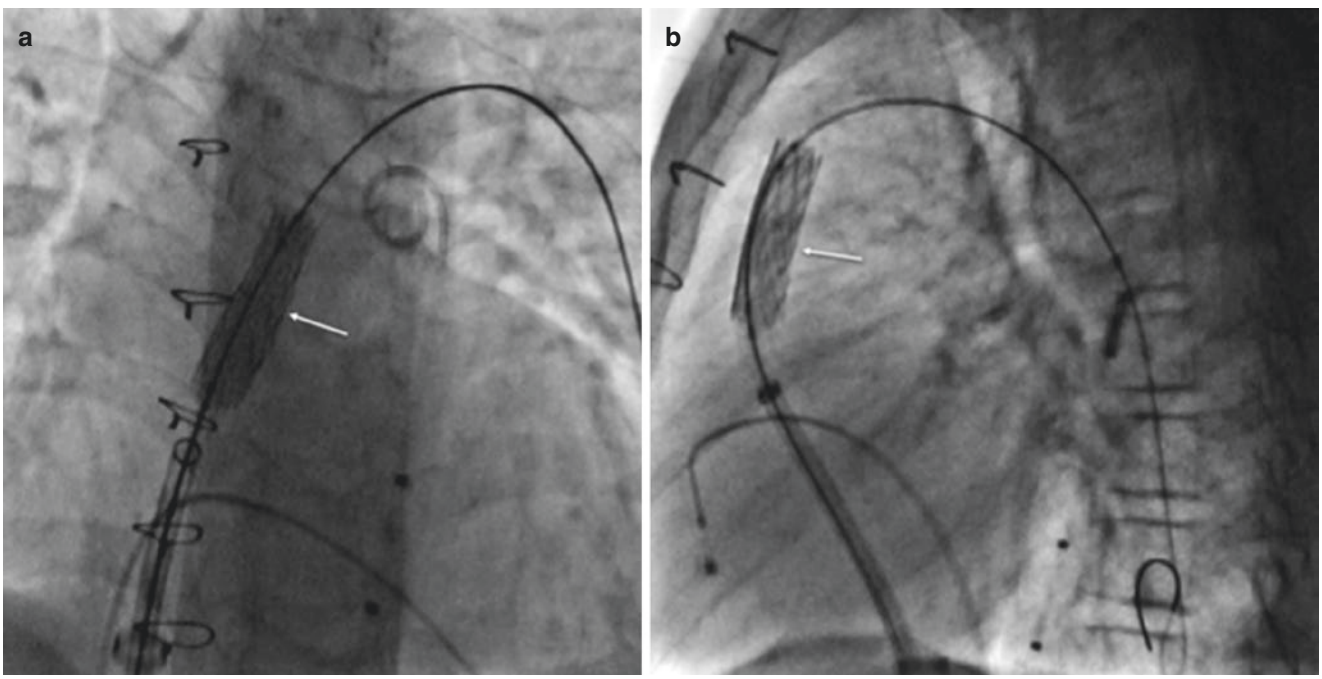



**Fig. 27.9** Cine angiographic/fluoroscopic images in a 13-year-old male patient with dysfunctional 22 mm homograft in two views (frontal: left anterior oblique with cranial angulation and straight lateral). (**a**, **b**) Inflation of the inner balloon (arrow)

- The valve is then delivered across the pre-stented RVOT over a stiff guide wire (Meier wire or Lunderquist wire); the flex wheel (Fig. 27.3) can be used to help maneuver the valve into correct position. With the release button deployed, the Flex catheter is retracted to the double marker to completely uncover the balloon. Fluoroscopy is used to make sure that the valve is in the middle of the stent (landing zone). Distal or proximal deployment of the valve may result in valve embolization.

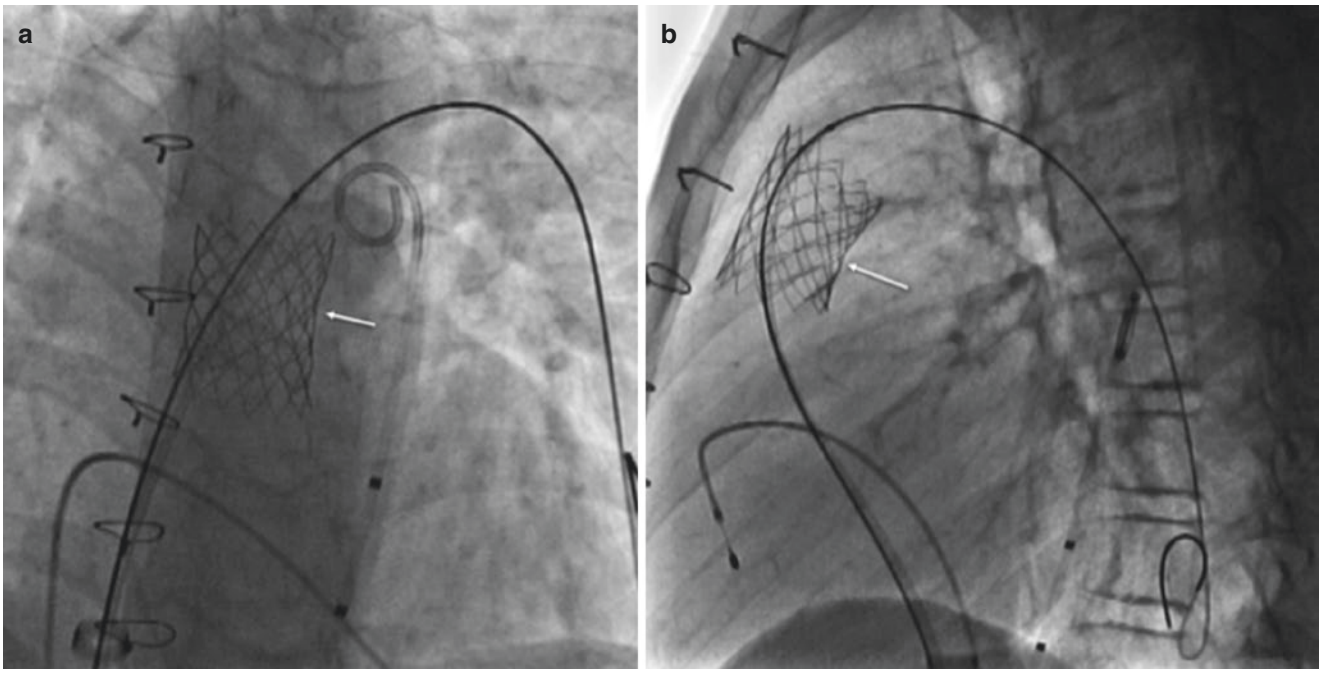



**Fig. 27.10** Cine angiographic/fluoroscopic images in a 13-year-old male patient with dysfunctional 22 mm homograft in two views (frontal: left anterior oblique with cranial angulation and straight lateral). (**a**, **b**) Stent is fully deployed in the homograft (arrows)

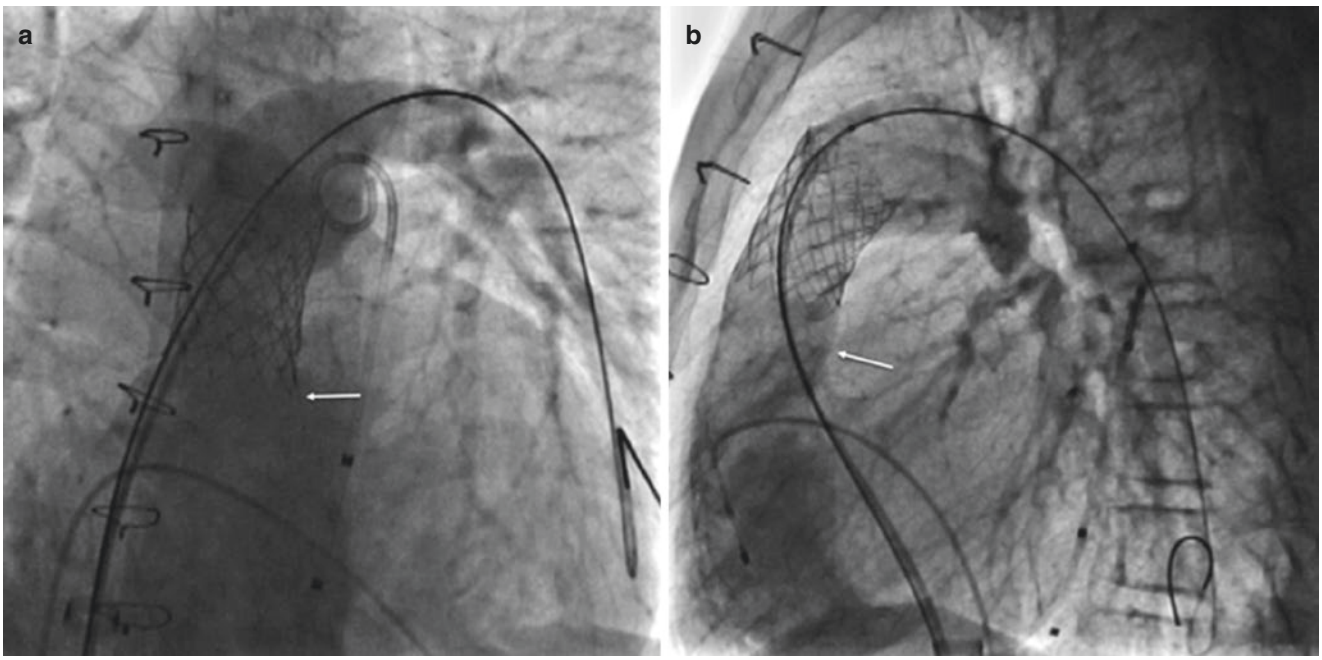



**Fig. 27.11** Cine angiographic/fluoroscopic images in a 13-year-old male patient with dysfunctional 22 mm homograft in two views (frontal: left anterior oblique with cranial angulation and straight lateral). (**a**, **b**) Angiogram demonstrating good position, free regurgitation (arrow), and no complications

- The valve is deployed slowly (with chance of readjustment of position, should the valve move distal or proximal). It is crucial to extrude the entire volume in the inflation device (it is volume dependent and not pressure dependent). Inflation is held for few seconds, and then the balloon is deflated. (Figs. 27.12a, b and 27.13a, b demonstrate balloon/stent/valve inflation).
- Valve performance is then evaluated by hemodynamic assessment (catheter pullback) and by angiography (Fig. 27.14a, b) to look for any valvular regurgitation.

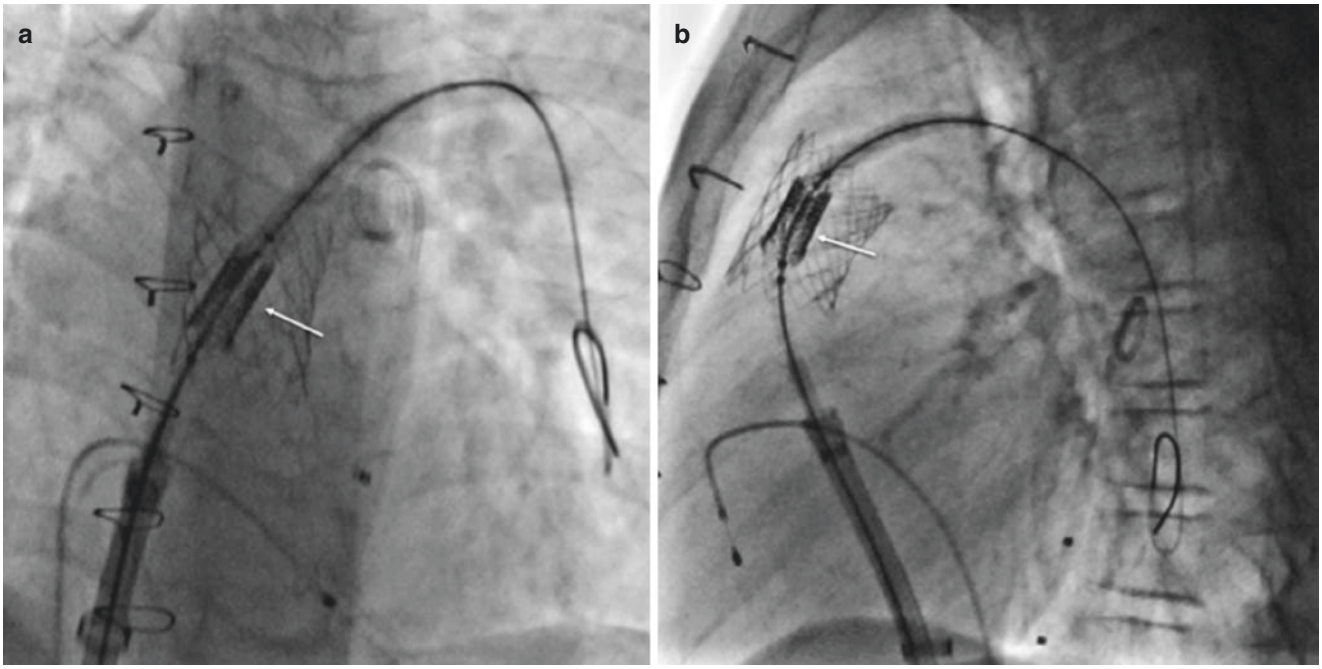



**Fig. 27.12** Cine angiographic/fluoroscopic images in a 13-year-old male patient with dysfunctional 22 mm homograft in two views (frontal: left anterior oblique with cranial angulation and straight lateral). (**a**, **b**) Positioning of a 23 mm Edwards SAPIEN valve in the middle of the landing zone (arrows)

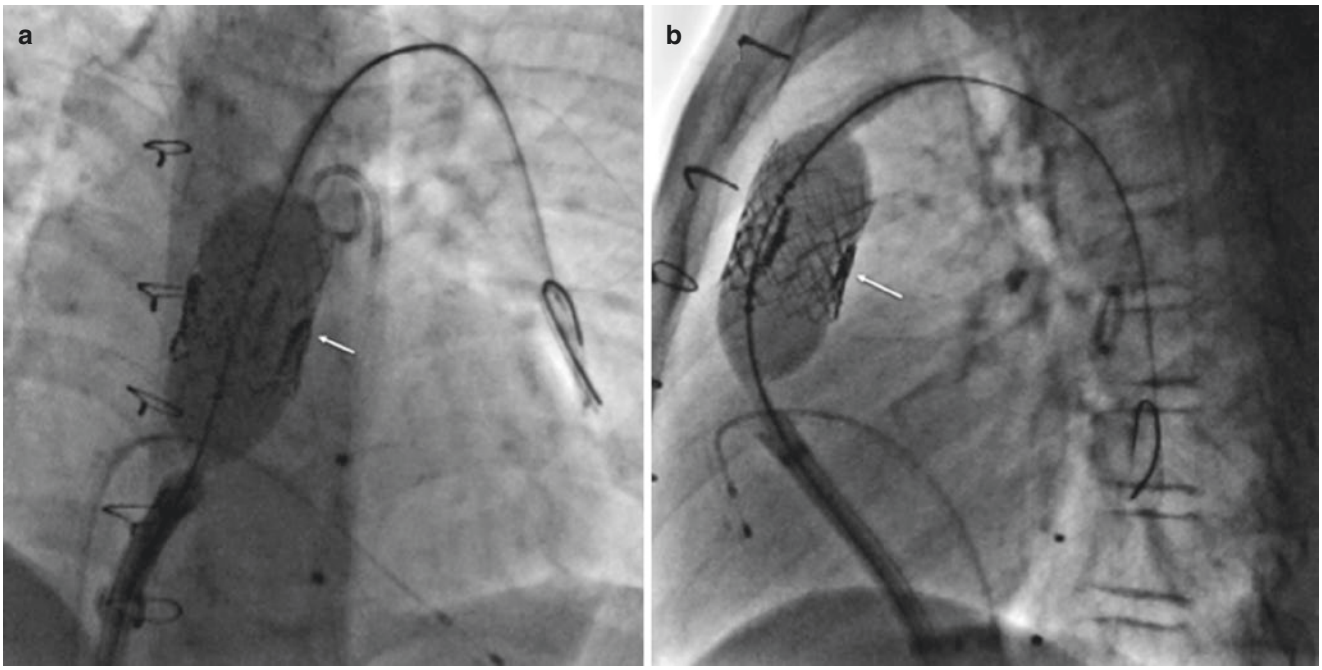



**Fig. 27.13** Cine angiographic/fluoroscopic images in a 13-year-old male patient with dysfunctional 22 mm homograft in two views (frontal: left anterior oblique with cranial angulation and straight lateral). (**a**, **b**) Full inflation of the balloon/valve assembly (arrows)

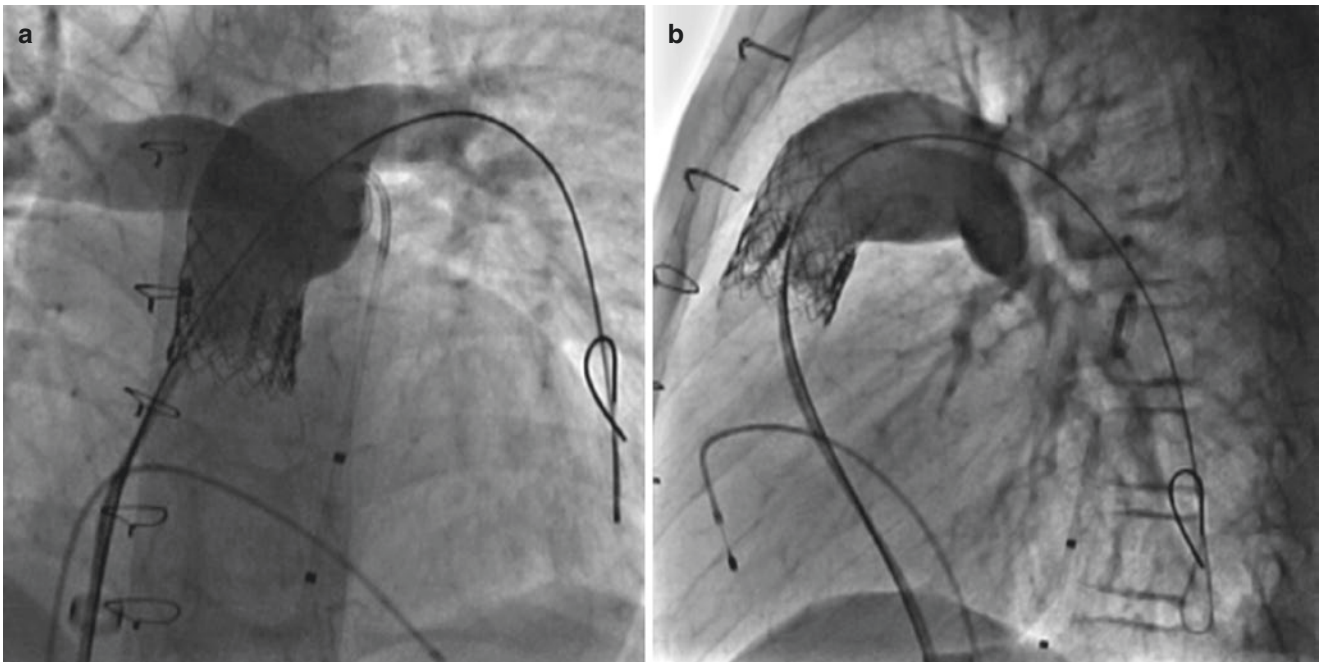



**Fig. 27.14** Cine angiographic/fluoroscopic images in a 13-year-old male patient with dysfunctional 22 mm homograft in two views (frontal: left anterior oblique with cranial angulation and straight lateral). (**a**, **b**) Angiogram after valve has been deployed showing good valve position, no regurgitation, and no complications

- Ascending aortic root or selective left coronary angiography in steep caudal angulation is performed to assess the flow in the left main coronary artery (Fig. 27.15).
- Further, we have been employing intracardiac echocardiography to assess RVOT function before and after valve insertion. We have found this tool to be helpful in mainly assessing the exact degree of pulmonary insufficiency after removal of guide wire. Figure 27.16a–c demonstrates images of the conduit after stenting showing the degree of regurgitation using color Doppler and continuous wave Doppler and Fig. 27.16d–f in the same patient after implantation of a SAPIEN valve demonstrating trivial if any regurgitation.
- Given the large size of the sheath, it is recommended that venous hemostasis be achieved utilizing 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 use the "figure of 8" suture effectively. The Vicryl 0 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.

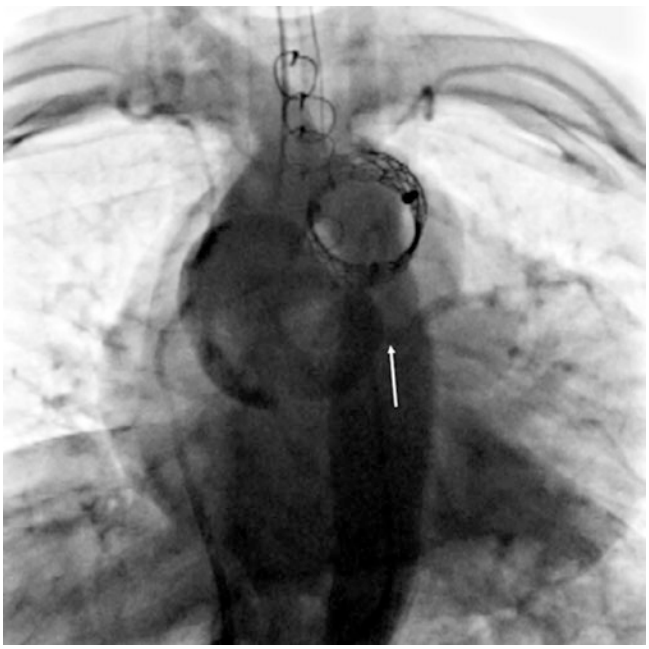


**Fig. 27.15** Cine angiographic/fluoroscopic images in a 13-year-old male patient with dysfunctional 22 mm homograft in two views (frontal: left anterior oblique with cranial angulation and straight lateral). Aortic root angiogram after valve has been deployed in step caudal angulation showing normal flow in the left main and anterior descending coronary arteries (arrow)

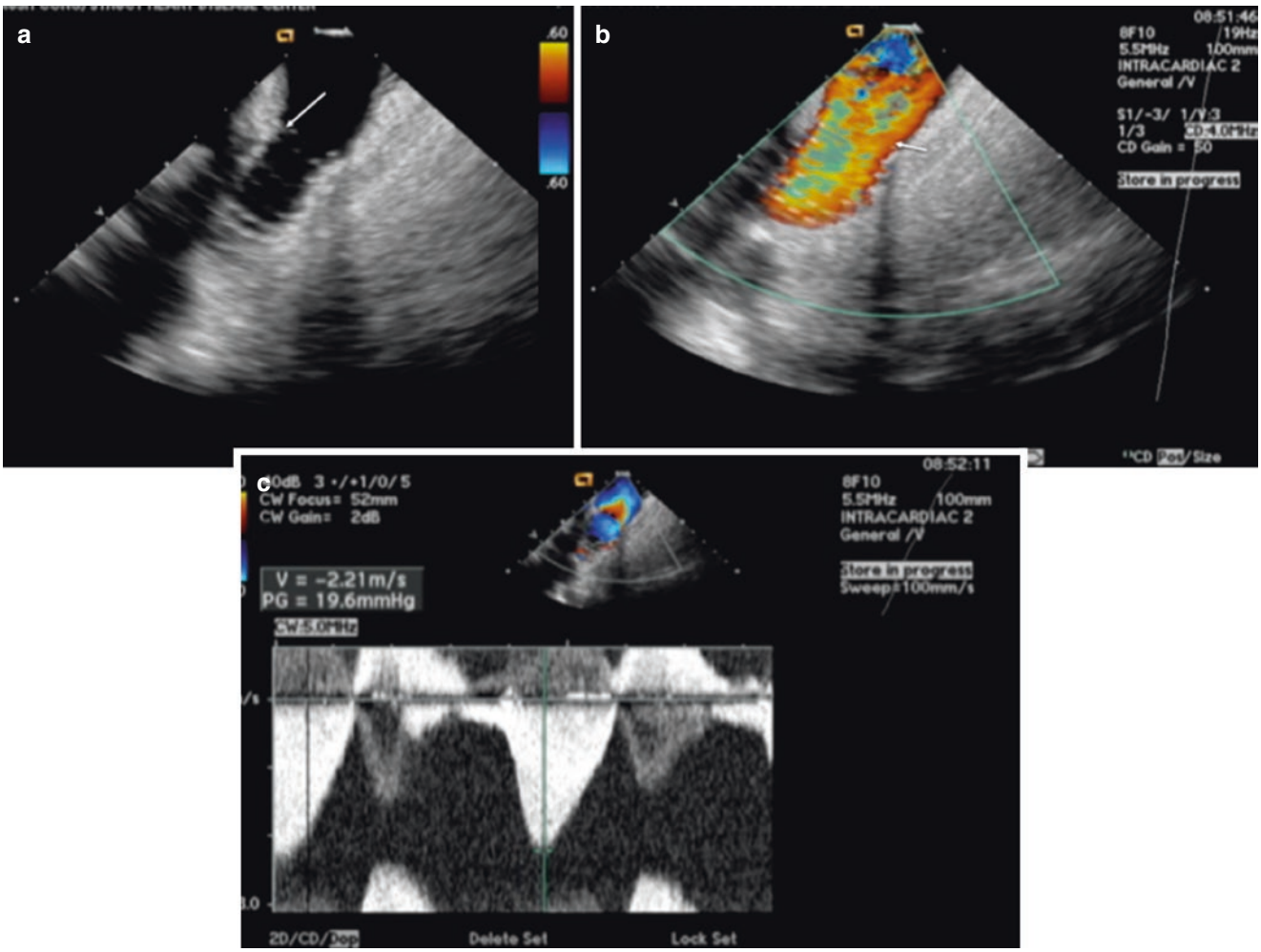



**Fig. 27.16** Intracardiac echocardiographic (ICE) images in a patient with dysfunctional homograft who already had a stent implanted in the homograft as a landing zone pre-valve (**a**, **b**, **c**) and post-valve (**d**, **e**, **f**) implantation. (**a**, **b**) Views from the right ventricular outflow tract without and with color Doppler showing the stent (arrow) and free regurgitation (arrow). (**c**) Continuous wave Doppler in the right ventricular outflow tract showing peak gradient of about 20 mmHg with reversal of flow indicating significant regurgitation. (**d**, **f**) After 23 mm valve has been implanted showing the valve leaflets (arrow) and trivial if any regurgitation (arrow). (**g**) Continuous wave Doppler showing peak gradient of 14 mmHg and no flow reversal

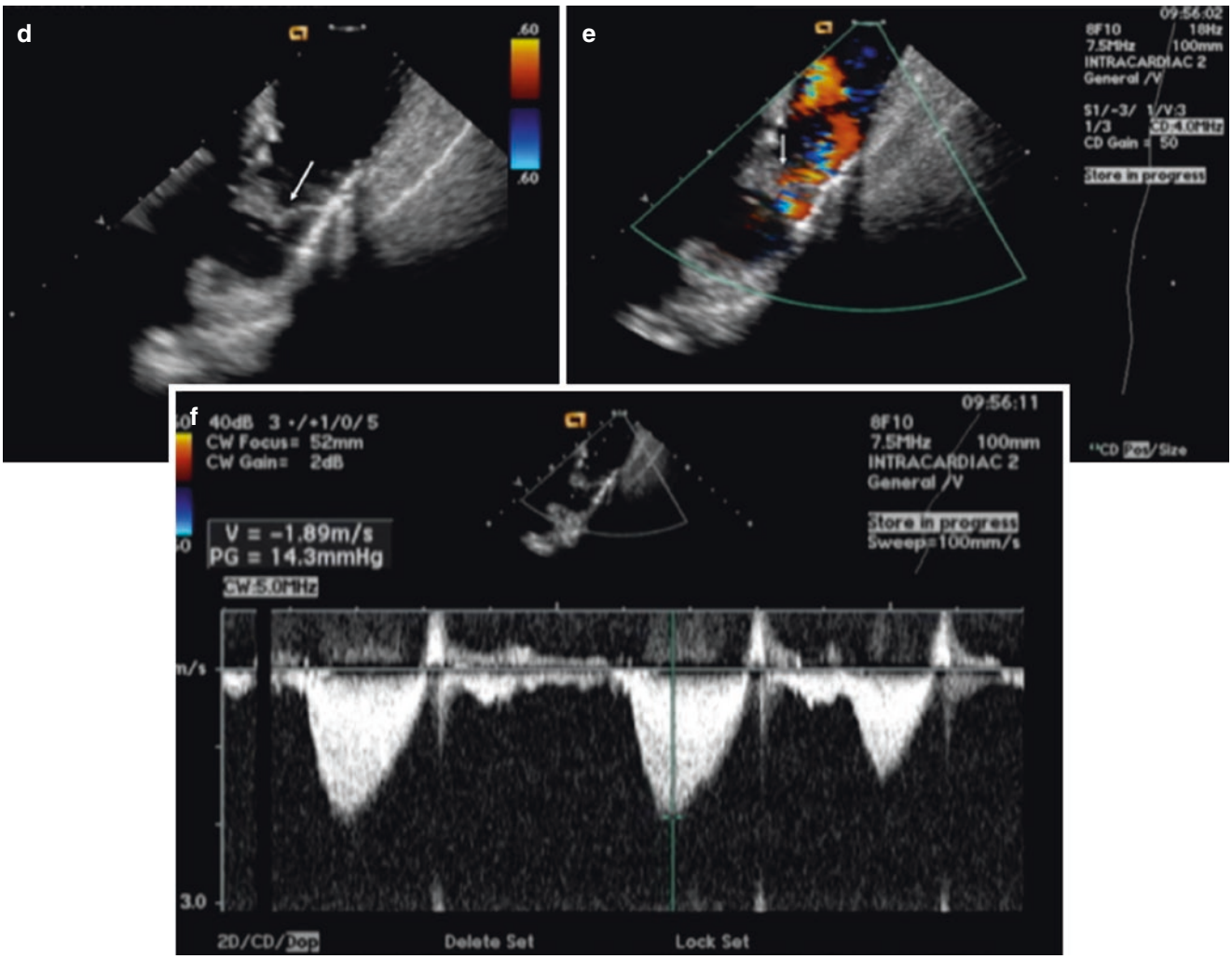



**Fig. 27.16** (continued)

## 27.5 Post-procedural Care

Patients are usually kept for observation overnight and discharged home the following day on 81 mg aspirin for 1 year and are asked to observe SBE prophylaxis precautions indefinitely. 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.

## 27.6 Follow-Up

Follow-up examination and echocardiography are performed at 1, 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 adherence to SBE prophylaxis is enforced.

# Percutaneous Tricuspid Valve Implantation (PTVI)

# 28

Andreas Eicken and Peter Ewert

## 28.1 Introduction

Tricuspid valve dysfunction may lead to surgical tricuspid valve replacement if a valve plasty is not successful. Usually, in patients with congenital heart disease, a biological valve prosthesis is chosen in tricuspid position, which has a limited durability. Once a repeated tricuspid valve replacement is indicated, percutaneous tricuspid valve implantation (PTVI) is a feasible and safe alternative to repeated cardiac surgery. The previously implanted bioprosthesis serves as a perfect landing zone for the percutaneous valve (valve-in-valve implantation). So far, two commercially available percutaneous valves have been used in tricuspid position, the Melody valve (Medtronic Inc., Minneapolis, MN) and the Edwards Sapien XT/Sapien 3 valves (Edwards Lifesciences LLC, Irvine, CA). Both valves have been described in detail previously and received FDA approval for implantation in pulmonic position (Melody valve and Sapien XT valve). Since until today there is no approval for percutaneous tricuspid valve implantation, this intervention is performed as an off-label procedure, which needs to be discussed with the patients or their guardians. Our own experience in Munich comprises 19 percutaneous tricuspid valve implantations.

## 28.2 Patient Selection and Pathophysiology

A bioprosthesis in tricuspid position may develop progressive stenosis, regurgitation or a combination of both. Echocardiography (transthoracic or transesophageal) (Fig. 28.1 and Video 1) allows grading of valve regurgitation and assessment of the mean Doppler diastolic inflow gradient through the tricuspid valve. In tricuspid valve dysfunction, right atrial and right ventricular enlargement may develop. If left untreated, right heart failure (atrial flutter/fibrillation, liver congestion, effusions) with reduced exercise capacity and reduced NYHA functional class may result. At cardiac catheterization a diastolic inflow gradient >5 mmHg (atrial a-wave to right ventricular end-diastolic pressure) may be present indicating valve stenosis, or a prominent right atrial v-wave is the hallmark of severe tricuspid valve regurgitation. Serial examinations (clinical, echocardiography, cardiac MRI with indexed right atrial and right ventricular volumes and right ventricular ejection fraction, exercise test with $VO_2$ max) help to determine the optimal time for PTVI.

## 28.3 Procedure

Femoral venous access is the preferred route for percutaneous tricuspid valve implantation. However, the right jugular vein may also be used. During continuous sedation, the femoral vein and artery are cannulated. Heparin (in adults 5000 units, in children 100 units/kg up to a maximum of 5000 units) is given to keep the activated clotting time above 200 s. A dose of a first- or second-generation cephalosporin (e.g. cefuroxime) is given followed by two additional doses 8 and 16 h later after successful PTVI. A complete hemodynamic assessment with pressure registration in the right atrium, right ventricle, aorta ascendens and left ventricle is performed. If no shunt is present, oximetry may not be

The original version of this chapter was revised. A correction to this chapter can be found at https://doi.org/10.1007/978-3-319-72443-0_46

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_28) contains supplementary material, which is available to authorized users.

A. Eicken (✉)
Department of Pediatric Cardiology and Congenital Heart Disease
German Heart Center Munich, Technische Universität München, Munich, Germany
e-mail: eicken@dhm.mhn.de

P. Ewert
Klinik für Kinderkardiologie und angeborene Herzfehler
Deutsches Herzzentrum München, Munich, Germany



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_28

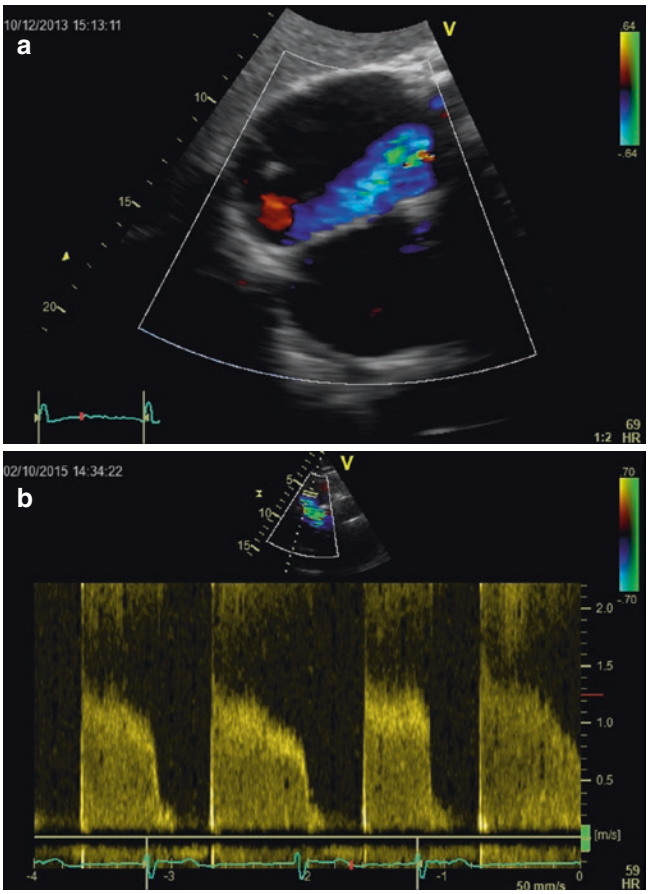



**Fig. 28.1** A 73-year-old female patient with Ebstein's anomaly of the tricuspid valve (TrV) presented in NYHA III functional class. After a surgical tricuspid valve plasty, a DDD-pm was inserted in 1995, in 2006 a bioprosthesis (Carpentier-Edwards Perimount 33 mm) was implanted. Now (2015) this prosthesis showed severe regurgitation with subsequent enlargement of the right atrium (**a**). Additionally, a mean Doppler diastolic inflow gradient of 5–6 mmHg developed over time (**b**), and the indication for bioprosthetic tricuspid valve replacement for regurgitation and stenosis was assessed. A surgical replacement of the TrV was thought to be associated with an increased risk by the surgical team. A percutaneous tricuspid valve implantation (PTVI) was scheduled

**Video 1** A 73-year-old female patient with Ebstein's anomaly of the tricuspid valve (TrV) presented in NYHA III functional class. After a surgical tricuspid valve plasty, a DDD-pm was inserted in 1995, in 2006 a bioprosthesis (Carpentier-Edwards Perimount 33 mm) was implanted. Now (2015) this prosthesis showed severe regurgitation with subsequent enlargement of the right atrium (**a**). Additionally, a mean Doppler diastolic inflow gradient of 5–6 mmHg developed over time (**b**), and the indication for bioprosthetic tricuspid valve replacement for regurgitation and stenosis was assessed. A surgical replacement of the TrV was thought to be associated with an increased risk by the surgical team. A percutaneous tricuspid valve implantation (PTVI) was scheduled (AVI 1517 kb)

necessary. A superstiff guide wire (i.e. Meier wire, Boston Scientific Corp, Quincy MA, USA; Amplatz ultrastiff 0.035′, Boston Scientific Corp; Lunderquist 0.035′, Cook Medical, Bloomington IN, USA) is positioned preferably

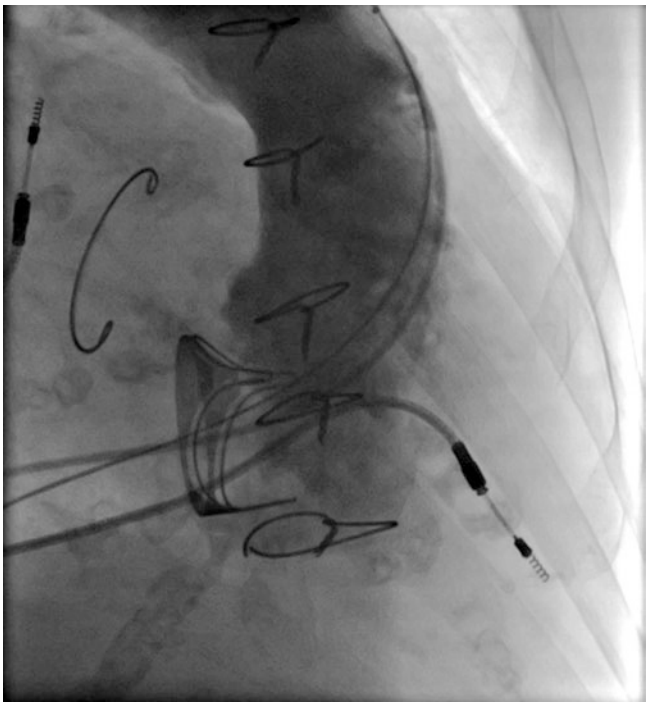


**Fig. 28.2** Angiography into the right ventricle with a 6F multitrack catheter (NuMED, Hopkinton NY, USA) over a superstiff guide wire (0.035′ Meier wire, Boston Scientific Corp, Quincy MA, USA)

**Video 2** Angiography into the right ventricle with a 6F multitrack catheter (NuMED, Hopkinton NY, USA) over a superstiff guide wire (0.035′ Meier wire, Boston Scientific Corp, Quincy MA, USA) (WMV 2659 kb)

distally into the left pulmonary artery. Following this, an angiography into the right ventricle is performed (Fig. 28.2 and Video 2). If the wire tip is located within the right ventricle, a safe and controlled tricuspid valve delivery is harder to achieve, since the wire position is not as stable as in a pulmonic position.

Then, although a valve-in-valve delivery can be performed without a "balloon test" if the bioprosthesis is well known, we tend to perform a balloon test for the final decision which percutaneous valve is best suited. The balloon should be at least 1–2 mm larger than the suspected internal diameter of the bioprosthesis. The balloon is advanced over the wire into the tricuspid valve and inflated until a waist is visible (Fig. 28.3 and Video 3). In general, if the bioprosthesis is well known and a Sapien valve is chosen, pre-stenting is not necessary. The percutaneous valve should also be at least 1–2 mm larger than the known internal diameter of the "landing zone" bioprosthesis.

Then a valve is selected. The venous sheath is upsized (14 Fr for Sapien 23 and 26 mm valves and 16 Fr for the Sapien 29 mm valve). Care needs to be taken for correct orientation of the Sapien valve in tricuspid position, since in PTVI the valve needs to be positioned 180° opposite to the regular

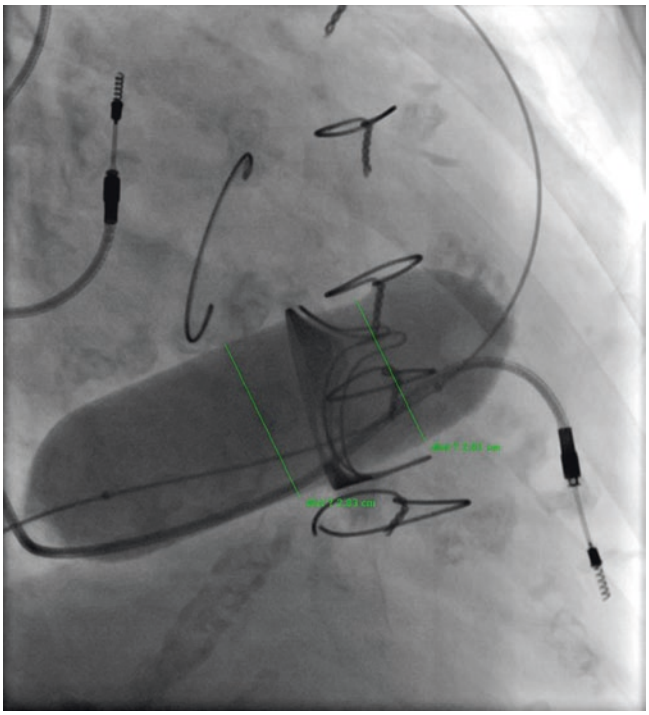


**Fig. 28.3** Balloon test with a 30 × 60 mm VACS balloon (Osypka Rheinfelden, Germany) in a 33 mm Carpentier-Edwards Perimount valve. An indentation is seen at 26 mm diameter

**Video 3** Balloon test with a 30 × 60 mm VACS balloon (Osypka Rheinfelden, Germany) in a 33 mm Carpentier-Edwards Perimount valve. An indentation is seen at 26 mm diameter (WMV 1359 kb)

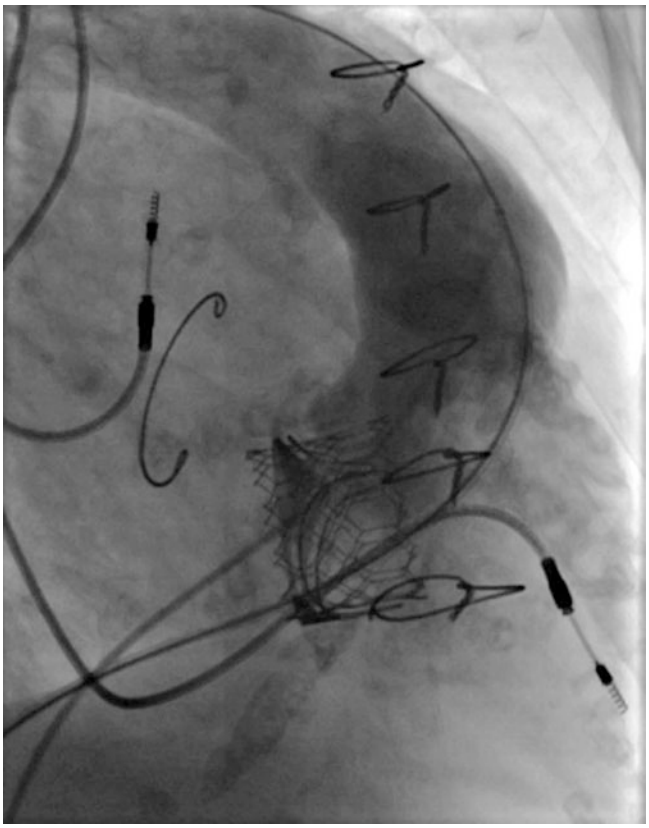


**Fig. 28.4** After delivery of a Sapien S3 29 mm directly into the Carpentier-Edwards 33 bioprosthesis, the proximal Sapien stent struts just peak into the right atrium, while most of the valve points towards the right ventricle

**Video 4** After delivery of a Sapien S3 29 mm directly into the Carpentier-Edwards 33 bioprosthesis, the proximal Sapien stent struts just peak into the right atrium, while most of the valve points towards the right ventricle (WMV 8516 kb)

**Video 5** After delivery of a Sapien S3 29 mm directly into the Carpentier-Edwards 33 bioprosthesis, the proximal Sapien stent struts just peak into the right atrium, while most of the valve points towards the right ventricle (WMV 1716 kb)

**Video 6** After delivery of a Sapien S3 29 mm directly into the Carpentier-Edwards 33 bioprosthesis, the proximal Sapien stent struts just peak into the right atrium, while most of the valve points towards the right ventricle (WMV 2619 kb)

orientation in a TAVI aortic procedure. The venous sheath is positioned into the inferior vena cava. Then the NovaFlex+ delivery system is inserted through the sheath via the loader until the valve exits the sheath. The balloon is then pulled back until the valve is positioned properly on the balloon between the two markers and the system is fixed. The valve is then advanced into the bioprosthesis. Once the Sapien valve is located across the bioprosthesis, slow balloon inflation is done by the second operator to ensure optimal positioning of the valve across the bioprosthesis. The first operator can manipulate the valve position during slow inflation by pushing or pulling on the guide wire and by keeping the valve in a horizontal orientation. This enables optimal positioning. At final valve delivery, the proximal Sapien stent struts should peak into the right atrium and most of the Sapien valve should point towards the right ventricle (Fig. 28.4 and Videos 4, 5, and 6). A final angiogram is done to document the result. Finally, right atrial, right ventricular and aortic pressures are assessed.

A transoesophageal echocardiogram may be done to depict valve function and additionally to assess the diastolic Doppler inflow gradient through the newly implanted valve (Fig. 28.5).

If a Melody valve is used, the vein needs to be dilated with a 22 Fr dilator to accommodate the 22 Fr delivery system. If the valve diameter is not known, a balloon test together with selective RCA depiction is indicated preceding PTVI (Figs. 28.6 and 28.7). For the Melody valve, prestenting is usually done to create a safe landing zone for the valve (Figs. 28.8 and 28.9) (Video 7). If the landing zone is short, the Melody valve may be folded and thus shortened to prevent coronary arterial compression.

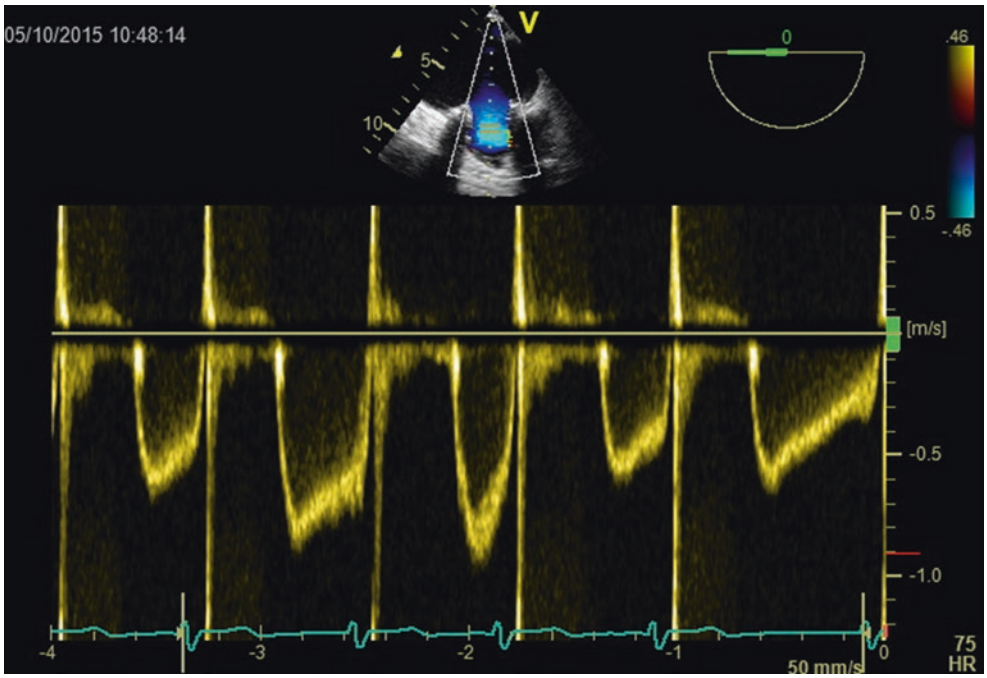



**Fig. 28.5** Transoesophageal echocardiography directly after PTVI with a Sapien S3 29 mm into a Carpentier-Edwards Perimount 33 mm valve, showing unrestricted diastolic TrV inflow and depicting the absence of any tricuspid regurgitation. Follow-up now 18 months post PTVI still shows excellent valve function, no regurgitation and no increased Doppler inflow gradient

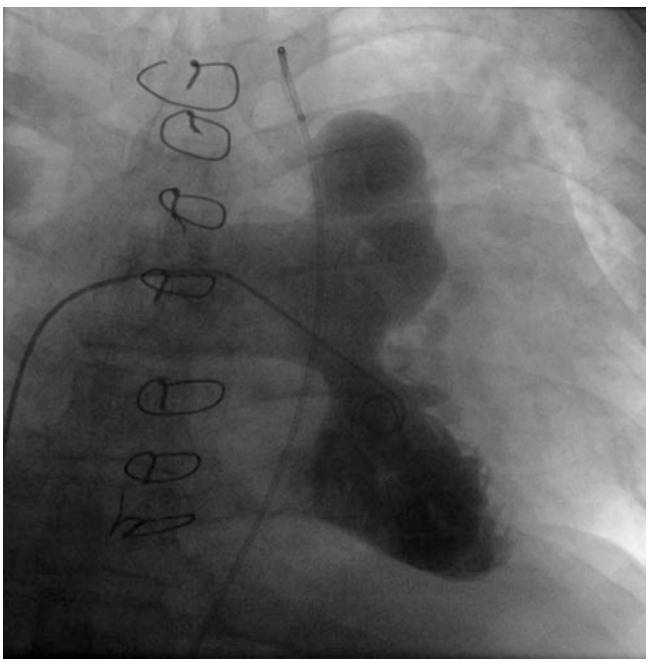


**Fig. 28.6** Thirty-eight-year-old patient with tricuspid atresia after a Fontan RA-RV anastomosis (Björk modification) with a homograft done in 1994. In 2007 the stenotic and calcified homograft was exchanged for a 25 mm homograft. Now the patient presents in NYHA III with atrial fibrillation and maximal "tricuspid" regurgitation with severe right atrial enlargement. RAP is $v = 21$, $m = 18$ mmHg. RVP is 27/5/11; hence, severe stenosis and regurgitation of the homograft are assessed. Repeated surgery was classified to be at an increased risk

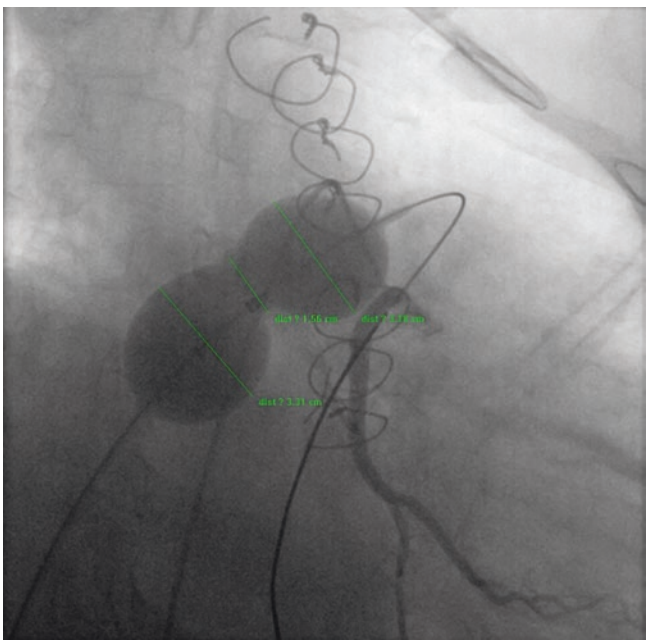


**Fig. 28.7** A balloon test was done with a 34 mm sizing balloon (St. Jude Medical St Paul, MN, USA) on a 0.035 superstiff guide wire. It shows a waist at 16 mm together with a rather close neighbourhood of the right coronary artery

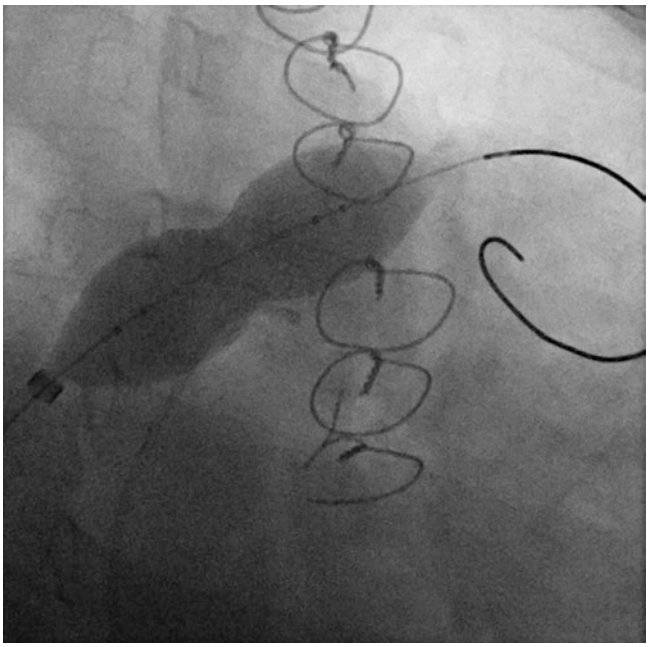


**Fig. 28.8** An EV3 MaxLD 26 mm (Medtronic Inc., Minneapolis, MN) stent was mounted on a 22 mm balloon-in-balloon catheter (Numed, Hopkinton NY, USA) and implanted into the stenotic homograft

The implantation of multiple covered CP stents (NuMED, Hopkinton NY, USA) may help to downsize the internal tricuspid valve diameter of a large bioprosthesis, since the outer diameter of a Melody valve is only 24 mm on a 22 mm BiB delivery system. In most of our patients, the Melody valve was delivered with a 22 mm delivery system into the tricuspid valve bioprosthesis. Folding of the distal and proximal Melody stent struts shortens the valve and adds another 1–2 mm of valve profile (Fig. 28.10). It may be cumbersome to load the Melody valve into the delivery system after folding. Dilatation of the ensemble helps to solve this issue (Figs. 28.11 and 28.12) (Video 8). Delivery of the Melody valve on a 24 mm balloon-in-balloon catheter results in an external diameter of 26 mm but is not generally recommended because valve regurgitation may result at this valve diameter. However, good valve function is documented in patients in whom a Melody valve was dilated to 24 mm internal valve diameter, as in our patient example (Figs. 28.13 and 28.14) (Video 9).

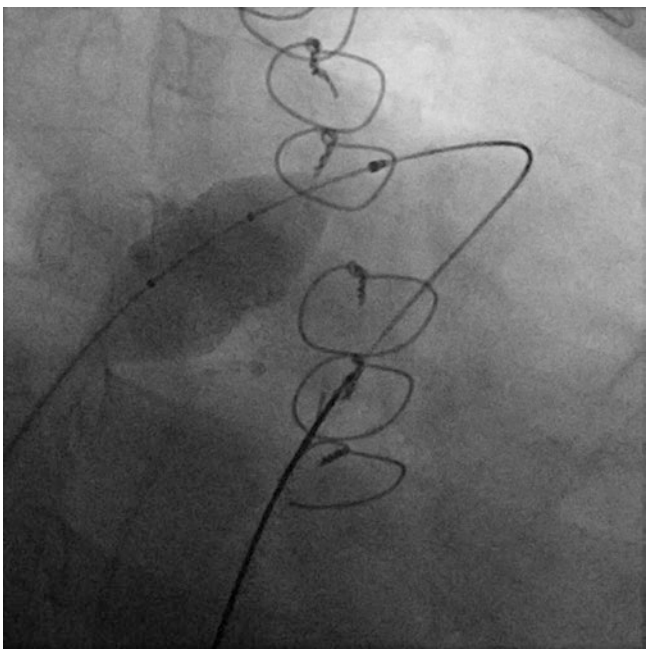


**Fig. 28.9** The stent was dilated with a 22 × 20 mm Atlas high-pressure balloon (Bard Tempe, AZ, USA)

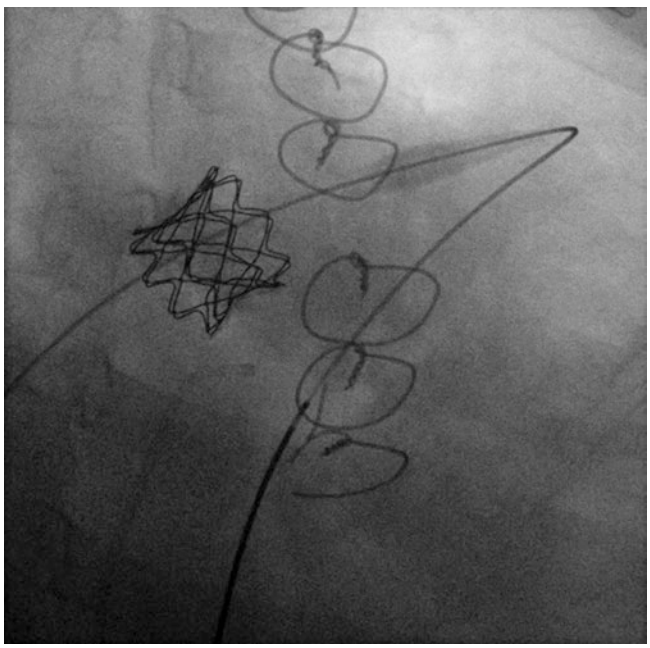


**Fig. 28.10** A double-folded Melody valve was crimped on a 22 mm delivery system and implanted into the MaxLD stent

**Video 7** An EV3 MaxLD 26 mm (Medtronic Inc., Minneapolis, MN) stent was mounted on a 22 mm balloon-in-balloon catheter (Numed, Hopkinton NY, USA) and implanted into the stenotic homograft (AVI 442662 kb)

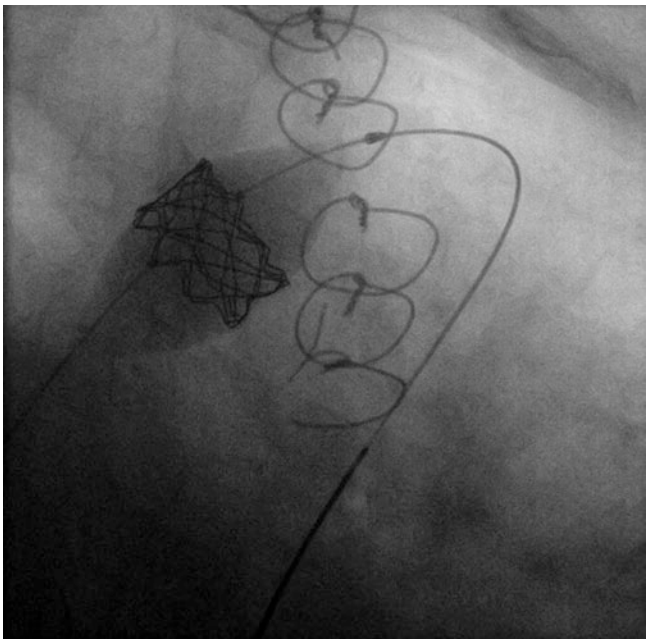


**Fig. 28.11** The Melody valve was dilated with a 24 × 20 mm Atlas balloon

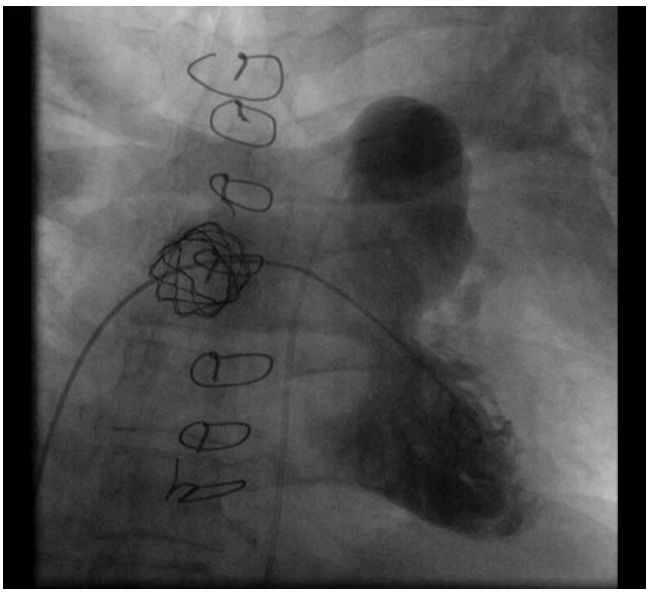


**Fig. 28.12** After the intervention, no residual tricuspid regurgitation is present

**Video 8** After the intervention, no residual tricuspid regurgitation is present (AVI 707144 kb)

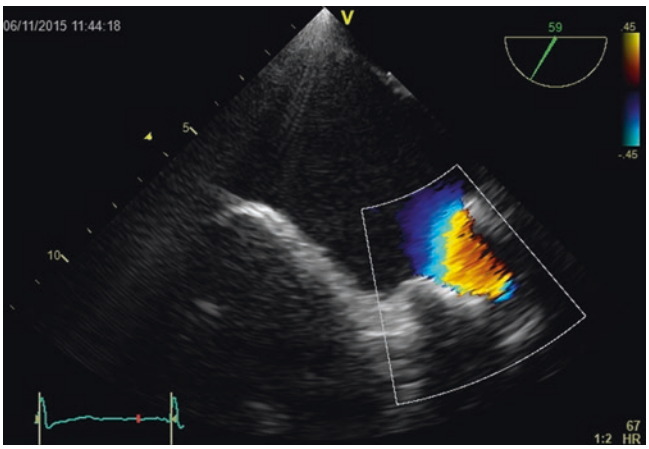



**Fig. 28.13** Transoesophageal echocardiography shows normal diastolic TrV inflow and absent tricuspid regurgitation

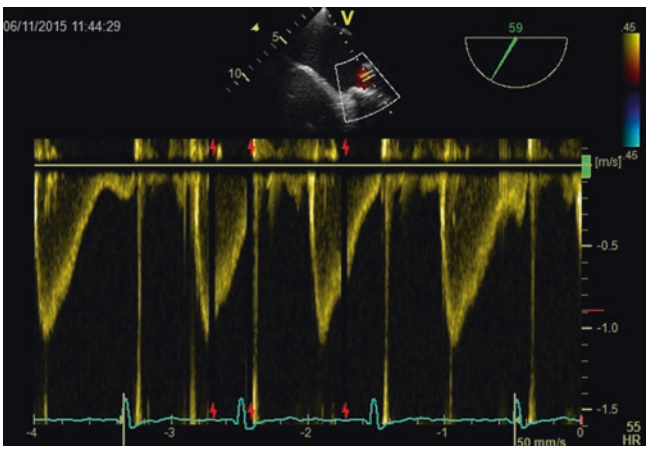



**Fig. 28.14** The Doppler inflow signal has normalized after Melody valve implantation into the stenotic homograft. A repeated TEE 14 months later shows good Melody valve function

**Video 9** Transoesophageal echocardiography shows normal diastolic TrV inflow and absent tricuspid regurgitation (AVI 5956 kb)

## 28.4 Post-procedural Care

After PTVI we keep our patients on heparin (10.000 units/$m^2$, aPTT 40–60) until the next day. Then aspirin (100 mg/d) is initiated for 6 months, and two doses of a cephalosporin are given. On the next day, echocardiography is performed. After 6 months an exercise test with assessment of $VO_2$ max is scheduled together with a cardiac MRI examination.

# Part VIII

# Step-by-Step Procedures: Principles of Hybrid Approach

# Hypoplastic Left Heart Syndrome: The Giessen Hybrid Approach

29

Dietmar Schranz and Hakan Akintuerk

The treatment of newborns with hypoplastic left heart syndrome (HLHS) follows a well-established classical three-step algorithm for most institutions worldwide. In Giessen, the hybrid approach was developed as a primary palliation for newborns with HLHS and later on for patients with left heart complex (HLHC). The hybrid procedure has moved from a rescue approach to an alternative modality of a Norwood palliation.

The "Giessen hybrid" stage I consists of surgical bilateral pulmonary artery banding combined with percutaneous stenting of the arterial duct and atrial septum manipulation, if necessary.

The physiological objectives of the hybrid approach are similar as for the classical Norwood procedure by alternative techniques in controlling pulmonary blood flow, whereas unobstructed systemic perfusion is maintained via an open arterial duct.

This strategy involves off-pump bilateral pulmonary artery banding (bPAB) and interventional stenting or continuous prostaglandin therapy leaving patent the ductus arteriosus; the hybrid approach is mostly, but not exclusively, performed in the neonatal period (hybrid stage I).

Aortic arch reconstruction using cardiopulmonary bypass combined with a superior cavo-pulmonary connection summarized as comprehensive stage II or as indicated a biventricular correction is deferred until the age of 4–5 in univentricular palliation or to the end of infancy for cardiac repair.

The hybrid approach could be established as a highly effective treatment in particular for newborns with cardiovascular collapse and even for premature babies or neonates too small for gestational age. However, the team has to be familiar and presupposed with the hybrid approach, which includes the technical skills and the required materials in the stock of the catheter laboratory.

The here presented data and technique of hybrid stage I therapy performed for all types of HLHS, HLCC, and variants was applied in a standard manner with very few variations based on our learning curve and due to novel developments of material for duct stenting and IAS manipulation.

## 29.1 Diagnostic Tools Prior to Giessen Hybrid

*Fetal echocardiography*, the outlook for newborns with hypoplastic left heart (HLH), has substantially improved. One decisive reason is an established fetal diagnostic and prenatal therapeutic program. The overall hospital survival on an intention-to-treat basis is meanwhile above 90%. In summary, prenatal diagnostics allows to offer hybrid approach with low procedural mortality and with high success rate. However, an obstructive interatrial septum associated with lymphangiectasia is still in most patients non-treatable even if a biventricular repair is performed by cardiac transplantation.

*Neonatal two-dimensional echocardiography* allows detailed anatomic diagnosis of HLHS with its four anatomical variants (MS, mitral stenosis; MA, mitral atresia; AS, aortic stenosis; AA, aortic atresia) and the different forms of HLHC including the analysis of their cardiovascular function. Magnetic resonance imaging (MRI) and heart catheterization are added to answer special anatomical and functional details. Especially, *cardiac magnetic resonance imaging (CMRI)* performed in only sedated newborns as an elective, noninvasive approach becomes in our institution an important tool to analyze exactly the aortic arch and the junction of duct to descending aorta as well as to exclude functional and myocardial perfusion deficits.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_29) contains supplementary material, which is available to authorized users.

D. Schranz (✉) · H. Akintuerk
Pediatric Heart Center, Justus-Liebig University Clinic, Giessen, Germany
e-mail: dietmar.schranz@paediat.med.uni-giessen.de



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_29

### 29.1.1 Surgical and Interventional Aspects of the Hybrid Stage I

The theoretical aspects of the surgical-interventional approach are just described elsewhere.

In terms of the physiopathology, hybrid approach consisting of bilateral pulmonary banding (bPAB), percutaneous duct stenting, or in some by long-term prostaglandin infusion, atrio-septostomy including stent placement is performed for lung protection, preserving adequate systemic perfusion and unloading of the atrium, respectively.

*Bilateral pulmonary arterial banding* is performed by median sternotomy followed by subtotal thymectomy and limited cranial pericardiotomy of 3–4 cm in length. Two pieces of a 3.5 mm PTFE tube are cut in bands with a width of about 1–2 mm to perform a bilateral PAB in patients with body weight of above 3 kg and 3 mm PTFE bands in lower weight, respectively. One of the most important surgical features of an effective bilateral PAB is depending on the **width** of the band! A band with bandwidths of >2 mm has the potential to induce a hypoplastic main pulmonary branch, in opposite to a small band (less <2 mm), which create a poststenotic vessel dilatation! Additionally, the bands have to be fixed at the adventitia by using a 6/0 prolene suture to avoid distal migration with the risk of bifurcation obstruction.

## 29.2 *Stenting of the Ductus Arteriosus* (SDA)/Manipulation of the Interatrial Septum (IAS)

In our institution, IAS manipulation and SDA are carried out by percutaneous transcatheter procedure, predominantly as an elective approach, when surgical PAB is always bilaterally placed.

### 29.2.1 IAS Manipulation and Duct Stenting Step by Step

#### 29.2.1.1 Preparation for the Transcatheter Part of Hybrid Procedure

The newborn's food is stopped mostly 4–6 h prior to the catheter procedure; instead continuous infusion of 10% glucose with physiologically substituted electrolytes is applied as a continuous fluid substitution. The newborn has to arrive the cath. lab. relaxed, best sleeping, and if necessary, by additional glucose10–20% application, dropped in the mouth before sedation, is given (Fig. 29.1a).

The baby is clinically monitored by noninvasive blood pressure measurement, performed at the right arm and continuous oxygen saturation measurement obtained from the left hand (Fig. 29.1b). Prostaglandin infusion (usually dosage of 2–5 ng/kg × min) is not stopped, if self-expandable stents are utilized (Fig. 29.1c). In respect to the prostaglandin infusion with its increased apnea incidence, sedation and analgesia are performed only in a low single dosage of 0.2–0.5/mg × kg of diazepam (rarely midazolam as a second-line tranquilizer) and ketamine, respectively. At first, 1 mg of both drugs is intravenously applied; in most newborns, the application starts with diazepam as a lipid formula to avoid pain at side of the peripheral intravenous cannula (Fig. 29.1d).

#### 29.2.1.2 In-Cath. Echocardiography Prior to Percutaneous Transcatheter Approach (Fig. 29.2)

When the newborn is sleeping, fixed and monitored, the cardiac function and anatomical structures are proved by ECHO, which should include the (retrograde) flow in the aortic arch, the coeliac trunk, as well as the anterior cerebral artery. The responsible physician has to be familiar with all anatomical and functional cardiac details before the catheter approach is

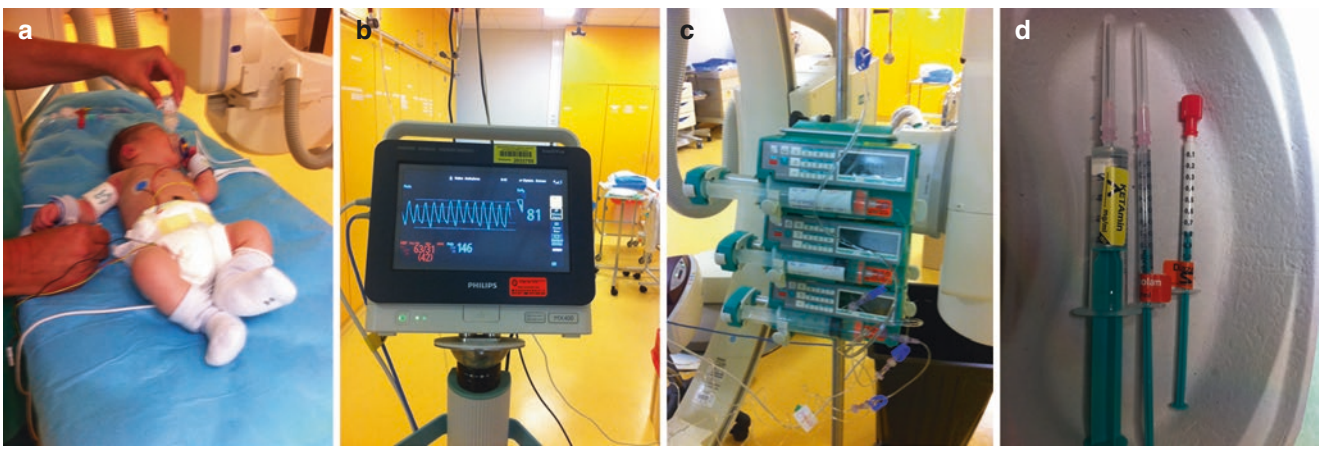



**Fig. 29.1** The baby is fully monitored (**a**, **b**). PGE infusion is continued (**c**). Drugs are prepared for the procedure (**d**)

started: (a) to be aware of all potential complications; (b) to avoid any unnecessary diagnostic catheter-based procedures, i.e., utilizing to much amount of contrast medium or radiation time for diagnostic skills; and (c) to focus on the hybrid stage I completion as duct stenting and interatrial septum (IAS) manipulation, if last necessary. The sequence of the transcatheter procedure depends on the hemodynamics and in term if an elective or high urgency approach is performed. In respect to an elective cardiac catheterization, the IAS manipulation is performed at first.

#### 29.2.1.3 Material and Puncture Side for Transcatheter Part of the Hybrid Approach (Fig. 29.3)

#### 29.2.1.4 Atrial Septum Manipulation

The (in-)adequacy of the atrial septal communication is determined by echocardiography and confirmed by invasive hemodynamic data measurements. In case of a HLHS, the goal is to create an unrestrictive atrial communication; in a HLHC with borderline left heart structures, but with the

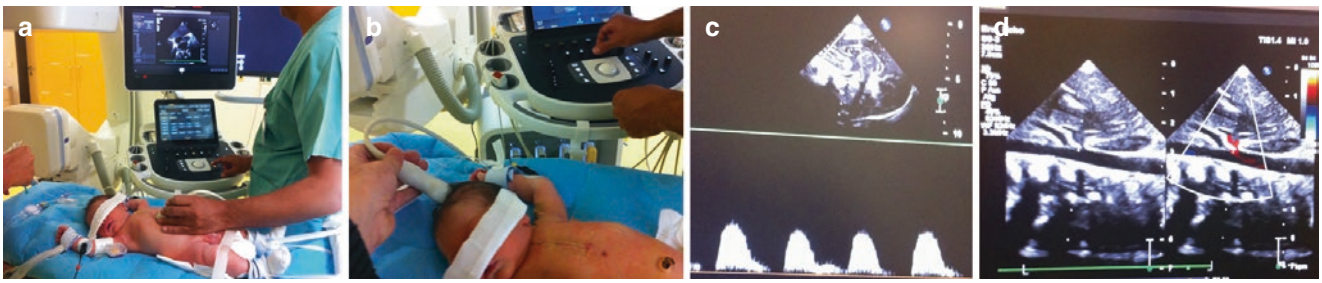



**Fig. 29.2** The newborn is sedated and placed properly over the catheterization table (**a**). Echo is performed (**b**–**d**). See text for details

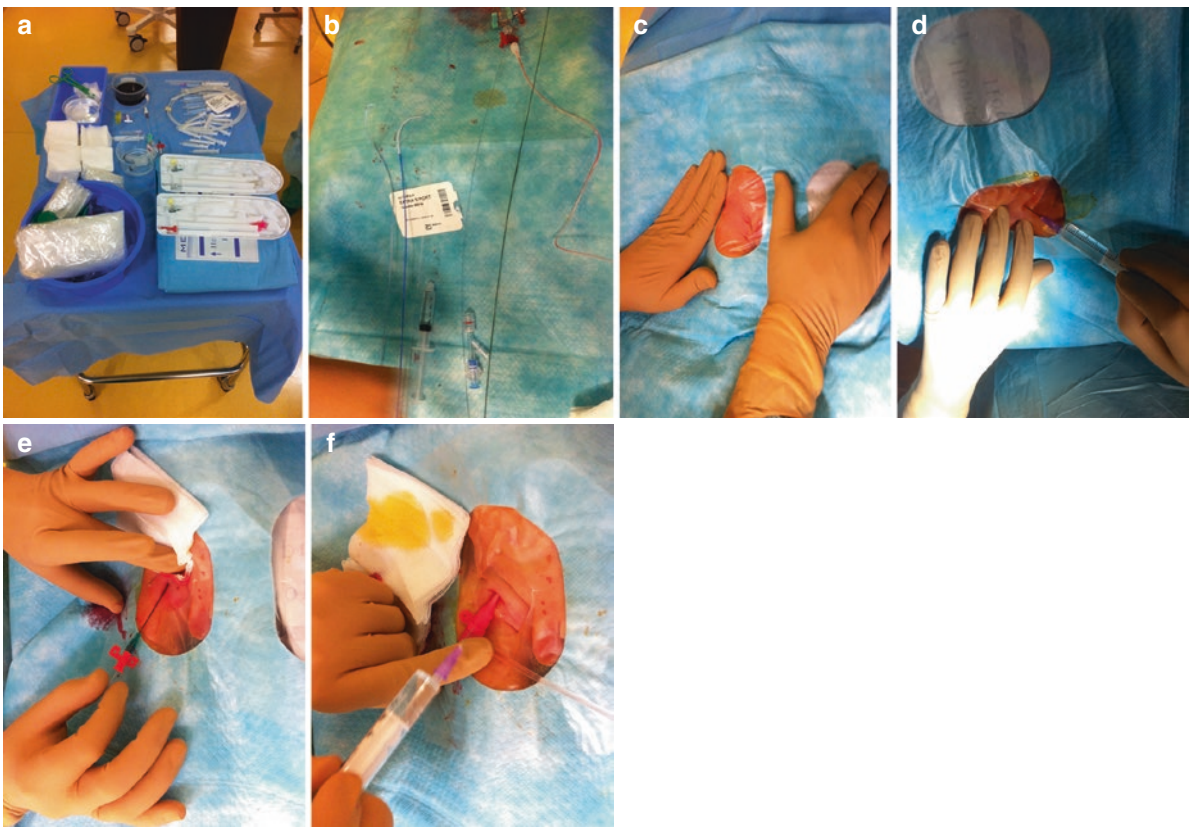



**Fig. 29.3** (**a**, **b**) The material needs to be prepared in advance: in addition to the usual used catheter set, a 4F (femoral artery access) and maybe 6F Terumo sheath (femoral vein access) are used for a provided SAD and Rashkind procedure, respectively. 4F multipurpose and 4F (2.5) right Judkins catheter, balanced middle weight and S'port super stiff 0.014 inch wires, hemostat-ventil, and Luer lock 5 cc syringe are used for probing the atrial septum, the aortic arch, and the arterial duct as well as angiographic evaluation. (**c**–**f**) The inguinal region is prepared by disinfection, local anesthesia is applied in small dosage before vessel puncture by "Seldinger" technique, and local anesthesia augmentation is performed after sheath placement

potential for further grown-up, the IAS need to be remain restrictive to guarantee a sufficient preload, without inducing a significant post-capillary hypertension despite bilateral PAB protection.

If in a HLHS patient, the atrial septal communication is documented as to restrictive (gradient across the septum <7 mmHg), a static IAS dilatation and/or a Rashkind procedure is performed (Fig. 29.4a,b).

The decision to place a stent within the IAS should be preferentially made in advance; stent placement following already manipulated IAS increased the risk of stent embolization. In respect to the risk of embolization, stenting of the atrial septal stent is currently also performed by a self-expandable stent with an open-cell design. Currently, we usually use the SinusSuperFlex (SSF)-DS (15 × 8 mm); a shorter or longer stent length (12 or 18 × 8 mm) is utilized depending on the size of left atrium (Video 1). In a dilated left atrium, a longer (15 or 18mm) self-expandable stent is used to minimize the risk of embolization.

Stenting of the IAS is preferentially performed if an atrial septum defect needs to be newly created by Brockenbrough or alternatively by high-frequency technique. Even an obstructed total anomalous venous return can be connected by catheter technique based on individualized decision making. Depending on the LA size and considering a floppy IAS, the ensemble of delivery system with its SSF-DS should be advanced far within the left atrium guided by a stiff support 0.0014 inch wire (Fig. 29.4c); considering an additional artificial septum pushing by the self-expandable stent delivery system, the stent should be partially developed and pulled back in ensemble with the delivery system to the atrial septum for further stent release

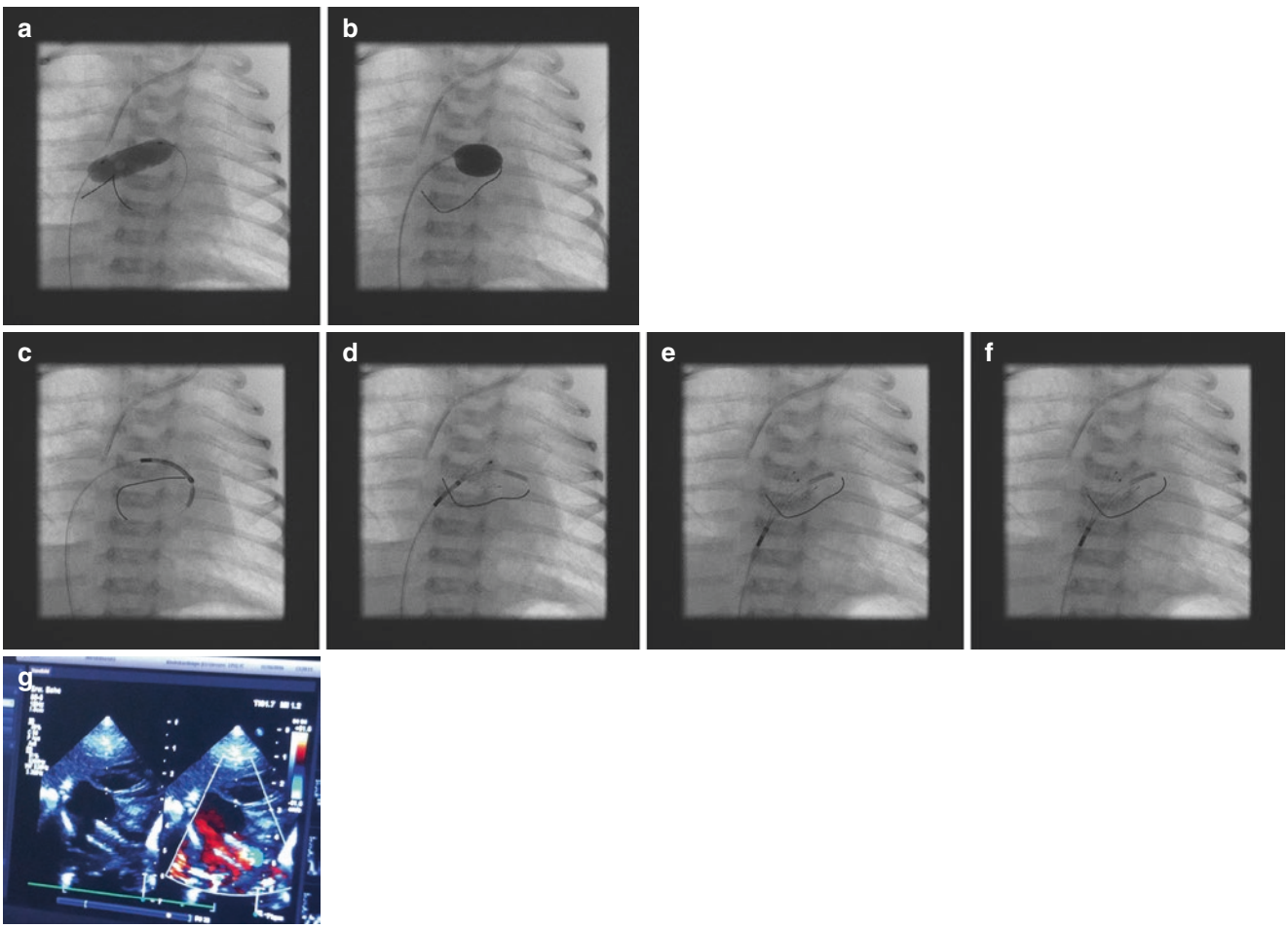



**Fig. 29.4** Shown is an inflated 12 × 20 mm VACS II low-pressure balloon within the IAS (**a**) as well as a 1-mL-filled Rashkind balloon (**b**). (**c**–**f**) Depicts the sequential placement of a SSF-DS within the interatrial septum. (**g**) Depicts the position of the fully self-expanded stent in the atrial septum by a subcostal view. The color Doppler demonstrates the unobstructed flow through the stent

(Fig. 29.4d, e); in doubt, an echocardiographic control of the position should be performed before the stent is fully expanded (Fig. 29.4f). The efficacy and exact position of the interatrial stent are best documented by echocardiography as shown on Fig. 29.4g.

#### 29.2.1.5 Arterial Duct Stenting by Arterial Access

Duct morphology and in particular the junction with the descending aortic arch are firstly delineated by single but repetitive hand injections of contrast medium. The angiographies are performed in right anterior oblique (RAO) 30° and lateral 90° plane, respectively. If an aortic coarctation is present, then balloon angioplasty can be performed (Fig. 29.5).

Choice of stents is largely influenced by the ductal anatomy and the morphology of the duct-aortic junction. According to the anatomic evaluation, choosing stent size and positioning within the duct is also based on right lateral oblique 30° and 90° lateral angiographies (Fig. 29.6).

Currently, SinusSuperFlex-DS stents (OptiMed, Karlsruhe Germany) with a certificated CE mark are delivered through 4F femoral artery sheath; the width of 7, 8, and 9 should be in any case 1–2 mm lager than the smallest measured duct diameter and for avoiding stent migration in the systemic circulation usually bigger than the diameter descending aorta (DAO). Stent length should cover the pretended duct tissue (Video 2). Additional narrowed aortic isthmus or aortic coarctation might be pretreated, but only by ballooning or after DA stenting by stent placement within the CoA in neighborhood or through the open cell struts of the SSF-DS, if necessary (Video 3).

Considering the persistence of a parallel circulation after hybrid stage I, routinely the patients are discharged home on chronic treatment with 1 × 0.1 −0.2 mg/kg bisoprolol and most but not all by lisinopril in the same dosage, respectively; medication of furosemide is routinely avoided. The main two indications to combine these both drugs are to reduce oxygen consumption by avoiding unnecessary high heart rate and consecutively breath rate and to reduce systemic vascular resistance without endangering perfusion pressures; blocking of the neurohumoral activation reduces diastolic left–right shunt across the stented duct (Fig. 29.7).

During the interstage prior to comprehensive stgae II, outpatient follow-up is routinely performed at 1–2-week intervals or earlier depending on the clinical condition; it includes historical information of the parents in particular how the baby is breathing during sleep, weight gain development, systolic and diastolic, never mean blood pressure measurements at the right arm and of the leg, which is not used by catheterization before, pulse oximetry at arm and one leg (HLHC!), respectively; after these mandatory information echocardiographic data are obtained. Patients with a hybrid approach are not referred for elective complete invasive hemodynamic and angiographic evaluation, but if a hemodynamic issue is suspected. CMRI is used in addition to the echocardiography assessment but as an elective imaging

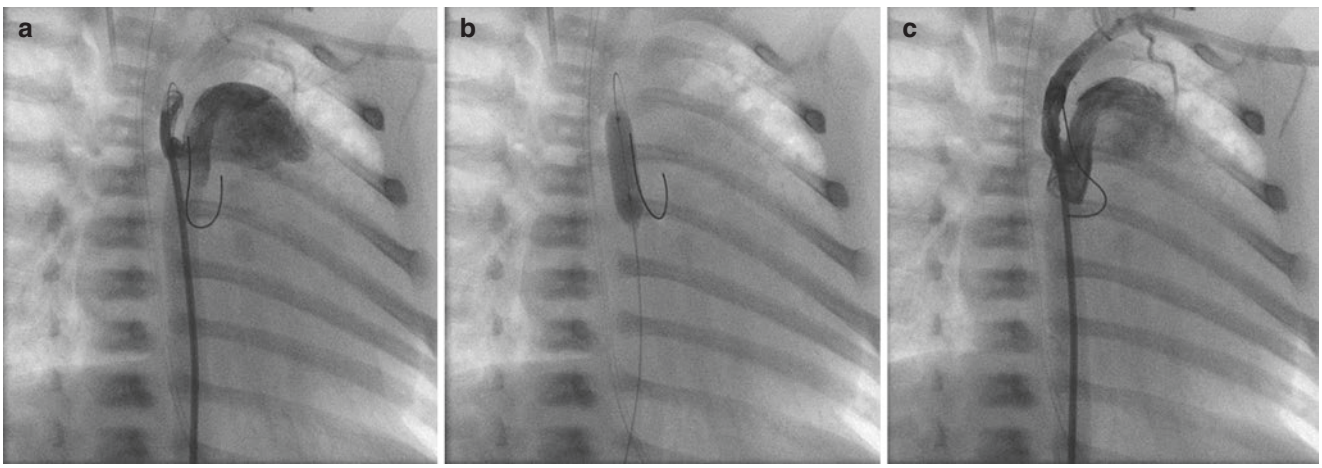



**Fig. 29.5** (**a**) Shows the duct descending aortic junction by contrast injection through a 4F multipurpose catheter positioned in a slightly obstructed aortic coarctation (CoA). Prior to duct stenting, the coarctation was dilated by a 6 × 20 mm Tyshak Mini balloon (**b**); on (**c**) the immediate result is depicted

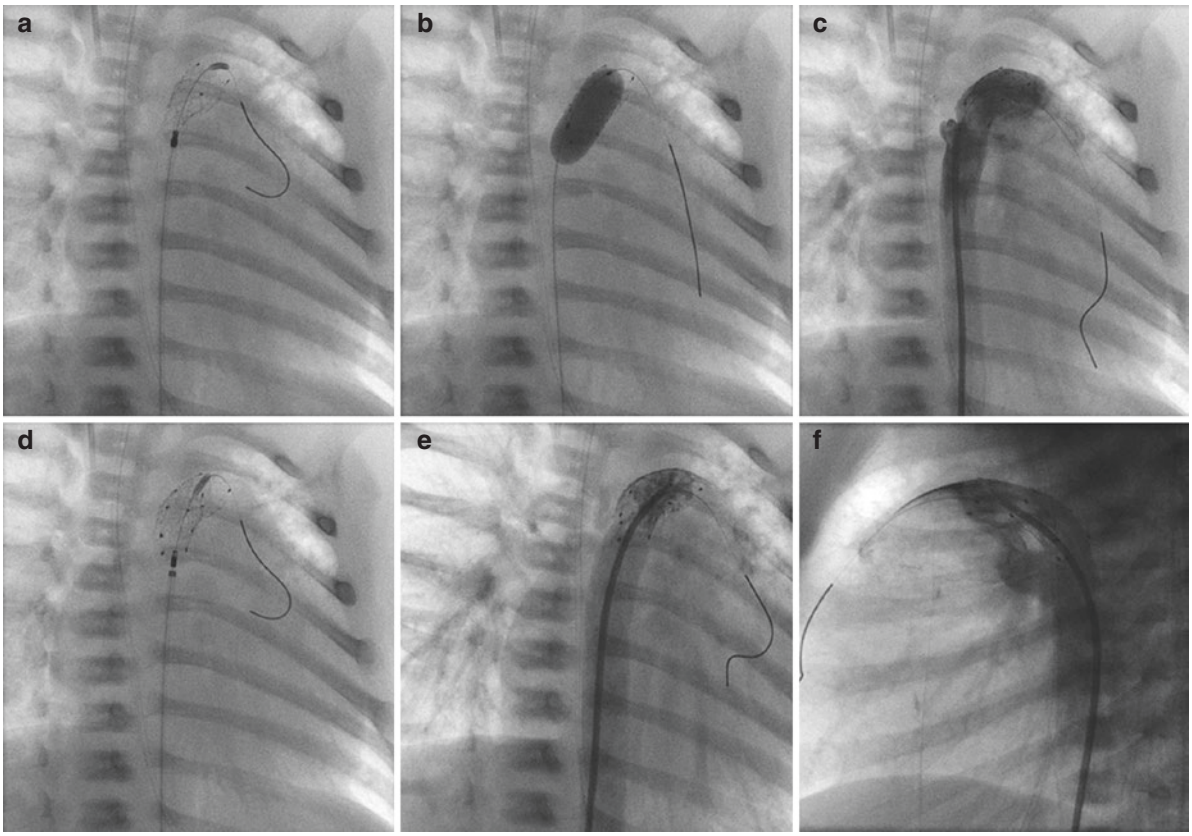



**Fig. 29.6** (**a**) Demonstrates stented duct with a SSF-DS 8 × 18 mm in RAO30°; the self-expandable stent is fully expanded at the pulmonary side, because of the low radial power obstructed at the distal part of the stent; (**b**) shows the dilatation of the obstructed stent part by a 8 × 20 mm Mini Tyshak balloon, and the immediate result by contrast-medium injection through a readvanced 4F multipurpose catheter (**c**); based on this angiography, the decision was made to implant a second (8 × 15 mm) stent in telescope technique; without complete over-stenting the connecting DAO (**d**, **e**) and with a favorable result demonstrated by an additional angiography in LAO 90°(**f**)

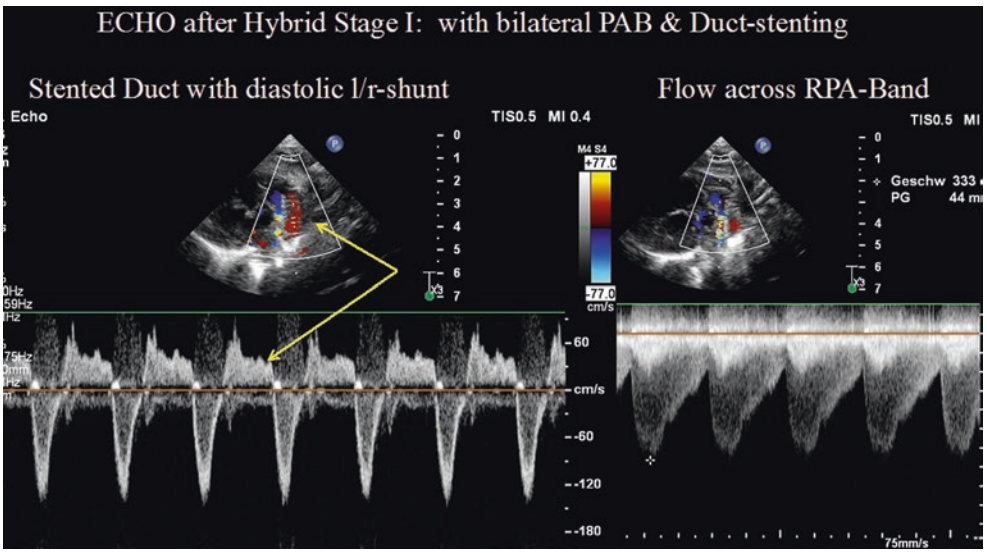



**Fig. 29.7** Summarizes important components of a successfully stage I approach: the flow through the stented duct is unobstructed; the flow is slightly higher because of the noncompliant stent within the duct; the amount of the diastolic left-to-right shunt (yellow arrows) depends on the efficacy of the bilateral PAB, the width of the stented duct, and the "Windkessel" of the pulmonary trunk–DAO–DA unity as well as by the presence of a pulmonary valve regurgitation, and if, by its degree. The here depicted CW Doppler flow obtained through the banded right pulmonary artery demonstrates in consideration of the systemic blood pressure and the gradient across the stented duct two facts: at first, the efficacy of the surgical band which is related to the presence (height) and absence of the diastolic flow pattern and, second, by the systolic gradient, which represents the degree or significance of an equilateral post-capillary obstruction

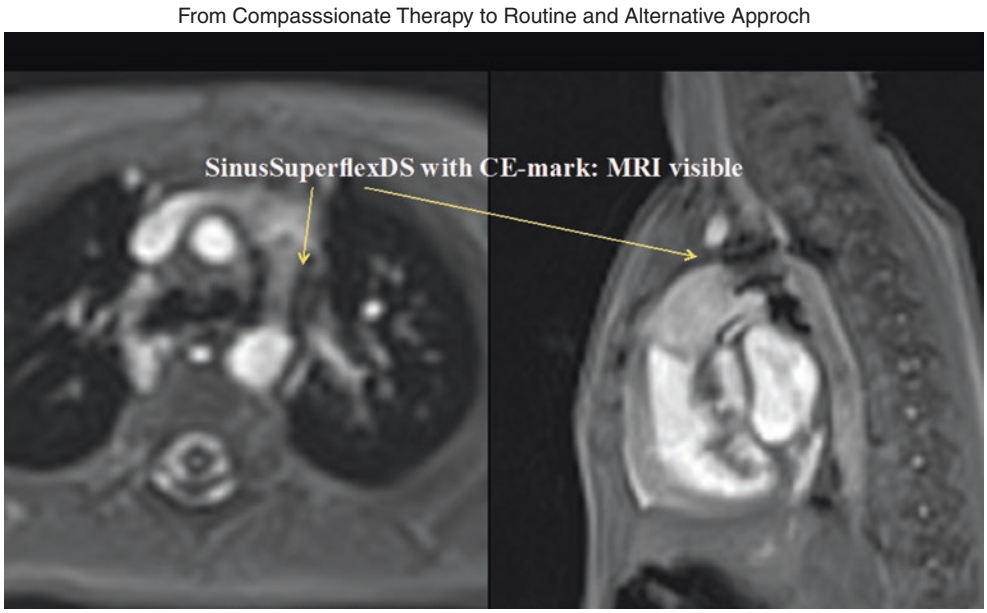



**Fig. 29.8** Shows magnetic resonance imaging of the visible self-expandable SinusSuperFlex-DS positioned in the arterial duct in sagittal and coronary plane. Not at least based on the technical improvements, the hybrid developed from a rescue to a first-line approach in neonates with HLHS

only with a sedated and spontaneous breathing patient; general anesthesia is routinely avoided (Fig. 29.8).

Since 2002, the hybrid stage II was first time described by Akintuerk et al., and several modifications were performed and adapted mostly based on the patient's anatomical condition and in particular because of technical improvements. See Butera et al. Chap. 28.

*Perspective:* The surgeon will focus on a comprehensive stage II, while stage I and III are performed as a transcatheter approach in an only sedated patient. In Columbus and Giessen, the surgeons and pediatric cardiologists made the decisions to change their classical program for favoring hybrid stage I approach in most with HLHS or univentricular variants. In Giessen the hybrid stage I approach is additionally used for newborns with HLHC to postpone high-risk operations from the neonatal period to late infancy with augmenting the surgical options and to avoid an initial decision for a univentricular strategy.

**Video 1** Represents a movie on which an enormous enlarged left atrium is shown by hand-injected contrast medium through a stiff-wire stabilized 4F right Judkins catheter (AVI 1107 kb)

**Video 2** Shows the echocardiographic result immediately after stent placement within the duct. The color Doppler demonstrates an unrestrictive flow through the stented duct (AVI 3883 kb)

**Video 3** Indicates a high-risk HLHS patient with an aortic atresia and extreme tiny ascending aorta, in whom stenting of the atrial septum, the arterial duct, as well as the coarctation became necessary to complete stage I hybrid approach; meanwhile the 4-year-old boy received a complication-free stage II and Fontan completion with a wonderful neurological outcome (AVI 1453 kb)

# Hybrid Approach: Ventricular Septal Defect Closure

# 30

Gareth J. Morgan

## 30.1 Introduction

The hybrid approach to closure of ventricular septal defects is applicable in a wide range of muscular ventricular septal defects. The key anatomical relationships to consider on the right ventricular aspect are the tricuspid valve, the moderator band, proximity to the free wall of the right ventricle and the pulmonary valve in the case of outlet defects. On the left side, obstruction of the aortic valve or left ventricular outflow tract may be an issue in outflow defects. The mitral valve is rarely a problem, but in cases where the mitral valve has abnormal attachments or where the anatomy is that of a repaired AVSD with a residual VSD, the left-sided valve structures can become relevant.

## 30.2 Clinical Scenarios, Indications and Patient Selection

1. *The patient is too small to consider transcatheter closure of the septal defect.* Transcatheter VSD closure is usually reserved for patients greater than 10 kg. This weight is not usually achieved by children with the physiological handicap of heart failure until after 1 year of age.
2. *Relative contraindication to cardiopulmonary bypass.* This may be due to an ongoing neurological concern, renal dysfunction or thrombotic/thrombophilia tendency. In the vast majority of cases, hybrid VSD closure can be 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, anterior mid-muscular and those closely associated with the moderator band, may be more amenable to a hybrid approach (Fig. 30.1).

## 30.3 Case Example

*Clinical scenario*: Postoperative perimembranous VSD closure. Haemodynamically significant additional muscular VSD. Patient weight, 4.8kg. Unable to progress from ITU/respiratory support care despite maximal anti-failure treatment. Chest radiograph is consistent with a large left-to-right shunt. Echocardiogram shows a dilated left side of the heart and significant left-to-right flow across the ventricle septum. The tricuspid valve regurgitant velocity suggests an RV pressure which is approx. 75% systemic.

*Anatomy*: Surgically repaired perimembranous VSD. Separate muscular defect positioned apical to the moderator band and measuring 5 mm on transthoracic echocardiogram.

*Treatment options*: Before embarking upon any definitive therapy, appropriately aggressive medical management should be implemented to ensure that a definitive procedure is necessary at this stage.

Option 1: Surgical device closure: The patient is within 2 weeks of his/her initial cardiopulmonary bypass run. The position of the defect in this weight of infant is likely to provide a major challenge to the surgeon.

Option 2: Percutaneous transcatheter device closure. Although a theoretically feasible option, the practicalities at this weight in this clinical scenario are likely to be prohibitive. Assuming that the defect could be crossed from the left side with a wire and catheter, manipulating a stiff delivery sheath through the right side of the heart and accurate device delivery would be very challenging.

The original version of this chapter was revised. A correction to this chapter can be found at https://doi.org/10.1007/978-3-319-72443-0_46

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_30) contains supplementary material, which is available to authorized users.

G. J. Morgan (✉)
The Heart Institute, Children's Hospital of Colorado, University of Colorado, Denver, CO, USA



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_30

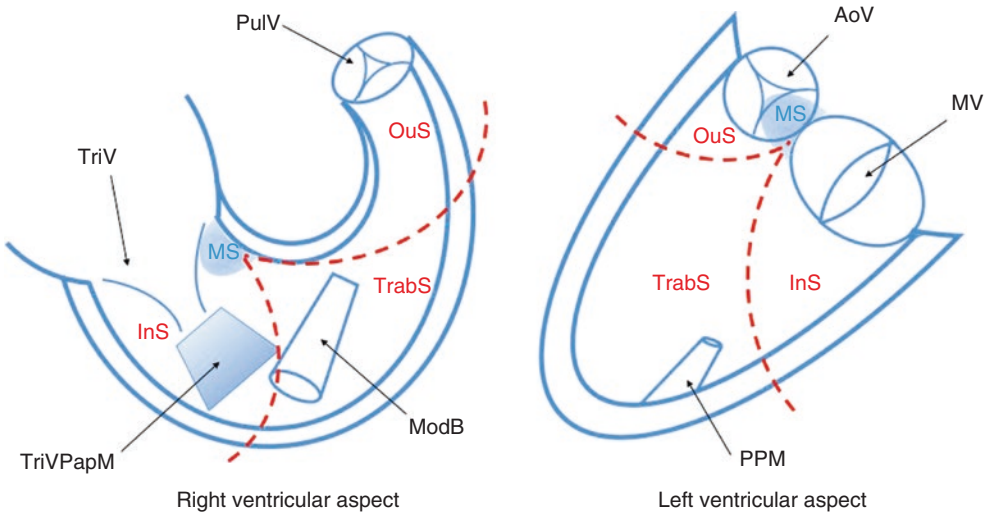



**Fig. 30.1** The right ventricular aspect of the ventricular septum is more complex than the left, partly explaining why disc conformation on the left side is rarely an issue but always a concern for the right-sided disc. The diagrams show the potentially problematic structures in each corresponding portion of the septum on either ventricular aspect. *TriPapM* tricuspid valve papillary muscles; *InS* inlet septum; *TrabS* trabecular septum; *OuS* outlet septum; *ModB* moderator band; *PPM* posterolateral papillary muscle; *MS* membranous septum)

Option 3: Hybrid periventricular VSD device closure. Given the patient weight, the position of the defect and the clinical condition, this may be an attractive option, as discussed below.

*Pre-procedural imaging:* (a) High-quality transthoracic echocardiogram alone; (b) if the key features cannot be delineated, then transoesophageal echocardiography (TOE) may help to delineate the anatomy further (Fig. 30.1, right panel). 3D echo imaging can add useful information in these cases; (c) angiographic delineation of the ventricular septum may be particularly useful in larger patients with complex multiple defects.

*Key features*: (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 mitral valve apparatus. (3) Proximity to the apex and the cavity size on either side of the defect (i.e. how much practical space there is 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 recognized to ensure that the correct defect will be crossed.

## 30.4 Technique (Step by Step)

1. The ideal place to perform hybrid procedures is in a fully specified hybrid operating facility. A 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 mandatory. Depending on patient size and defect position, epicardial echocardiography can also provide imaging guidance (Fig. 30.2, panel b).
2. When cardiac position and connections are normal, a sternotomy is usually the optimal approach; however, thoracotomy or subxiphoid approach may be used in cases where the anatomical orientation is favourable.
3. After locating and delineating the defect on echocardiography, 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 to allow the operators to manipulate the catheters and sheaths needs to be considered. Practically this is easily done by indenting different parts of the RV free wall with a finger whilst observing the echo image.
4. Prior to puncturing the RV, the occlusion device should be 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 (Fig. 30.3 panel a+b). In particular cases the use of a single disc "duct-type" device may be particularly important if the defect is close to the free wall of the right ventricle or abutting the moderator band; the lack of RV disc should allow better conformation of the disc and reduce the chance of the device interfering with and damaging the ventricular free wall (Fig. 30.4 and 30.5)
5. Once the RV wall position is identified and the device prepared, a purse string is placed on the RV free wall. Hundred

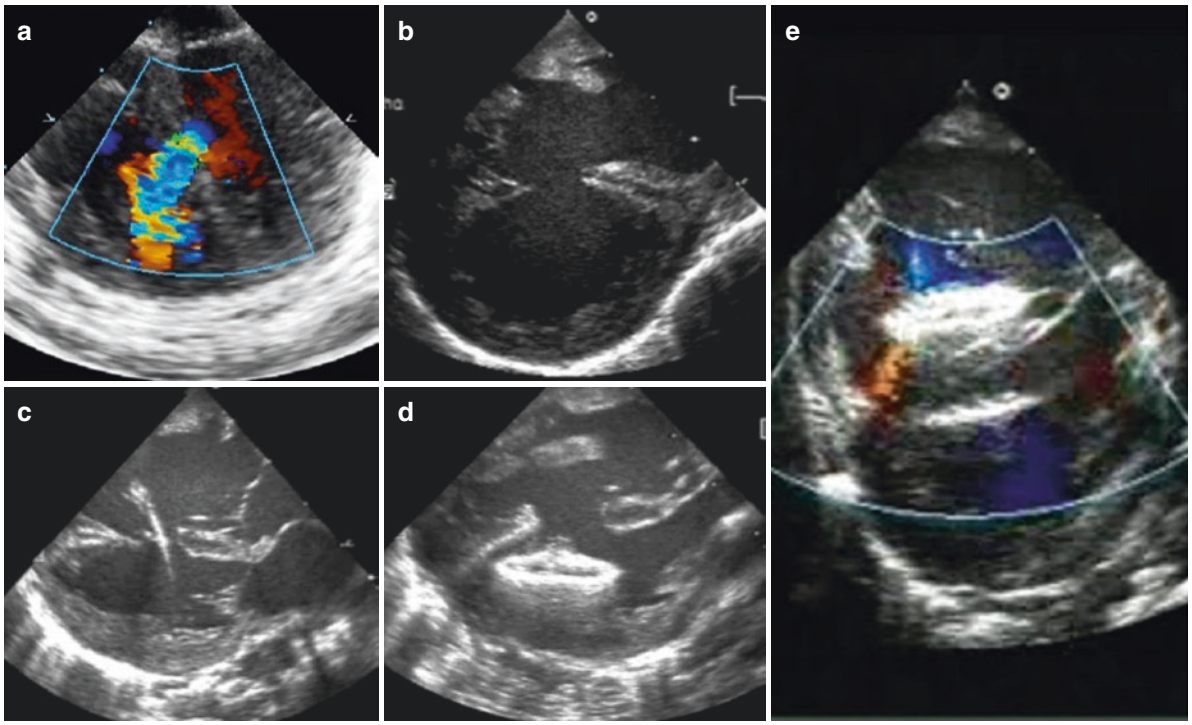



**Fig. 30.2** Panel (**a**) shows a mid-muscular VSD delineated with epicardial echo in a 4.8 kg patient. Epicardial echo can be useful as the probe can be used to mimic the angle and direction desired for the wire and sheath passage. Panel (**b**) shows the corresponding TOE view with 2D imaging and colour flow Doppler. Panel (**c**) shows the sheath across the defect, and panels (**d**) and (**e**) show the deployment of the left and right ventricular discs, respectively

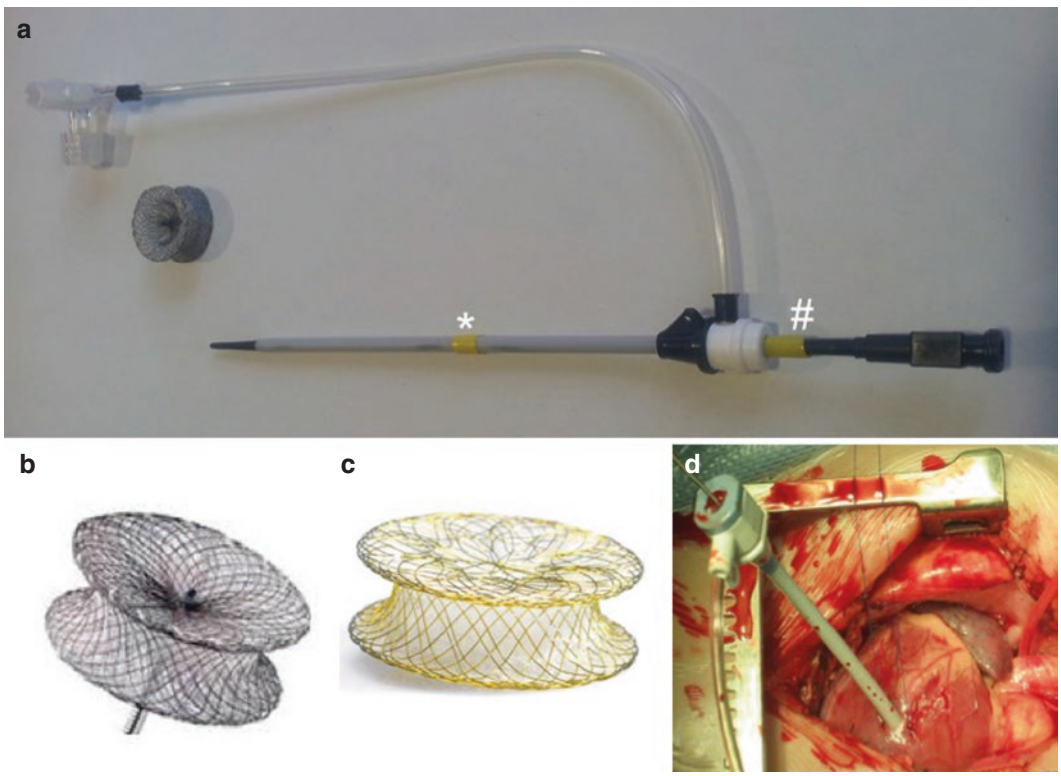



**Fig. 30.3** Panel (**a**) illustrates preparation of the short delivery sheath with a rubber shod placed to lessen the chance of the stiff dilator being pushed towards the posterior wall of the left ventricle (#) and another placed (*) to guide and monitor the distance the sheath is inserted through the free wall of the RV. Panel (**b** and **c**) show the AGA muscular VSD device and the Occlutech muscular VSD device, both of which can be used in the method above. Panel (**d**) shows a typical surgical view of the sheath placed in the RV with a purse string suture in place (Panel **d** image courtesy of Prof Neil Wilson, University of Colorado)

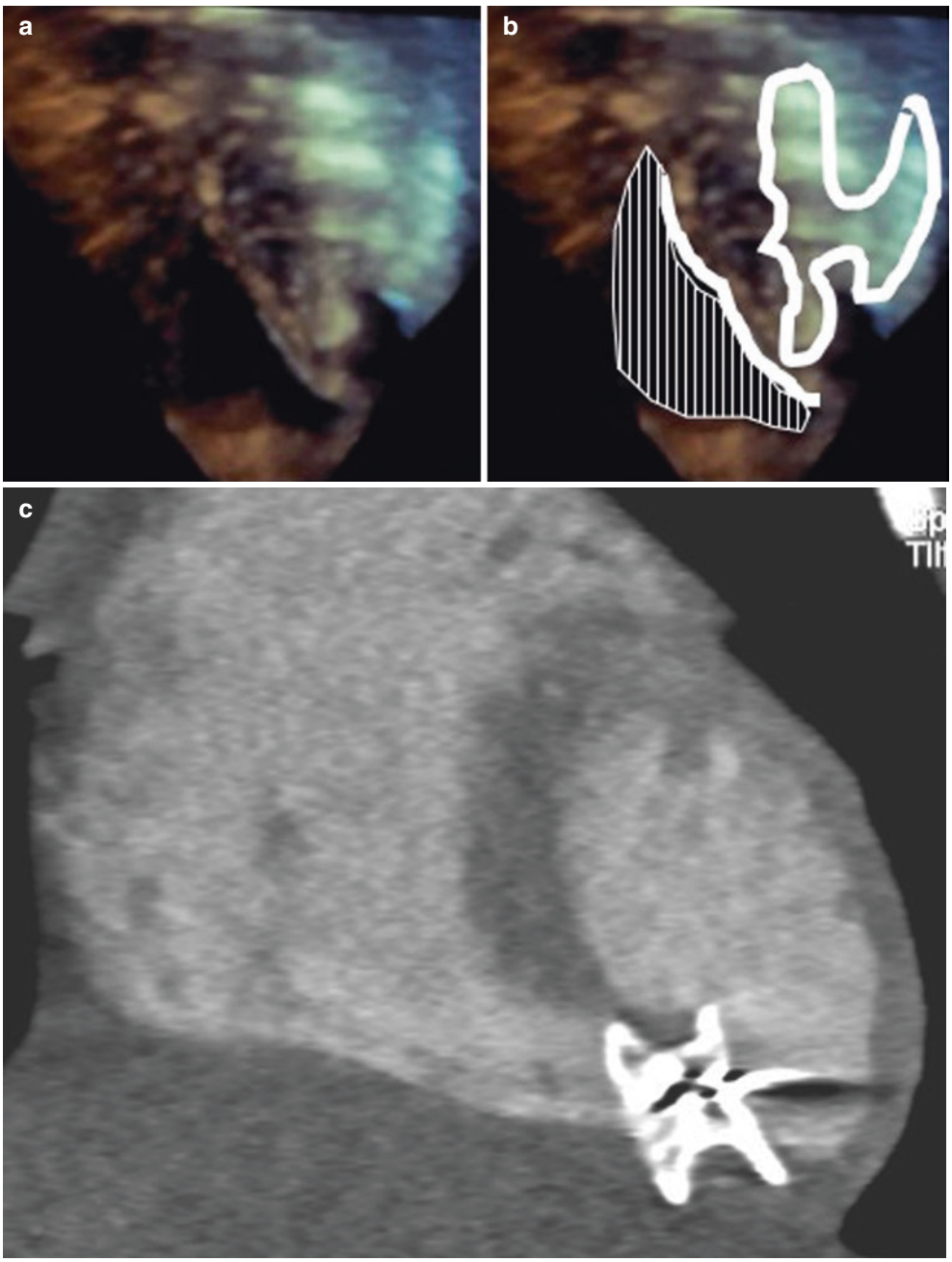



**Fig. 30.4** Images of an apically placed occluder close to the right ventricular free wall with a related pericardial effusion. Panel (**a**) is a still frame from live 3D echo imaging showing the device, the external surface of the RV and a pocket of pericardial fluid. Panel (**b**): Schematic overlay for illustrative purposes. The striped area denotes the effusion; the thick white line defines the epicardial aspect of the RV; and the irregular shape represents the apposed aspect of the muscular VSD device. CT imaging (Panel **c**) was carried out in an attempt to rule out perforation and help to assess risk of future perforation

units per kilogram of heparin should be administered at this stage. Under TOE guidance, the RV is punctured using an 18-gauge needle and a 035″ Terumo J-tip hydrophilic wire guided across the defect and into the LV cavity. The wire will ideally be directed out the left ventricular outflow tract to minimize the possibility of interference with the mitral papillary muscles and direct the sheath away from the posterior wall of the LV (Fig. 30.5). A short sheath (approx. 10 cm), large enough to accommodate the prospective device (usually between 6 French and 10 French), is then advanced over the wire and across the VSD to sit in the LV cavity (Fig. 30.2, panel c; Fig. 30.3 panel d). A perpendicular approach from the free wall to the ventricular septum is important in order not to distort the anatomy and enable successful deployment.

6. Under careful echocardiographic guidance, the LV disc should be deployed in the mid-cavity and the entire apparatus withdrawn to appose the LV disc onto the septum (Fig. 30.2, panel d). Then, the waist of the device and subsequently the RV disc should be uncovered by withdrawal of the sheath. It may take several attempts to correctly conform the RV disc; therefore, care should be

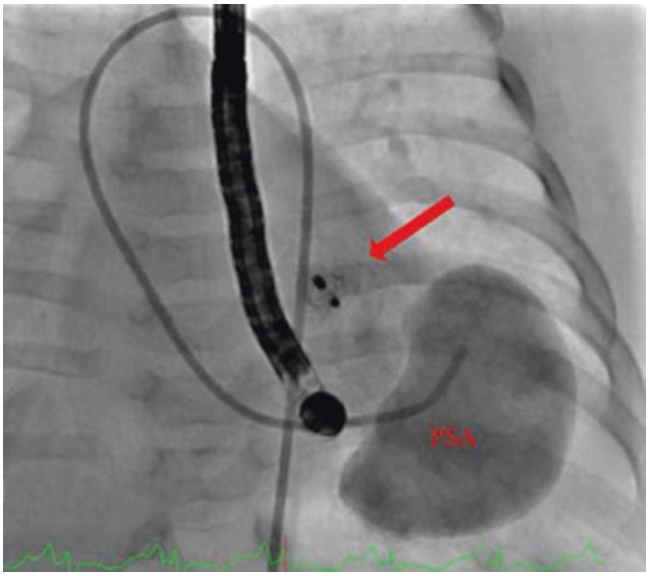



**Fig. 30.5** LV pseudoaneurysm secondary to hybrid VSD closure. During the procedure the wire was noted to have excessive contact with the posterior wall of the LV. This led to the development of a large pseudoaneurysm within 2 weeks of the procedure which was initially treated with percutaneous occlusion but was later surgically resected. The red arrow indicates the muscular VSD device, remote from the pseudoaneurysm which is labelled "PSA" (Image courtesy of Dr. Damien Kenny, Dublin, Ireland)

taken not to pull the sheath out of the RV during the initial deployment. Indeed the RV disc may not completely conform on the RV septal aspect due to trabeculations, moderator band and limited chamber size near the apex. The operators must then decide whether the RV disc has formed adequately to allow defect occlusion and device stability even if it looks constrained (Fig. 30.2, panel e).

7. The device can be released in the standard manner. Echocardiography may be supplemented by angiography at any stage; however, this is rarely necessary with high-quality echo imaging.
8. The sheath can then be withdrawn and the purse string closed.

**Video 1** Hybrid muscular VSD closure. TEE monitoring of the procedure showing the various steps (video courtesy of Dr. Butera Gianfranco, Milan, Italy) (MOV 31404 kb)

# Hybrid Approach: Stent Implantation

# 31

Ralf J. Holzer and Jeffrey Dayton

## 31.1 Introduction

Stent delivery using a hybrid approach is a type of procedure that utilizes both surgical and interventional techniques to combine the advantages of each approach. Hybrid procedures have increased in frequency over the last two decades and are performed by the majority of congenital cardiac centers, as documented by a CCISC survey conducted by Dan Gruenstein in 2013. Intraoperative stents have been placed in a variety of locations, even though pulmonary arteries are the most common destination for this therapy.

Hybrid stent therapy can be performed under direct vision using endoscopic (± fluoroscopic) guidance as needed (on cardiopulmonary bypass), which Holzer and colleagues reported to be used in about 75% of cases of hybrid pulmonary artery stent therapy. Alternatively, a direct approach through a purse string with angiographic and fluoroscopic guidance can be used. Each of these approaches has their own specific advantages and disadvantages which need to be carefully considered to the transcatheter and surgical alternatives available to treat a vascular lesion. Table 31.1 lists the various therapeutic strategies to treat vascular lesions with the specific advantages, disadvantages, and complications which need to be taken into account when deciding upon the best strategy for each patient. In many occasions, the ultimate decision of surgical patch augmentation versus intraoperative stent placement may only be made in the operating room once the surgeon inspects the lesion and considers the additional surgical requirements. As such, hybrid therapy complements, rather than competes, with the more traditional surgical and transcatheter therapies.

One common element of the decision-making process is to involve the cardiothoracic surgeon from the start, so that there is an agreement on the type of approach that should be used. Specific prerequisites for a hybrid approach under direct vision include detailed pre-procedural imaging data, whether this is CT, MRI, or cardiac catheterization. Once on cardiopulmonary bypass with the vessel opened, it is often much more difficult to determine the exact vascular dimensions and stent lengths that are needed (as well as the anatomy of side branches), and as such any patient that may be considered for intraoperative stent placement should have adequate imaging data obtained prior to going to the operating room. In general, stent implantation under direct vision is usually performed as a concomitant procedure when a patient requires other cardiac surgical interventions and very rarely would this ever be done as the sole intended procedure. In contrast, hybrid therapy using angiographic guidance can be performed on the beating heart when the specific approach is considered more suitable for a specific patient. Often, these types of hybrid procedures result from an abnormal exit angiography that identifies a vascular anomaly.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_31) contains supplementary material, which is available to authorized users.

Ralf J. Holzer (✉)
Professor of Clinical Pediatrics Division Chief Pediatric Cardiology, Director Pediatric Cardiac Catheterization
Weill Cornell Medicine,
NewYork-Presbyterian Komansky Children's Hospital,
New York, NY, USA
e-mail: rjh3001@med.cornell.edu

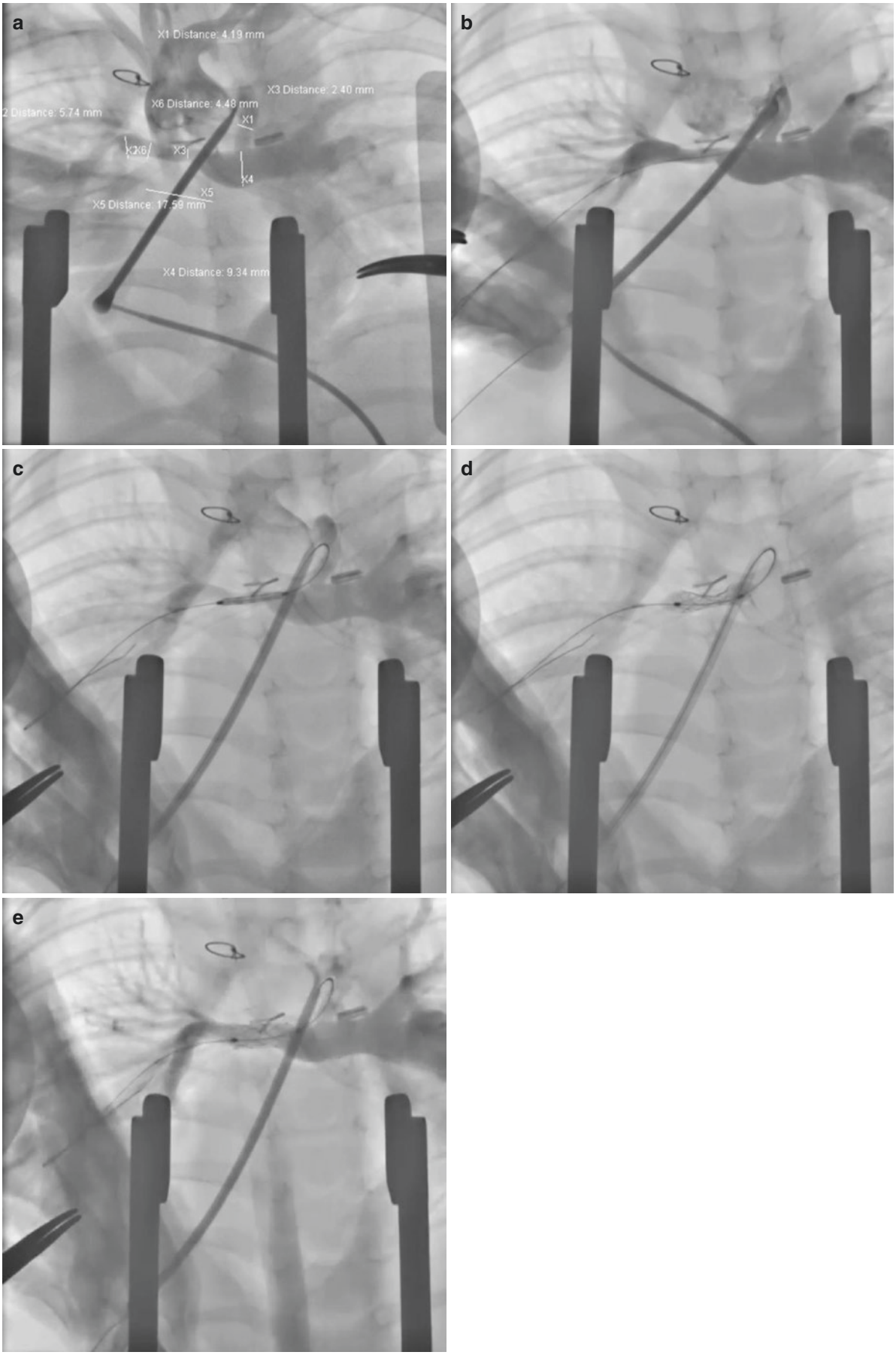
J. Dayton
Assistant Professor Clinical Pediatrics
Weill Cornell Medicine,
NewYork-Presbyterian Komansky Children's Hospital,
New York, NY, USA
e-mail: jed9031@med.cornell.edu


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_31

**Table 31.1** Advantages and disadvantages of the various therapeutic options available to treat vascular lesions: transcatheter approach, hybrid stent delivery using direct puncture with angiographic guidance, hybrid stent delivery under direct vision with endoscopic/fluoroscopic guidance, and surgical patch plasty

| | Advantages | Disadvantages |
|---|---|---|
| Transcatheter stent | • Suited for vascular kinks<br>• Suited for external compression<br>• No need for cardiopulmonary bypass<br>• No need for surgical dissection<br>• Biplane imaging for optimum guidance | • Technically more challenging (long sheaths, stiff wires, curves, bends)<br>• Longest case times<br>• Longest fluoroscopy times and radiation exposure<br>• More prone to hemodynamic instability<br>• Least control of vascular complications<br>• Edges of stents sometimes "stick out" and create difficulties at future cath<br>• Use of adult-sized stents limited in small children<br>• Stent-specific disadvantages (in-tent stenosis, need for redilatation in children, etc.) |
| Hybrid stent with angiography (Fig. 31.1 and Fig. 31.3) | • Avoids curves and bends<br>• No long sheaths or stiff wires<br>• More likely to be able to use "adult" stents in small patients<br>• Suited for vascular kinks<br>• Suited for external compression<br>• Shorter case times than transcatheter approach<br>• Less fluoroscopy time than transcatheter approach<br>• Often less (but not zero) needed for extensive surgical dissection<br>• No cardiopulmonary bypass<br>• Reduced hemodynamic instability<br>• Better control of vascular complications (in the OR, open chest, CPB standby) | • A surgical dissected vessel is more prone to tear during stent expansion than a vessel embedded in scar tissue<br>• Less suited for small vessels<br>• Stent-specific disadvantages (in-tent stenosis, need for redilatation in children, etc.)<br>• Fresh suture lines and purse strings can be focal points for dissections during angiography |
| Hybrid stent under direct vision (Fig. 31.2) | • Avoids curves and bends<br>• No long sheaths or stiff wires<br>• Suited for vascular kinks<br>• Use of "adult" stents irrespective of patient size<br>• Suited for external compression<br>• Short case times<br>• No/little fluoroscopy time<br>• Reduced hemodynamic instability<br>• Better control of vascular complications<br>• Ability for stent modification (e.g., shortening of closed cell design stents)<br>• Stent can be "molded" to vessel wall | • Difficulty to assess distal wire position<br>• Distal end of stent not visible during inflation unless fluoroscopy is used<br>• More difficult to assess vascular dimensions<br>• A surgical dissected vessel is more prone to tear during stent expansion than a vessel embedded in scar tissue<br>• Requires detailed pre-procedural imaging data<br>• Less suited for small vessels<br>• Stent-specific disadvantages (in-tent stenosis, need for redilatation in children, etc.)<br>• Need for cardiopulmonary bypass |
| Surgical patch plasty | • No long sheaths or stiff wires<br>• No need for fluoroscopy<br>• Patient/vessel size does not matter<br>• Reduced hemodynamic instability<br>• Great control of vascular complications<br>• Avoids stent-specific disadvantages<br>• Usually "smooth" vessel wall without stent struts creating future obstacles | • Need for extensive surgical dissection (with issues such as phrenic nerve injury and bleeding)<br>• Need for cardiopulmonary bypass<br>• Less suited for kinks<br>• Less suited for external compression |

**Fig. 31.1** An 18-month-old (8.9 kg) male child with complex congenital heart disease who had previously undergone placement of a central shunt as well as being 1 day post-op patch augmentation of the right pulmonary artery. It was felt that given the origin of the shunt, a direct puncture from the aorta would provide the best access for possible RPA stent placement, which was then performed through a median sternotomy using a hybrid approach. (**a**) After median sternotomy, the shunt was visualized and a purse string placed at the opposite site of the aorta in direct proximity to the shunt. Subsequently a sheath was inserted and an angiography through the sidearm of the sheath (hand injection) performed, documenting a hypoplastic and narrow proximal RPA measuring 2.4 mm at the narrowest. (**b**) Using the initial angiography as a roadmap, a 014″ coronary wire was manipulated with the aid of a Judkins right coronary catheter into the RPA and an angiography performed to document the anatomy with the wire in place. (**c**) A 6 mm × 15 mm Palmaz Blue stent was advanced over the wire and an angiography performed through the sheath to document stent positioning. (**d**) Once adequate stent position was confirmed, the stent was expanded. (**e**) An angiography was performed after stent expansion (as a hand injection through the sidearm of the sheath) documenting the stent in appropriate position


a
X1 Distance: 4.19 mm
X3 Distance: 2.40 mm
X6 Distance: 4.48 mm
2 Distance: 5.74 mm
X1
X2
X6
X3
X4
X5
X5 Distance: 17.59 mm
X4 Distance: 9.34 mm
b
c
d
e

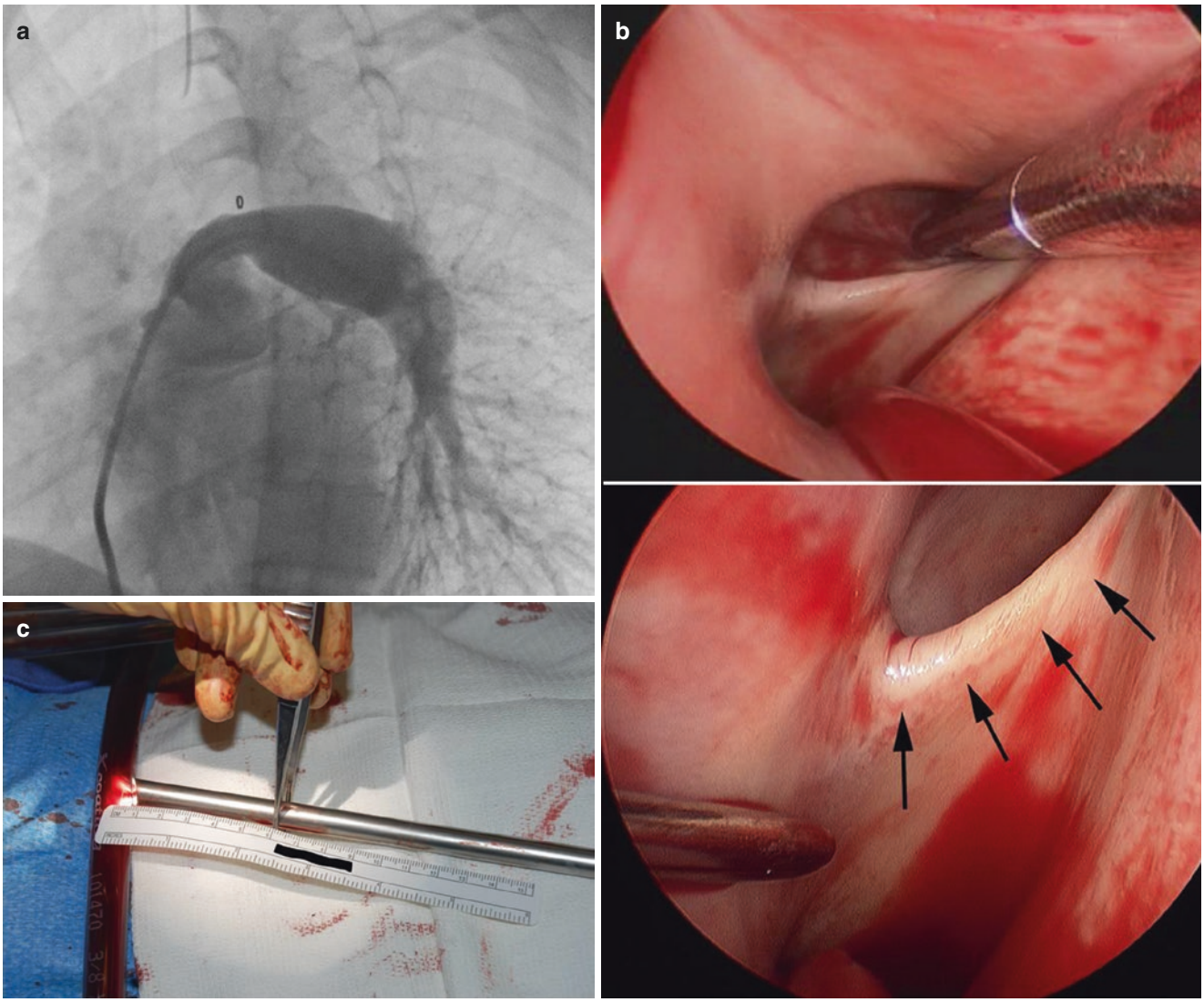



**Fig. 31.2** Adult patient with status post repair of tetralogy of Fallot with a proximal left pulmonary artery (LPA) stenosis (kink) who required surgical pulmonary valve replacement and who was considered for intraoperative stent placement (under direct vision) at the same time. (**a**) Preoperative cardiac catheterization with angiography documenting what appeared to be a proximal kink of the left pulmonary artery. (**b**) Intraoperative inspection of the LPA narrowing that documented a ridge/kink at the entrance to the left pulmonary artery. Hagar dilators were utilized to evaluate the dimensions and confirm consistency with the measurements obtained in the cardiac catheterization laboratory. (**c**) An endoscope was utilized to inspect the vascular anatomy. (**d**) The endoscope was utilized to evaluate the origin of the side branches and measure the distance based on the depth of insertion of the endoscope. The endoscope was also utilized to guide placement of the guide wire. In contrast to transcatheter stent placement, a soft wire is sufficient to guide intraoperative stent placement. Once the wire is placed, the balloon-mounted stent is advanced over the wire into position. Adequate position is confirmed purely by the stent aligning adequately proximally and knowing the length of the stent and distance to origin of pulmonary side branches. If the available stent lengths are inadequate, closed cell design stents can be cut to the appropriate length. During stent expansion, visualization of the side branches is obscured. If there are concerns that a stent may potentially be advanced too distally into a smaller part of the vessel, then expansion can be performed under fluoroscopy, which will clearly document if there is a distal obstruction to stent expansion. (**e**) Once the stent is expanded, careful evaluation is performed of the stent position. As once can see in the top image, there is sufficient distance to the side branches (no jailing). Any stent mesh that protrudes proximally can be folded over the ridge (middle image). The bottom image shows the placed stent looking from the main pulmonary artery (MPA)

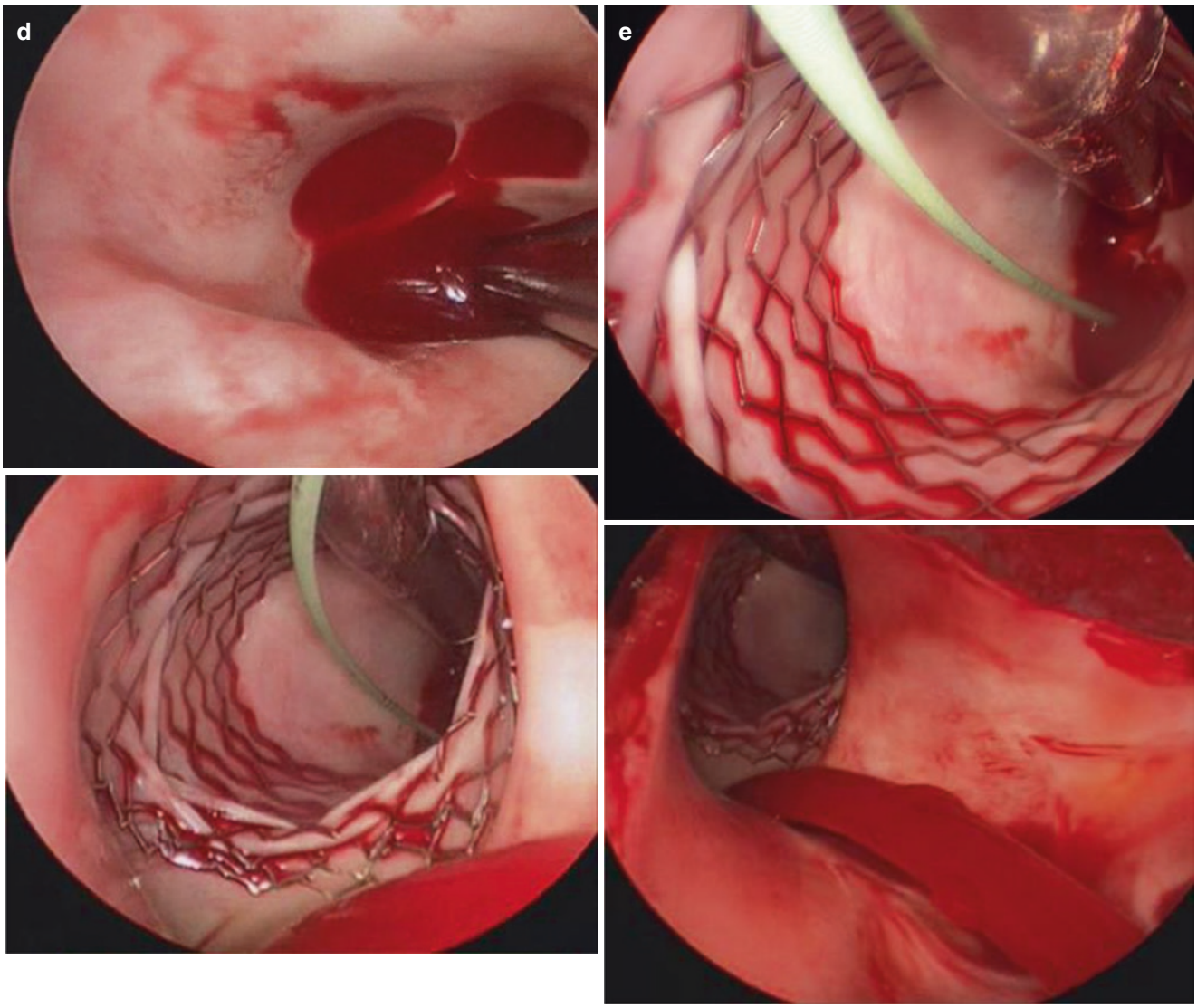



**Fig. 31.2** (continued)

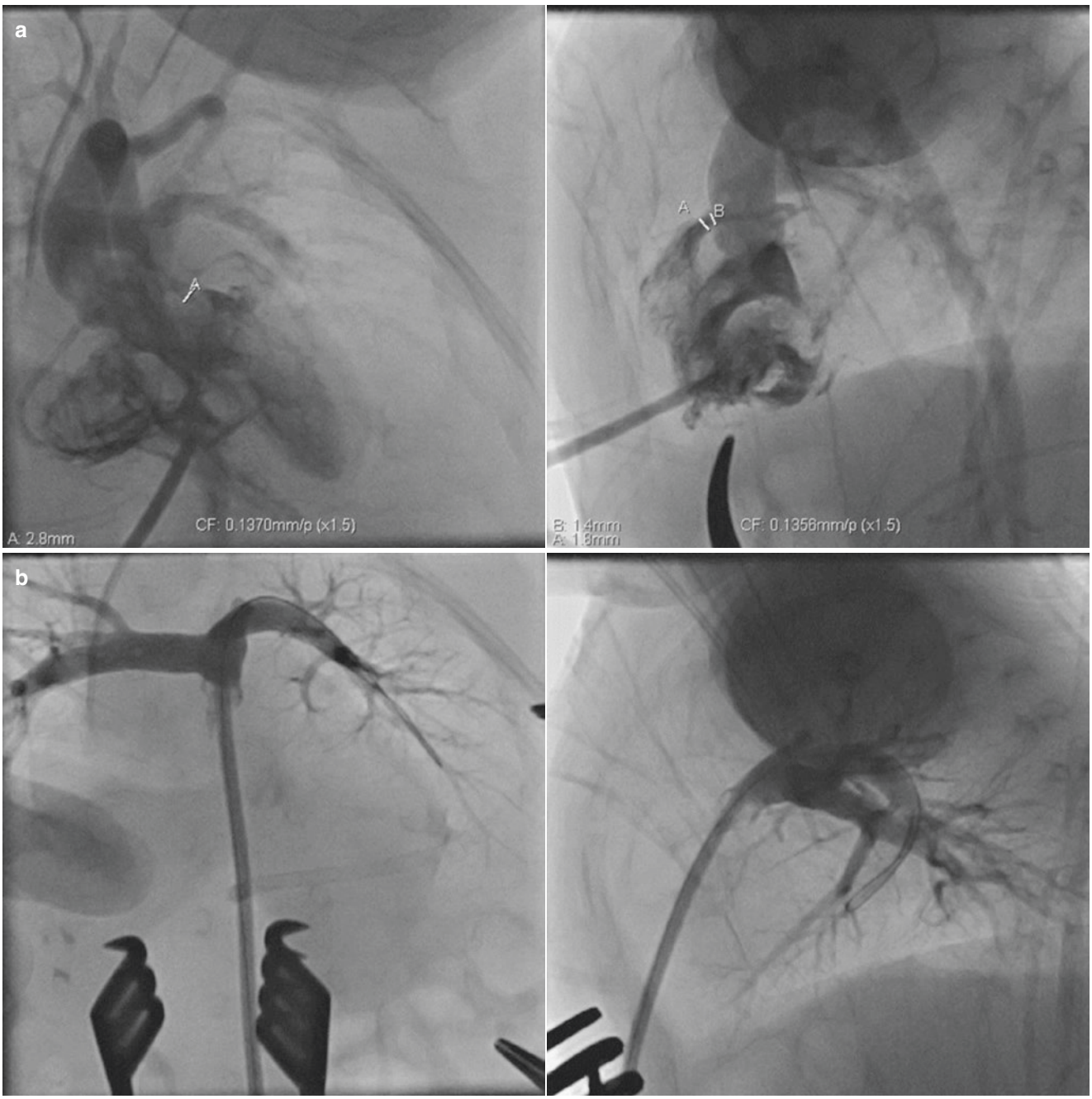



**Fig. 31.3** Neonate with ventricular septal defect and pulmonary valve stenosis and small main pulmonary artery. It was felt that intraoperative stent placement across the pulmonary valve and main pulmonary artery (MPA) should be performed. All images obtained are antero-postero projection on the left and lateral projection on the right. (**a**) Baseline right ventricular angiography after placement of a hemostatic sheath through a purse string at the RV free wall. One can see that the sheath is pointed in the wrong direction (away from the right ventricular outflow tract (RVOT)). (**b**) After readjusting the sheath position, one can see that the sheath is now positioned directly underneath the pulmonary valve. (**c**) A 014″ wire is then inserted through the sheath into the left pulmonary artery and a 4 mm pre-mounted coronary stent advanced over the wire across the RVOT and into the MPA. (**d**) A hand injection is performed through the sheath confirming adequate stent position. (**e**) The stent is subsequently expanded. (**f**) A final angiography is performed through the short sheath to confirm adequate stent position and lack of vascular injury prior to removing the guide wire

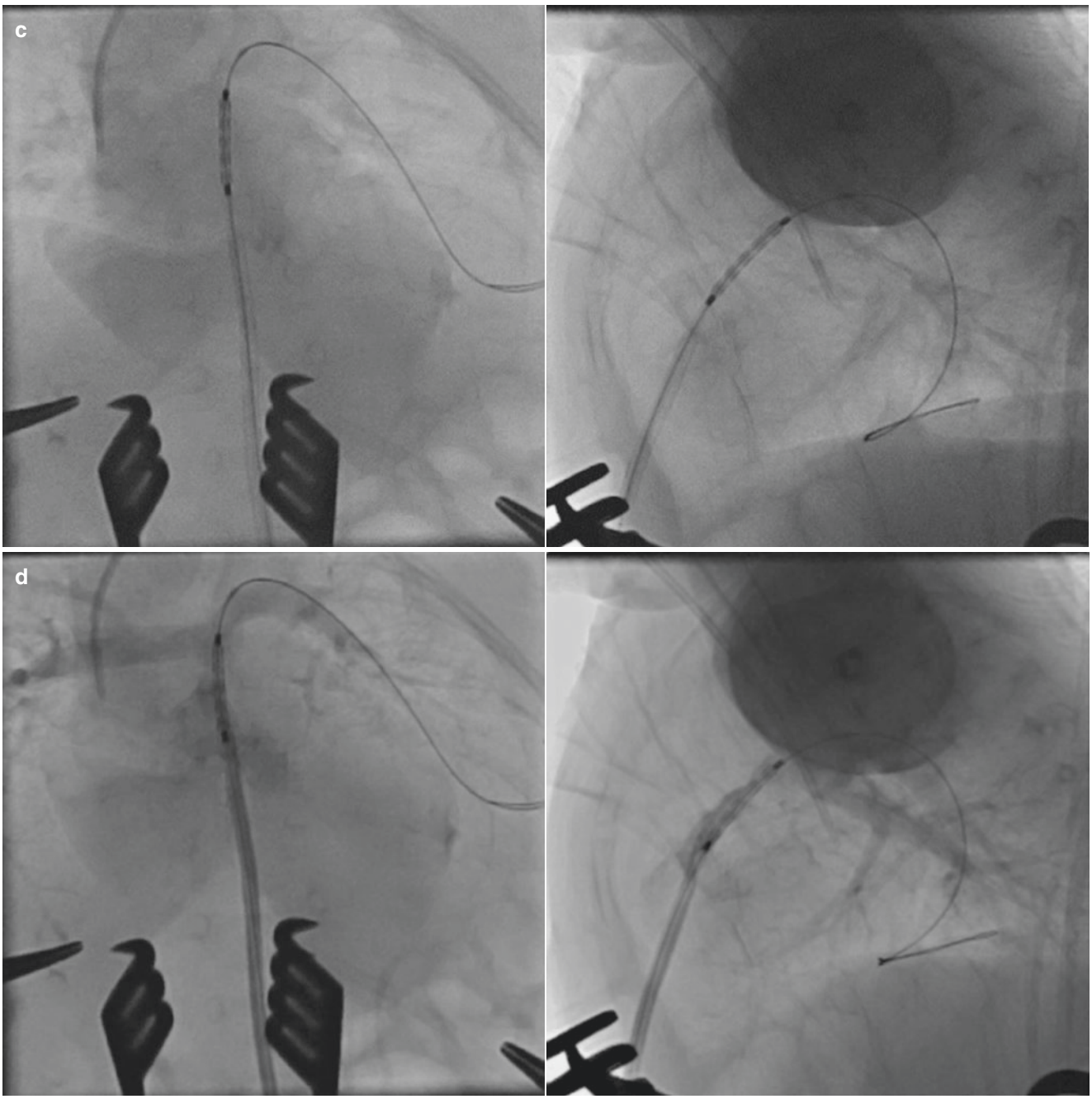



**Fig. 31.3** (continued)

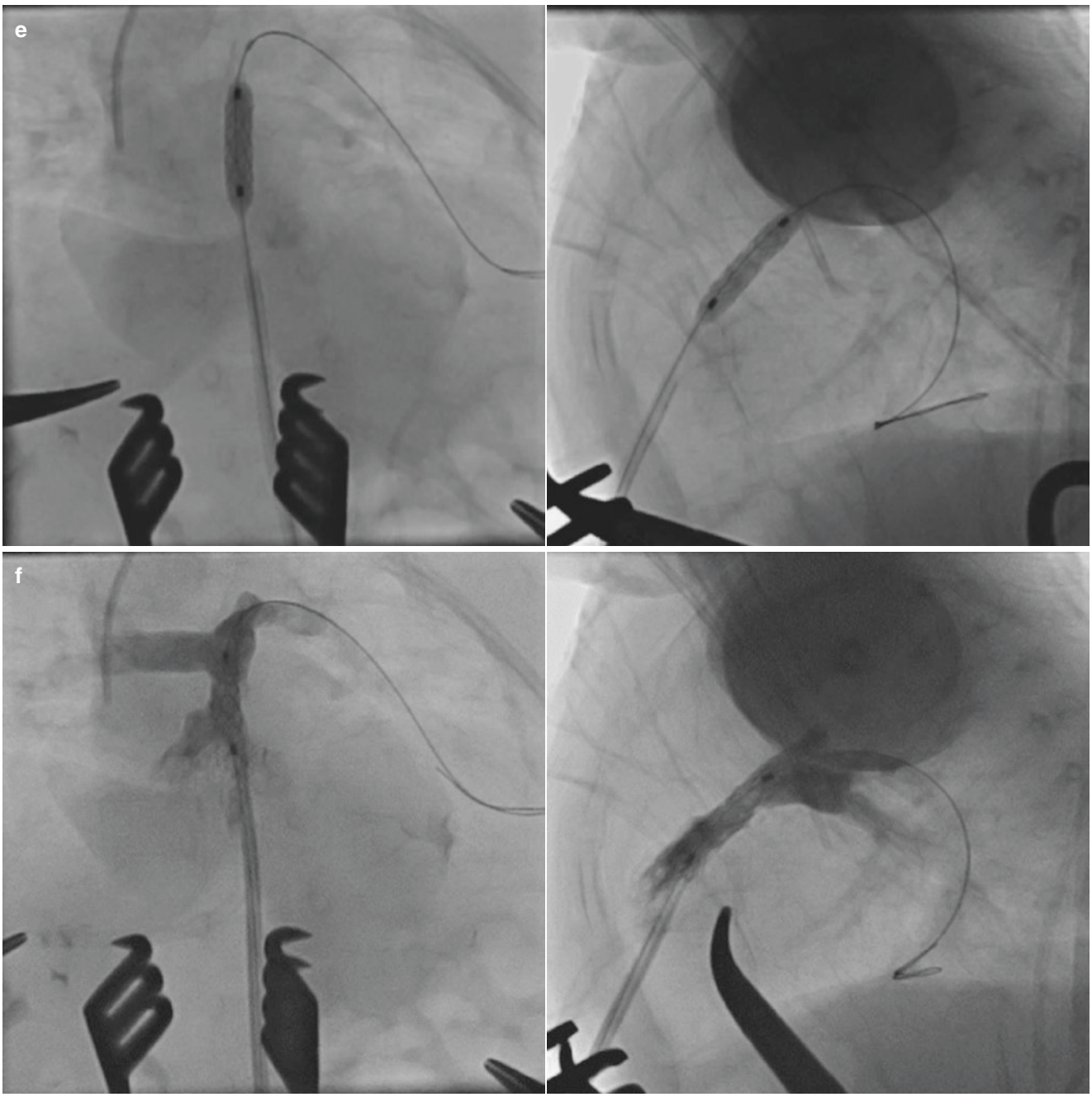



**Fig. 31.3** (continued)

**Video 1a** Five-month-old infant with tetralogy of Fallot that has previously undergone placement of a 3.5 mm right modified Blalock-Taussig shunt (RMBTS) and presented with desaturations into the 60th. The infant had previously developed femoral artery occlusion after cardiac catheterization, and therefore it was felt that a direct approach using carotid cutdown was the best approach for this patient. Right carotid cutdown was performed and a 6Fr sheath inserted. (**a**) Baseline angiography through the sidearm of the sheath, documenting the shunt to measure 2–2.5 mm in diameter with an additional proximal and minor distal narrowing. Prior to entering the shunt, a decision was made to use two coaxial coronary stents (an 18 mm × 4 mm Multilink Vision as well as a 15 mm × 4 mm Multilink Vision). It is crucial to have those stents ready to go in case more profound desaturations are encountered when crossing the shunt (MOV 67 kb)

**Video 1b** Five-month-old infant with tetralogy of Fallot that has previously undergone placement of a 3.5 mm right modified Blalock-Taussig shunt (RMBTS) and presented with desaturations into the 60th. The infant had previously developed femoral artery occlusion after cardiac catheterization, and therefore it was felt that a direct approach using carotid cutdown was the best approach for this patient. Right carotid cutdown was performed and a 6Fr sheath inserted. (**b**) A 014″ choice PT wire is advanced through the sheath and shunt into the right pulmonary artery and wire position confirmed using a hand injection through the sidearm of the hemostatic sheath. (MOV 206 kb)

**Video 1c** Five-month-old infant with tetralogy of Fallot that has previously undergone placement of a 3.5 mm right modified Blalock-Taussig shunt (RMBTS) and presented with desaturations into the 60th. The infant had previously developed femoral artery occlusion after cardiac catheterization, and therefore it was felt that a direct approach using carotid cutdown was the best approach for this patient. Right carotid cutdown was performed and a 6Fr sheath inserted. (**c**) Subsequently the 18 mm × 4 mm Multilink Vision stent is advanced though the shunt using repeated small hand injections to confirm the position (MOV 257 kb)

**Video 1d** Five-month-old infant with tetralogy of Fallot that has previously undergone placement of a 3.5 mm right modified Blalock-Taussig shunt (RMBTS) and presented with desaturations into the 60th. The infant had previously developed femoral artery occlusion after cardiac catheterization, and therefore it was felt that a direct approach using carotid cutdown was the best approach for this patient. Right carotid cutdown was performed and a 6Fr sheath inserted. (**d**) Once appropriate position is confirmed, the stent is expanded in a controlled manner (MOV 1548 kb)

**Video 1e** Five-month-old infant with tetralogy of Fallot that has previously undergone placement of a 3.5 mm right modified Blalock-Taussig shunt (RMBTS) and presented with desaturations into the 60th. The infant had previously developed femoral artery occlusion after cardiac catheterization, and therefore it was felt that a direct approach using carotid cutdown was the best approach for this patient. Right carotid cutdown was performed and a 6Fr sheath inserted. (**e**) After stent expansion, an angiography is performed through the sidearm of the sheath to confirm adequate stent position as well as a lack of any vascular injury (MOV 237 kb)

**Video 1f** Five-month-old infant with tetralogy of Fallot that has previously undergone placement of a 3.5 mm right modified Blalock-Taussig shunt (RMBTS) and presented with desaturations into the 60th. The infant had previously developed femoral artery occlusion after cardiac catheterization, and therefore it was felt that a direct approach using carotid cutdown was the best approach for this patient. Right carotid cutdown was performed and a 6Fr sheath inserted. (**f**) Subsequently the 14 mm × 4 mm Multilink Vision stent is advanced though the shunt coaxial into the previous stent using repeated small hand injections to confirm the position (MOV 184 kb)

**Video 1g** Five-month-old infant with tetralogy of Fallot that has previously undergone placement of a 3.5 mm right modified Blalock-Taussig shunt (RMBTS) and presented with desaturations into the 60th. The infant had previously developed femoral artery occlusion after cardiac catheterization, and therefore it was felt that a direct approach using carotid cutdown was the best approach for this patient. Right carotid cutdown was performed and a 6Fr sheath inserted. (**g**) Once appropriate position is confirmed, the second stent is expanded in a controlled manner (MOV 1556 kb)

**Video 1h** Five-month-old infant with tetralogy of Fallot that has previously undergone placement of a 3.5 mm right modified Blalock-Taussig shunt (RMBTS) and presented with desaturations into the 60th. The infant had previously developed femoral artery occlusion after cardiac catheterization, and therefore it was felt that a direct approach using carotid cutdown was the best approach for this patient. Right carotid cutdown was performed and a 6Fr sheath inserted. (**h**) After expansion of the second stent, a final angiography is performed through the sidearm of the sheath to confirm adequate stent position as well as a lack of any vascular injury (MOV 254 kb)

**Video 2a** Five-month-old infant with hypoplastic left heart syndrome undergoing comprehensive stage II palliation. Exit angiography revealed lack of flow to the left pulmonary artery, and therefore intraoperative hybrid stent placement using a direct approach under angiographic guidance was recommended (videos from Dr. John Cheatham, Nationwide Children's Hospital, Columbus, OH). (**a**) Initially a Berman angiographic catheter is inserted through a purse string into the superior caval vein and an angiography performed, which in this case documented lack of flow to the left pulmonary artery (MP4 10900 kb)

**Video 2b** Five-month-old infant with hypoplastic left heart syndrome undergoing comprehensive stage II palliation. Exit angiography revealed lack of flow to the left pulmonary artery, and therefore intraoperative hybrid stent placement using a direct approach under angiographic guidance was recommended (videos from Dr. John Cheatham, Nationwide Children's Hospital, Columbus, OH). (**b**) To obtain more distal landmarks and size of the vessel, the catheter is further advanced carefully into the left pulmonary artery and an angiography repeated in distal position. This is then followed by advancing a short 7Fr sheath into the SVC. It is often difficult to advance the sheath into a distal LPA position, and therefore frequently the balloon-mounted (19 mm Genesis XD) stent is carefully advanced directly into the LPA without the use of sheath cover. This has the advantage of not distorting the anatomy once the stent has been placed in LPA position. It helps gently inflating the balloon to cover the sharp edges of the stent (MP4 5834 kb)

**Video 2c** Five-month-old infant with hypoplastic left heart syndrome undergoing comprehensive stage II palliation. Exit angiography revealed lack of flow to the left pulmonary artery, and therefore intraoperative hybrid stent placement using a direct approach under angiographic guidance was recommended (videos from Dr. John Cheatham, Nationwide Children's Hospital, Columbus, OH). (**c**) The stent is then expanded under fluoroscopic guidance and a final angiography performed as a hand injection through the sidearm of the sheath, documenting appropriate stent position without any vascular injury. There was minor jailing of the left upper lobe branch (MP4 4568 kb)

**Video 3** Video complementing the still images (Fig. 31.2) in an adult patient with status post repair of tetralogy of Fallot with a proximal left pulmonary artery (LPA) stenosis (kink) who underwent for intraoperative stent placement (under direct vision) at the same time. The video documents the initial evaluation of the pulmonary artery anatomy and side branch location, followed by documentation of the result after stent implantation (MOV 7065 kb)

# Part IX

# Step-by-Step Procedures: Miscellanea

# Retrieval Techniques

# 32

Duarte S. Martins, Inês C. Mendes, João R. Silva, and Rui Anjos

## 32.1 Introduction

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. In most cases, unintentional embolization of devices occurs during interventional procedures and can be retrieved at the same occasion. However, in a considerable number of cases, lost objects can be incidental findings on imaging studies in asymptomatic patients.

Successful endovascular retrieval has been described in over 90% of cases. A small number of patients will require a hybrid or surgical approach. Unsuccessful retrievals requiring surgical approach, frequently with cardiopulmonary bypass, are more frequent with large devices, usually atrial septal defect (ASD) or patent ductus arteriosus (PDA) closure devices.

The most frequently embolized material requiring retrieval is by far catheter fragments as the result of fractured long-term venous lines. Many other embolized objects have been reported in the literature, including therapeutic devices, coils, stents, venous filters, fractured balloons, guidewires or pacing wires. Generally embolization and migration occur into the systemic veins, right heart or pulmonary arteries. Occasionally devices can migrate into the left heart and the aorta or distally in systemic arteries.

As a rule, embolized catheters and devices should be retrieved in order to avoid severe and potentially fatal complications including arrhythmias, cardiac and vessel perforation, myocardial infarction, vascular occlusion with ischemia or congestion and secondary infection. Nevertheless, in particular cases, leaving embolized foreign bodies in place may be acceptable, like very small fragments positioned in difficult access locations with low hazard potential.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_32) contains supplementary material, which is available to authorized users.

D. S. Martins · I. C. Mendes · J. R. Silva · R. Anjos (✉)
Pediatric Cardiology, Hospital de Santa Cruz, Centro Hospitalar de Lisboa Ocidental, Lisbon, Portugal
e-mail: ranjos@netcabo.pt


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_32

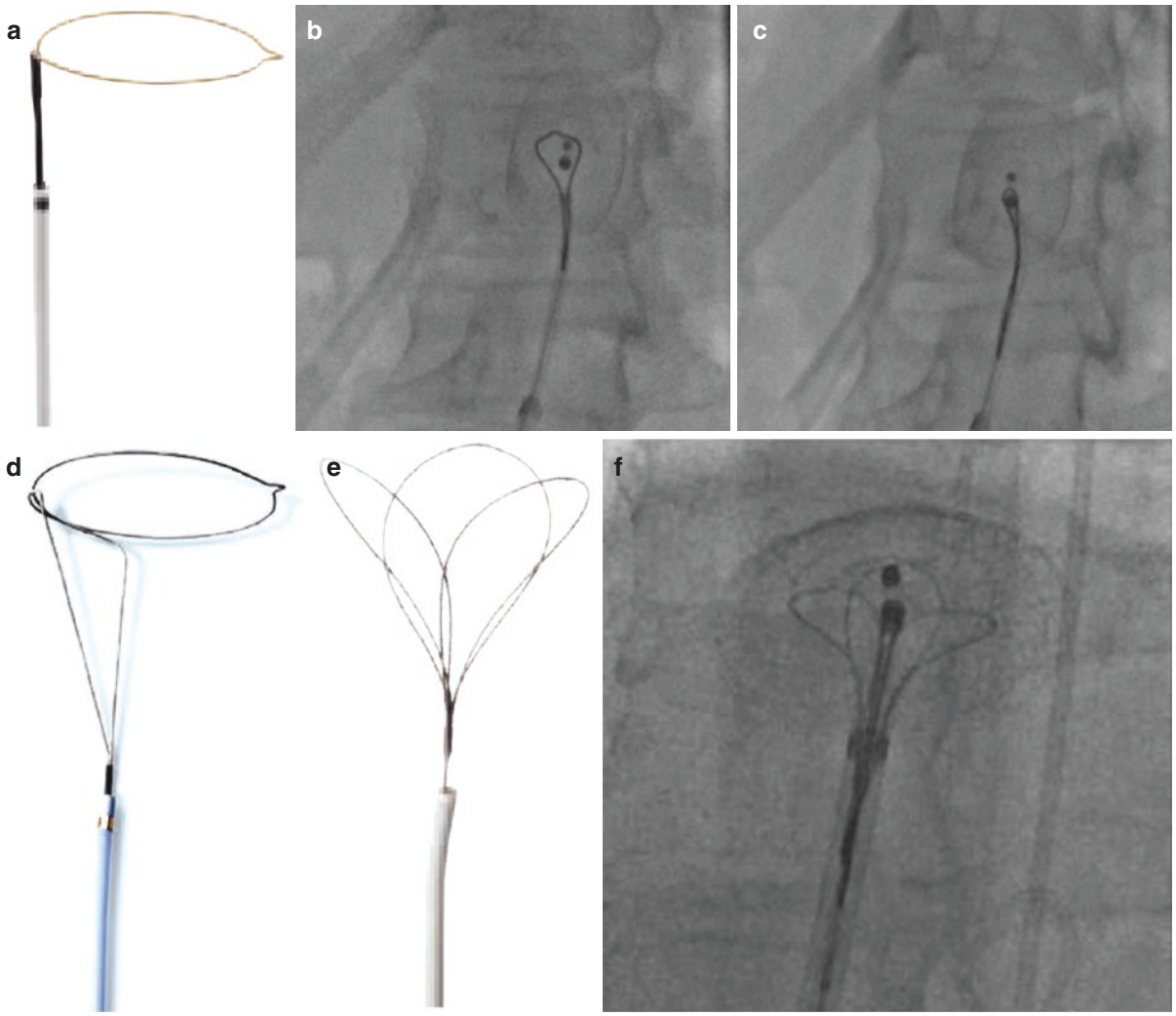



**Fig. 32.1** Retrieval devices overview

**Snares**: These devices are the most widely used type of retrieval device accounting for up to 90% of the devices used in retrieval procedures. It consists of a single or multiple wire loop that closes upon itself when pulled into a catheter grasping the foreign body

- *Single loop snares:* The simplest type and most frequently used. There are many variations with the loop generally coming at an angle with the wire. One of these is the Merit Medical ONE Snare® (Fig. 32.1a) which also displays a small fold in the loop, theoretically providing a better grasping of the foreign body. Single loop snares open their loop when completely outside the snare catheter (Fig. 32.1b) which is gradually closed around the foreign body as it is pushed into the catheter (Fig. 32.1c, Video 1a). Snare sizes typically range from 5 to 35 mm advancing through 4–6 Fr catheters.
- *Multiple loop snares:* With several loops in order to maximize the chances of capturing the foreign body. The PFM Multi-Snare® (Fig. 32.1d) provides two orthogonal loops with diameters ranging from 5 to 40 mm, introduced through 4–6 Fr catheters. More complex snares exist with 3-loop options such as the Merit Medical EN Snare® (Fig. 32.1e, f, Video 1b) and 4-loop options such as the Cook Indy OTW Vascular Retriever® (Fig. 32.1g).

**Graspers and forceps**: These devices are specially designed for the retrieval of small linear objects such as coils, catheters or guidewires. They are useful for small vessels with insufficient space for the unfolding of a snare loop. One such example is the Cook Vascular Retrieval Forceps® (Fig. 32.1h) which functions by opening and closing of a metal jaw against a fixed guidewire. Graspers can also be used to retrieve devices which are too large to be retrieved with a snare, as large ASD devices.

**Baskets**: These devices consist of a helical wire mesh that expands when exposed from the catheter. They are particularly useful for engaging spherical or ovoid foreign bodies (such as duct occluders and vascular plugs). The basket is exposed side by side with the foreign body and then rotated in order to draw the said body inside its mesh. Once captured inside the mesh, the foreign body can be secured by closing the basket as it is pulled inside its catheter (Video 1c). One such example is the Andramed AndraBasket® (Fig. 32.1i) composed of four helical wires, with diameters from 12 to 30 mm and profile from 2 Fr to 7 Fr. The Cook basket (Dotter Intravascular Retrieval Device®) is a larger and stronger device, with an 8 Fr shaft, requiring a 9–11 Fr sheath.

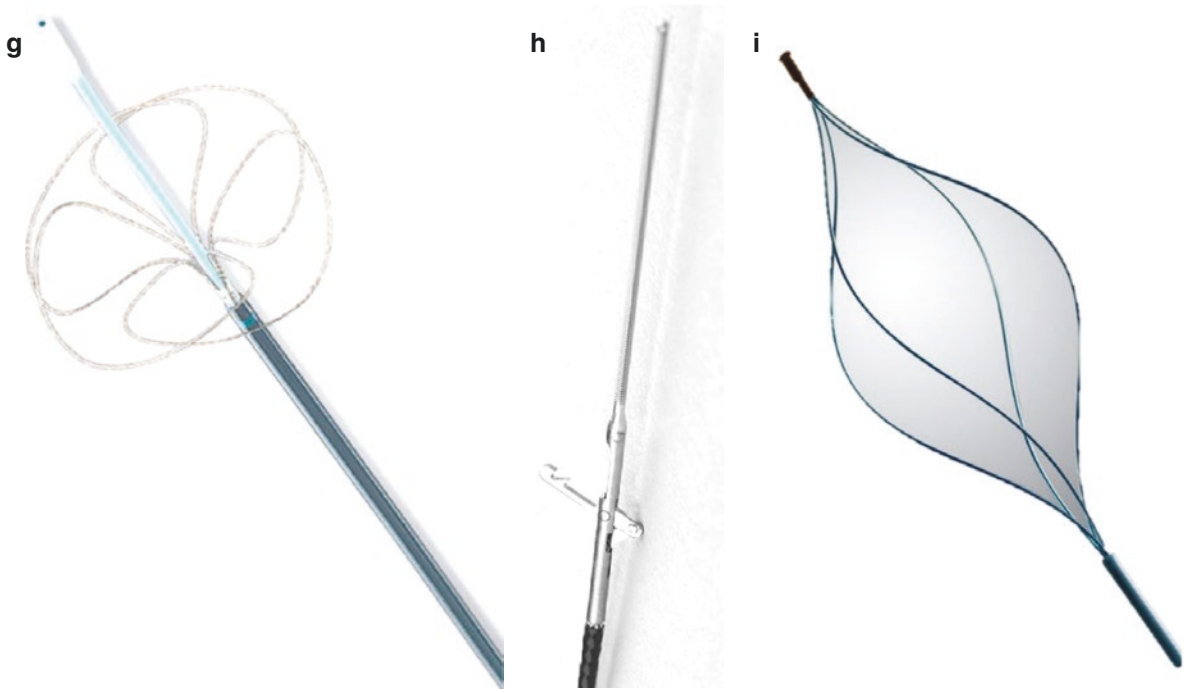



**Fig. 32.1** (continued)

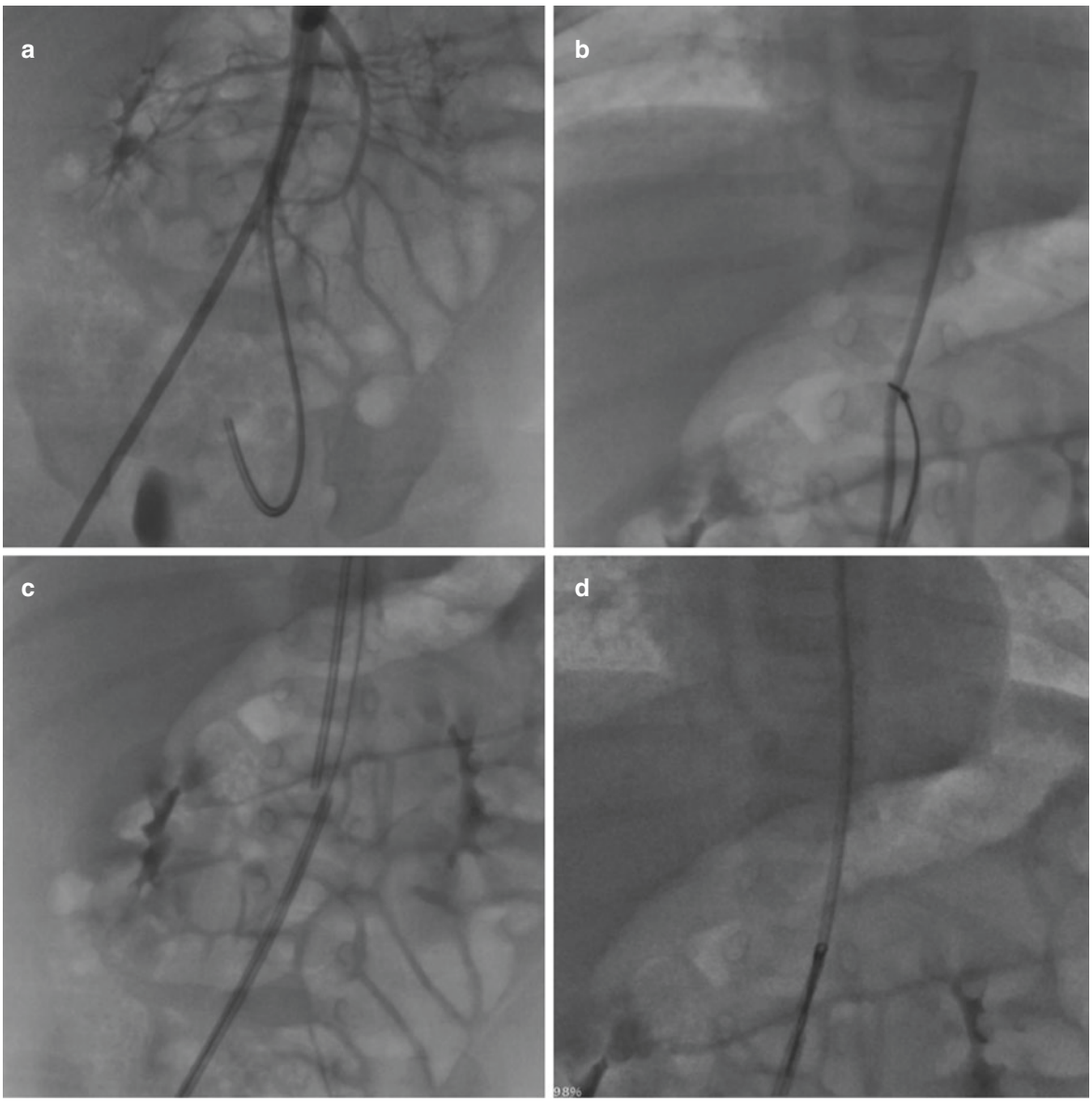



**Fig. 32.2** Step-by-step retrieval technique of an embolized catheter or coil

Fracture and embolization of the distal portion of a 3,5F umbilical arterial catheter upon manual removal

1. Obtain informed consent.
2. Obtain vascular access according to location of the device and planned procedure, and administer a loading dose of heparin (100 UI/kg).
3. Confirm position of the catheter tip by fluoroscopy and angiography (Fig. 32.2a, Video 2a).
4. Prepare the retrieval device and guiding catheter or sheath. Usually small linear objects are easily captured using a small snare (in this case, a 10 mm single snare was used) and retrieved into a long sheath or guiding catheter to avoid vessel damage. For such a small catheter, a 5 Fr sheath may suffice, while 5 Fr catheters should be retrieved into a 6–7 Fr sheath or guiding catheter, and 7 Fr catheters should be retrieved into a 9 Fr guiding catheter or an 8–9 Fr valved sheath. If using a guiding catheter, it may be helpful to use a haemostatic valve with a side port (such as Tuohy-Borst).
5. Advance the retrieval system proximally to the catheter tip. If using large systems in relatively small vessels, it may be helpful to obtain position with a small diagnostic catheter and then exchange it over the wire to avoid vessel damage or dislodgement of the catheter tip (causing it to embolize distally). Open the snare loop, rotate it (by turning the torqueing device) so that the catheter tip is contained inside, and then run it along the catheter long axis if necessary, to release it from the wall of the vessel, in case of long-standing embolization. Close the loop by advancing the snare catheter over the snare achieving a stable grasp (Fig. 32.2b). Secure the grasp by advancing and tightening the torquer over the snare to the proximal end of the snare catheter, keeping tension between the two.
6. Make sure the foreign object is free moving before retrieving it, to avoid vascular damage. In this case, the proximal end of the catheter was still inside the umbilical artery. The snare catheter tip was advanced into the left subclavian artery, freeing the end of the catheter (Video 2b).
7. Successful retrieval into the sheath can be facilitated by improving the alignment between sheath and foreign body (Fig. 32.2c) and carefully advancing the snare proximally along the foreign body so as to grasp it closer to the tip of the sheath aiding manipulation (Fig. 32.2d).
8. A nice technique for the retrieval of hollow object such as a catheter tip is to advance a guidewire into its lumen (Fig. 32.2e, Video 2c). The guidewire directs the catheter tip straight into the sheath, allowing safe removal (Fig. 32.2f, Video 2d).

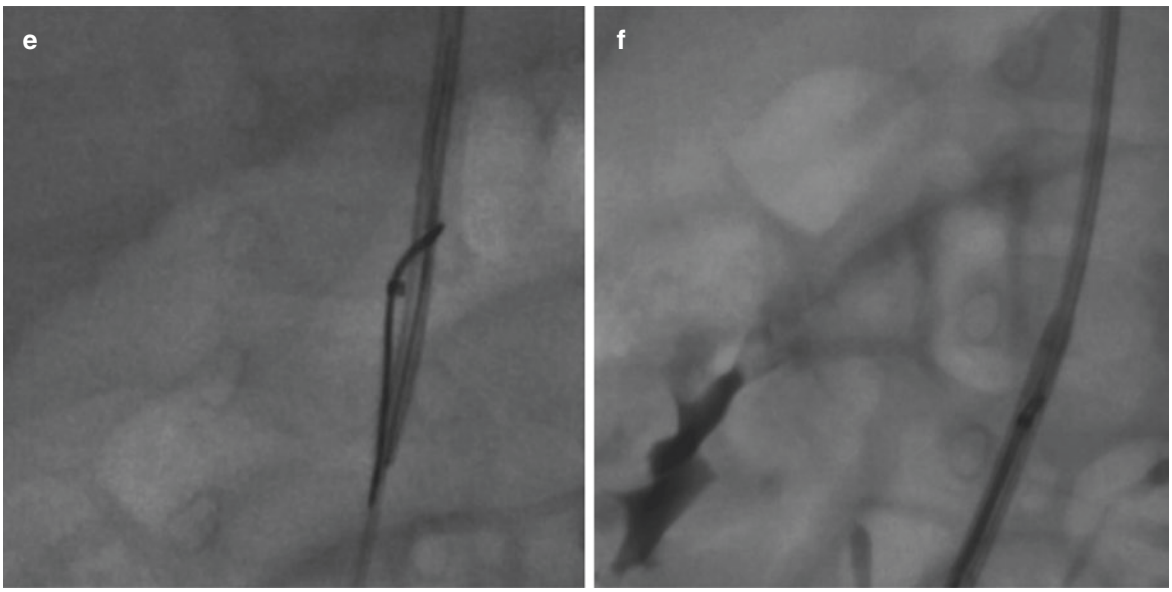



**Fig. 32.2** (continued)

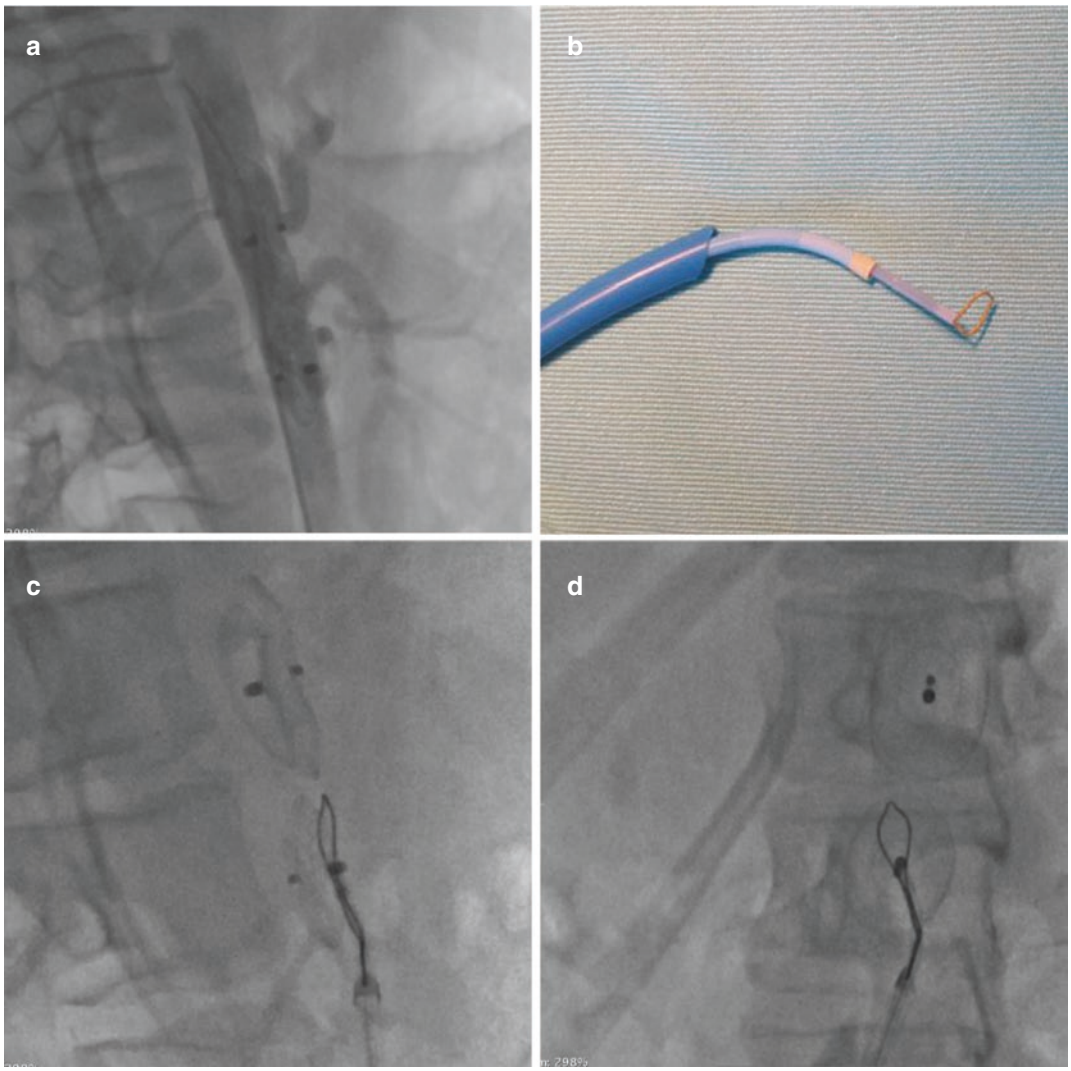



**Fig. 32.3** Step-by-step retrieval technique of an occluder device (ASD)

In this case, two embolized ASD occluder devices (Lifetech CeraFlex ASD occluder® 10 and 14 mm) were found in the abdominal aorta at predischarge echo scan. The patient was completely asymptomatic

1. Obtain informed consent.
2. Obtain vascular access according to location of the device and planned procedure, and administer a loading dose of heparin (100 UI/kg).
3. Confirm position of the embolized devices by fluoroscopy and angiography (Fig. 32.3a, Video 3a).
4. Prepare the retrieval device. A good option for large devices is to use a multiple coaxial telescopic system, formed by a snare, a guiding catheter and a large valved sheath such as displayed in Fig. 32.3b. The sheath should be at least 2 Fr larger than the delivery sheath for the same device (if the embolized device is the largest device delivered through a given sheath size, it might be a good idea to go 3 Fr larger). Sheath bevelling of 30–40° as seen in the image is only possible in non-armoured sheaths as the Cook Mullins Check-Flo® and provide a larger sheath distal end area. Increasing the distal sheath mouth area is performed at the expense of both sheath resistance and safety (care should be taken not to manipulate the sheath without the distal protection of the dilator or guiding catheter). As for the guiding catheter, a curved tip such as Judkins right catheters is helpful to direct the snare.
5. Advance the coaxial system into the vessel taking care not to expose the bevelled tip of the sheath to the arterial wall. The sheath should be placed close enough to stabilize the system and allow quick retrieval while directing the snare and guiding catheters.
6. It is generally preferable to aim at the right disc attachment pin as it gives a small yet stable snaring position while allowing the natural folding of the device into the retrieval sheath. This can usually be caught by unfolding the snare proximally and gently retrieving it (Fig. 32.3c). Confirm correct apposition of the snare and pin using fluoroscopy at an approximately right angle (Fig. 32.3d).
7. Use the angle of the guiding catheter to guide the closing snare. Pull the snare into the guiding catheter stabilizing the grip on the device tip (Fig. 32.3e, f, Video 3b). Remember to keep tension in the snare—a torquer may help secure the snare against the proximal end of the catheter.
8. Gently pull the device-snare-guiding catheter into the sheath (Fig. 32.3g, Videos 3c and 3d).
9. Sometimes correct apposition of the snared device and the sheath might pose a challenge, as was the case in the second device retrieved (Fig. 32.3h, Video 3e). A second snare may help correctly direct the ASD occluder device (Fig. 32.3i).
10. If insertion into the sheath is still difficult despite correct apposition, it may be helpful to simultaneously gently advance the sheath over the device-snare-guiding catheter system (Fig. 32.3j, Video 3f). Increasing the sheath size is another alternative.

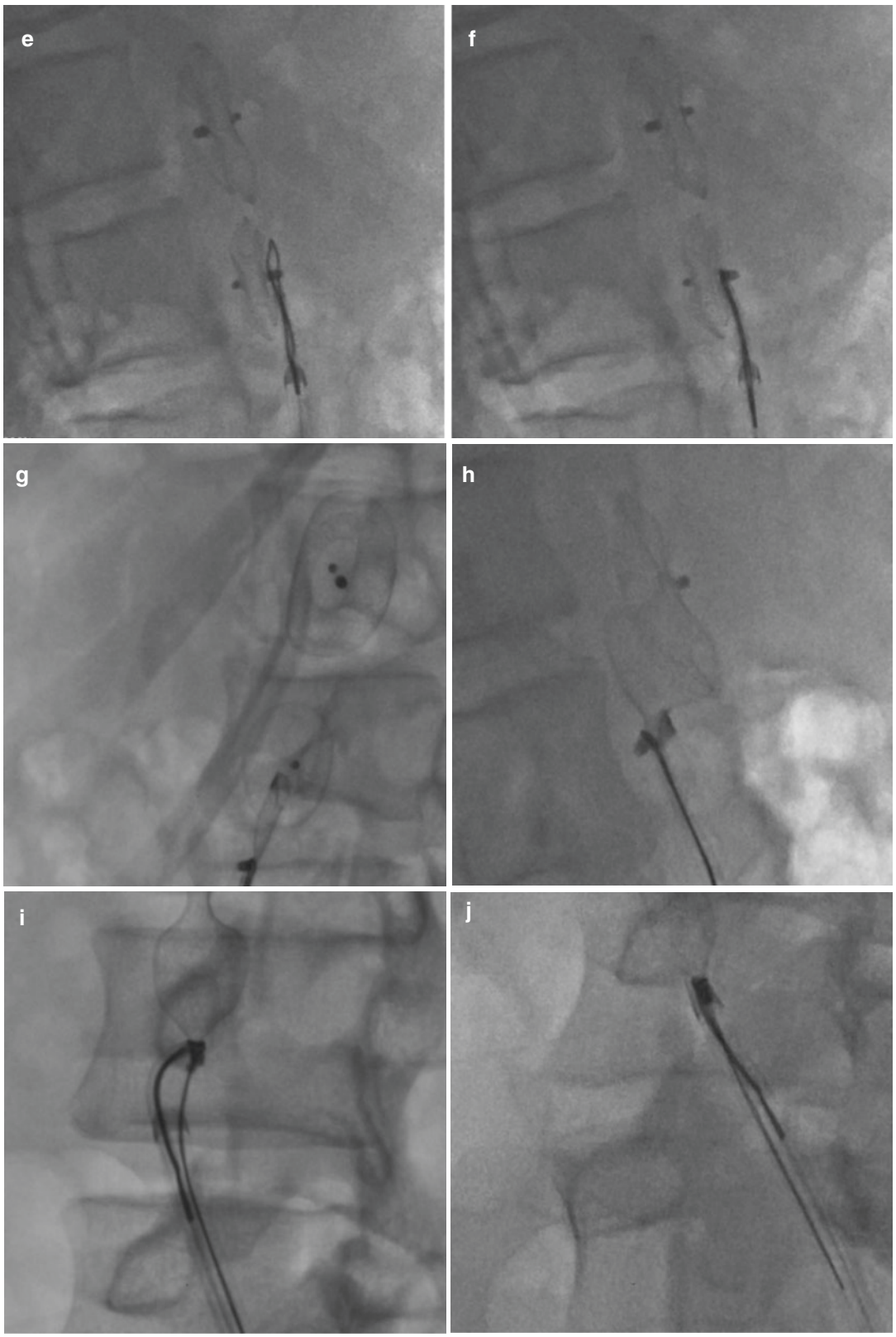



**Fig. 32.3** (continued)

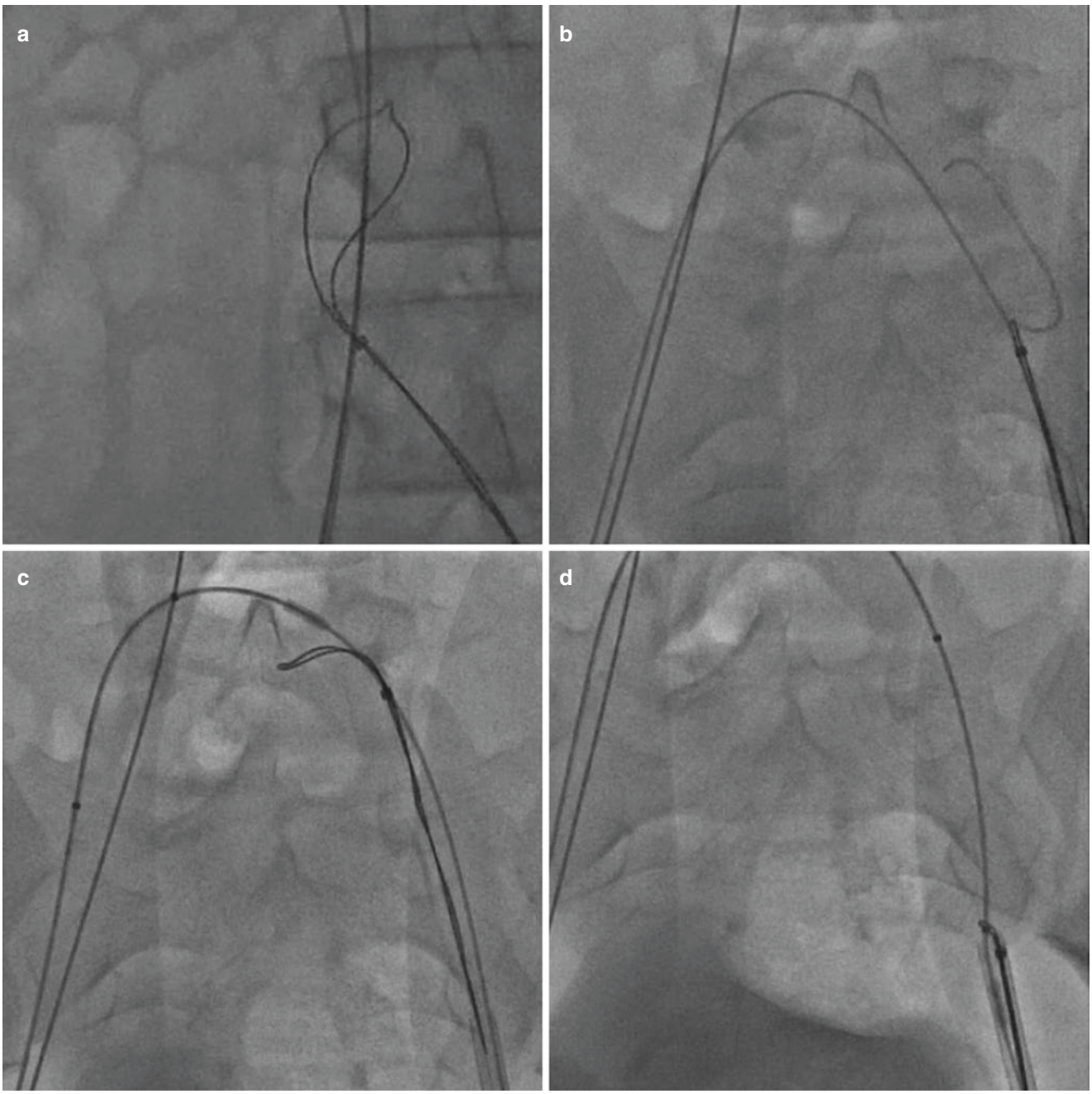



**Fig. 32.4** Step-by-step retrieval technique of a circumferentially ruptured balloon (see also Fig. 3.11 Chap. 3)

1. Avoid losing guidewire position as it will prevent vascular embolization of the distal part of the balloon.
2. Obtain contralateral femoral venous access, inserting an introducer 2 Fr larger than the sheath required for balloon insertion.
3. From this contralateral access, snare the guidewire distally to the balloon (Fig. 32.4a, Video 4a) forming a loop that avoids embolization of the distal end of the balloon. Extract the distal end of the guidewire through the contralateral sheath (Fig. 32.4b, Video 4b).
4. Advance again the snare over the guidewire to capture the balloon (Fig. 32.4c, Video 4c).
5. Cut the balloon shaft and remove the proximal part of the balloon catheter conserving the guidewire loop. The balloon can now be easily pulled inside the contralateral sheath (Fig. 32.4d, Video 4d). A diagnostic catheter such as a multipurpose catheter inserted over the wire on the proximal end of the balloon is helpful in assisting contralateral removal by pushing the balloon into the contralateral sheath.

**Video 1a** (M4V 4 kb)

**Video 1b** (MOV 4 kb)

**Video 1c** (MOV 4 kb)

**Video 2a** (M4V 4 kb)

**Video 2b** (M4V 4 kb)

**Video 2c** (M4V 4 kb)

**Video 2d** (M4V 4 kb)

**Video 3a** (M4V 4 kb)

**Video 3b** (MOV 4 kb)

**Video 3c** (M4V 4 kb)

**Video 3d** (M4V 4 kb)

**Video 3e** (M4V 4 kb)

**Video 3f** (M4V 4 kb)

**Video 4a** (MOV 4 kb)

**Video 4b** (MOV 4 kb)

**Video 4c** (MOV 4 kb)

**Video 4d** (MOV 4 kb)

# Pericardiocentesis

# 33

Maarten Witsenburg

## 33.1 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.

## 33.2 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. 33.1). Right atrial collapse in late diastole, increased

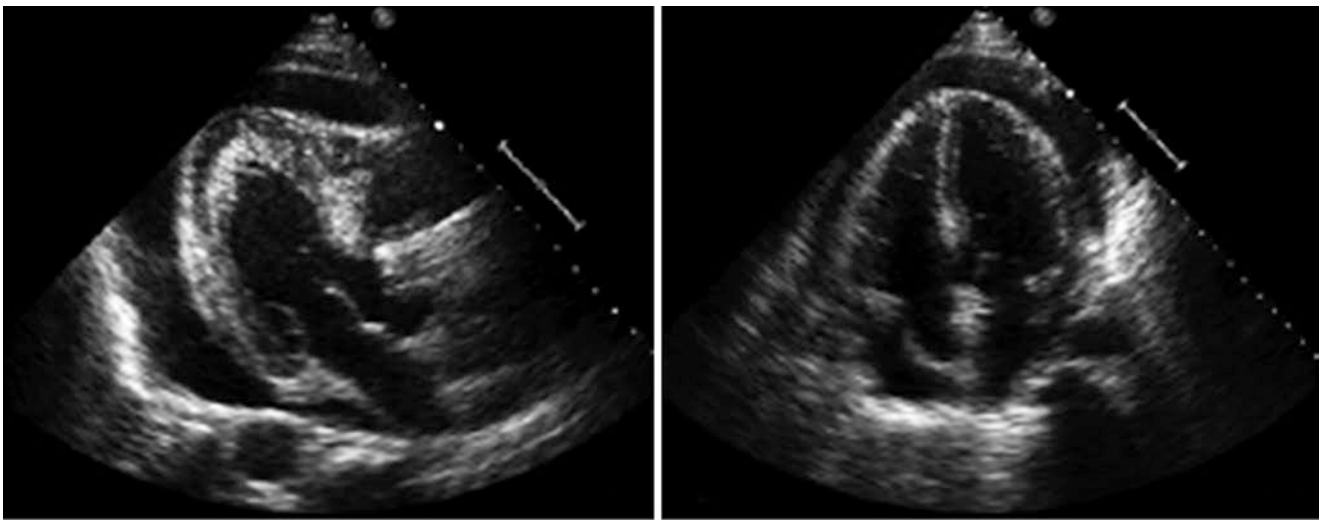


**Fig. 33.1** Echocardiographic four-chamber and long-axis view showing moderate pericardial effusion

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


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_33

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.

## 33.3 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

## 33.4 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.

## 33.5 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.

## 33.6 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.

## 33.7 Access and Drainage

A pericardial puncture set is prepared (Fig. 33.2).

Positioning the patient in a 30° head-up angle may help pooling the effusion at the inferior site of the heart. With the help of echocardiography, the location of the effusion is reconfirmed and the appropriate puncture site is marked. The patient is draped and cleansed with an aseptic solution. The skin and subcutaneous tissue are infiltrated with a local anaesthetic. The needle (appropriately long for patient size) is slowly advanced through the skin at an angle of 15–30° pointing at the left shoulder (Fig. 33.3). Mild negative pressure with a 5–10 mL Luer-Lok syringe is applied. The patient monitor is continuously checked for arrhythmias. Passing the parietal pericardium into the pericardial space may be felt as a pop, and then it should be possible to gently aspirate fluid. When necessary, the access can be echocardiographically confirmed by injecting some agitated saline.

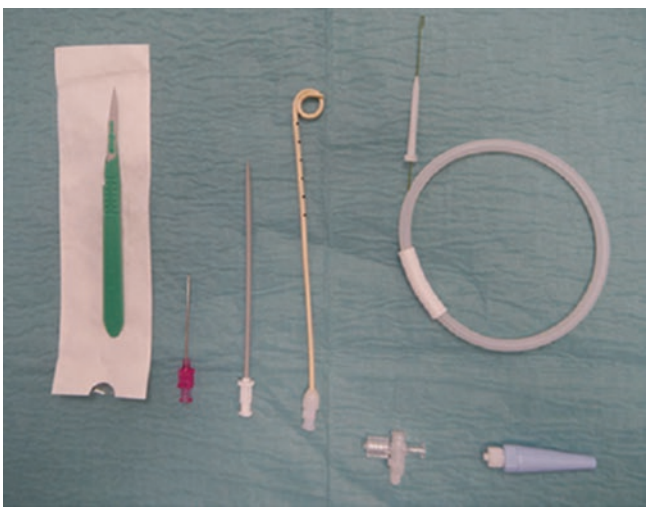


**Fig. 33.2** Paediatric pericardial drainage set with scalpel (William Cook Europe, Bjaeverskov, Denmark)

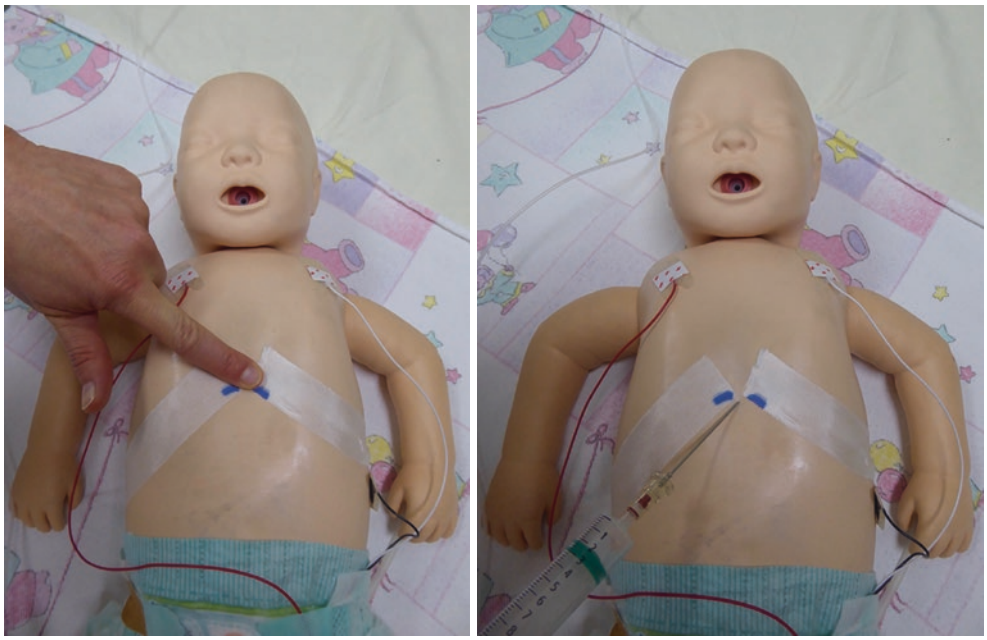


**Fig. 33.3** Demonstration of pericardial puncture approach on a dummy infant. After identifying the lower end of the sternum, this site is marked. The needle is inserted just below the sternum and slowly advanced through the skin at an angle of 15–30° pointing at the left shoulder

(Alternative but nowadays less frequently used techniques include ECG monitoring from the aspiration needle, pressure monitoring from aspiration needle, contrast injection and/or observation of wire curve once introduced during fluoroscopy).

Depending on the urgency and indication for pericardial drainage, some more fluid is aspirated and a J-wire is inserted. The wire advance should not be forced against resistance. The entry site is dilated with a 6–8 F dilator and a (pigtail) catheter with multiple side holes is advanced for continuing drainage. Fluid is collected for laboratory analysis and culture. A three-way stopcock is connected with a 20–50 mL syringe and collection bag. The drain can be sutured if continued drainage is expected. In case of haemorrhagic fluid aspiration, a rapid comparison of the fluid and whole blood haematocrit may confirm the proper drainage site.

## 33.8 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.

# Endomyocardial Biopsies

# 34

Davide Marini and Andrea Wan

## 34.1 Indications

Endomyocardial biopsy of the right ventricle (EMB) remains the gold standard for in vivo diagnosis of rejection in cardiac allograft patients. EMB may also have a role in the diagnosis of other myocardial diseases such as cardiac tumours, myocarditis or other infiltrative cardiovascular diseases such as amyloidosis, sarcoidosis, Fabry disease and arrhythmogenic right ventricular dysplasia (ARVD).

## 34.2 Patient Selection

Despite the increasing accuracy of non-invasive techniques to detect myocardial inflammation, surveillance EMB remains an important diagnostic tool for rejection surveillance after heart transplantation. EMB may also be warranted after reduction of immunosuppression and for monitoring after treatment of acute rejection episodes.

Clear indications for EMB in the diagnosis of other myocardial diseases are difficult to define. In 2007, the American Heart Association (AHA), the American College of Cardiology (ACC) and the European Society of Cardiology (ESC) published guidelines (Table 34.1) regarding the role of EMB in the management of cardiovascular diseases. Given the limited data available, these recommendations are based mostly on expert opinion. The main value of EMB is in the diagnosis of myocardial diseases that have unique treatments and prognoses which cannot be diagnosed by less invasive tests.

## 34.3 Procedure

In adolescents and adults with normal ejection fraction, EMB can be performed using local anaesthesia and/or sedation. Infants and young children require general anaesthesia, endotracheal intubation and mechanical ventilation. Standard monitoring includes electrocardiographic rhythm, blood pressure and pulse oximetry.

The use of heparin prophylaxis might be avoided if the approach is exclusively venous and there are no intracardiac shunts. Some centres use 50 UI/Kg of heparin to prevent venous thrombosis and vascular occlusion. The old catheterization reports should be reviewed to exclude known vascular access issues. The most common venous access points are the internal jugular vein and the femoral vein. The procedure is routinely done with fluoroscopic guidance. Single plane or biplane may be used depending on operator preference and available facilities. The addition of echocardiography guidance may be useful especially when certain areas are targeted for sampling (i.e. cardiac tumour).

In general, five to ten samples, each 1–2 mm in size, should be obtained from different regions of the RV septum. The minimum number of samples needed will depend on the testing required for each clinical scenario.

## 34.4 Materials

Endomyocardial biopsies of the right ventricle should be performed with a bioptome through a long sheath which has been positioned in the right ventricle. In general, the following materials are required:

- A short introducer
- An end-hole catheter
- A standard guidewire
- A long sheath (shorter than the selected bioptome) (Fig. 34.1)
- A bioptome (Figs. 34.1, 34.2, and 34.3)
- Containers for the specimen

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_34) contains supplementary material, which is available to authorized users.

D. Marini (✉) · A. Wan
Division of Cardiology, Department of Pediatrics,
The Labatt Family Heart Centre, The Hospital for Sick Children,
University of Toronto School of Medicine, Toronto, ON, Canada
e-mail: davide.marini@sickkids.ca



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_34

**Table 34.1** Class of recommendation and level of evidence in 14 clinical scenarios

| 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 haemodynamic 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, which responds to usual care within 1–2 weeks | IIb | B |
| 10 | Heart failure of >3 months' duration associated with a dilated left ventricle, without new ventricular arrhythmias or second- or third-degree heart block, which 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 |

Some common bioptomes available on the market are listed in Table 34.2. With good positioning when sampling, a 5 or 6 fr bioptome should usually be sufficient to obtain adequate samples (Figs. 34.4, 34.5, 34.6, 34.7, 34.8, and Videos 1–4).

## 34.5 Potential Complications

Potential major complications include cardiac perforation with pericardial tamponade requiring pericardiocentesis, haemopericardium, AV block, pneumothorax and tricuspid regurgitation. Of note, the risk of EMB is highest in young patients with poor ventricular function, a dilated thin-walled ventricle and high right ventricle pressure.

In cases of pericardial tamponade, immediate pericardiocentesis should be performed, promptly returning the blood drained from the pericardium through the vascular introducer. The standard manoeuvres of resuscitation, surgical repair of the lesion and implantation of a ventricular assist device may be required.

**Acknowledgements** We thank our technologists Dariusz Mroczek and Renato Prosio for providing images and videos.

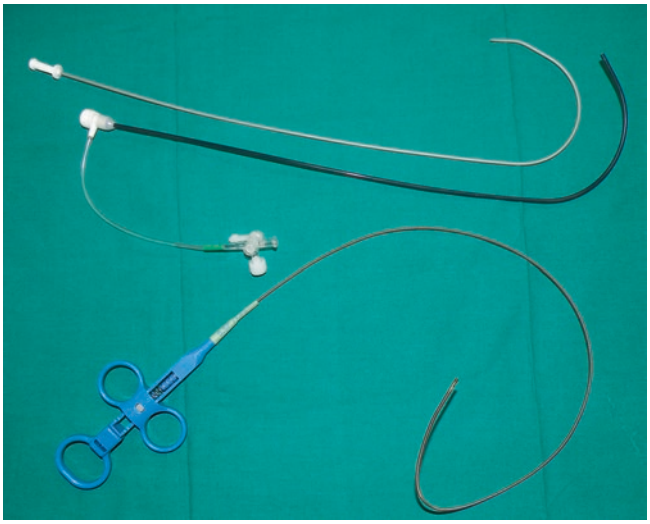


**Fig. 34.1** The bioptomes commonly employed are single use. There are two different types of bioptomes available. One type has a pre-shaped distal end and the other has a flexible shaft and unshaped distal end. In this picture, we show a 50 cm long 5.5 fr flexible bioptome (Cordis®) that may be used in a 6F 40 cm long pre-shaped Flexor® Check-Flo® Introducer® (Cook®). This combination can be used to perform biopsies from the jugular vein. In infants and small children, this combination could be long enough to be used from the femoral vein

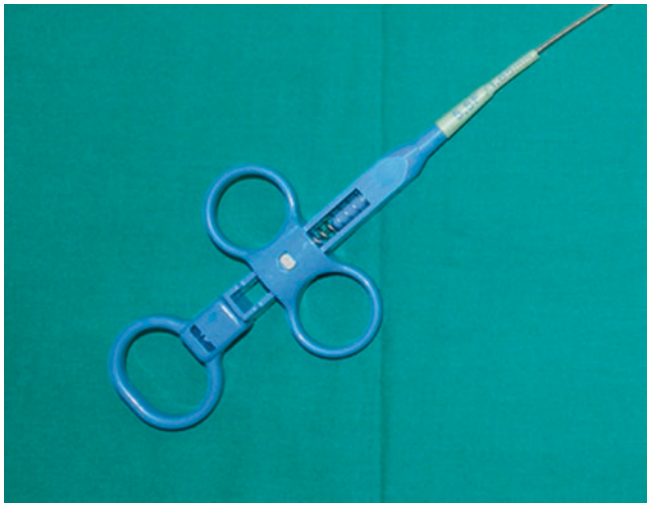


**Fig. 34.2** Modern bioptomes have a 3-pull ring handle with a spring so that samples can be held easily while being removed. The central ring of the handle can rotate to accommodate any hand position. The two anterior rings are grabbed with the forefinger and the middle finger, and the posterior ring with the thumb. The jaws of the bioptomes are opened by pushing the two anterior rings forward; pulling them back to the posterior ring closes the jaws

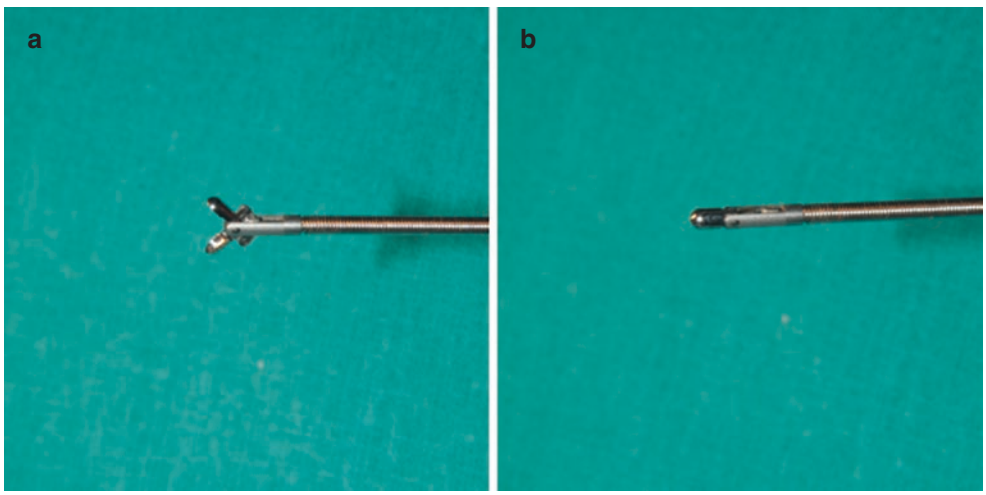



**Fig. 34.3** In these pictures, we show a bioptome with forceps completely open (Panel **a**) and closed (Panel **b**). Please note that the bioptome has sharpened cusps designed to pinch rather than to cut the myocardial muscle

**Table 34.2** Common bioptomes of different sizes and shaft lengths

| Model | Size (fr) | Shaft length (cm) |
|---|---|---|
| Cook® | 3.0, 5.2 | 60 and 120 |
| Cordis® | 5.5, 7.0 | 50 and 104 |
| Sparrow Hawk® | 5.0, 6.0, 7.0 | 50 and 105 |
| Novatome™ | 6.0, 7.0, 8.0, 9.0 | 50 and 100 |
| Argon® | 5.0, 5.5, 6.0, 7.0,7.5 | 50 and 105 |

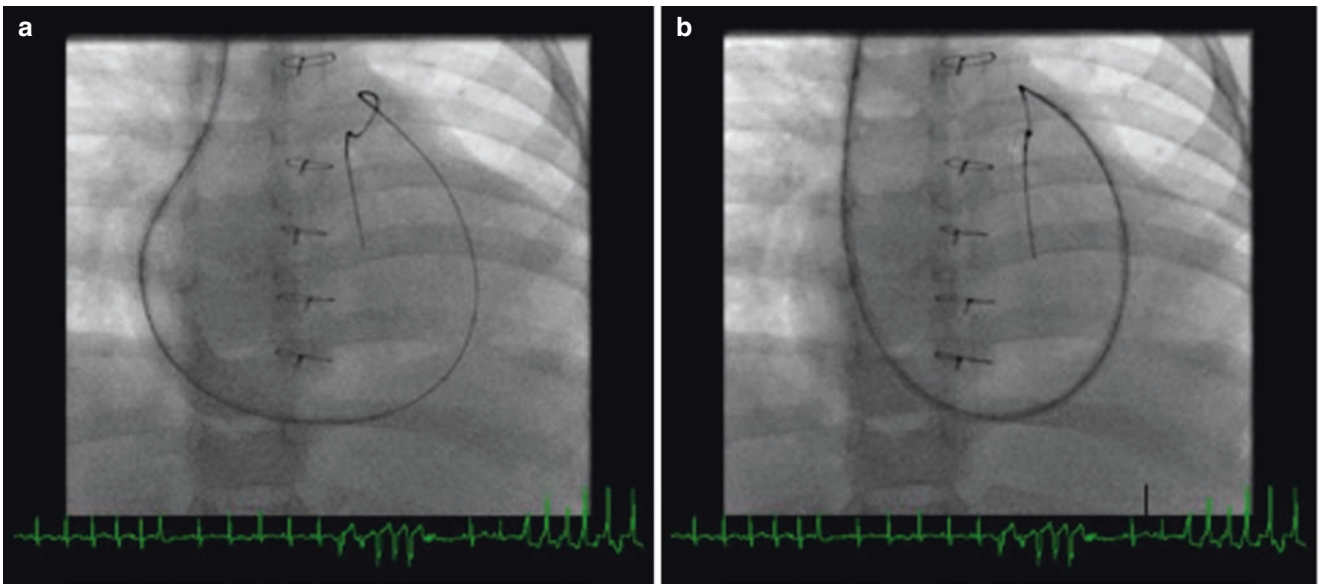



**Fig. 34.4** A standard guidewire has been placed in a distal pulmonary artery branch via a standard end-hole catheter from an internal jugular vein approach (Panel **a**). The long sheath is advanced gently over the wire (Panel **b**). The wire and dilator are then removed, leaving the tip of the long sheath in the right ventricle

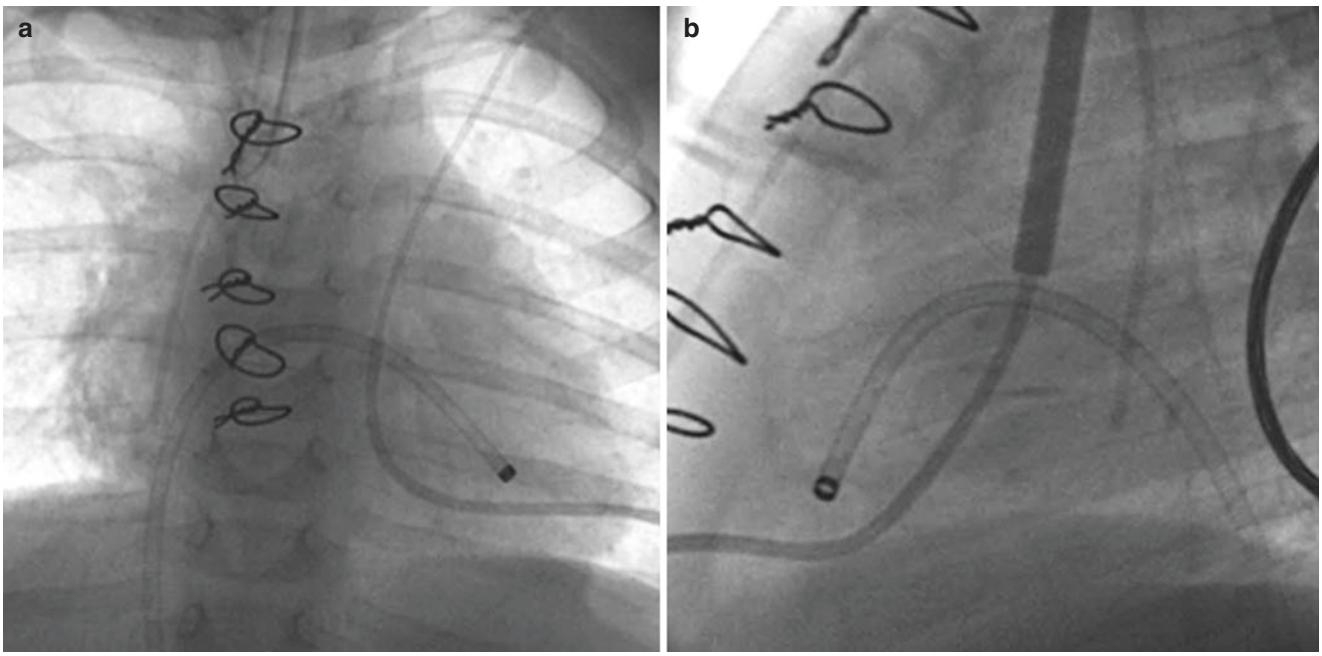



**Fig. 34.5** Panels (**a** and **b**) show the position of a long sheath in the right ventricle which was inserted via the femoral vein in anteroposterior and lateral projections, respectively. The samples should be taken from the subapical interventricular septum to avoid cardiac perforation

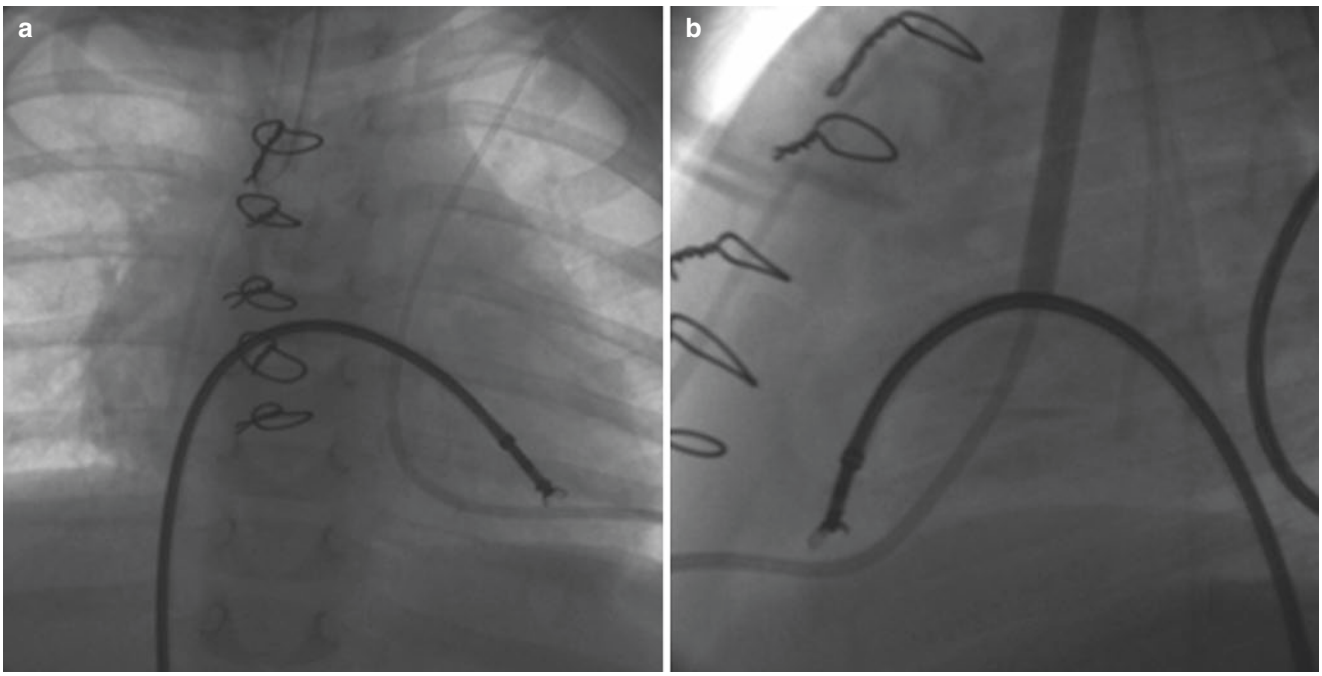



**Fig. 34.6** Panels (**a** and **b**) show in anteroposterior and lateral projections the open jaws of the bioptome, facing the interventricular septum and ready to pinch the muscle

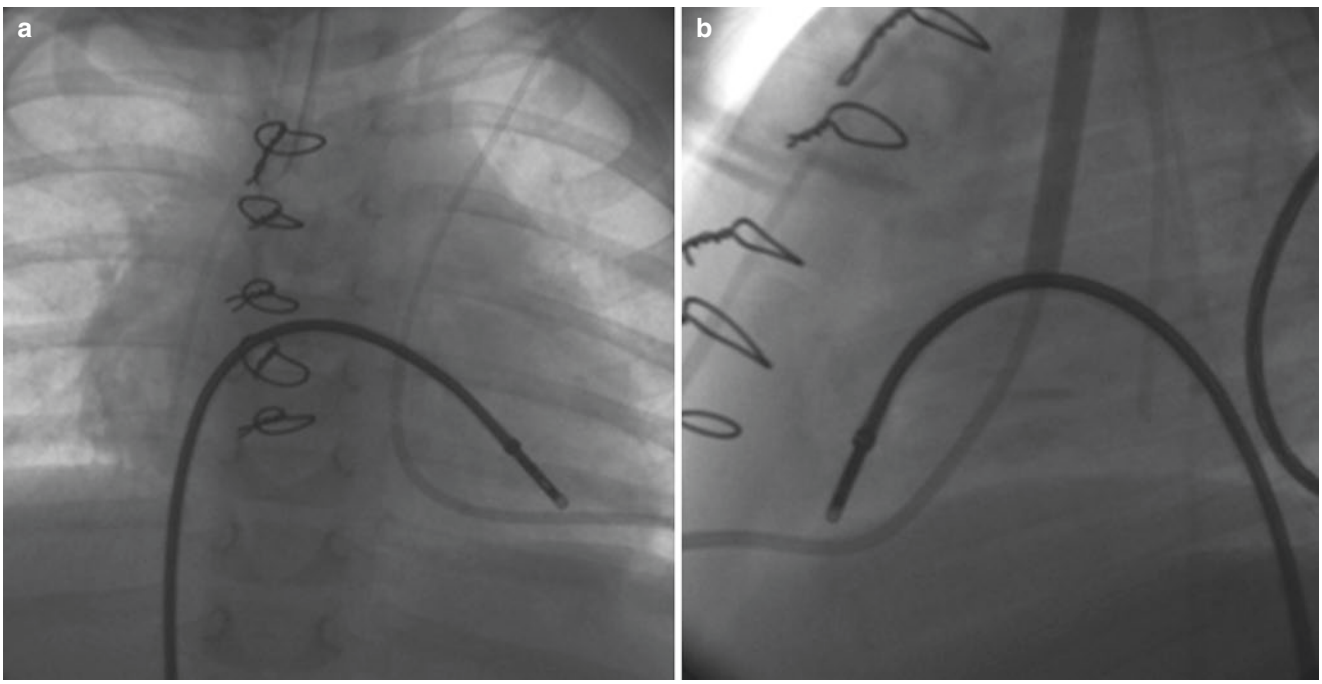



**Fig. 34.7** Panels (**a** and **b**) show in anteroposterior and lateral projections, respectively, the bioptome with the jaws closed, immediately after the sample has be taken. The bioptome can now be retrieved from the long sheath

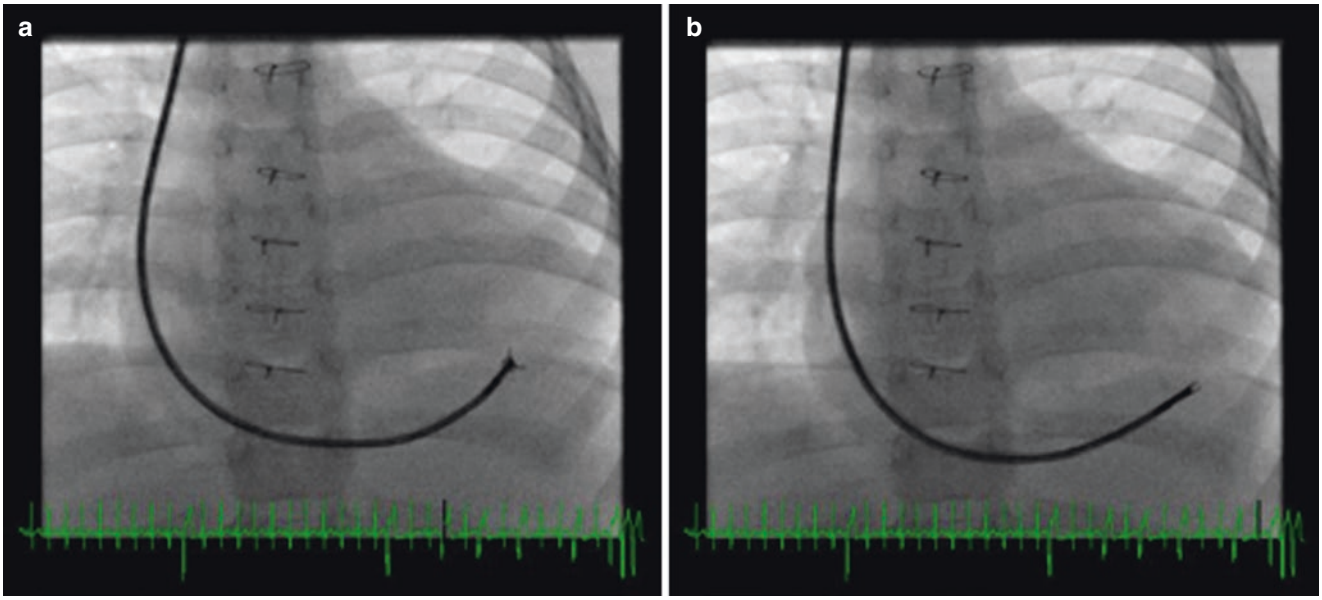



**Fig. 34.8** Panel (**a** and **b**) show in anteroposterior projection the bioptome inserted from the jugular vein with the jaws open and closed, respectively

**Video 1** The long sheath is advanced from the jugular vein over a wire previously placed in the distal pulmonary artery. The wire and dilator are then removed, leaving the long sheath in the right ventricle. Good guidewire position will allow for smooth advancement of the sheath into the right ventricle. In patients after cardiac transplantation, difficulties in advancing the long sheath through the superior vena cava may suggest stenosis at the anastomotic site (MOV 4949 kb)

**Video 2** Before the insertion of the flexible bioptome through the long sheath, it may be useful to curve the distal end of the bioptome to direct the tip towards the interventricular septum. The bioptome should be advanced under fluoroscopic surveillance generally in anteroposterior and 90° lateral (when biplane is used) views. Care should be taken to keep the end of the long sheath stable as the bioptome is advanced. When the tip of the bioptome is near the end of the long sheath, the bioptome handles should be pushed forward, allowing the jaws to open as the tip is carefully advanced out of the long sheath. Once the jaws of the bioptome are seen to be fully open, the bioptome should be advanced slowly to engage the right ventricular myocardium. When the bioptome jaws are engaged in the myocardium, mild resistance may be felt and premature ventricular beats may be provoked. There is also a change in the curve of the bioptome/long sheath due to the mild pressure that is applied. Too much pressure may result in myocardial perforation; however, the jaws of the bioptome must remain engaged with the myocardium to obtain a good sample. The handles of the bioptome are then brought together, closing the jaws and 'pinching' off a sample of tissue (MOV 9484 kb)

**Video 3** The bioptome may then be retrieved, always keeping the jaws closed. Mild resistance may be felt on retrieval as the jaws 'pinch off' a sample. Care should be taken to maintain a stable position of the long sheath. Advancing the long sheath gently as the bioptome is retrieved may help maintain a good position in the RV (MOV 991 kb)

**Video 4** Samples should be taken from different parts of the interventricular septum. This may reduce the risk of myocardial perforation from repeated sampling at the same site, as well as increase the chance of obtaining a diagnosis in disease processes that do not affect the myocardium uniformly. Repositioning the long sheath safely can be achieved by advancing a balloon catheter (i.e. Berman catheter) through the long sheath (MOV 1606 kb)

# Evaluations Before Partial and Total Cavopulmonary Connections

35

Gabriella Agnoletti

- Fontan circulation is the therapeutic option for a large variety of complex heart diseases not suitable for biventricular repair.
- Fontan circulation is usually obtained by two-stage procedures: partial (superior) cavopulmonary connection (PCPC) followed by later total cavopulmonary connection (TCPC or Fontan completion).
- According to the native heart disease, we can face several clinical scenarios: duct-dependent pulmonary circulation, duct-dependent systemic circulation, protected pulmonary vascular bed, and non-protected pulmonary vascular bed.
- The large majority of patients need a neonatal palliation before PCPC (pulmonary banding or BT shunt or hybrid treatment for hypoplastic left heart syndrome).
- Almost all candidates to PCPC and TCPC are evaluated with a catheterization.

Pre-PCPC catheterization aims at evaluating:

- Pulmonary arterial pressure (PAP)
- Size and anatomy of pulmonary arteries
- Associated anomalies

In the presence of BT shunt without an additional pulmonary blood flow, an arterial access is generally needed. This approach will provide a complete evaluation. An angiography in the right subclavian artery, at the origin of the shunt, will show the anatomy of pulmonary arteries. If we want to avoid the arterial approach, the ascending aorta can also been reached via a venous femoral or jugular access. This approach needs more catheter manipulation and can prolong the duration of the exam.

Rarely the shunt has to be entered with a Judkins right catheter to perform an exhaustive evaluation. To enter the shunt, a guidewire (e.g., Terumo, coronary, etc.) can be used and support the catheter as far as both pulmonary arteries. We will then measure pressures and perform an angiography.

Sometimes the shunt cannot be entered; in this case, PAP can be measured via wedge pulmonary vein pressure. Also in this case, we will need a venous access.

In the presence of an antegrade pulmonary blood flow, a ventricular angiography is performed to evaluate both pulmonary outflow and the anatomy of pulmonary arteries. Pressures will be measured into the ventricle and in pulmonary arteries. In the presence of a tight pulmonary banding or severe pulmonary stenosis, it might be impossible to reach the pulmonary artery. In this case, as before, PAP will be measured via wedge pulmonary vein pressure.

Pre-TCPC catheterization aims at evaluating:

- The PCPC anatomy and the pressure in the superior vena cava (SVC)
- PAP
- Size and anatomy of pulmonary arteries
- Associated anomalies

To reach the pulmonary vascular bed, an internal jugular access is generally obtained. We will measure PAP, and if it is elevated (≥12 mmHg), we will also measure the wedge atrial pressure. We will then perform an angiography to evaluate PCPC and the anatomy of pulmonary arteries.

In the presence of a left superior vena cava, a bilateral PCPC is performed. At time of catheterization, both anastomoses have to be evaluated. In fact, in some case, mostly when SVCs have a different size, the competitive flow can prompt thrombosis of the smallest SVC and/or one pulmonary artery.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_35) contains supplementary material, which is available to authorized users.

G. Agnoletti (✉)
Department of Pediatric Cardiology, Hospital Città della Salute, Torino, Italy
e-mail: gagnoletti@cittadellasalute.to.it



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_35

If an antegrade additional pulmonary blood flow is present, the pulmonary artery can be entered via the femoral venous access. If necessary (occlusion of both femoral veins), the femoral arterial access can also be used to reach pulmonary arteries via the ventricle. In the presence of retrograde additional pulmonary blood flow (e.g., BT shunt), an aortography can also be performed. Sometimes we can notice the preferential perfusion of the right pulmonary artery via SVC and of the left pulmonary artery via the ventricle.

To obtain a reliable measurement of PAP, concomitant anomalies have to be looked for in PCPC patients. Main confounding factors that lower PAP are veno-venous collateral vessels and pulmonary fistulae. On the contrary, restrictive atrial septal defect, ventricular dysfunction, aortic coarctation, obstruction to pulmonary venous return, and additional pulmonary blood flow tend to increase PAP.

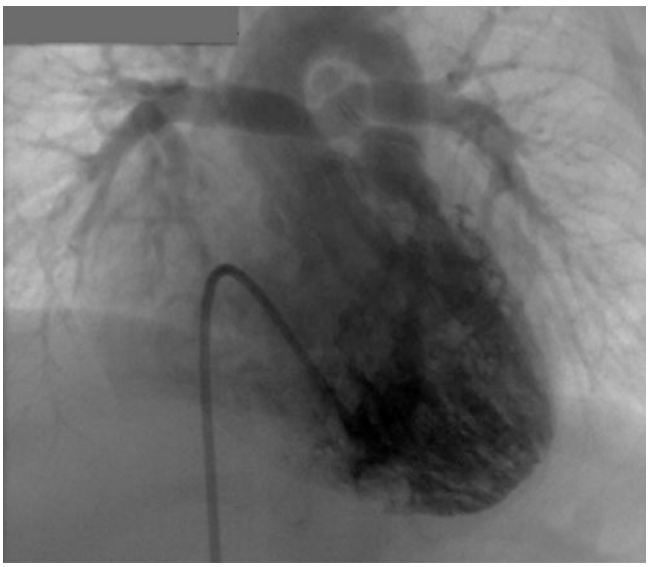


**Fig. 35.1** Patient 1: Double outlet right ventricle with malposition of the great arteries. The patient underwent Rashkind procedure at birth and pulmonary arteries banding at 1 month. The figure shows the ventricular angiography at the age of 6 months. A 5F Berman catheter via the femoral vein access is used. Pulmonary banding is in place and pulmonary arteries have good size

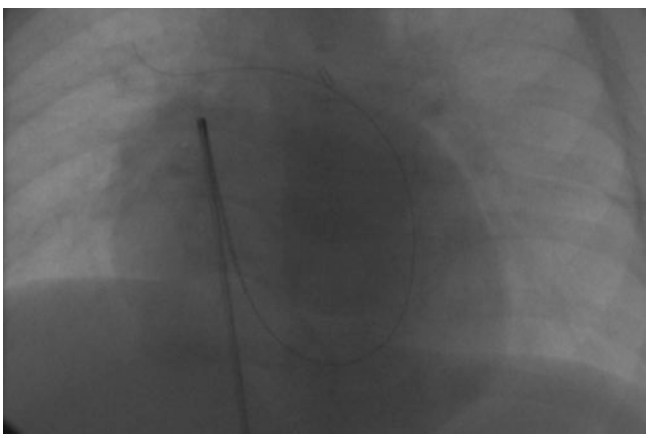


**Fig. 35.2** Patient 1: Pulmonary banding is tight and difficult to pass; a flow-directed catheter on a coronary guidewire couldn't reach the pulmonary trunk. Pulmonary pressure was then obtained via wedge pulmonary vein

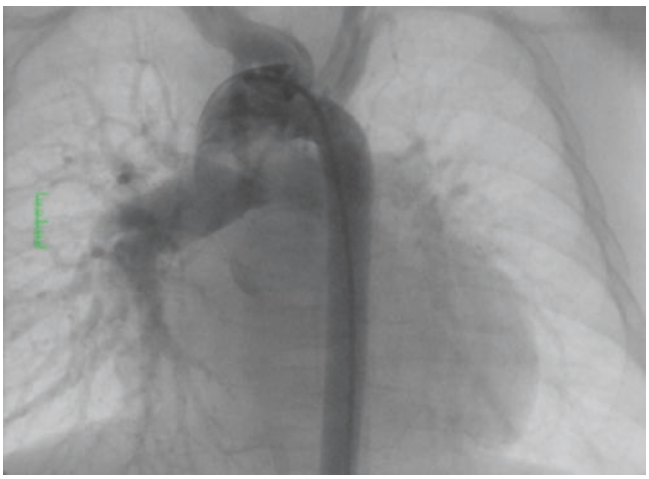


**Fig. 35.3** Patient 2: Tricuspid atresia with pulmonary stenosis. The patient underwent Rashkind procedure at birth and BT shunt at 5 days. This figure shows the aortography at the age of 5 months. A 4F pigtail catheter is used. Aortography rules out the presence of aortopulmonary collateral vessels and shows the origin of the BT shunt. Pulmonary arteries are dilated; a washout is evident in the left pulmonary artery

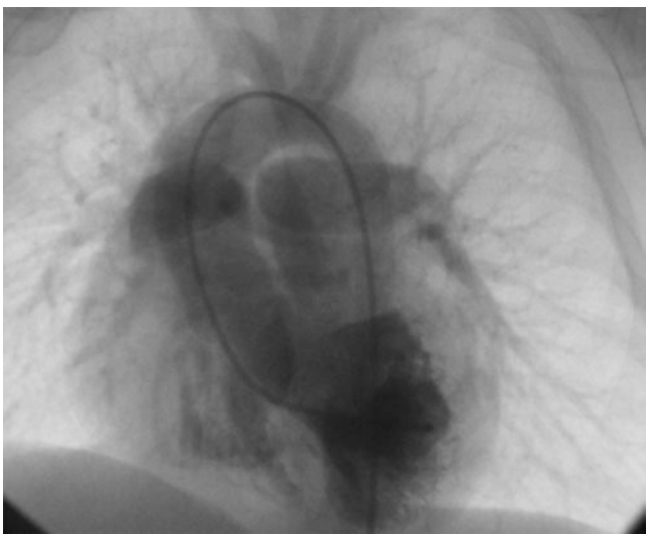


**Fig. 35.4** Patient 2: The figure shows that, at ventricular angiography, there is an abundant antegrade pulmonary blood flow

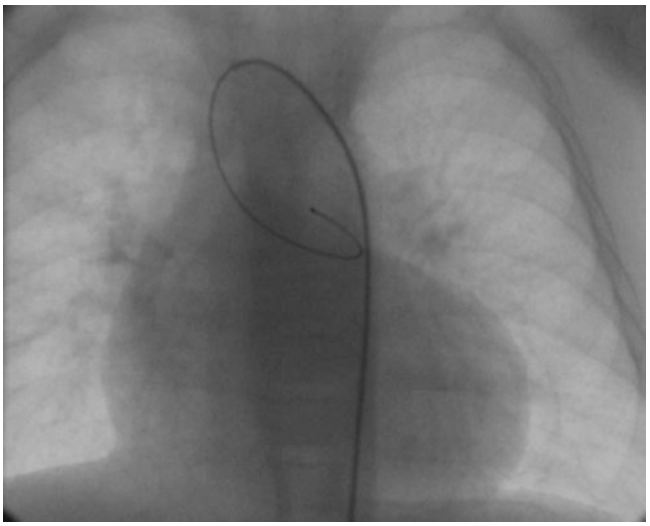


**Fig. 35.5** Patient 2: The BT shunt is entered using a 4F Judkins right catheter supported by a Terumo guidewire

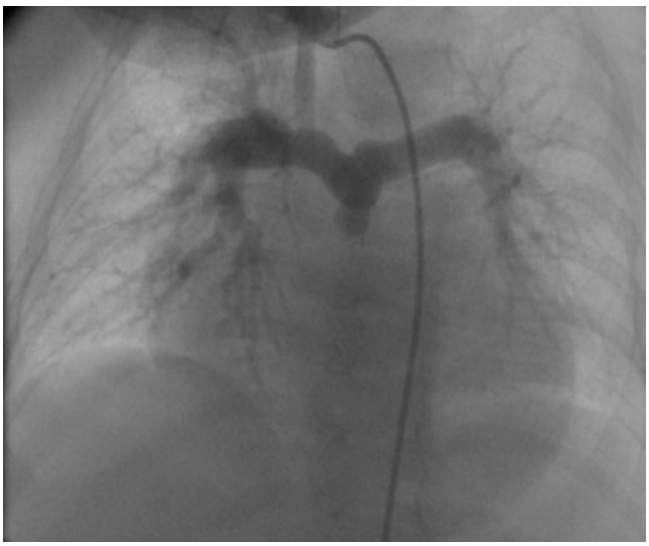


**Fig. 35.6** Patient 3: Pulmonary atresia with intact ventricular septum. This patient underwent BT shunt at 6 days. The nonselective injection of BT shunt, performed with a 4F Judkins right, allows to evaluate the proximal anastomosis and to visualize both pulmonary arteries

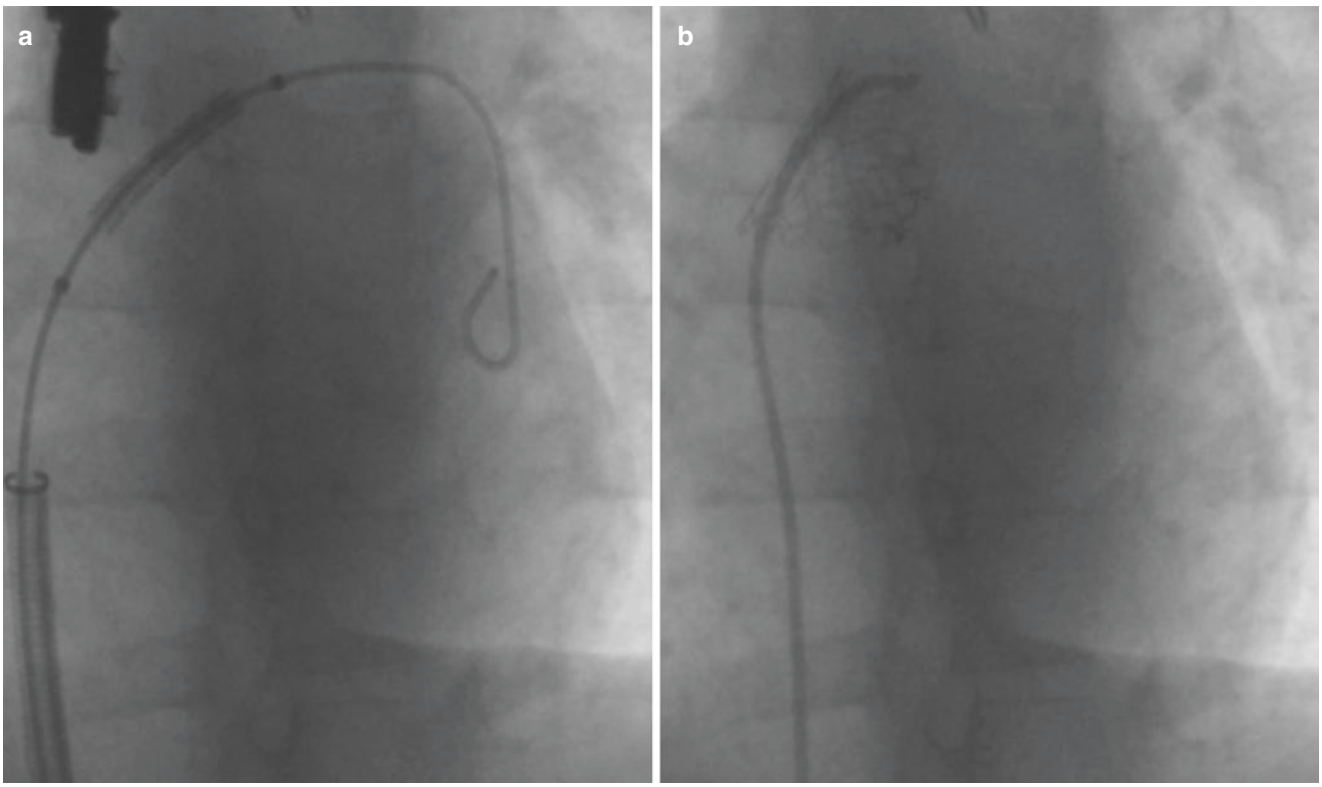



**Fig. 35.7** Patient 1: This patient underwent bilateral PCPC at the age of 6 months. Catheterization at the age of 3 years reveals the presence of a restrictive atrial septal defect with a pressure gradient of 5 mmHg. An 18 mm Genesis XD stent mounted on a 14 mm Tyshak balloon is implanted using a 7F Mullins long sheath. The stent is through the atrial septum (**a**). Final result (**b**)

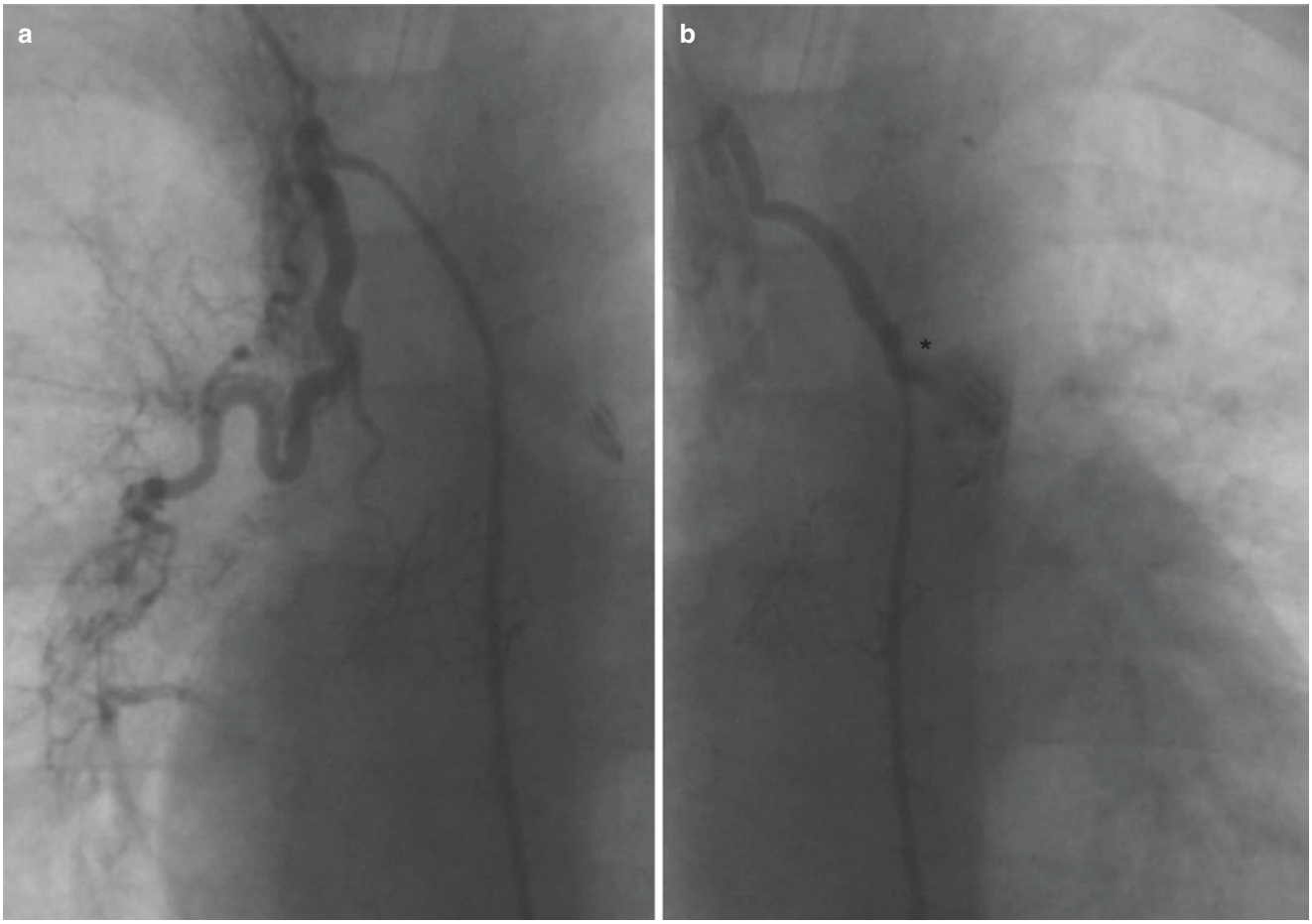



**Fig. 35.8** Patient 1: During the same catheterization, performed at the age of 3 years, a selective injection shows a large aortopulmonary collateral vessel (**a**). After embolization with particles, the right pulmonary vascular bed is no more visible and contrast reflows into the aorta (**b***)

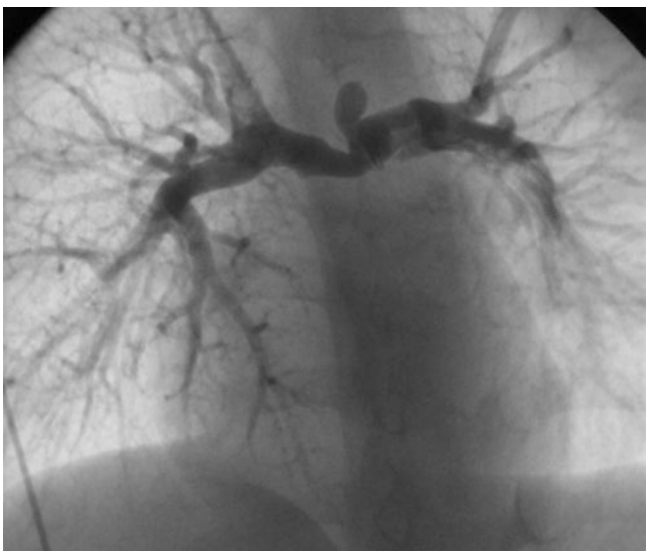


**Fig. 35.9** Patient 1: During the same catheterization, injection in the SVC with 5F NIH catheter shows bilateral pulmonary arteries stenosis

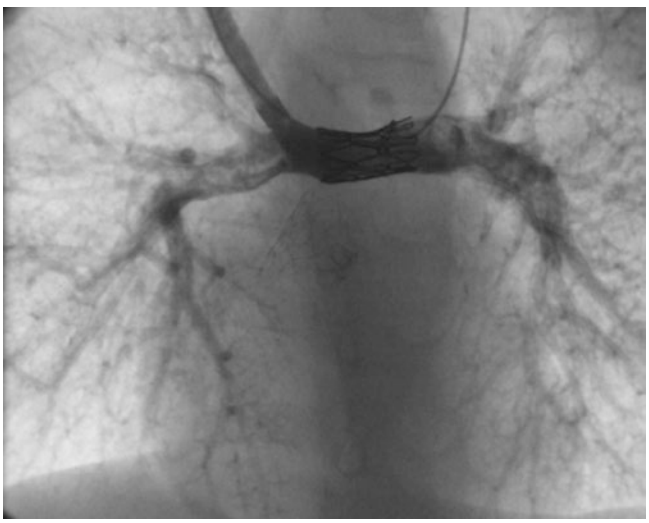


**Fig. 35.10** Patient 1: Due to the presence of stenosis at the bifurcation, pulmonary artery stenting is performed using a 16 mm CP stent mounted on a 12 mm BiB balloon. The stiff guidewire is stabilized in the left SVC

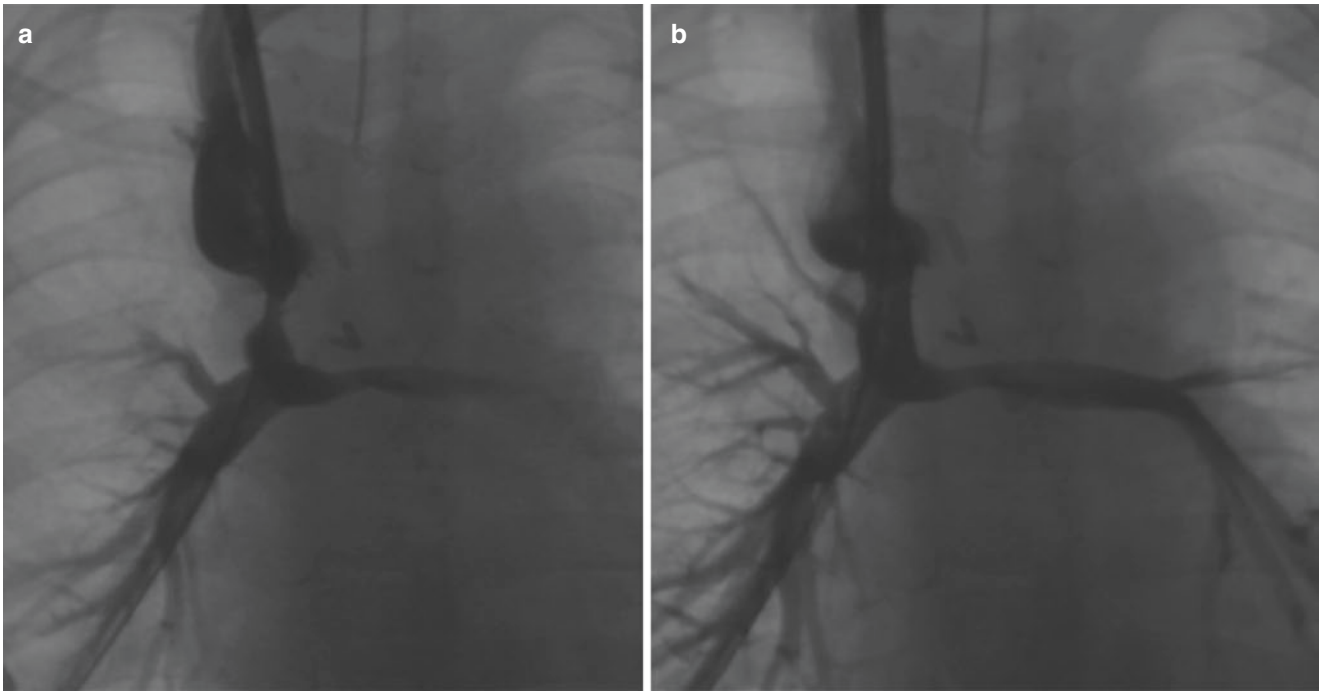



**Fig. 35.11** Patient 4: Hypoplastic left heart syndrome. The patient underwent Norwood 1 at 20 days and PCPC at 7 months. Catheterization, performed at the age of 20 months, shows a severe stenosis of SVC (**a**) that is treated with an 16 mm IntraStent mounted on 8 mm Cristal Balloon (**b**)

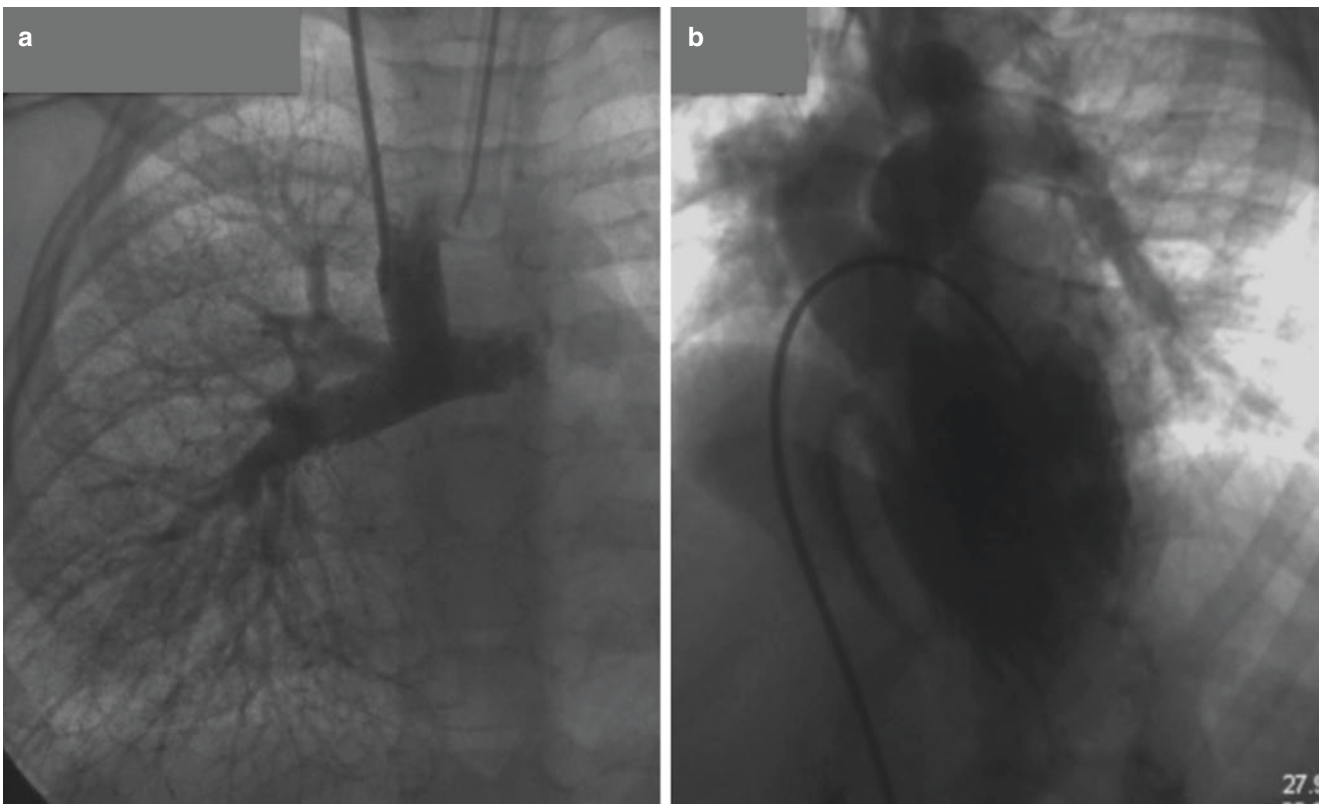



**Fig. 35.12** Patient 5: Tricuspid atresia. This patient underwent PCPC at the age of 5 months. During catheterization, performed at 5 years, injection in SVC shows an evident washout of the left pulmonary artery (**a**). Ventricular angiography put in evidence a preferential perfusion of the left pulmonary artery from the native outflow (**b**)

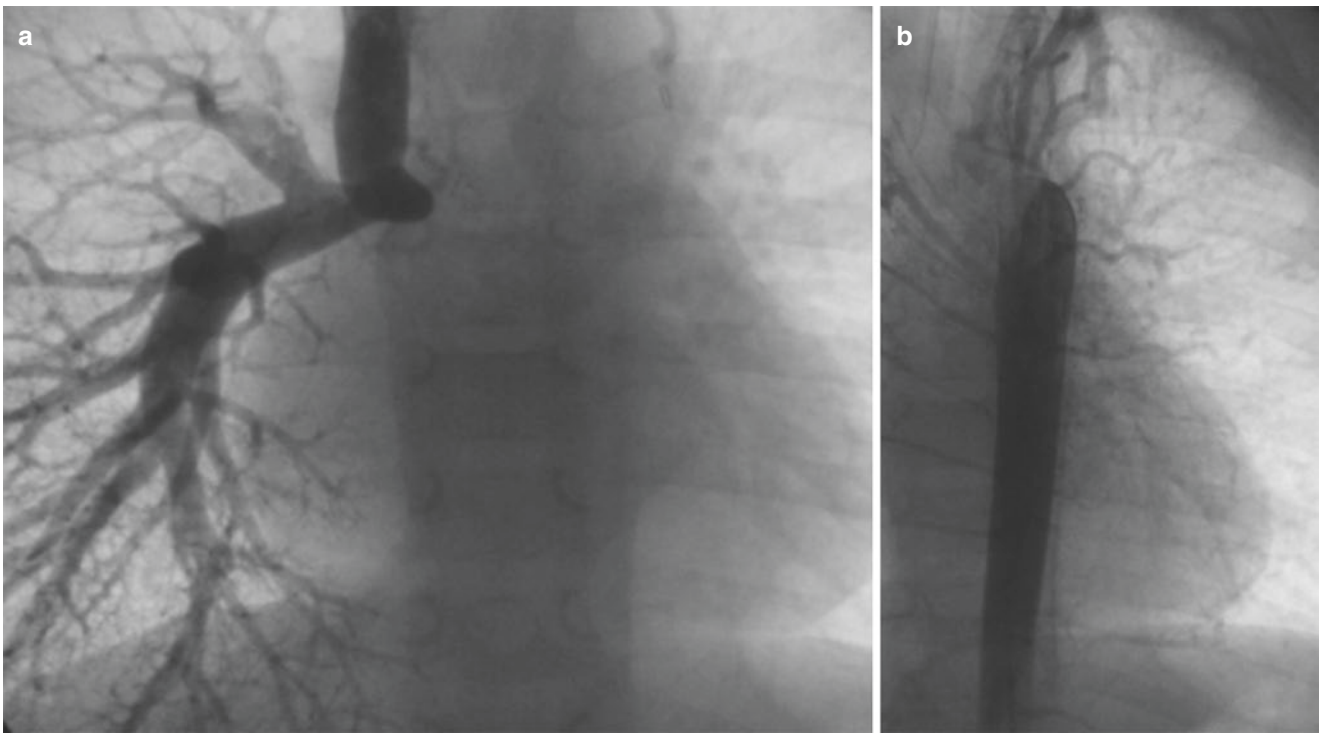



**Fig. 35.13** Patient 6: Hypoplastic left heart syndrome. This patient underwent hybrid treatment at the age of 5 days and PCPC associated with plasty of the left pulmonary artery at 1 year. Catheterization performed after PCPC, at 4 years, put in evidence the complete occlusion of the left pulmonary artery (**a**). After percutaneous recanalization (balloon and thrombus aspiration), the vessel is stented with 2 19 mm Genesis XD mounted on a 12 mm Tyshak balloon. Aortography shows the presence of several, bilateral aortopulmonary collaterals (**b**)

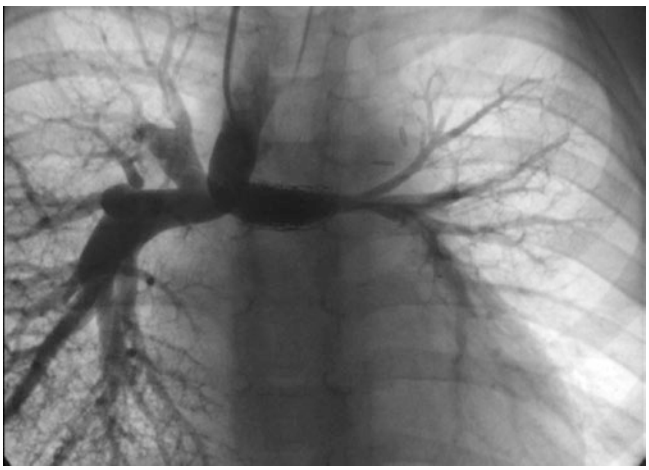


**Fig. 35.14** Patient 6: The angiography performed 7 months after the pulmonary recanalization shows patent vessels without intrastent restenosis

**Video 1** The video shows the selective angiography in the BT shunt in patient 3. We can evaluate the anatomy of the pulmonary arteries, the distal anastomosis between the shunt and the right pulmonary artery. The origin of the BT is not evaluable; for this reason in some case, a nonselective injection in the subclavian artery is needed (Fig. 35.6) (MOV 3574 kb)

**Video 2** The video shows the angiography in the ventricle of patient 2. We can see an abundant antegrade pulmonary blood flow through the native outflow (MOV 4946 kb)

**Video 3** The video shows the angiography in the SVC of patient 1 6 months after pulmonary stenting. The stent is patent and pulmonary arteries and left SVC have good size (MOV 7778 kb)

**Video 4** The video shows the angiography in the SVC of patient 4. We can see the multiple venous collaterals above the stenosis of the SVC (MOV 1863 kb)

**Video 5** The video shows the angiography in the SVC of a patients with pulmonary atresia with intact ventricular septum at time of pre-TCPC. The patient underwent BT shunt at 6 days and PCPC at 5 months. We can see good size of SVC, pulmonary arteries and good peripheral pulmonary vascolarization; in the levophase, we can see the normal pulmonary veins return and normal aorta anatomy (MOV 7826 kb)

# Imaging and Transcatheter Treatments in PA/VSD/MAPCAs

# 36

D. Porras

## 36.1 Introduction

Angiography and transcatheter interventions play an integral role in the management of tetralogy of Fallot with pulmonary atresia and major aortopulmonary collaterals (TOF/PA/MAPCAs). The role of angiography starts with a complete diagnostic investigation that delineates the anatomy of each source of pulmonary blood flow, as well as the vascular health of each segment of the lung. Transcatheter interventions are also used to eliminate unnecessary or potentially harmful sources of pulmonary blood flow, to maximize the health of the distal vasculature, and to optimize the anatomy after surgical repair has been completed. In addition, the majority of these patients will have a right ventricle-to-pulmonary artery conduit as part of their surgical repair, and this will require transcatheter interventions to treat conduit dysfunction in most patients. In this chapter, we will focus on the role of cardiac catheterization in the diagnostic evaluation of TOF/PA/MAPCAs and the role of transcatheter interventions as part of an integrated strategy to manage this complicated disease.

## 36.2 The Role of Cardiac Catheterization and Angiography in the Diagnostic Evaluation of TOF/PA/MAPCAs

The main goal of the management of a patient with TOF/PA/MAPCAs is to be able to perform a full repair that results in cardiopulmonary physiology that is as close to normal as possible. This, in general terms, is defined as a two-ventricular circulation with no residual intracardiac shunts, with antegrade flow to all lung segments, and with normal or near-normal pulmonary artery and right ventricular pressures. Therefore, it is paramount to incorporate as much pulmonary vascular surface area to the repair and to do this in a timely fashion in order to avoid the development of vascular disease (1). In order to appropriately plan the best-suited surgical approach for a particular patient, the goals of the initial diagnostic evaluation are to:

1. *Characterize the sources of pulmonary blood flow*: by definition, patients with TOF/PA/MAPCAs have several sources of pulmonary blood flow. In general, major aortopulmonary collaterals (MAPCAs) are enlarged bronchial arteries that preserve primitive connections to the native pulmonary arteries (PAs) (2). It is important to delineate the anatomy of each of these sources of pulmonary blood flow, including their anatomic relationship to other anatomic structures. It is especially useful for the surgeons if the anatomy of each source can be delineated in relation to other anatomic structures that are easily identifiable in the operating room, like the pulmonary veins, the carina, the mainstem bronchi, etc. Therefore, the description of an APC should include:
   (a) Origin: from what vessel does it originate (descending thoracic aorta, subclavian artery, internal mammary artery, thyrocervical trunk) and from what part of that vessel (anterior/posterior, left/right, proximal/distal). In the case of the aorta, it is important to specify if it originates from the underside of the distal aortic arch, in which case the vessel may be a ductus arteriosus and its patency may respond to prostaglandins. When it is unclear if it is a ductus arteriosus (PDA) or not, but the origin of it is anatomically consistent with a PDA, they are sometimes called "ductlike collaterals."

   Since the majority of true APCs are enlarged bronchial arteries, their origins and trajectories follow

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_36) contains supplementary material, which is available to authorized users.

D. Porras (✉)
Division of Invasive Cardiology, Department of Cardiology, Boston Children's Hospital, Boston, MA, USA
e-mail: diego.porras@cardio.chboston.org



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_36

certain predictable patterns. The majority arise from the descending thoracic aorta, near the carina and the mainstem bronchus ipsilateral to the arch sidedness. However, they can also originate from the brachiocephalic artery, from the subclavian arteries and their branches, and even from the abdominal aorta and its branches (2).

(b) Level of origin: if it originates from the descending thoracic aorta, at what level does it originate and close to what structures (see Fig. 36.1)? For example, one description could be APC that originates from the anterior/rightward aspect of the proximal descending thoracic aorta, immediately under the carina. Describing the level of thoracic vertebra at which the APC arises is of limited utility, since this is not a landmark that can be identified in the operating room.

(c) Trajectory (Fig. 36.1): this includes the detailed description of the course of each collateral, especially in relation to other anatomic structures. For example, APC travels under the right mainstem bronchus, slightly posterior to it, and then makes a loop around the mainstem bronchus near the origin of the upper lobe bronchus, from posterior to anterior, connecting to the native right upper lobe pulmonary artery at this level.

(d) Branching: many APCs give off several branches (Fig. 36.3f). It is important to describe this in detail so that the surgeon can be aware of where these branches originate and whether each one needs to be unifocalized separately.

(e) Segments supplied by each APC, as detailed below and in Figs. 36.1, 36.2, 36.3, 36.4, 36.5, and 36.6).

2. *Establish the presence or absence of central mediastinal pulmonary arteries* (Figs. 36.2, 36.3, and 36.4): central mediastinal PAs, even when diminutive, have enormous growth potential once exposed to higher

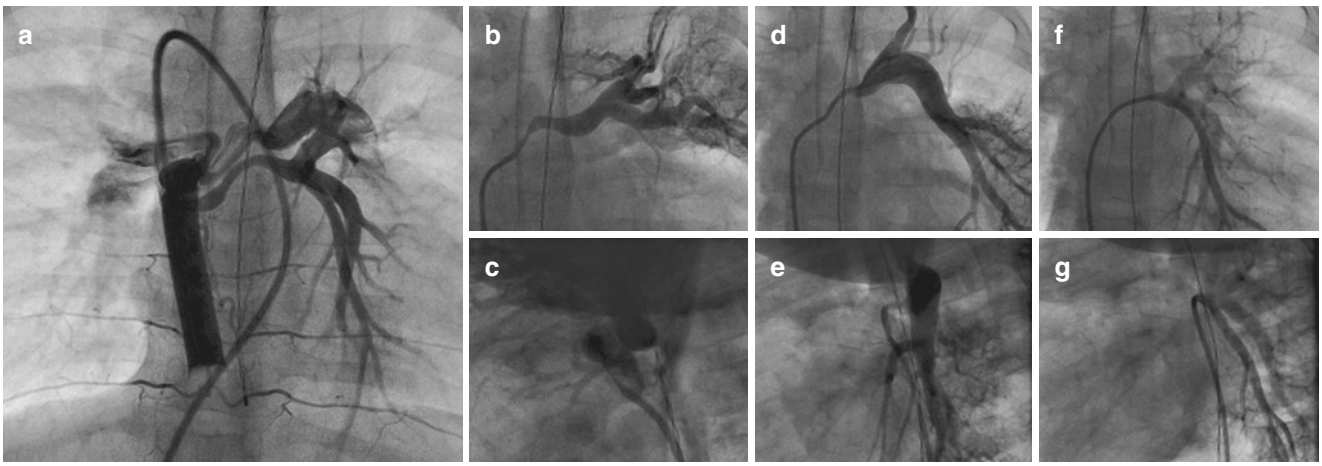



**Fig. 36.1** Defining major aortopulmonary collaterals. (**a**) Balloon occlusion aortogram: a Berman catheter is advanced from the femoral vein, through the RA, RV into the aorta, and around the aortic arch into the descending thoracic aorta, positioned behind the cardiac silhouette. The balloon, which is distal to the angiographic holes in the catheter, is inflated to occlude the distal thoracic aorta, and a power injection of approximately 1 cc/kg is given at a rate of 1 cc/kg/s. The resulting angiogram gives adequate opacification of the collaterals proximal to the balloon and avoids runoff of contrast to undesired parts of the aorta. In this example, three different major collaterals that supply the left lung are seen arising from the thoracic aorta, below the level of the carina. There are also two collaterals going toward the right lung. (**b**–**g**) Detailed delineation of the vessels supplying the left lung: in order to delineate the anatomy of each collateral, including which segments of the left lung each supplies and whether these segments have dual supply, selective injections in straight frontal (**b**, **d**, **f**) and lateral (**c**, **e**, **g**) projections are necessary. (**b**, **c**) show that the collateral that arises from the anterior aspect of the thoracic aorta, below the carina, travels in anterior to the left mainstem bronchus, then over it. The relationship of this vessel to the left upper pulmonary vein can also be seen in this angiogram, since the vein is lightly opacified in the levophase of the injection. The relationship of each collateral to these structures (carina, mainstem bronchus, pulmonary veins), which are easily identifiable for the surgeon, is important in the description of the anatomy of these collaterals. The anatomy of this particular collateral suggests that it connects to the native proximal LPA (it travels from anterior to posterior and right to left as it goes over the left mainstem bronchus). However, it only supplies the anterior segment of the left upper lobe and the superior lingula. (**d**, **e**) show a collateral that arises at the same level of the descending thoracic aorta, but from the lateral aspect. It travels under the left mainstem bronchus and connects to what appears to be part of the native left lower lobe pulmonary, supplying some of the basal segments of the left lower lobe (lateral, medial, and anterior basal left lower lobe segments). It also gives a branch proximally that supplies the apical left upper lobe and one that supplies the inferior lingula. (**f**, **g**) show injection in a collateral that arises lower in the thoracic aorta, from its posterior and leftward aspect, and supplies the posterior and medial basal left lower lobe segmental branches, as well as the superior segment of the left lower lobe. With this kind of detailed analysis, we can account for the supply to each segment of the left lung, and we can also say that all of these three collaterals are the single source of pulmonary blood flow for their corresponding segments. Therefore, unifocalization of all of them is paramount in preserving the maximum amount of lung vasculature possible at the time of repair. A similar delineation is carried out for the collaterals supplying the right lung (not shown)

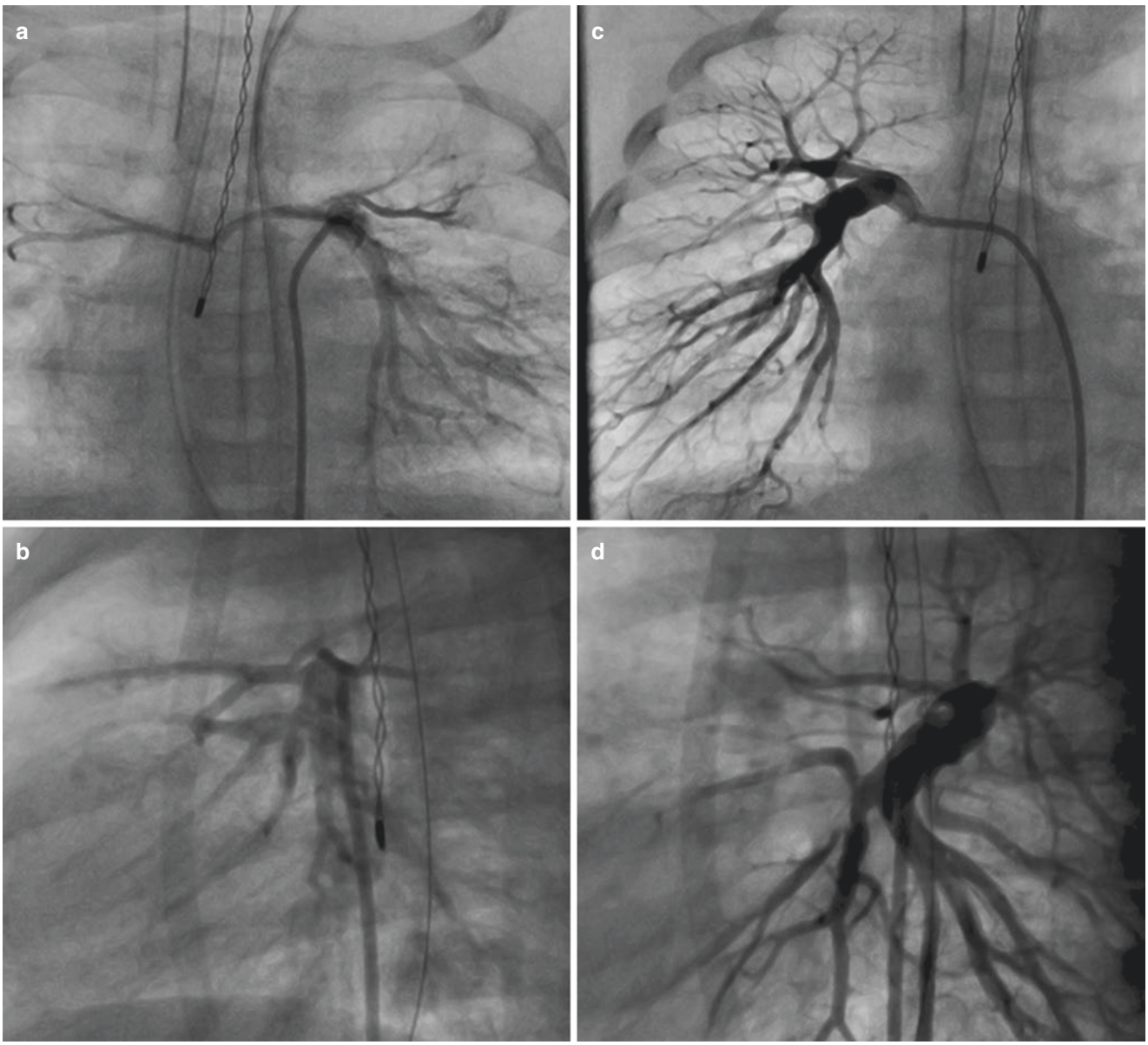



**Fig. 36.2** Determination of the presence of native mediastinal pulmonary arteries. (**a**, **b**) show the frontal and lateral projections of a selective injection into a collateral that supplies the entire left lung vasculature. The native mediastinal LPA is seen filling retrograde and then connecting to the MPA and RPA. These mediastinal branch pulmonary arteries are in continuity and are quite small, measuring approximately 1.5 mm in diameter. It is also evident in (**a**) that the mediastinal RPA only supplies a few segments of the right lung, at least under the present conditions. Further exploration is necessary to determine the sources of pulmonary blood flow to the rest of the right lung. (**c**, **d**) show a selective injection into a collateral that arises from the rightward aspect of the descending thoracic aorta, under the carina. This collateral supplies flow to the majority of the right lung vasculature. Therefore, in this case, this vessel must be unifocalized to the native branch pulmonary arteries, in order to preserve flow to the majority of the right lung

pressure and flow. Therefore, whenever possible, it is important to incorporate them into the repair as early as possible. In many cases, the central mediastinal PAs may not be readily identifiable, and certain techniques are required to establish their presence. These include selective injections into APCs (Figs. 36.1, 36.2, and 36.3) and pulmonary venous wedge injections (Figs. 36.4 and 36.6). It is also important to delineate the anatomy of the central mediastinal PAs if they are present, including their size, presence of discrete stenoses, and/or atretic segments (continuity vs. lack of continuity) (Figs. 36.4 and 36.6).

3. *Characterize how each segment of each lung is supplied and which segments have single* vs. *dual blood supply* (Fig. 36.5): each segment of each lung should be accounted for. Some segments may not have direct flow at

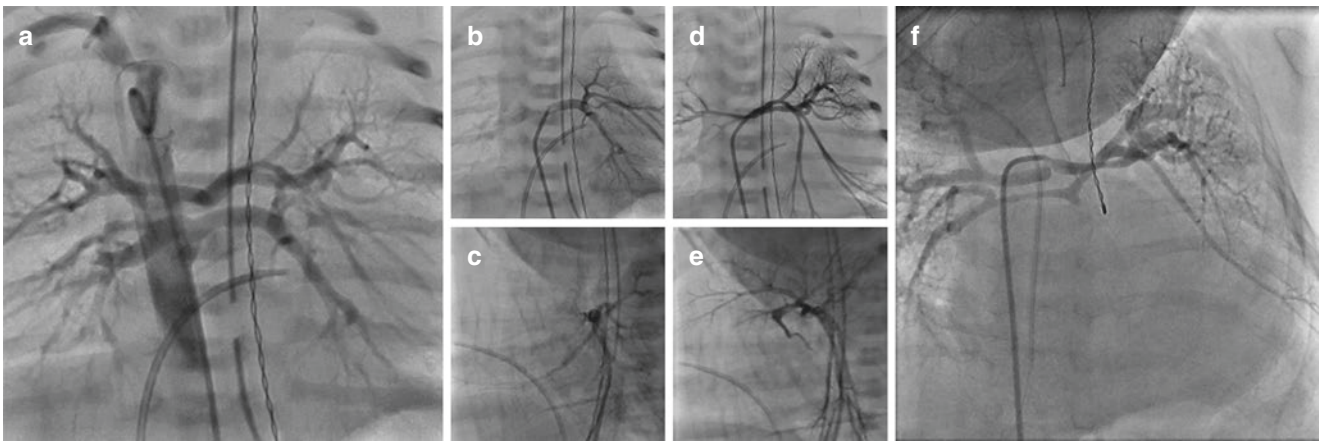



**Fig. 36.3** How do you define native mediastinal branch pulmonary arteries? (**a**) AP projection of a descending thoracic aortogram showing at least three vessels that could potentially be the native mediastinal branch pulmonary arties. (**b**, **c**) show selective injection in the lowest collateral seen in (**a**). The lateral projection (**c**) shows that the vessel that goes leftward stays posterior and inferior to the left mainstem bronchus. This vessel supplies the superior, lateral, and medial segments of the left lower lobe, but is not connected to the central mediastinal pulmonary arteries. (**d**, **e**) show injection into the collateral that arises from the anterior aspect of the descending thoracic aorta, immediately below the carina. This vessel provides flow to the rest of the left lower lobe, the lingula, and the entire left upper lobe. On the lateral, it can be seen that it also connects to what may be a mediastinal LPA, because it is seen traveling anteriorly over the left mainstem bronchus and going across the midline to the mediastinal RPA. This is confirmed in (**f**), with cranial angulation. This angulation makes it clear that this collateral supplies the mediastinal LPA which is in continuity with the MPA and the mediastinal RPA. The retrograde filling of the LPA to the MPA and then toward the RPA forms the classic "seagull" appearance in the cranial angulation, which is characteristic of mediastinal pulmonary arteries that are in continuity. In this particular case, the same collateral bifurcates early into a branch that goes rightward and is the sole supply to the majority of the right upper lobe, without connecting directly with the native RPA

the time of the catheterization and will therefore not fill from APC injections or aortagrams because the APCs supplying them have become atretic. Pulmonary venous wedge injections are useful in identifying these segments, by forcefully filling them with contrast in retrograde fashion (Fig. 36.6). Once identified, their anatomy, especially in relation to other pulmonary arteries and APCs, should be clearly delineated since they will need to be found and unifocalized in future surgeries. Once each segment has been identified, it is important to determine how each segment is supplied:

(a) Single supply: only supplied by a single APC and does not connect in any way to the rest of the native pulmonary arterial tree. Once the APC feeding it is interrupted, no other blood flow will go to this segment unless it is surgically unifocalized.

(b) Dual supply: these are segments that are supplied by the central mediastinal PAs, which at the time of the catheterization are supplied by other collaterals. In addition, these segments are supplied by one or more separate collaterals, meaning that once the collaterals feeding the central mediastinal PAs are interrupted and the central mediastinal PAs are given a new source of blood flow (RV-PA conduit or shunt), these segments will continue to receive flow from other APCs. This flow will, therefore, be redundant and competitive, which can lead to hypertension and lack of growth of the proximal vessel. Therefore, sources of dual supply should be eliminated before the surgery.

(c) Potential dual supply: these are segments that are supplied by native pulmonary arteries, which are in turn supplied by a single MAPCA. This means that once the central mediastinal PAs are given a new source of pulmonary blood flow (shunt or RV-PA conduit), the MAPCA, if not divided, will provide dual supply, which can be detrimental. This MAPCA cannot be interrupted before surgery, because at that time they are the only source of pulmonary blood flow to those segments. Therefore, they should be ligated during surgery or, if not possible, then interrupted using transcatheter techniques shortly after surgery.

4. *Delineate the anatomy of each lobar, segmental, and subsegmental pulmonary artery branch, including the presence or absence of discrete stenoses in the peripheral branches*: some of these vessels can be highly abnormal and can have discrete stenoses that will need to be addressed surgically or using transcatheter techniques in the future. Knowing this in advance will help guide decisions related to the type of surgical approach taken, as well as decisions regarding closure of the ventricular septal defect.
5. *Characterize the health of the vascular bed for each segment of the lung*: because each segment may be supplied

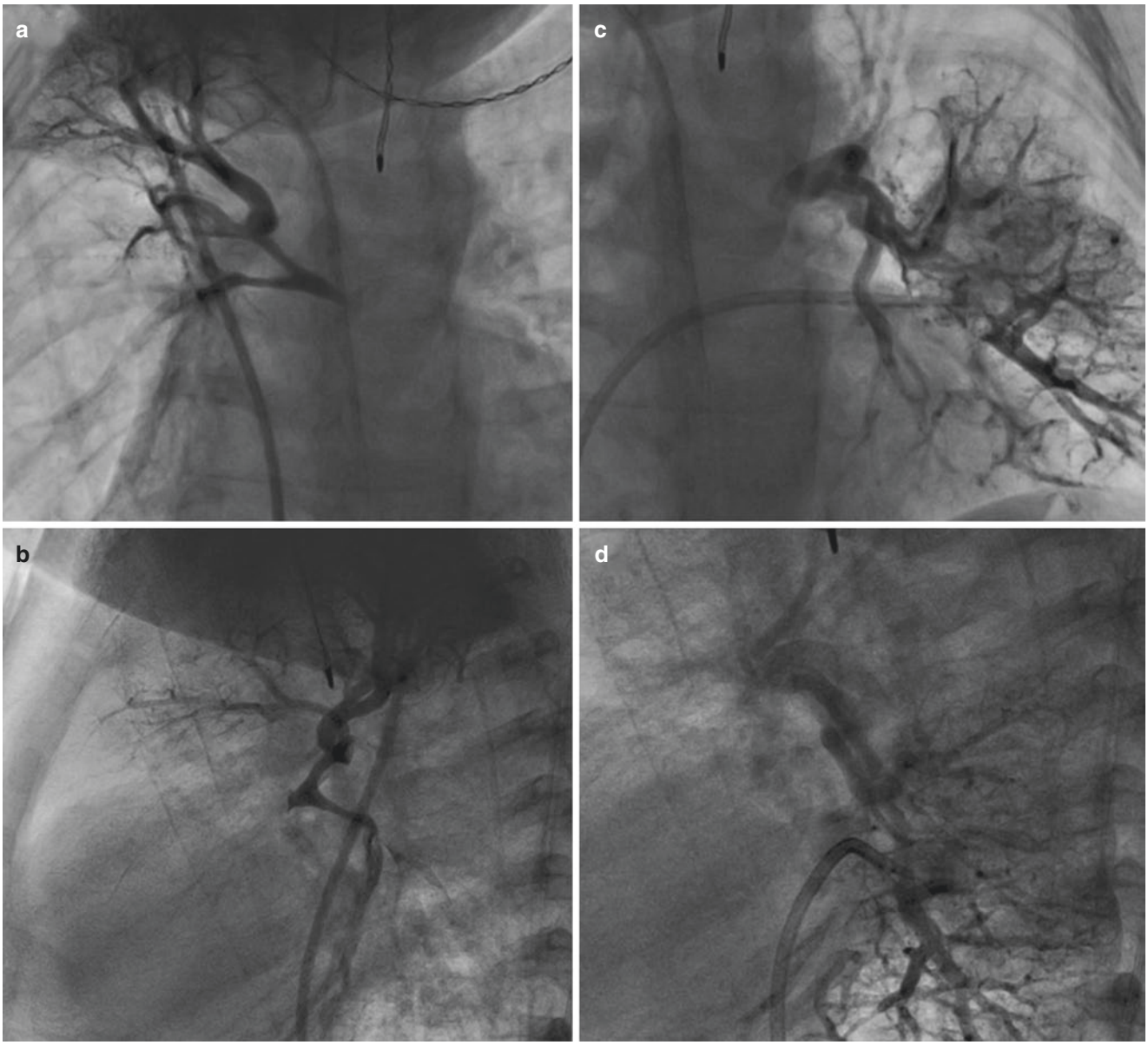



**Fig. 36.4** Present, but discontinuous native mediastinal pulmonary arteries. (**a**, **b**) show the frontal and lateral projections of a pulmonary venous wedge injection in the right upper lobe. This technique consists of advancing a wedge catheter into the distal pulmonary vein branches in retrograde fashion. Once the catheter is wedged, an injection of approximately 3 cc of contrast layered with approximately 7 cc of heparinized saline solution behind it is injected. This forces the contrast retrograde up the pulmonary veins, to the capillaries, to arterioles, and finally to the pulmonary artery. In this example, the right upper pulmonary artery is seen filling and connecting to the native mediastinal RPA which connects to the rest of the right pulmonary artery branches and also fills the proximal native mediastinal RPA to a diminutive MPA and a miniscule LPA that ends blindly almost immediately. When a similar injection is carried out in the left lower lobe (shown in **c** and **d**), the native LPA is seen filling retrograde, traversing over the left mainstem bronchus, and then ending blindly. This patient, therefore, has mediastinal pulmonary arteries, but they are not in continuity because the native mediastinal LPA is interrupted between its proximal portion, just after the takeoff from the MPA, and the hilum of the left lung

by different vessels under different conditions, the health of each segment can be highly variable. Ideally, the health of each segment would be determined by understanding the resistance within each segment. Practically, this is not possible, because in order to know the resistance, one would have to know not only the pressure drop across the segment (which can often be determined) but also the exact amount of flow going through the segment (which is impossible to determine using currently available techniques). Therefore, this is usually a very subjective assessment, and it is based on the angiographic appearance of the vascular bed, the subjective assessment of

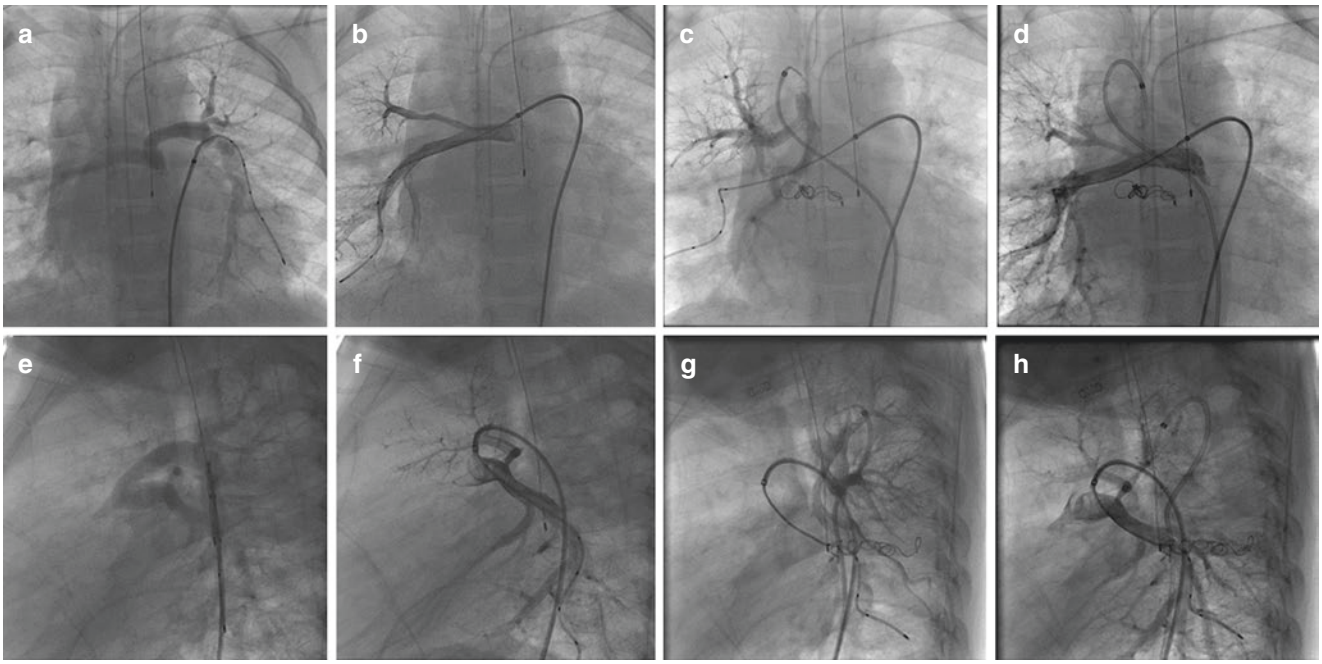



**Fig. 36.5** Delineation of dual vs. single supply. Top panels (**a–d**) show frontal views of selective angiograms. Bottom panels (**e–h**) show the corresponding lateral view for each injection. (**a**, **e**) Collateral from the leftward aspect of the descending thoracic aorta connects to the native left lower lobe pulmonary artery, which fills the mediastinal pulmonary arteries in retrograde fashion. On the lateral projection (**e**), it is clear that the vessels that cross the midline are the mediastinal PAs (LPA, MPA, RPA) because they travel anteriorly and form the classic seagull appearance. (**b**, **f**) The long sheath has been advanced to the mediastinal RPA, and injection shows that this vessel supplies the anterior and apical segments of the right upper lobe, the medial segment of the right middle lobe, and the medial and anterior segments of the right lower lobe. It may also supply the lateral and posterior segments of the right lower lobe. However, these segments appear to "wash out" with non-contrasted blood, suggesting that they have dual supply from another collateral. (**c**, **g**) Collateral from the right subclavian artery supplies the posterior right upper lobe and gives another branch that supplies the superior segment of the right lower lobe and the posterior and the lateral segments of the right lower lobe. (**d**, **h**) Angiogram in native RPA while the collateral from the right subclavian artery is balloon occluded confirms that the lateral and posterior segments of the right lower lobe are supplied both by the native mediastinal RPA and by the collateral from the subclavian artery, since now these segments opacify fully without washout (compare to **b**, **f**). This is important for surgical planning, because it means that the branch of the collateral from the subclavian that supplies the posterior right upper lobe must be unifocalized to the native RPA because it is its only direct supply. However, the branch that supplies the lateral and posterior segments of the right lower lobe can be ligated

flow through that particular vascular bed, and the pressure in that segmental artery. Some of the angiographic findings consistent with pulmonary hypertension and vascular disease include:

(a) Paucity of distal vessels: described as a tree in winter, as opposed to a tree in summer which would be the appearance of a healthy pulmonary vascular bed.
(b) Tortuosity of the branches.
(c) Pulsatility of distal branches.
(d) Rapid tapering of distal branches: healthy vascular beds are characterized by smooth, subtle tapering of the vessel size as it goes from proximal to distal. Diseased pulmonary vascular beds are characterized by the proximal vessels being somewhat dilated and then rapidly becoming narrow (rapid taper).
(e) High mean pressures in the distal vasculature: this obviously depends on the amount of flow going into that particular vascular bed. However, in general, segments should have mean pressures under 20 mmHg. Segments that have mean pressures above 30 mmHg have a high likelihood of having rapidly progressive vascular disease. However, it is important to keep in mind that this is entirely dependent on the amount of flow. For example, a segment with a mean pressure of 25 mmHg may be quite healthy if it is receiving a very large amount of flow and it is still able to accommodate it without significant hypertension. On the other hand, a very diseased segment that has the same pressure but is receiving a negligible amount of flow would be unsalvageable.

It should also be noted that in recent years, CTA has become widely available and used as the initial preoperative diagnostic modality in these patients. CTA does provide excellent anatomic data and can be used as the only

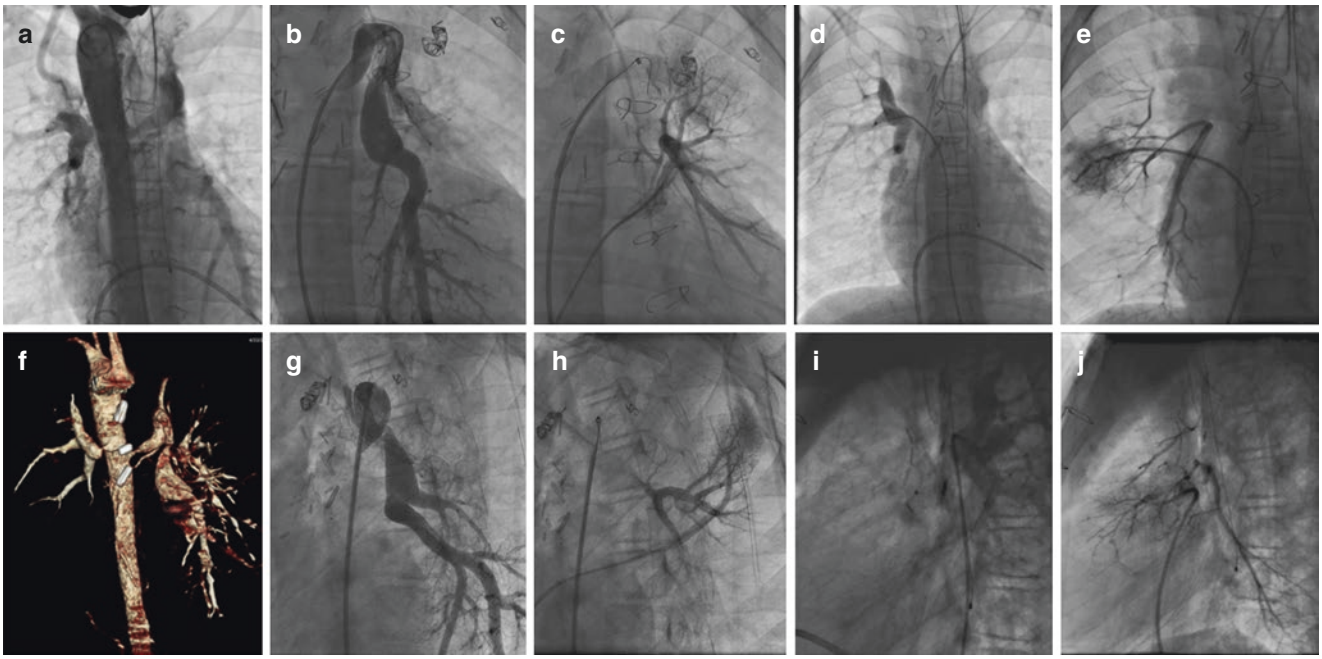



**Fig. 36.6** Pulmonary venous wedge injections to delineate the anatomy of vessels that don't have antegrade flow. (**a**) Anteroposterior projection of aortogram in a patient with TOF/PA/MAPCAs, showing two large APCs from the descending thoracic aorta, one to each lung. (**f**) shows the same information from a three-dimensional reconstruction from a CTA. In both images there appear to be entire segments of both lungs that are not perfused. (**b**, **g**) Injection into the collateral to the left lung shows that it provides flow to medial and posterior segments of the left lower lobe, but no flow to the upper lobe or any anterior segments of the left lung. (**c**, **h**) AP and lateral projections of a pulmonary venous wedge injection performed in a branch of the left upper pulmonary vein. Contrast is seen filling the superior segment of the left lower lobe in retrograde fashion. It also fills the lingular branches, all of which are missing segments in the injection of the collateral to the left lung (**b**, **g**). (**d**, **i**) AP and lateral projections of selective injection into collateral to the right lung showing that this collateral supplies apical and posterior right upper lobe, as well as the superior segment of the right lower lobe. (**e**, **j**) AP and lateral projections of pulmonary venous wedge injection into a branch of the right upper pulmonary vein, filling the right intermediate pulmonary artery, which is connected to part of the right lower and middle lobes as well as anterior right upper lobe. Putting all of this information together, it is evident that the vessels that are not receiving antegrade flow will have to be unifocalized to the vessels fed by the large collaterals in order to have sufficient pulmonary vasculature to be able to complete a full repair in the future

preoperative imaging modality in a subset of patients. Some criteria to select patients that can forego preoperative catheterization include (1):

1. CTA clearly demonstrates all MAPCAs provide dual supply, as defined above.
2. Normal PA arborization.
3. No lung segments receive sole blood supply from a MAPCA that is not otherwise connected to the rest of the pulmonary circulation.
4. Physiologic data is considered unnecessary: in young patients in whom the CTA clearly shows stenotic MAPCAs and pulmonary arteries that appear to be at low pressure based on appearance and clinical presentation, it may be determined that obtaining the physiologic information provided by the catheterization is unnecessary.

Similar level of anatomic definition, in terms of extravascular structures, can be obtained in the catheterization laboratory using 3D CT rotational angiography (see Fig. 36.7). This modality provides excellent definition of the anatomy and allows for creation of 3D models similar to those created with standard CTA. The disadvantage is that it is invasive and that it requires more radiation than a regular CTA. However, in patients undergoing cardiac catheterization for preoperative planning and interventions, it can help reduce the total amount of contrast and radiation and provides excellent anatomic definition. Because the patient is already in the catheterization laboratory, it also allows for further interrogation of areas of the anatomy that may be unclear, in addition to the hemodynamic and interventional advantages described in this chapter.

A reasonable strategy would be to obtain a cardiac CTA on all TOF/PA/MAPCAs as an initial study for preoperative planning and then use this information to decide whether a cardiac catheterization is necessary, based on the criteria described above. This would allow for excellent definition of the anatomy with less radiation and less contrast exposure while potentially avoiding an invasive procedure. Importantly, avoiding instrumentation of the femoral arteries during the

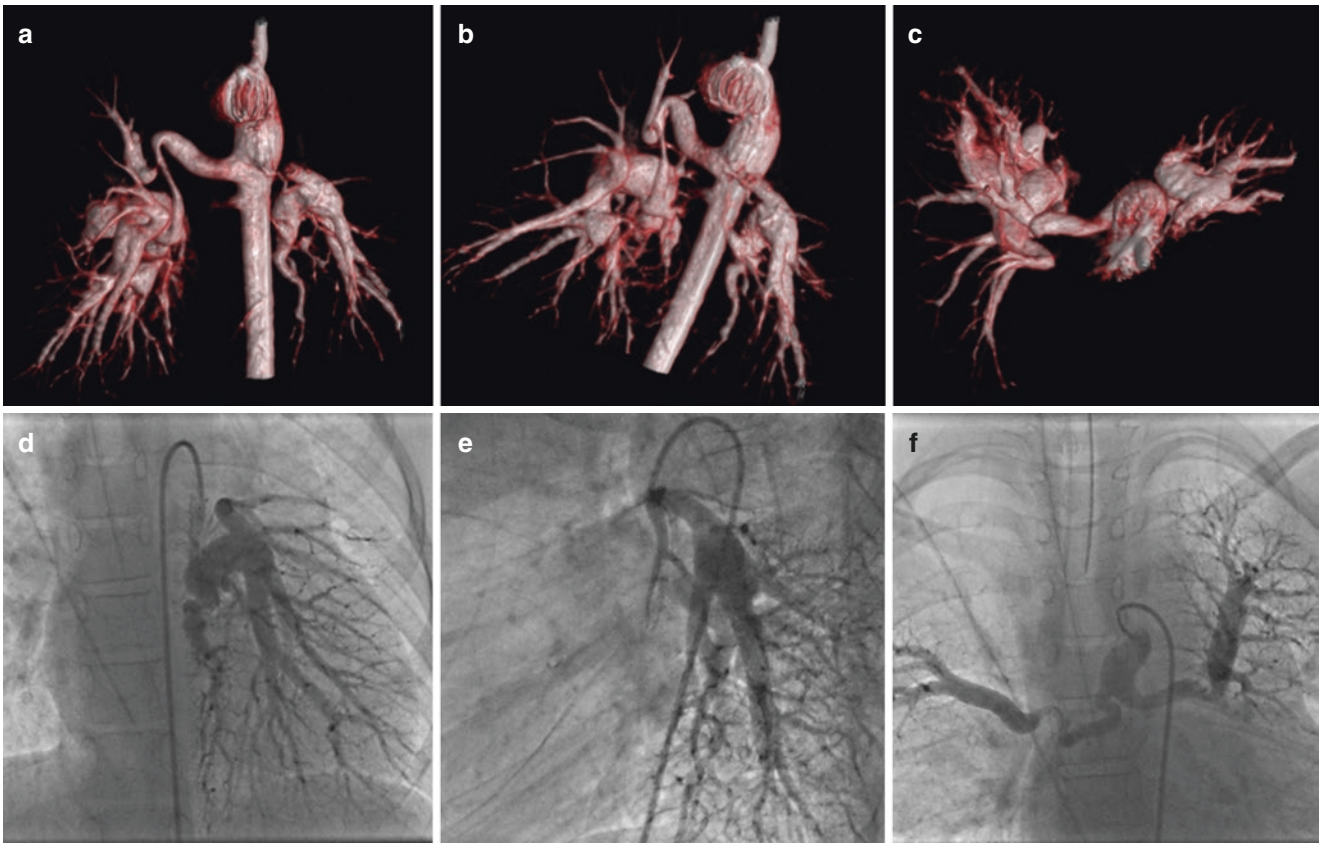



**Fig. 36.7** Occlusion of aortopulmonary collateral providing dual supply to the left lung. (**a**) AP view of an aortogram showing a collateral that connects to the previously unifocalized left lower lobe pulmonary artery. There is a catheter in the left lower lobe PA and a wire goes from this catheter, through the collateral into the descending thoracic aorta, proving that this collateral provides flow to the same vessels supplied antegrade by the LLPA which is now connected to an RV-PA conduit. (**b**) The wire in the LLPA catheter has now been moved to a lower lobe branch, and there is a wire through a long sheath in the descending thoracic aorta, through the collateral into a LLPA branch. Contrast is injected into the collateral which fills the entire left lower lobe and part of the proximal LPA in retrograde fashion. (**c**) A vascular plug is deployed in the collateral (still attached to the delivery cable), and an angiogram confirms adequate position. (**d**) After the device is deployed, repeat angiogram shows complete occlusion of the APC. (**e**) Angiogram after stenting the proximal LPA shows good antegrade flow into the left lower lobe, without any competitive flow ("washout") from the occluded collateral shown in (**a–d**)

neonatal period may reduce the risk of loss of patency of these vessels, in patients in whom multiple catheterizations may be needed throughout a lifetime. Of course, all of this needs to be weighed against the inherent risk related to incomplete diagnostic information. Therefore, the threshold to perform a cardiac catheterization to confirm, or further delineate the anatomy and physiology, should be low.

## 36.3 The Role of Transcatheter Interventions in the Management of Patients with TOF/PA/MAPCAs

Once the anatomy and physiology has been fully delineated and understood, transcatheter interventions can play an important ancillary role in the management of these patients, before and after surgery. Some of the interventions that are often performed in this patient population include:

1. *Balloon and stent angioplasty of APCs* (Fig. 36.8): it is important to remember that APCs are primitive sources of pulmonary blood flow that are programmed to go away. The risk of losing blood flow to a segment supplied solely by a highly stenotic APC is high. Therefore, if these are identified, and surgical incorporation of the vessel is not feasible in a timely manner, consideration should be given to balloon and stent angioplasty of these vessels in order to preserve flow to these segments and, ideally, to augment flow and promote growth of the distal vasculature to increase the chances of surgical success once the segments are brought into the pulmonary circulation. These vessels are highly abnormal and can be resistant to high-pressure balloons. Therefore, cutting balloons are often required to achieve any kind of meaningful result (3). Once the vessel has undergone successful angioplasty, stenting can ensure a longer-lasting result. It is important to avoid primary stenting of these vessels, since they have

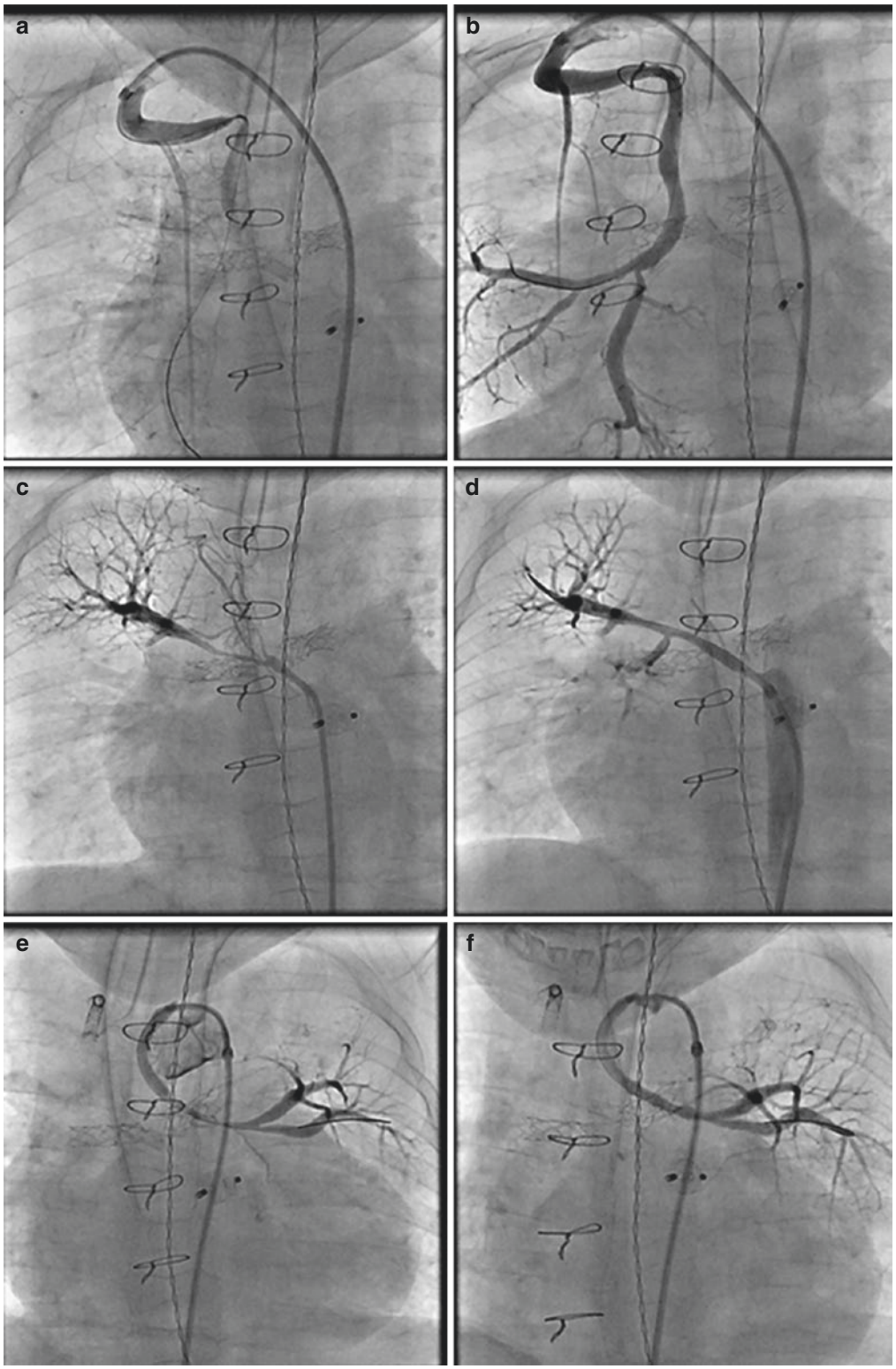



**Fig. 36.8** Stenting of stenotic aortopulmonary collaterals. In some patients with multiple, small, or obstructed aortopulmonary collaterals, it may be necessary to stent the APCs in order to maintain flow to the segments supplied by them and get some growth of the distal vasculature before they can be successfully unifocalized at a later surgery. In this patient, three APCs were the sole supply to segments in both lungs. Panels (**a**, **c**, **e**) show each collateral before stenting, and panels (**b**, **d**, **f**) show the corresponding collateral after stenting

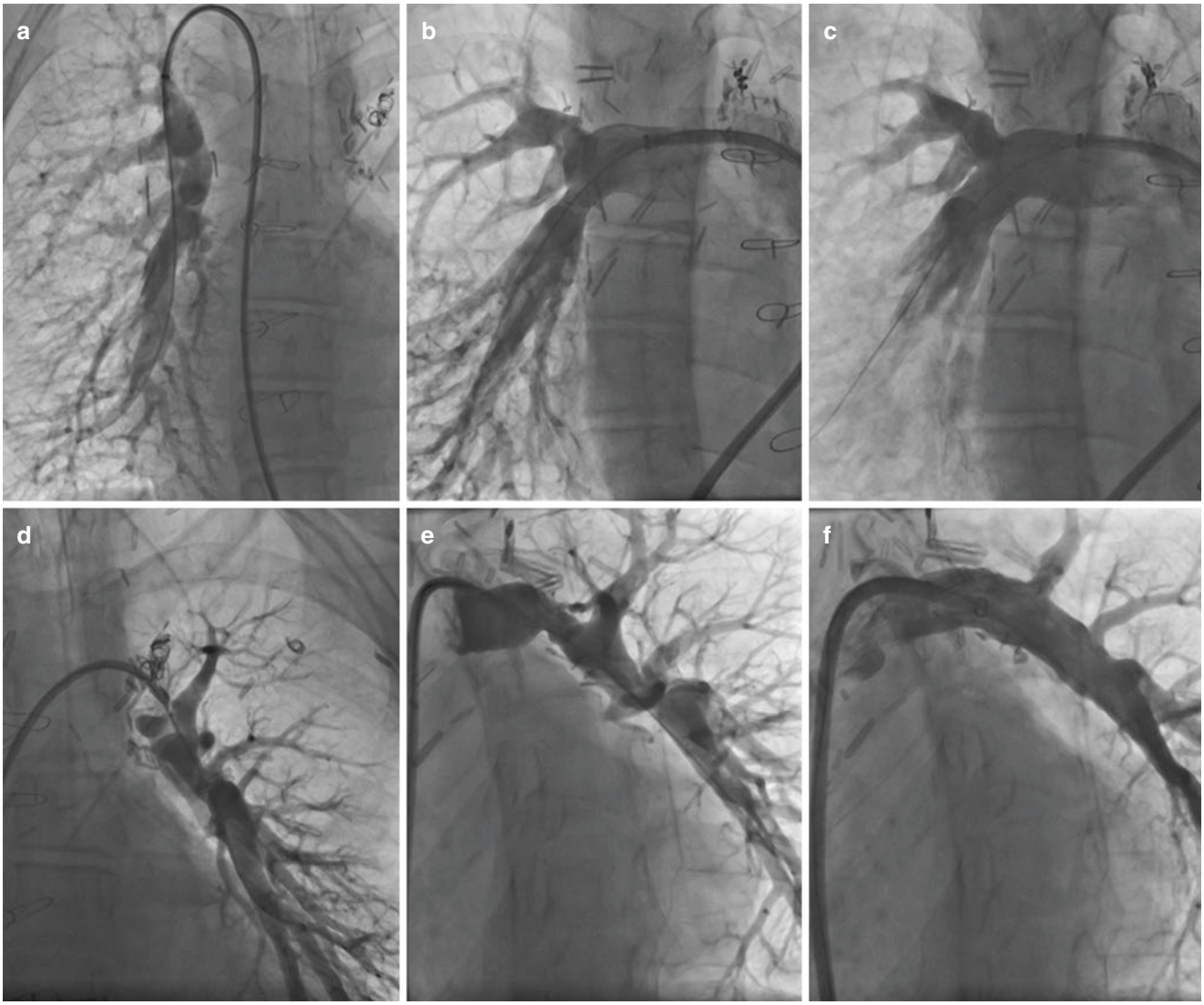



**Fig. 36.9** Balloon and stent angioplasty as a part of the management of patients with repaired TOF/PA/MAPCAs. This patient is the same patient shown in Fig. 36.6. (**a**, **d**) AP projections in the shunted, unifocalized pulmonary arteries to the right (**a**) and left (**d**) lungs. (**b**, **e**) After another surgery consisting of unifocalization of these arteries to an RV-PA conduit with closure of the VSD and takedown of the shunts, angiograms into the RPA (**b**) and LPA (**e**) show good distal vasculature and some areas of discrete stenosis, mainly of the proximal left pulmonary artery, the left lower pulmonary artery, the RULPA, and the right intermediate PA. (**c**, **f**) After balloon and stent angioplasty, the vessels have a much better caliber and no discrete stenoses

highly resistant stenoses, and it is unlikely for the stent to open the stenosis sufficiently without the prior use of a cutting balloon. Finally, it is important to perform the angioplasty in a very gradual manner, to ensure that the distal pressure in the vessel does not rise to unsafe levels that can lead to acute pulmonary hemorrhage and/or pulmonary hypertension in that segment.

2. *Balloon and stent angioplasty of pulmonary artery branches (*Fig. 36.9*)*: this includes unifocalized APCs, as well as reconstructed or native branch PAs, lobar, segmental, and subsegmental branches of the pulmonary arteries. These interventions are reviewed elsewhere, and some examples are given in the figures in this chapter.
3. *Establishment of antegrade flow across a functionally or anatomically atretic pulmonary outflow* (Fig. 36.10): in some cases there may be platelike atresia of the pulmonary valve or functional atresia of the valve (see Fig. 36.10), which prevents opening of the valve because of the combination of sub-PS and pressurized MPA due to APC flow. In these cases it is possible to establish antegrade flow across the pulmonary outflow by balloon and stent angioplasty, either using percutaneous or hybrid transcatheter techniques (4). Once this antegrade flow is established, the segments that have dual blood supply from APCs may be occluded to promote antegrade flow through the native pulmonary arteries. Over time, this will

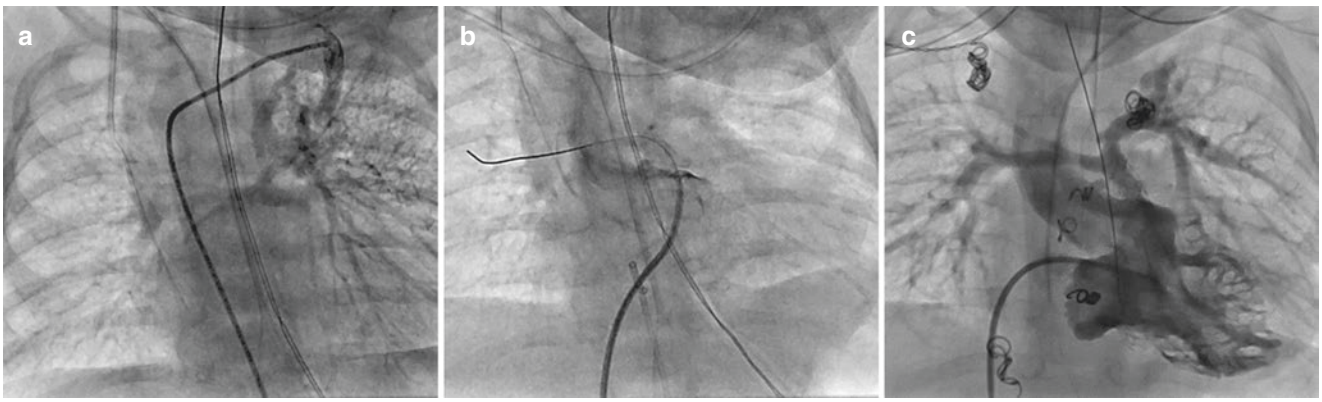



**Fig. 36.10** Opening and stenting of a functionally atretic right ventricular outflow tract followed by coiling of APCs allows for transcatheter establishment of antegrade pulmonary blood flow, with the goal of establishing pulmonary artery growth in preparation for future full surgical repair. (**a**) AP projection of selective angiogram into an APC that supplies the left pulmonary artery branches and the central mediastinal pulmonary arteries in retrograde fashion. (**b**) Even though there was no visible antegrade flow across the pulmonary valve, probing the outflow tract resulted in a wire crossing the valve and going into the RPA. (**c**) The valve and infundibulum were balloon dilated and stented and the APCs were coil occluded. Follow-up angiogram in the RV shows good antegrade flow across the stented pulmonary outflow with continuous mediastinal PAs of reasonable size supplying all segments of both lungs. Over time, the expectation would be that the pulmonary arteries will grow sufficiently to allow a single-stage full repair of tetralogy of Fallot

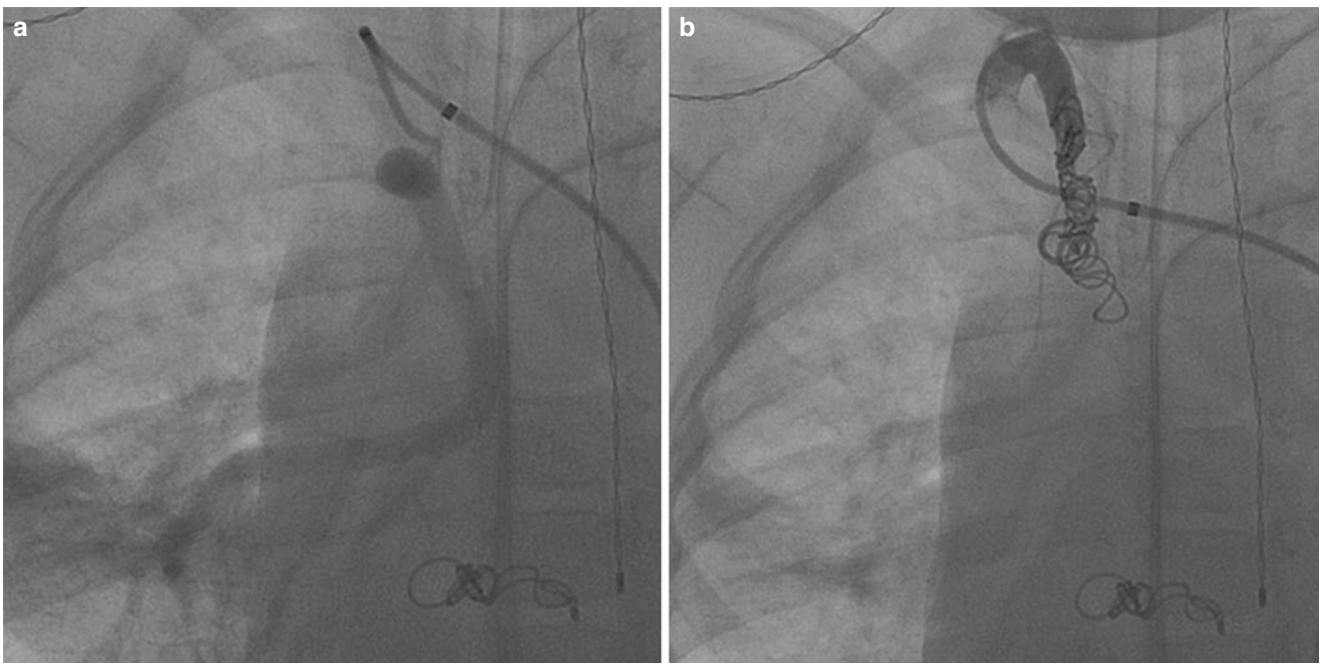



**Fig. 36.11** Coiling of collaterals that supply vasculature that is at high pressure and will not be unifocalized. (**a**) Selective injection into collateral from the right thyrocervical trunk to the right middle lobe. The pressure in this lobe was significantly elevated and the vasculature appeared very hypertensive and damaged. Therefore, the vessel was coil occluded (**b**) to avoid unnecessary inefficient pulmonary blood flow that provides no physiologic benefit to the patient and may be at risk for causing hemoptysis in the future. (**c**, **d**) Small APC providing single supply to small portions of the right lung. The vasculature is quite abnormal and the collateral is too remote and small to be unifocalized, so it was coil embolized (**d**)

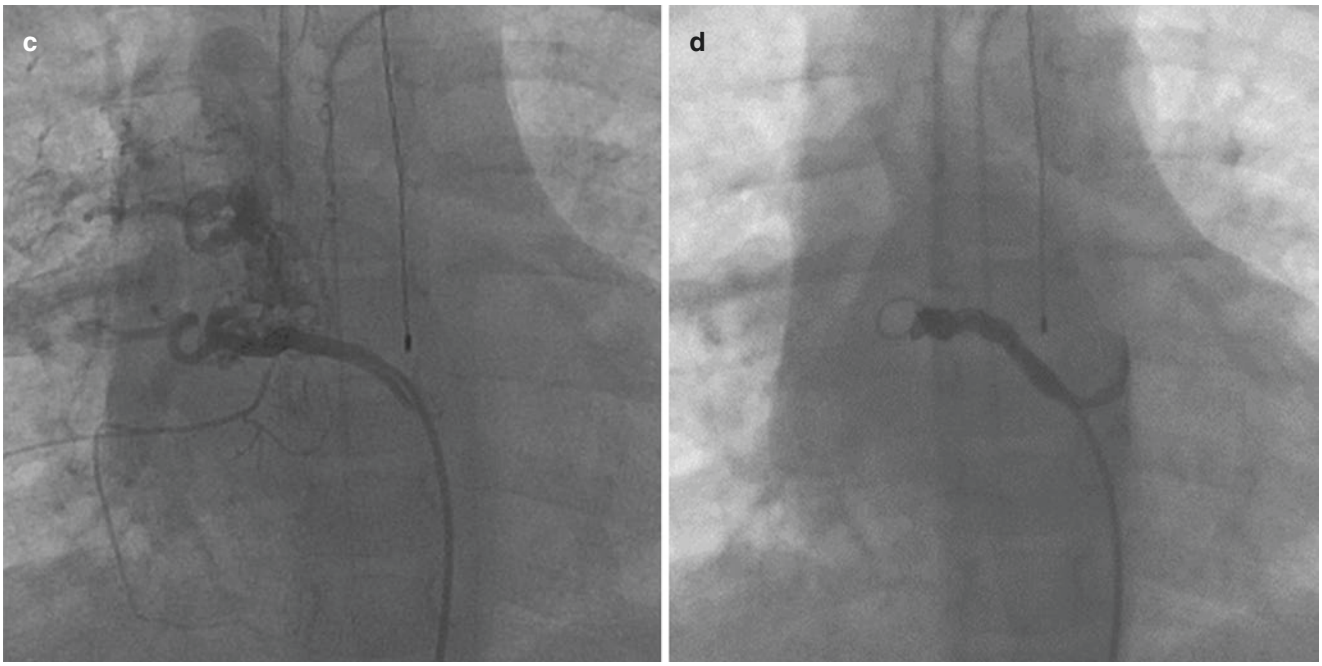



**Fig. 36.11** (continued)

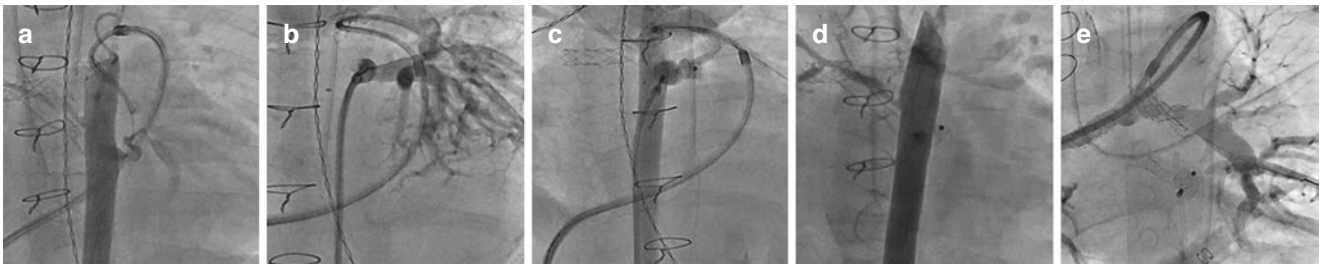



**Fig. 36.12** Three-dimensional CT rotational angiography to delineate the anatomy of major aortopulmonary collaterals. (**a**–**c**) Three-dimensional reconstruction obtained from CT rotational angiography in the catheterization laboratory. This kind of image can be obtained by injecting half-diluted contrast through a pigtail catheter while performing rapid pacing of the ventricle. The rapid pacing decreases the cardiac output during the injection so that the contrast opacifies the area of interest, but is not "lost" to other parts of the circulation, while the image is being obtained. The image is obtained by a rapid >180 degree rotation of the C-arm around the area of interest (see Video 1). The data from this injection and angiography is processed immediately by specialized software to produce a CT angiography and automatic 3D reconstruction of the area of interest. This 3D reconstruction can then be adjusted to produce images like the ones seen on (**a–c**). This model can be manipulated to see different angles and understand the anatomy of the vessels in three dimensions. In this patient, there are two large APCs with severe stenoses along their length. The APC to the right lung gives off two branches. The first branch is highly stenotic and supplies the posterior right upper lobe. The second branch continues caudally and posteriorly and supplies a dilated right lower lobe branch, which connects to the rest of the right lower lobe branches. It also connects, in retrograde fashion, to the right intermediate PA which gives off right middle lobe branches anteriorly (**c**) and then a right upper lobe branch that supplies the apical and anterior segments of the RUL and finally has a blind end anteriorly at the hilum. The CT images can also be used to improve understanding of the relationship of each vessel to other structures like the airway, pulmonary veins, etc. For example, in this case, the native RPA which fills retrograde from the right lower lobe could be seen ending blindly immediately in front of the right mainstem bronchus, at the level of the right upper lobe bronchus takeoff (not shown). Evaluating the anatomy of the APC to the left lung on the 3D model, it is noted that the APC arises at the same level as the larger APC to the right lung but straight anteriorly and is markedly stenotic along its length. In similar fashion to the APC to the right lung, it connects to a dilated branch to the left lower lobe, which in turn connects to the rest of the left lower lobe and, in retrograde fashion, fills the native left lower lobe pulmonary artery, ending blindly as it turns over the left mainstem bronchus, at the hilum. It is important to keep in mind that only the vessels that receive contrast will be seen in the 3D model. In this case, it would be easy to miss the supply to the lingula or the left upper lobe on the 3D model, for example. In (**a–c**), no vessels are seen supplying any of the lingula or the left upper lobe. (**d**, **e**). Follow-up selective injections of the collateral to the left lung seen in **a–c** show that, if contrast is forcefully injected, the lingula is also supplied by this collateral. (**f**) The supply to the left upper lobe was found by injecting a separate collateral that arose from the underside of the arch (and therefore was not filled by the descending thoracic aorta injection). This is important, because this collateral is the single supply to the left upper lobe and, therefore, will need to be found and unifocalized to the native LPA during surgery. The collateral that arises from the underside of the arch (**f**) also bifurcates and gives a branch to part of the right upper lobe that was also supplied by the larger APC to the right lung. Because this collateral represents dual supply, it may be ligated during surgery and does not need to be unifocalized. This case illustrates the importance of accounting for flow to each and every segment of both lungs. It is important to note that some segments may not be receiving flow under normal circumstances, but these vessels can still be found with techniques like selective injections, like in this case, or other techniques illustrated in prior images like pulmonary venous wedge injections and balloon occlusion injections

promote growth and may facilitate a full single-stage surgical repair.

4. *Coil or device embolization of APCs* (Figs. 36.11 and 36.12): as described above, APCs that provide dual supply should be occluded to prevent competitive flow, which may result in pulmonary hypertension in the affected segments, as well as underdevelopment of the native pulmonary arteries supplying those segments.
5. *Device closure of atrial or ventricular septal patch fenestrations and/or residual defects*: these techniques are also reviewed elsewhere and can be an important part of the management of these patients.
6. *Interventions to treat RV-PA conduit dysfunction*: including balloon and stent angioplasty of conduits, as well as transcatheter pulmonary valve replacement: also reviewed in other chapters and will not be addressed here.

**Video 1** Rotational angiography is obtained by injecting half-diluted contrast through a pigtail catheter while performing rapid pacing of the ventricle. The rapid pacing decreases the cardiac output during the injection so that the contrast opacifies the area of interest, but is not "lost" to other parts of the circulation while the image is being obtained. The image is obtained by a rapid >180 degree rotation of the C-arm around the area of interest. The data from this injection and angiography is processed immediately by specialized software to produce a CT angiography and automatic 3D reconstruction of the area of interest. This 3D reconstruction can then be adjusted to produce images like the ones seen in Fig. 36.12a–c and Video 2 (AVI 63498 kb)

**Video 2** Three-dimensional reconstruction from CT rotational angiography as explained in Video 1 and Fig. 36.12. This model has been cropped to include only areas of interest for this particular case. The manipulation of the model shown in this video exemplifies how it can be used to understand the 3D anatomy of the vasculature, including angles that could never be reached with conventional angiography (AVI 1222 kb)

# Stenting of the Right Ventricular Outflow Tract as Initial Palliation for Fallot-Type Lesions

37

Oliver Stumper, Daniel Quandt, and Gemma Penford

## 37.1 Introduction

The initial management of severely cyanosed patients with tetralogy of Fallot-type lesions remains challenging. True neonatal repair of these lesions remains the exception (1–3). The creation of a Blalock-Taussig (BT) shunt is well established but continues to have a high early and late complication rate and mortality (4). Earlier attempts at transcatheter interventions (either balloon pulmonary valvuloplasty or stenting the right ventricular outflow tract) were rather high risk or yielded unpredictable results (5–8). It is only recently that several groups have revisited stenting of the right ventricular outflow tract (9–12) in the initial palliation of symptomatic patients with Fallot-type lesions.

At Birmingham Children's Hospital we have performed close to 100 of such procedures until the end of 2015, and this report summarizes our experience focusing on technical tips and advice.

## 37.2 Patient Selection and Imaging

Tetralogy of Fallot is amongst the most common cyanotic congenital cardiac lesion. Some children may progress to acquired pulmonary atresia within a few weeks after birth. Others remain stable until weighing more than 5 kg in body weight when one-stage complete repair should be the preferred treatment option. Yet, associated cardiac lesions such as a complete AVSD, hypoplastic pulmonary arteries, double outlet right ventricle arrangement, or abnormal systemic or pulmonary venous return or the co-existence of associated syndromes or co-morbidities may make initial palliation preferable, rather than an attempt at early complete repair.

The diagnosis of Fallot is universally made on cardiac ultrasound. CT angio may be useful in patients with suspected coronary artery anomalies or collaterals. If the patient requires palliation prior to complete surgical repair, cardiac ultrasound is very accurate in defining the relative contribution of subvalvar, valvar, and supravalvar obstruction and assessment of the branch pulmonary arteries. The most reliable views to assess these are subcostal images of the RVOT and parasternal short-axis views. Diagnostic cardiac catheterization in the initial diagnosis of Fallot-type lesions is nowadays rare—its mayor impact being the delineation of the peripheral pulmonary arteries, the exclusion of associated collateral arteries, and anomalous coronary distribution.

## 37.3 Catheter Stenting of the RV Outflow Tract

### 37.3.1 Patient Selection

Indications for consideration of RVOT stenting in symptomatic Fallot-type lesions are all cases in whom one-stage complete repair is considered high risk or where there are significant associated cardiac lesions or morbidities which make delay of bypass surgery preferable.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_37) contains supplementary material, which is available to authorized users.

O. Stumper (✉)
The Heart Unit, Birmingham Children's Hospital, Birmingham, UK

Birmingham Children's Hospital, Birmingham, UK
e-mail: oliver.stumper@nhs.net

D. Quandt · G. Penford
The Heart Unit, Birmingham Children's Hospital, Birmingham, UK
e-mail: Daniel.Quandt@kispi.uzh.ch; gemma.penford@doctors.org.uk


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_37

### 37.3.2 Procedural Preparation

Cases should be discussed in a multidisciplinary team meeting. Detailed informed consent has to be obtained. All patients should be cross-matched for blood products, and there has to be thorough briefing of the whole team prior to sending for the patient. Emergency drugs are prepared, and the cardiac surgical and intensive care team should be made aware about the timing of the procedure.

### 37.3.3 Cardiac Catheterization

The standard vascular approach should be from the right femoral vein. This largely facilitates entry into the right ventricular outflow tract using either right Judkins or Cobra preshaped catheters. The right internal jugular venous approach may be beneficial in neonates weighing less than 2 kg due to vessel size (Fig. 37.1). Ideally the patient should be draped to allow for peri-procedure ultrasound scanning from subcostal

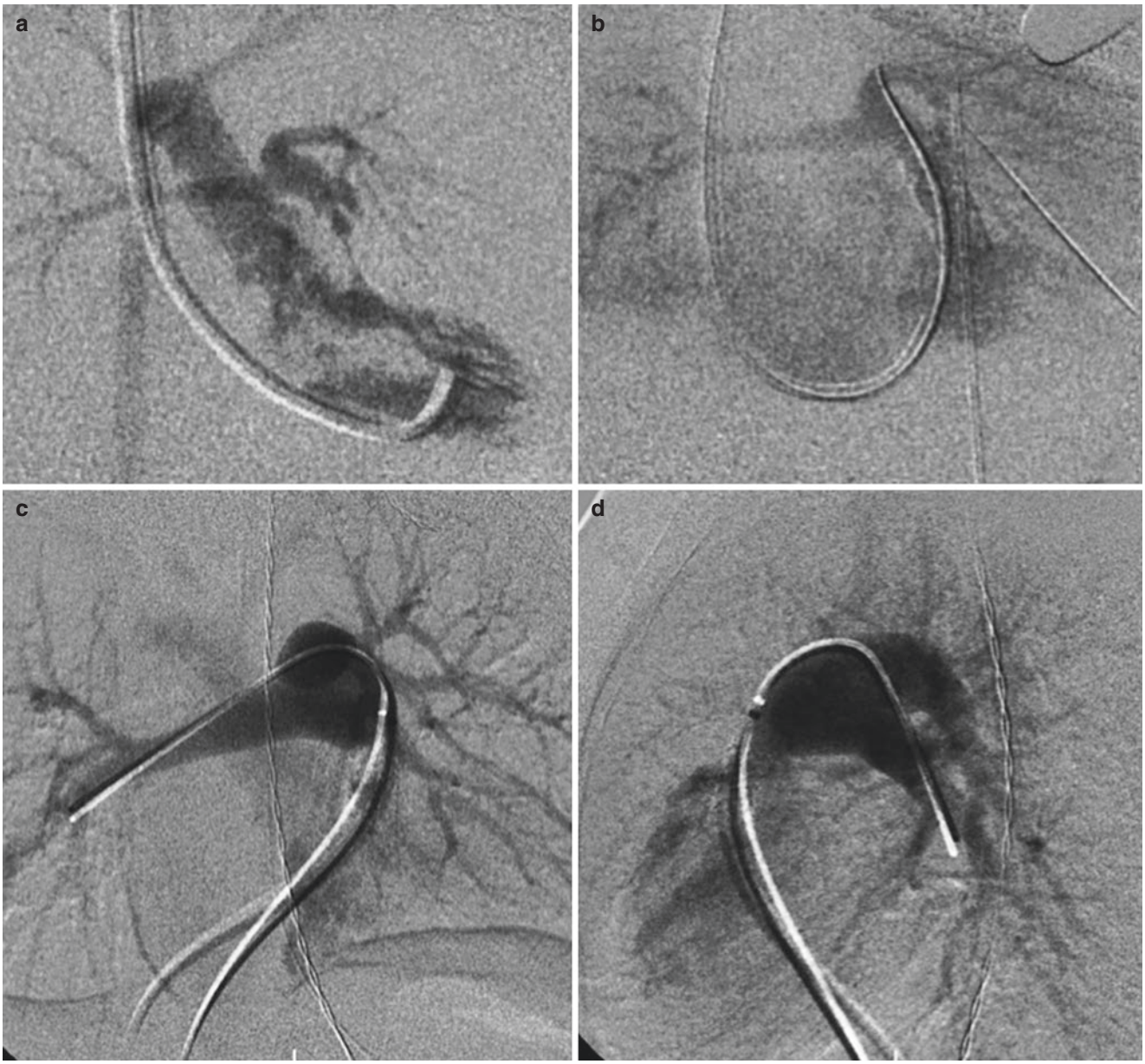



**Fig. 37.1** RVOT stenting in a 1.7 kg premature (28 weeks +5) baby at 3 weeks of age. Digital subtraction angiographic stills. (**a**) From a right internal jugular venous approach, a diagnostic angiogram is performed in 30 RAO + 20 cranial through a 4 French Cobra catheter. Note the very long-segment RVOT obstruction. (**b**) The 4 mm coronary stent was placed through a 4 F introducer sheath advanced to the right ventricle to allow for sidearm test injections—immediate result. (**c**) At 8 months of age (weight 5.3 kg), a further catheter was undertaken performing 30 RAO + cranial and lateral angiograms. Note the very good growth (post stenotic dilatation) of the branch pulmonary arteries. (**d**) Lateral angiogram after further stenting with a 6 mm biliary stent. The child underwent complete repair at 15 months, weighing 7.8 kg

and parasternal projections. A sterile ultrasound probe should be available for the operators to use.

Baseline angiograms are performed in 30 RAO + 20 cranial and lateral projections.

### 37.3.4 Measurements

Ultrasound measurements of the RVOT are generally the most reliable. Angiography is always likely to underestimate required stent length due to foreshortening. Echo measurements should be made to cover the entire length of the RVOT and should ideally be a sum of distances added together to pay tribute to the curved nature of the RV outflow tract (Fig. 37.2). During stenting of the RVOT, an attempt should be made to avoid crossing the pulmonary valve, unless there is significant supravalvar stenosis.

### 37.3.5 Procedure

Following the initial RV angiogram (see above), the catheter is withdrawn to the IVC. Reference angiographic stills are selected and displayed in the room. Repeat measurements are being made from the angiograms.

The appropriate kit is selected and prepared before proceeding any further. In children weighing more than 2.5 kg where only short-term palliation is required, a 5 mm coronary stent is chosen (4.5 or 4 mm in kids weighing less). In those who require medium- to long-term palliation, a peripheral vascular or biliary stent such as the Cook Formula (13), the Omnilink or Genesis stent is chosen—which allow for later over-dilatation.

The stent chosen for implantation dictates the required delivery system. Departmental preference is for either 4 or 5 F Cook Flexor sheaths or for a 6 F short (55 cm) right Judkins Guide catheter (Cordis) (Fig. 37.3). All is prepared and is introduced over a 0.035″ wire placed in the SVC.

A 4/5 F right Judkins or Cobra catheter is inserted into the delivery catheter and is then used to enter the pulmonary arteries directly under pressure monitoring. Position is confirmed on hand test angio. The appropriate wire for the stent system is placed in the distal (right) pulmonary artery, and the delivery sheath or guide catheter is advanced from the IVC to the branch PA over the diagnostic catheter and delivery guidewire. Next the diagnostic catheter is removed over the wire and the stent is delivered. Angio test injections are used to confirm stent position prior to (hand) inflation of the balloon. The lateral *X*-ray projection is particularly useful for placing stents where it was decided to spare the pulmonary valve (Fig. 37.4). Hand inflation of the balloon for placement of the stent is almost always sufficient and allows for fine positional adjustments during placement. A further balloon inflation across the pulmonary valve should be performed so as to dilate the most commonly stenotic pulmonary valve. Next, the delivery sheath or guide catheter is advanced slightly to oppose the proximal part of the stent, so as to prevent dislodgement of the stent whilst retrieving the deflated (negative suction!) delivery balloon. The delivery sheath/guide catheter is then withdrawn to the IVC, and further detailed cardiac ultrasound study is performed. This focuses on whether the proximal portion of the infundibulum is covered, assessment of ventricular function and exclusion of pericardial effusion.

RVOT stenting has evolved to be a valuable tool in patients with very complex anatomies bordering on

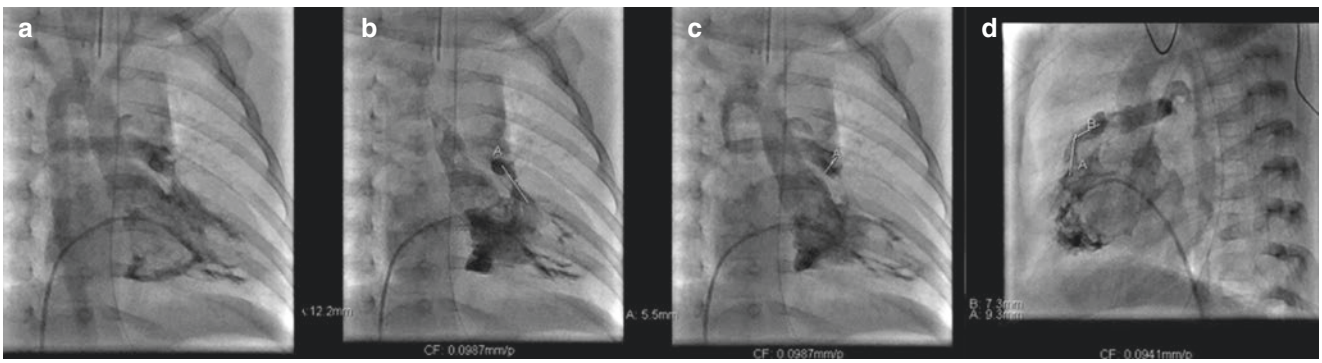



**Fig. 37.2** Still frame RV angiograms in a 3-month-old child with complete AVSD and Fallot with significant co-morbidities [see also attached Video file]. (**a**) RAO + cranial projection shows the anterior deviation of the outlet septum, a reasonably developed pulmonary valve annulus and decent branch pulmonary arteries. (**b**) The length of the RVOT is always underestimated using angiographic views. (**c**) Pulmonary valve and branch PA measurements are reliable. (**d**) In the lateral projection, the most reliable length measurement for the RVOT can be obtained by summation of at least two measurements, so as to allow for the curvature of the outflow tract. Always choose a stent slightly longer than the longest measurement taken—even those on ultrasound

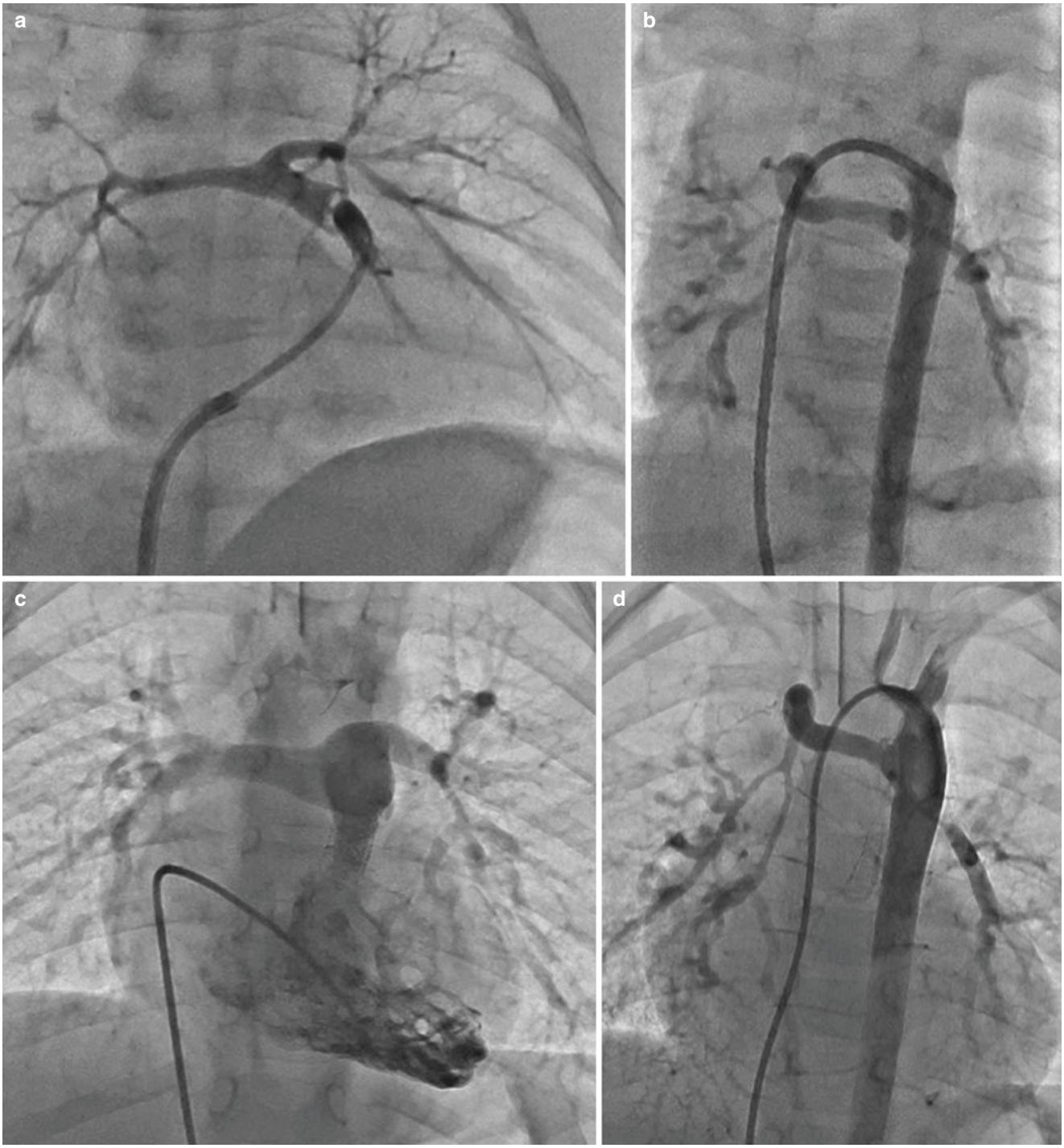



**Fig. 37.3** Serial angiographic still frames in a child with severe Fallot and stenosed MAPCAs from a left arch. (**a**) After the initial angiogram, decision was taken to proceed to RVOT stenting. A 6 F right Judkins Guide catheter was placed in the right atrium, and the distal right ventricular outflow tract was intubated using a 4 F Judkins right catheter. Note the severe pulmonary valve stenosis and the appearance of the peripheral pulmonary arteries suggestive of Alagille syndrome (negative genetics). (**b**) AP angiogram depicting the left aortic arch and the stenosed MAPCAs to both lung fields—attempts at surgical recruitment are not warranted. A 5 mm coronary stent was implanted at 4 weeks of age (weight 3.9 kg) with good effect. (**c**) RV angiogram at 3.5 years of life (14 kg—saturations 84%) after further dilatation of the initial stent at 9 months (7.5 kg) and implantation of a 7 mm Cook Formula stent at 19 months (10 kg). The central PAs have grown very nicely, but there remain bilateral hilar stenoses, which were addressed by balloon angioplasty. (**d**) The repeat arch angiogram documents further progression of the long-segment stenoses within the MAPCAs. At 4.5 years of age, the child is now listed for complete surgical repair and preceding catheter occlusion of the MAPCAs

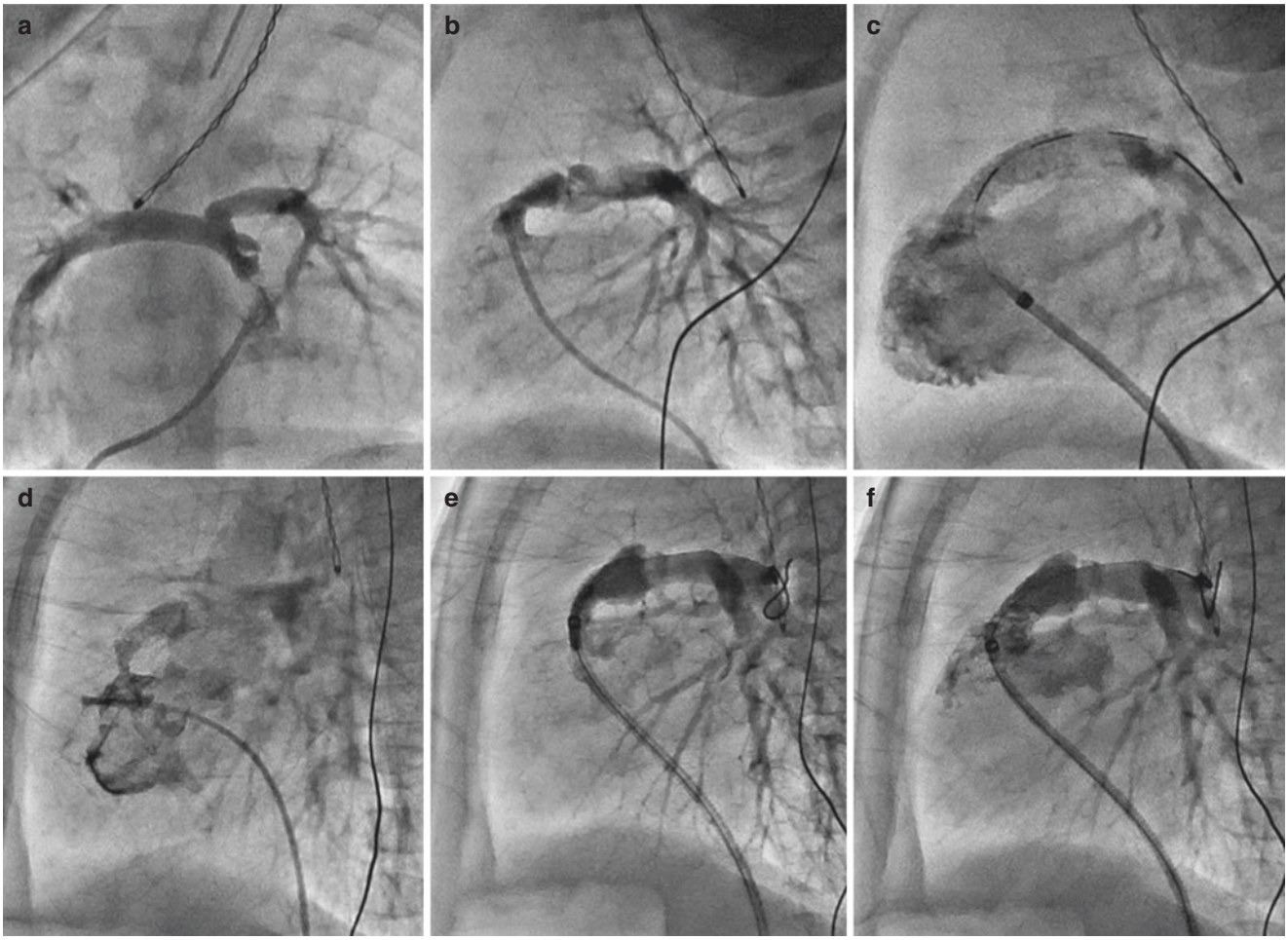



**Fig. 37.4** Sequence of angio stills in a 5-week-old boy (3.4 kg) with double outlet right ventricle (Fallot-type), complete AVSD and bilateral SVC with left SVC draining to an unroofed coronary sinus. (**a**) Four-chamber view documents tight infundibular and valvar pulmonary stenosis—well-developed central pulmonary arteries and peripheral arborization. (**b**) Lateral angiogram showing short infundibulum and significant anterior deviation of the outlet septum. (**c**) RVOT stent placement (5 × 20 mm coronary) via 4 F Flexor over 0.014" Thruway wire just crossing the pulmonary valve annulus. (**d**) Repeat lateral angio 4 months later (5.8 kg) shows proximal obstruction. (**e**) A further stent (6 mm Cook Formula) was placed proximally through a 5 F Flexor under repeat sidearm angio guidance. (**f**) Good final result after placement. Note the growth of the pulmonary valve annulus. Complete repair was performed at 13 months (9 kg)

pulmonary atresia with VSD (Fig. 37.5) and in our opinion is safer and more effective than PDA stenting in cases with duct-dependent pulmonary blood flow, as long as there is even a tiny residual right ventricular outflow tract with some anterograde flow (Fig. 37.6). Further, in our experience, it has become a low-risk effective initial palliation technique in selected univentricular patients with limited anterograde blood flow (Fig. 37.7).

### 37.3.6 Post Procedure Management

All patients receive antibiotics at induction of anaesthesia and two further doses after stent implantation. Prostaglandin infusions, when present, are stopped on placement of the stent. Patients who experience a rise in oxygen saturations of more than 20% are commenced on twice daily diuretics. Patients are kept on intravenous fluid management for at least 12 h and are started on single-agent antiplatelet therapy once oral intake is re-established. If clinically stable and feeding well, patients are discharged home 48 h post procedure with frequent outpatient review.

Re-intervention is considered in cases where further delay to repair is desirable or in those who develop significant recurrent outflow tract obstruction either due to not entirely covering the proximal portion of the outflow tract, tissue ingrowth into the stent or somatic outgrowth.

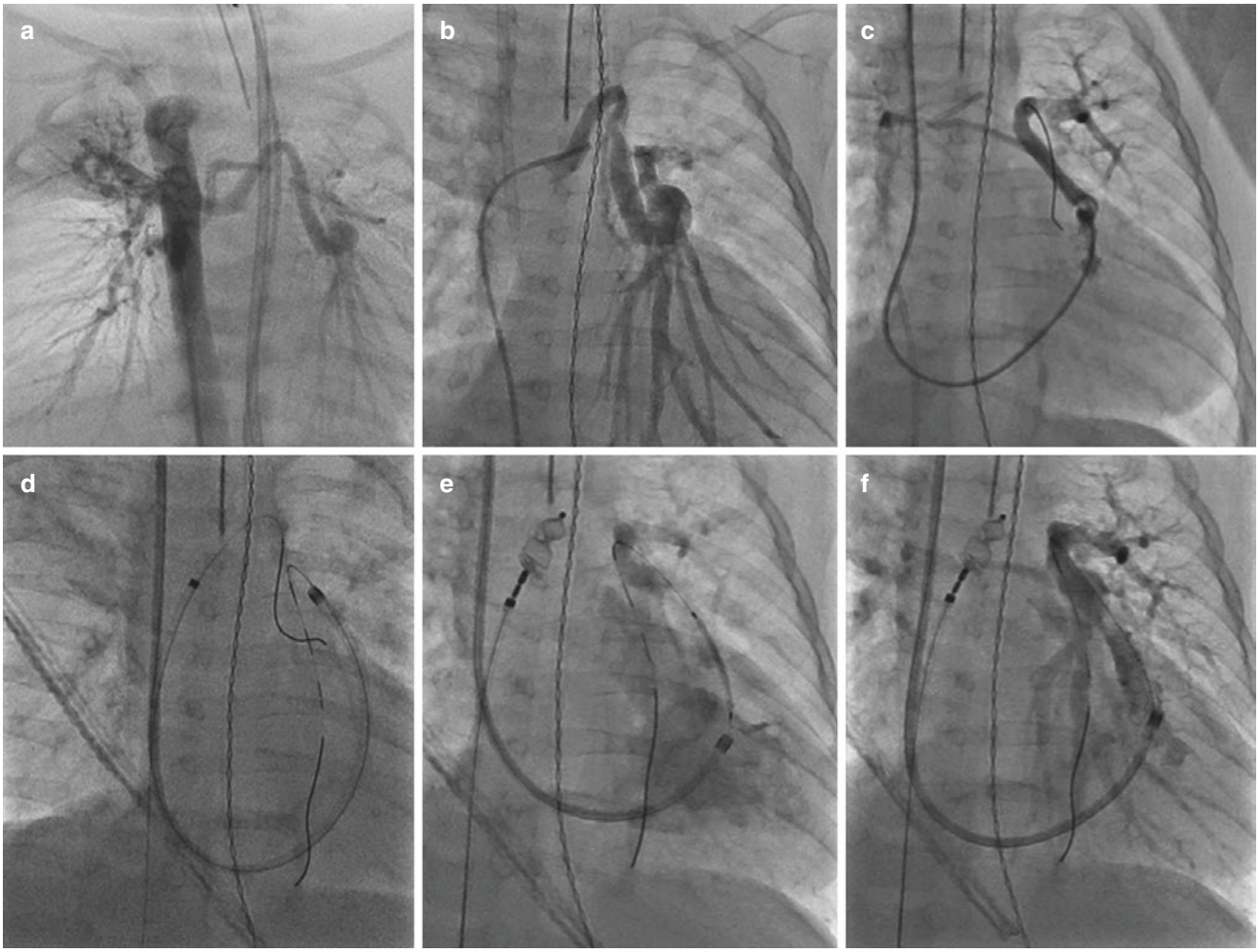



**Fig. 37.5** Still frames of RVOT stenting in a 5-month-old child with extreme tetralogy of Fallot and large MAPCAs to both lung fields. Weight 5.1 kg, on CPAP ventilation due to excessive pulmonary blood flow and airway compression by the MAPCAs. (**a**) Descending aortogram showing numerous collaterals to the right lung and at least two MAPCAs to the left lung. (**b**) Injection into the large MAPCA to the left lung does not show any significant stenosis and good peripheral arborization, suggesting early communication with the native left lung artery system. (**c**) Injection into a severely hypoplastic right ventricular outflow tract demonstrates a native confluent PA system with extreme hypoplasia of the right pulmonary artery. (**d**) During further simultaneous injections, it was confirmed that the left lung in fact had dual supply (native LPA and MAPCA) to all segments. The left MAPCA was test occluded. (**e**) Placement of a vascular plug to occlude the large left MAPCA and placement of a stent on the RVOT (crossing the hypoplastic pulmonary valve annulus) via a Flexor sheath over a coronary wire. (**f**) RVOT angiogram showing good flow to the entire LPA system and improved flow to the RPA. Subsequently the vascular plug was released. The patient weaned successfully from ventilation 5 days later. He is awaiting further catheter 6 months later (saturations 84%, weight 7.6 kg) with a view to dilate the stent further and ultimately to undergo recruitment of right MAPCAs and unifocalization to a restrictive valved conduit

### 37.3.7 Experience So Far

RVOT stenting is a complex procedure with a definite learning curve. Meticulous procedure planning and execution is essential. Early mortality rates of <2% compare favourably with either BT shunt procedures or early Fallot repair. In as yet unpublished studies, we could document that RVOT stenting provides better pulmonary arterial growth (Fig. 37.8) compared to BT shunt and that time to complete repair was reduced.

## 37.4 Summary

RVOT stenting in the initial palliation of patients with tetralogy of Fallot is displacing BT shunt surgery. Catheter procedures are complex and have to be well planned, and executed, ideally by an established team. Strict adherence to the above step-by-step guide is likely to result in success and short procedure times.

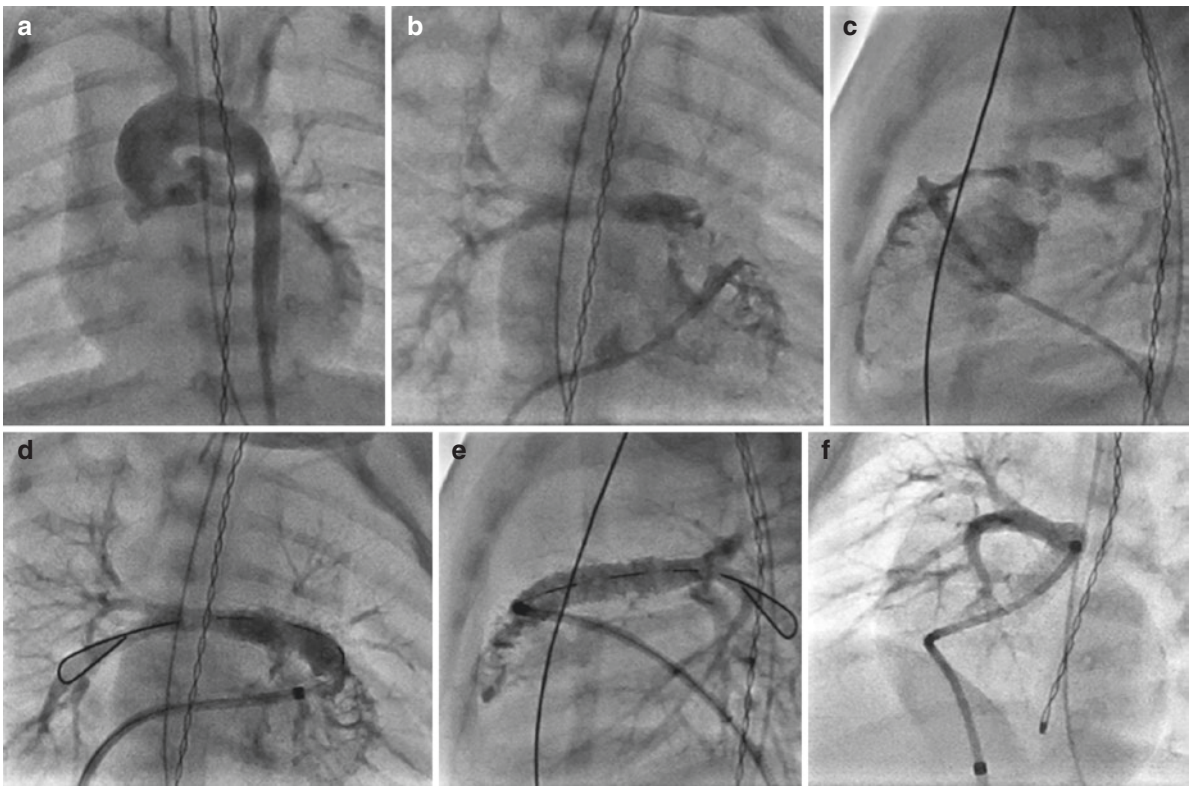



**Fig. 37.6** Sequence of angio still frames in a 2-week-old child who was initially thought to suffer from pulmonary atresia with a tortuous duct from the underside of the left aortic arch. (**a**) Retrograde aortogram in AP projection documents a left arch and a wide opening but rather tortuous duct to confluent PAs of reasonable size. (**b**) RV angio in 30 RAO + cranial documented some miniscule forward flow through the RVOT mainly to the right PA. (**c**) Lateral projection of the same angio. Decision was taken to stent the RVOT rather than to attempt stenting the wide and tortuous duct with the potential of acute instability and possible later branch PA stenosis. (**d**) RAO angio after 5 mm coronary stent placement through 4 F Flexor sheath over a 0.014" Thruway wire. Note the stent was placed across the pulmonary valve annulus but stops short of the bifurcation. (**e**) Lateral projection of the above. (**f**) Final angio in four-chamber angulation documents good flow to the right PA. The left PA was predominantly perfused through the patent duct, as confirmed on post-placement cardiac ultrasound. Prostin was stopped, and the child underwent complete repair at 6 months with transannular patch and surgical bifurcation plasty

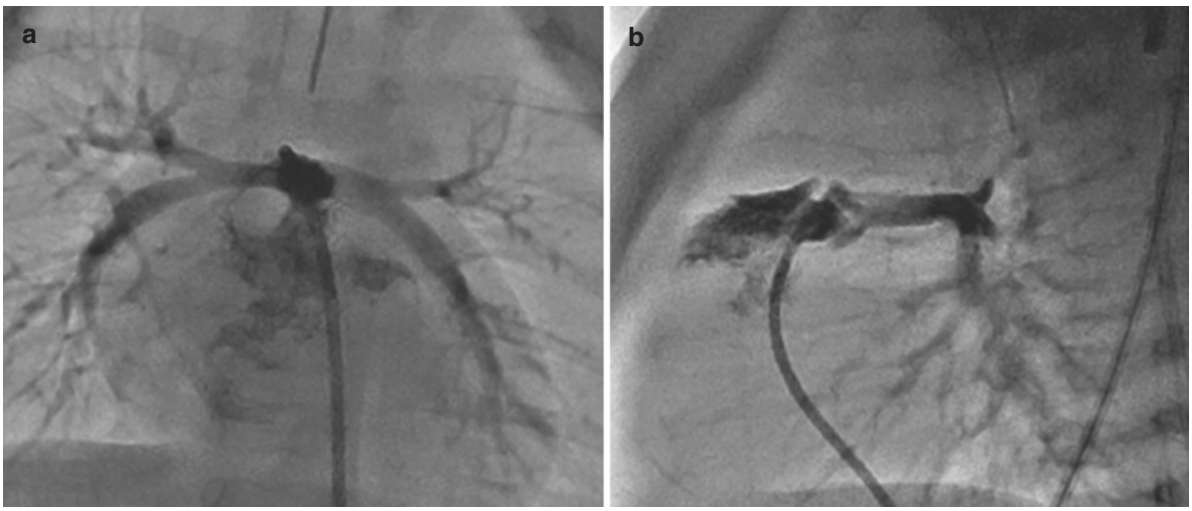



**Fig. 37.7** Sequence of angio stills in a neonate with right atrial isomerism, unobstructed total anomalous pulmonary venous return, complete AVSD and severe valvar and supravalvular pulmonary stenosis. (**a**) AP projection with cranial tilt showing symmetric branch pulmonary arteries of a reasonable size. (**b**) Lateral angiogram documents wide open subpulmonary area and severe valvar and the supravalvular pulmonary stenosis. (**c**) Four-chamber projection documents hypoplasia of the main pulmonary artery up to the bifurcation. (**d**) RVOT stent was placed across the pulmonary valve and the main pulmonary artery up to the bifurcation. No further intervention was required until the age of 9 months (bilateral CP shunts, PA augmentation and repair of TAPVC)

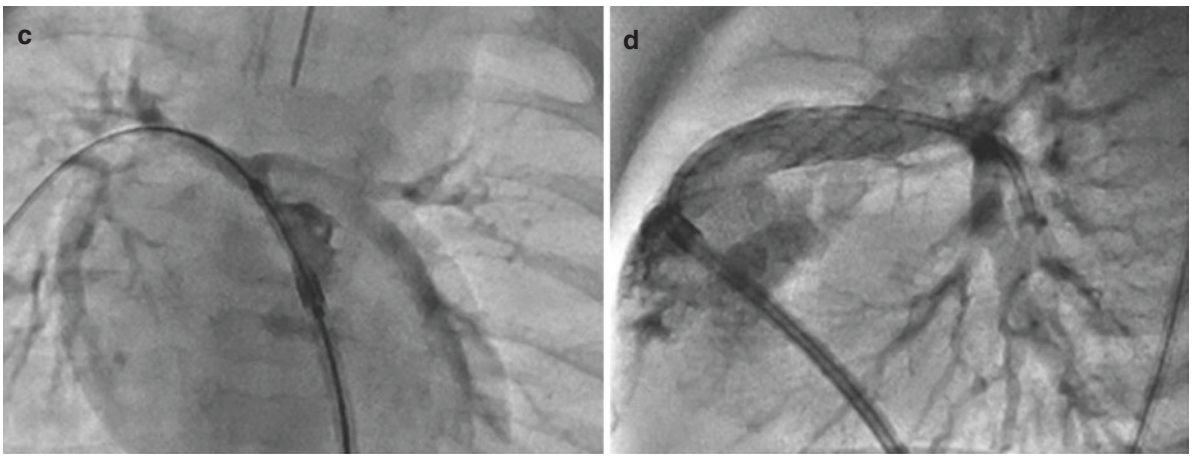



**Fig. 37.7** (continued)

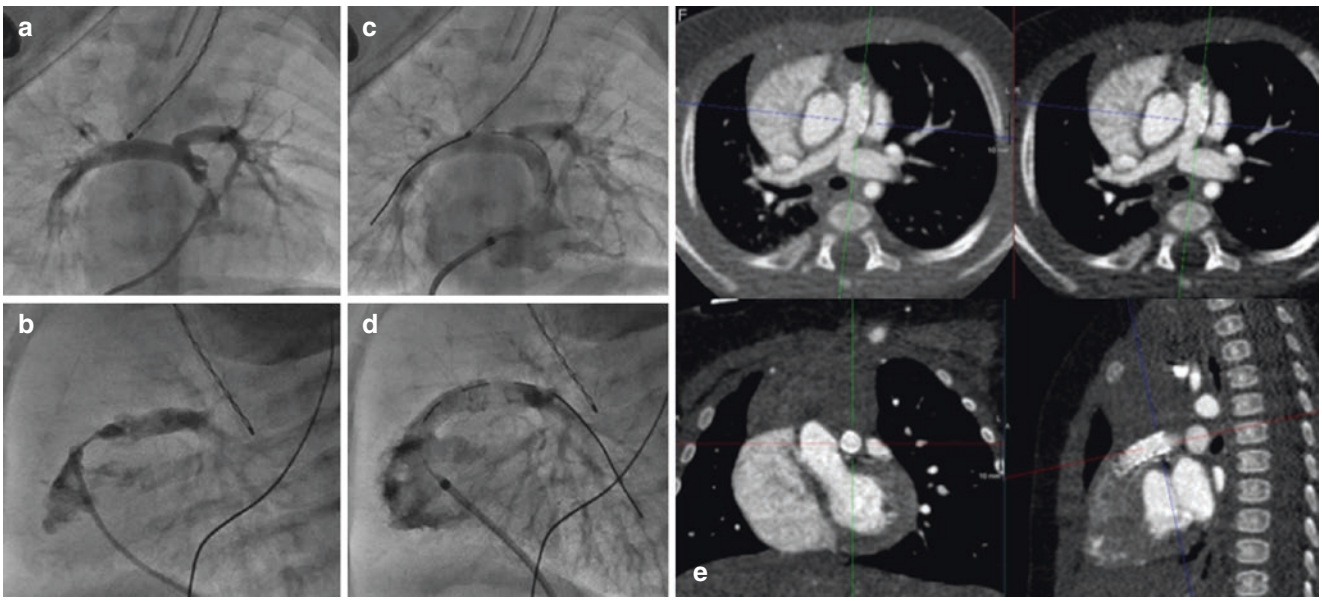



**Fig. 37.8** Image sequence in a neonate with severe Fallot undergoing RVOT stenting at 2.6 kg in weight. (**a**) Good-sized branch pulmonary arteries on four-chamber projection. (**b**) Tight right ventricular outflow tract obstruction on lateral projection. (**c**) Corresponding angio stills after placement of a 5 mm coronary stent (**d**) as above. (**e**) CT angio multiplanar reconstruction images of the branch pulmonary arteries and the stented right ventricular outflow tract prior to complete repair at 6 months of age

**Video 1** Step-by-step RVOT stenting procedure in a 3-month-old child with complete AVSD and tetralogy of Fallot and significant co-morbidities (MOV 171123 kb)

# Part X

# Role of Specific Imaging Techniques

# 3D Rotational Angiography for Percutaneous Interventions in Congenital Heart Disease

38

Gregor Krings

## 38.1 Introduction

3D rotational angiography (3DRA) is an evolving technique that delivers computed tomographic imaging at the heart catheterization suite. A 180 ° rotation of the frontal plane during an interval of 4–7 s produces a rotational angiography (RA) when contrast is injected simultaneously with vendor-specific differences in the post processing.

## 38.2 Rotational Angiography

In a historic publication, Professor Schad/Stuttgart (Schad 1964–1966) demonstrated the value of rotational angiographies in congenital heart disease with the patient being fixed in a cradle and moved around the static C-arm. It took 40 years until complex algorithm (Kehl/Vogt, Frauenhofer 1999) allowed for computed conversion of the single rotational frames into stacks known from CTA or MRA.

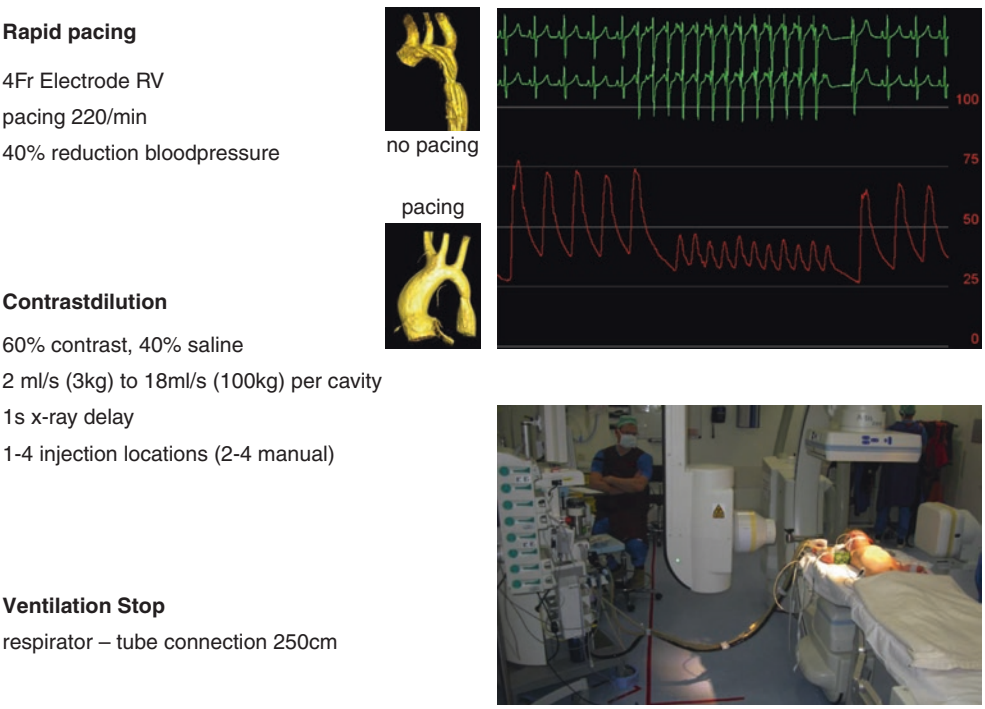


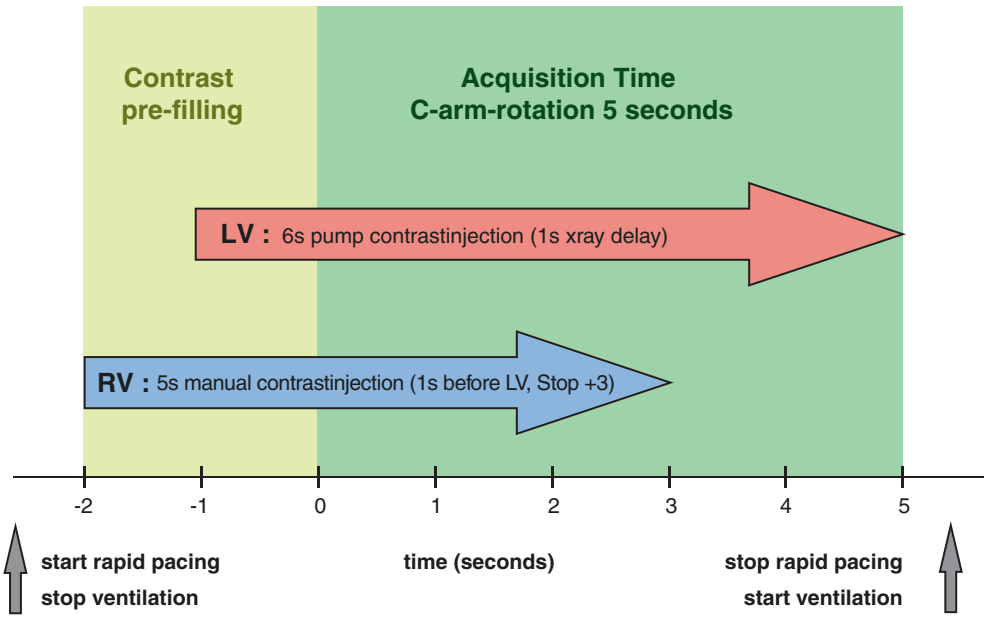

**Fig. 38.1** 3DRA workflow

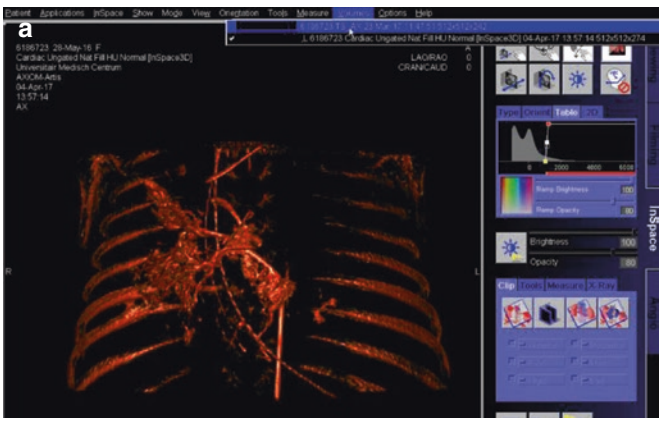
**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_38) contains supplementary material, which is available to authorized users.

G. Krings (✉)
University Medical Centre, Utrecht, The Netherlands
e-mail: g.krings@umcutrecht.nl

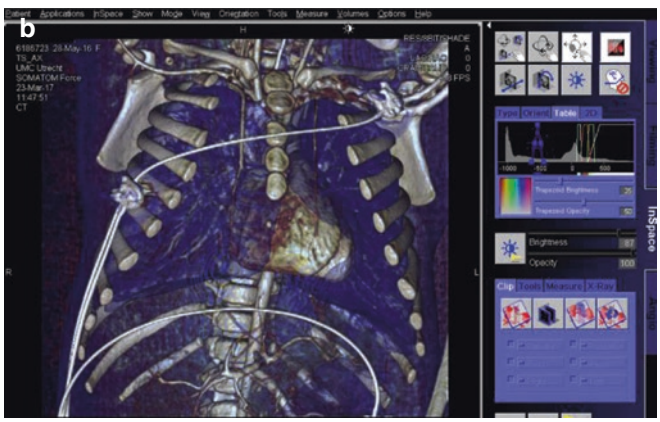

G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_38

**Fig. 38.2** 3DRA acquisition

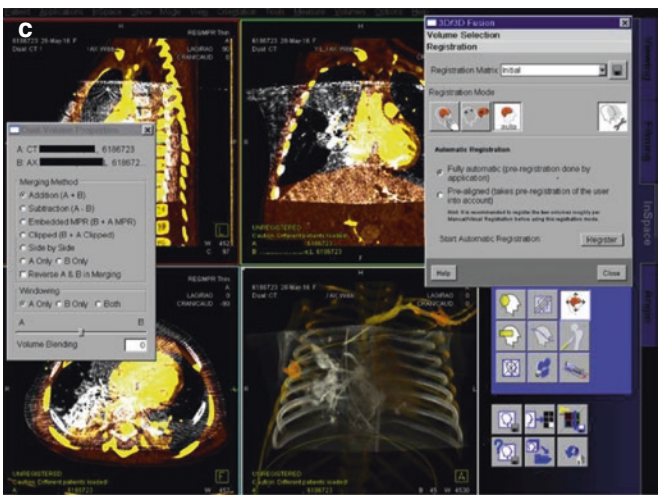


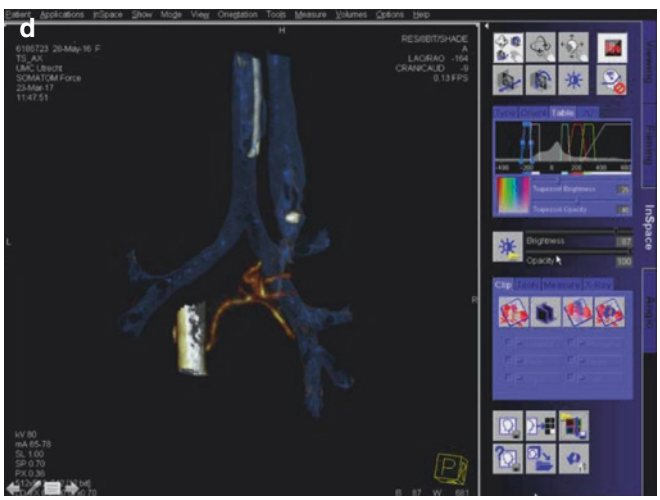

**actual 3DRA**
visualisation of right sided MAPCA's




**import of previous CTA**
on 3DRA workstation as second 3D dataset




**registration of 3DRA and CTA**
manual or automatic overlay




**result after post processing:**
3DRA and CTA information in one image

**Fig. 38.3** 3DRA: Fusion with previous CTA

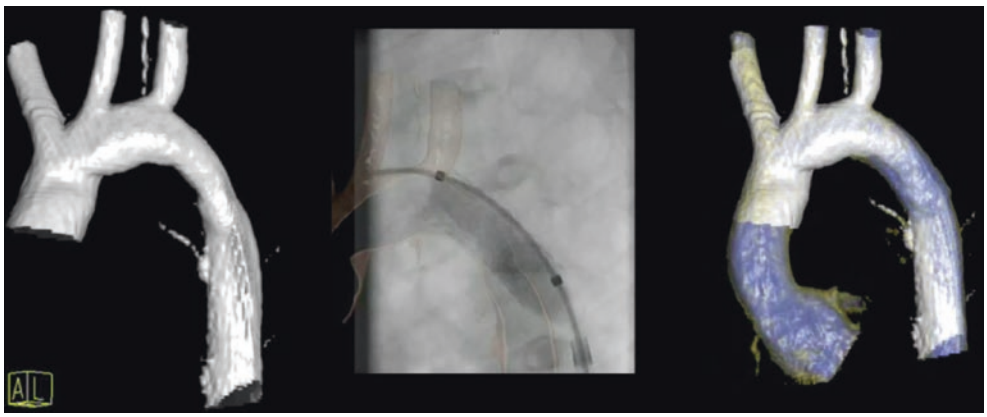


**Fig. 38.4** Case 1: Coarctation stenting

**3DRA in post surgical Re-CoA (8 kg weight)**

1. contrast injection in LV guarantees homogeneous contrast in Aortic Arch
2. 3D roadmapping reduces contrast during stent placement
3. 3DRA pre and post intervention, optimal stent position and dimension

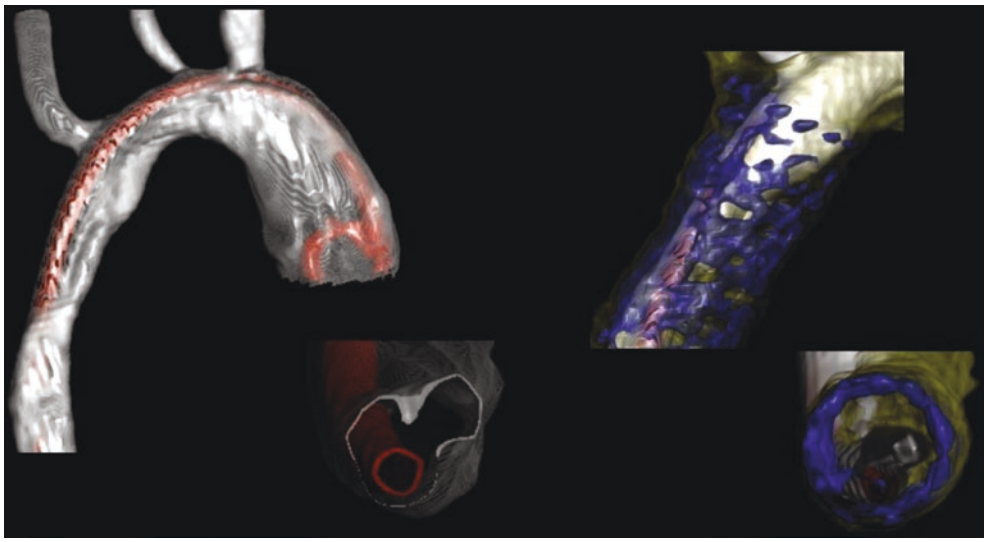


**Fig. 38.5** Case 2: CoA dissection

**3DRA post balloon dilatation in Re-CoA demonstrates dissection**

1. dissection visualized in different cross sectional planes and hollowed aorta
2. stent indication based on dissection pattern and residual gradient
3. 3DRA post intervention demonstrates stent position (Cook Formula 10x20mm)

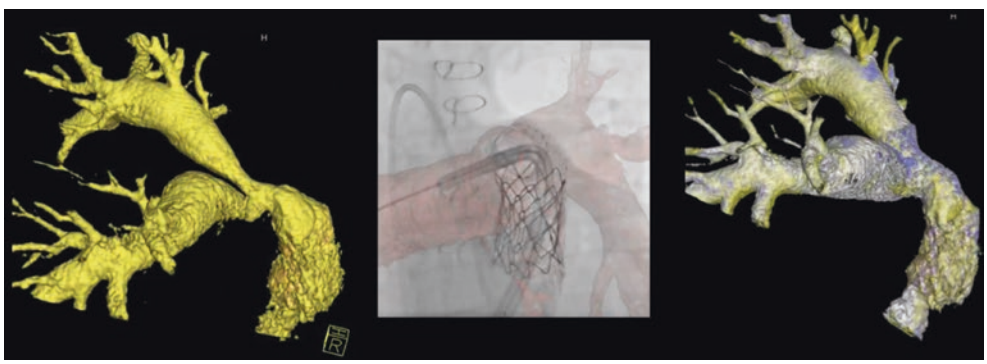
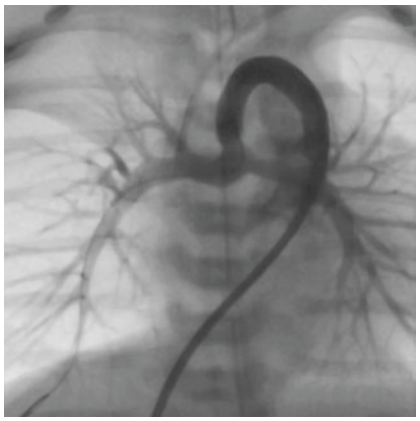


**Fig. 38.6** Case 3: Bifurcation stenosis

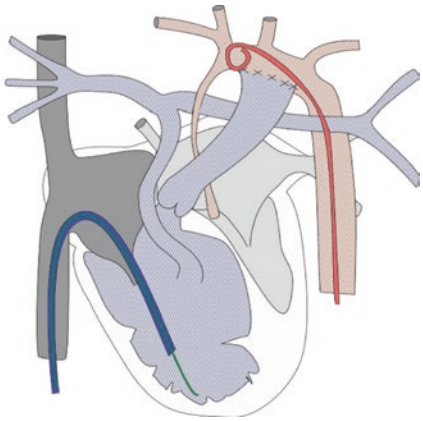
**3DRA in complex pulmonary artery bifurcation stenosis and PPVI**

1. visualisation of both PA branches and Melody valve in different cross sectional planes
2. Y stenting with telescope and anchor technique (ev3 Mega LD and ev3 Max LD)
3. 3DRA pre and post intervention demonstrate stent positions and dimensions



**conventional angiography**

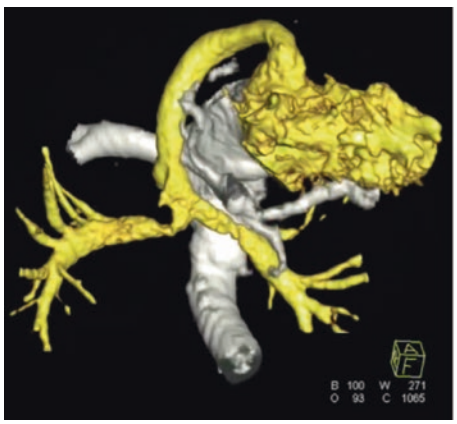
stenosis not visible



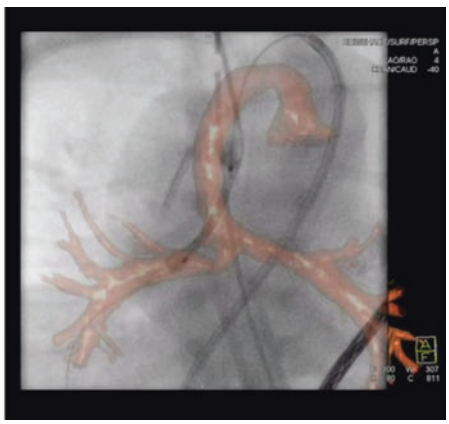
**3DRA**

injection protocol :
12 ml RV, 8 ml AoArch
pacing 220/min via long sheath

**Fig. 38.7** Case 4: Sano shunt stenosis. HLHS, Norwood I, 5 mm Sano shunt. 3 days post-op: oxygen saturation 50–60%



**3DRA with all-in-one visualization**
severe RPA stenosis only visible in virtual caudal angulation neo aorta with coronaries in silver systemic RV, Sano shunt and pulmonary arteries in gold



**3DRA and roadmap**
3D image reduced to relevant structures and colour optimised, 3D projected on 2D frontal plane for intervention anatomic shift can occur and be adapted manually

**Fig. 38.8** Case 4: Sano shunt stenosis. 3D visualizing of entire anatomy and guidance of stent implantation

## 38.3 Status Quo of 3DRA

The current systems use integrated workstations to automatically calculate and 3D reconstruct the scanned tissue within 10–20 s after rotation. The principle of contrast distribution in 3DRA is different from conventional angiography (CA) with a complex workflow including optional rapid pacing (Fig. 38.1), breath holding, and injection of diluted contrast simultaneously at multiple locations during 5–7 s. Post processing of the 3DRA dicom data is similar to CTA or MRA and allows for scissoring to hide irrelevant structures, threshold adaptation to visualize a specific range of Hounsfield units, use clipping planes and MIP views to measure, and virtually fly through anatomic structures. Projection of the post-processed 3D image to the CA screen enables for road mapping to guide complex procedures without the need for sequential localizing angiographies. Typical indications for 3DRA are aortic arch (AoS) stenosis, pulmonary artery or vein stenosis (PAS, PVS), interventions in pulmonary atresia (PA VSD), pulmonary valve implantation (PPVI), and interventions in single ventricle (SV) stages I–III.

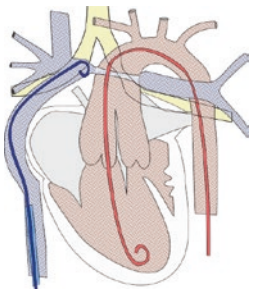
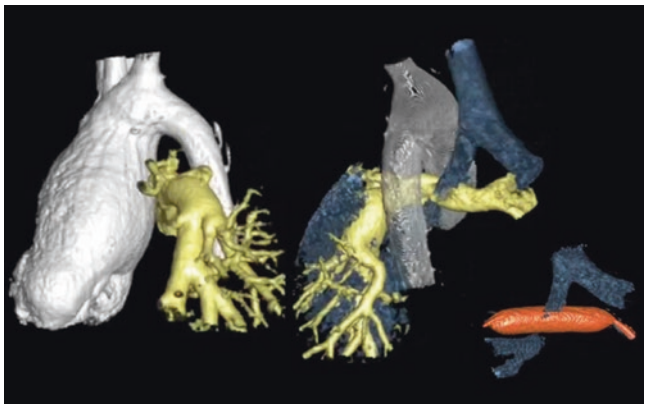


**Fig. 38.9** Case 5: LPA stenosis in TCPC. 3D visualizing of entire anatomy and guidance of airway interrogation

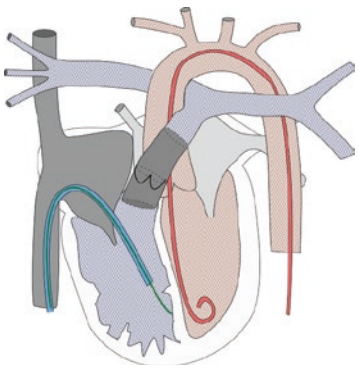
**3DRA**

injection protocol (50 kg):
25ml VCI and 25ml VCS,
50ml single Ventricle
internal pacemaker 180/min



**two 3DRA's : diagnostic and interrogation**

1. visualization of DKS, LPA and airway
2. LPA ballooning to check for bronchus compression



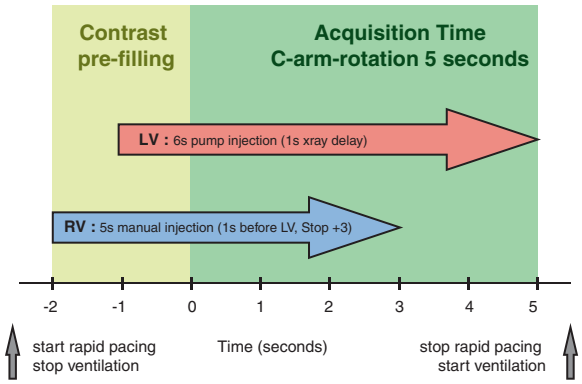
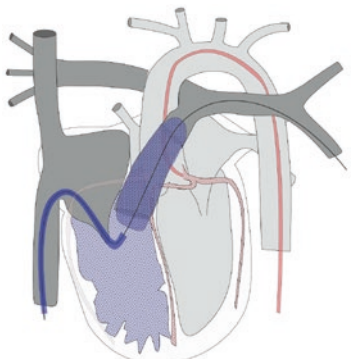



**Fig. 38.10** Case 6: PPVI "entire heart protocol"

**First 3DRA "Entire Heart Protocol" to visualize the entire anatomy in PPVI**

1. RV contrast manually via long sheath, RV rapid pacing via same long sheath, LV contrast via injector
2. prefilling RV -2 s and stop at 3s, LV -1s with continuation until 5s
3. first 3DRA can be skipped when previous detailed CTA or MRA data are available



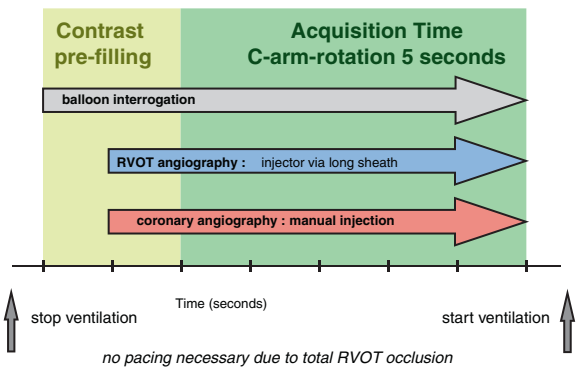



**Fig. 38.11** Case 6: PPVI "interrogation protocol"

**Second 3DRA "Interrogation Protocol" to visualize MPA and coronaries when ballooning**

1. low amount RV contrast manually via long sheath during balloon interrogation MPA and simultaneous selective coronary angiogram
2. no rapid pacing necessary due to total MPA occlusion causing no-output
3. 3DRA visualizes precisely coronary artery and balloon – MPA topography

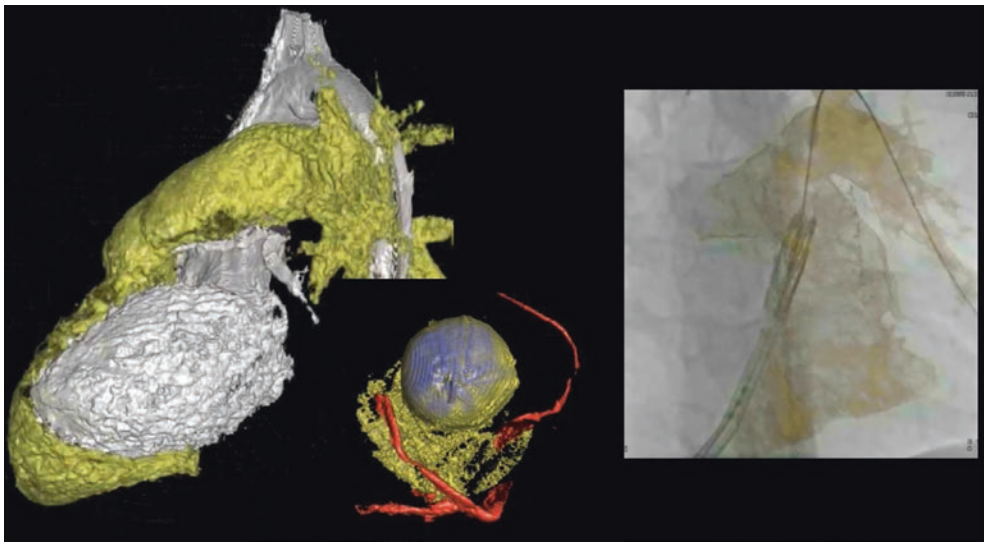


**Fig. 38.12** Case 6: PPVI based on 3DRA

**3DRA in standard PPVI :**

1. Entire Heart and Interrogation 3DRA protocol in Fallot after transanulary patch
2. 3D based prestenting and implantation of Sapien 3 valve

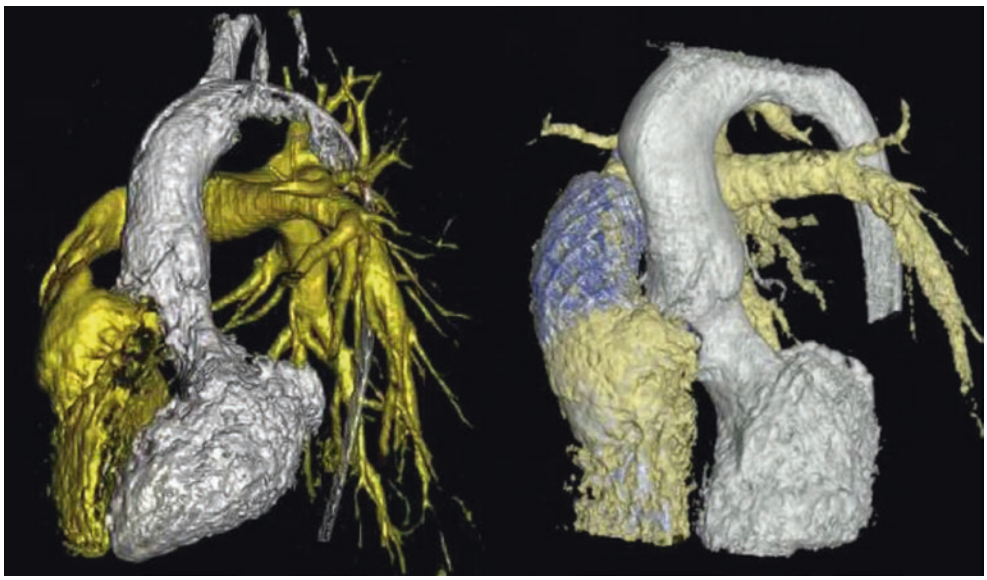


**Fig. 38.13** Case 7: 3DRA in complex PPVI

**3DRA pre and post PPVI in small child with severe retrosternal compression**

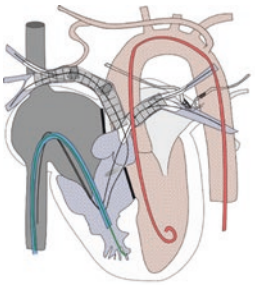
1. Fallot after transanulary patch, 22kg,
2. homograft failed twice due to retrosternal compression, sternum partly resected without improvement
3. 3DRA pre and post PPVI : prestenting with Andra XXL and 2 CP stents, implantation Melody 20mm

## 38.4 Multimodality

Previous CTA, MRA of 3DRA dicom data can be imported – the so-called merge or fusion – to generate overlay visualization and sometimes can substitute a 3DRA. Anatomical annotation of those data is based on bone structures.

## 38.5 Benefit of 3DRA

3DRA offers the ability to understand the entire anatomy in unrestricted angulations, thus delineating substrates not or hardly visible by CA. As with all kinds of new techniques, the benefit of 3DRA strongly depends on the user's confidence and trust in the resulting images. A validated algorithm



**3DRA**

36 ml RV (manual, long sheath)

36 ml LV (injector)

rapid pacing 220/min

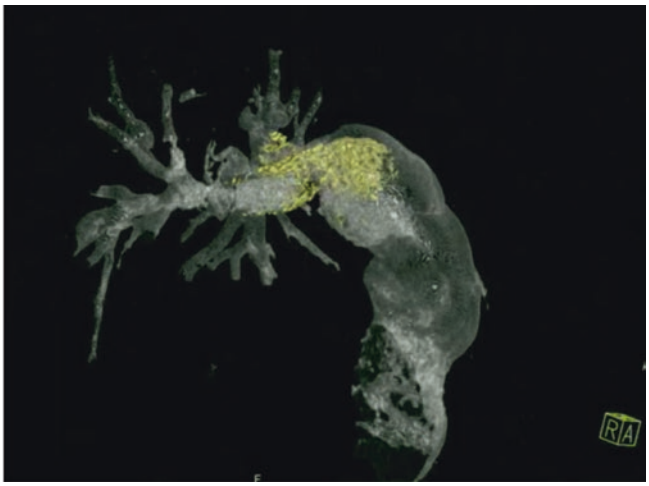



**Fig. 38.14** Case 8: Complex PA VSD. A 3-year-old child with PA VSD after Melbourne shunt, unifocalization, multiple stent interventions, and implantation of Contegra valve during correction, severe distal MPA, and bifurcation stenosis post surgery

**3DRA …**

produces detailed 3D imaging

during interventions

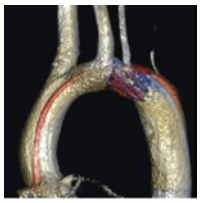
visualizes the entire anatomy

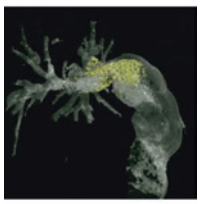
identifies critical interactions

enhances safety of interventions

offers guidance by 3D road mapping to

reduce contrast amount and radiation





**3DRA still needs …**

simplified workflow and workstations

biplane instead of monoplane 3D roadmapping

better correction of anatomical versus 3D shift

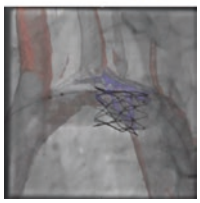


**Fig. 38.15** 3DRA conclusion

is essential covering contrast dilution and amount, injection locations, and timing as well as optimization of the systems radiation parameters. In an optimal setting, an "all in one run" scan can be performed with 0.5 mSv and a contrast amount equal to two CAs. In complex bi- and univentricular hearts, it will deliver information of the entire topography allowing for a safer procedure with less radiation and contrast compared to CA. Visualization of vessel-vessel as well as vessel-airway interaction will promote to defocus from the original target of the intervention and widen the horizon by identifying potential interactions to prevent complications. The following figures cover the workflow in general as well as its case-related adaptations in AoS, PAS, PVS, PA VSD, PPVI, and SV I–III.

*The images demonstrated are derived from a Siemens' syngo DynaCT with a biplane Artis zee system. The results reflect a 6-year period of use in congenital heart disease with use of 3DRA in 70% of all interventions. Toshiba, Philips, and GE offer comparable equipment.*

In conclusion 3DRA is a tool available during catheter interventions which enables to visualize all intrathoracic structures and their interactions. It helps to guide complex interventions, reduce radiation and enhance safety. Improvements of the available 3DRA systems are necessary in terms of post processing, 3D roadmapping with biplane projection and automized correction for anatomic shift (Fig. 38.15).

**Video 1** Case 1. 3D rotational angiography in post-surgical re-coarctation (MOV 1544 kb)

**Video 2** Case 2. 3D rotational angiography post-balloon dilatation in Re-CoA demonstrates dissection. A Cook Formula 10 × 20 has been implanted (MOV 4470 kb)

**Video 3** Case 3. 3D rotational angiography in complex pulmonary artery bifurcation stenosis and PPVI. Both PA branches and Melody valve are visualized in different cross-sectional planes. Two stents across the bifurcation are implanted by using Y stenting with telescope and anchor techniques. Ev3 Mega LD and ev3 Max LD have been implanted. Finally, 3DRA shows pre- and post-intervention stent positions and dimensions (MOV 37640 kb)

**Video 4** Case 4. Sano shunt anatomy and stenosis at the origin of the left pulmonary artery (MOV 3416 kb)

**Video 5** Case 4. 3D rotational angiography road map for stent implantation in Sano shunt stenosis. Note the anatomic shift 2D versus 3D, correction of shift performed manually (MOV 10521 kb)

**Video 6** Case 5. Left pulmonary artery stenosis in total cavopulmonary connection. It is possible to visualize DKS, left pulmonary artery (LPA), and airways. Furthermore, LPA ballooning is performed in order to check for bronchus compression (MOV 2600 kb)

**Video 7** Case 6. Percutaneous pulmonary valve implantation (PPVI). Entire heart protocol and interrogation protocol (MOV 36490 kb)

**Video 8** Case 7. Complex PPVI. Entire heart protocol and interrogation protocol (MOV 13243 kb)

**Video 9** Case 8. A 3-year-old child with PA VSD after Melbourne shunt, unifocalization, multiple stent interventions, and implantation of Contegra valve during correction. Severe distal MPA and bifurcation stenosis post surgery are seen by using 3d rotational angiography (MOV 10623 kb)

# Cardiac Magnetic Resonance

# 39

Francesca Romana Pluchinotta and Massimo Lombardi

## 39.1 Introduction

Imaging is important in the pre- and postoperative management of patients with congenital heart disease (CHD). From the fetal stage onward, imaging outlines anatomy and physiology, helps to refine management, evaluates the consequences of interventions, guides further procedures, and provides prognostic information in this unique set of patients.

Echocardiography remains the mainstay of cardiovascular procedure, as it is portable and noninvasive and provides immediate, high-resolution, anatomical and physiological information. However, echocardiography fails when acoustic windows are poor, particularly for the assessment of extra-cardiac vascular structures.

In the last decades, cardiovascular magnetic resonance (CMR) has progressively gained importance in the management of CHD patients. CMR is a particularly attractive imaging technique due to its excellent tissue characterization and its high spatial resolution in delineating extra-cardiac anatomy, including the great arteries and systemic and pulmonary veins. With CMR vascular and valvular flow can be assessed, shunts can be quantified, and ventricular volumes and myocardial function can be measured accurately with high reproducibility and regardless of ventricular morphology allowing for serial comparisons over time. Finally, CMR provides high-resolution, isotropic, three-dimensional datasets that allow for reconstruction of data in any imaging plane, giving complete visualization of complex cardiac anomalies.

The sequences used for assessing different conditions are summarized in Table 39.1.

### 39.1.1 Spin Echo Imaging Acquisitions

In spin echo imaging pulse sequence, also known as black blood images, the flowing blood appears dark. The contrast between myocardium, blood, and extravascular structures is enhanced. The sequence is less susceptible to artifacts caused by turbulent flow or metallic implants, such as coils, devices, or stents.

### 39.1.2 2D SSFP Imaging

In SSFP acquisition blood appears bright. Images can be displayed as still images or cine sequences. Cine images allow visualization of myocardial or valvular motion over the entire cardiac cycle. Cine can be performed in any plane to assess the dynamic function of any structure, including the outflow tracts, valves, great arteries, and veins. Furthermore, short-axis cine images acquired perpendicular to the long axis of the heart from the base to the apex can be used to accurately assess cardiac function and measure the ventricular volumes and myocardial mass.

### 39.1.3 Velocity-Encoded Phase Contrast MR

The velocity-encoded phase contrast sequences allow quantification of blood flow and velocity in moving tissue and provide estimation of stroke volume and cardiac output. Quantification of valvular regurgitation and/or stenosis and of pulmonary to systemic blood flow ratio (Qp/Qs) is an

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_39) contains supplementary material, which is available to authorized users.

F. R. Pluchinotta (✉)
Department of Pediatric Cardiology and Adult Congenital Heart Disease, Multimodality Cardiac Imaging Section—IRCCS Policlinico, Milan, Italy
e-mail: Francesca.Pluchinotta@grupposandonato.it

M. Lombardi
Multimodality Cardiac Imaging Section—IRCCS Policlinico San Donato, Milan, Italy


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_39

**Table 39.1** Reproduced with permission from Valsangiacomo Buechel ER, Grosse-Wortmann L, Gratz S, et al. Indications for cardiovascular magnetic resonance in children with congenital and acquired heart disease: an expert consensus paper of the Imaging Working Group of the AEPC and the Cardiovascular Magnetic Resonance Section of the EACVI. Cardiol Young. 2015 Jun;25(5):819–38

| | | | CMR sequence | | | | |
|---|---|---|---|---|---|---|---|
| Lesion | BB SE | 2D SSFP | PC flow | 3D CE-MRA | 3D SSFP | Perfusion imaging | LGE |
| Aortic arch anomaly | + | ++ | ++ | +++ | ++[a] | | |
| Pulmonary arteries | + | + | ++ | +++ | ++[a] | | |
| Pulmonary veins | + | + | + | +++ | ++[a] | | |
| Shunt lesions | ++ | ++ | +++ | ++ | | | |
| TOF | ++ | +++ | +++ | ++ | ++[a] | | ++ |
| Complex CHD | ++ | +++ | +++ | ++ | +++ | | |
| Single ventricles | ++ | +++ | +++ | ++ | ++[a] | | ++ |
| Intracardiac tumours | ++ | ++ | | | | +++ | ++ |
| Cardiomyopathies | + | ++ | ++ | | | ++ | +++ |
| Coronary arteries | | | | | + + | +++ | ++ |

+ may be used in the assessment of this lesion, but better alternatives available or does not provide additional information over other techniques; ++ useful technique, commonly applied in the assessment of the lesion; +++ needs to be part of any study in this lesion

[a]If contrast needs to be avoided. BB SE, Black blood spin-echo; 2D SSFP, two-dimensional steady-state free precession; PC flow, phase contrast flow; 3D CE-MRA, three dimensional contrast-enhanced magnetic resonance angiography; 3D SSFP, three-dimensional steady-state free precession; LGE, late gadolinium enhancement

important tool to assess disease progression and timing for surgery. While accuracy and reproducibility of phase contrast MR has been validated in numerous phantom experiences, velocity estimation might be underestimated, and this should be remembered when comparing with Doppler echocardiographic measures.

### 39.1.4 3D Dataset

The 3D capabilities of CMR play a key role in CHD. There are two methods of acquiring 3D data. One is the contrast-enhanced magnetic resonance angiography (CE-MRA) based on gadolinium injection, which is not cardiac gated representing therefore a systo-diastolic average across the cardiac cycle. The other method uses a 3D-balanced SSFP sequence which is respiratory and cardiac gated. This sequence does not necessarily required contrast; however quality is usually improved by contrast agent. Both acquisitions are isotropic and can be reformatted in any plan or used for volume rendering of complex structures.

### 39.1.5 Late Gadolinium Enhancement

Late gadolinium enhancement CMR is becoming an important part of imaging both in congenital and acquired cardiac diseases. The accumulation of gadolinium in the myocardium helps to differentiate between normal and diseased cardiac muscle and has been demonstrated to have an important prognostic value in patients with CHD.

### 39.1.6 Real Time

Real time is a fast imaging sequence which is largely used for magnetic resonance-guided cardiac catheterization. Although the temporal and spatial resolution achievable in CMR is not near that of conventional *X*-ray techniques, MR offers excellent soft tissue contrast and flexible slice positioning that are useful to guide cardiac catheterization. In addition, the advantages of reducing radiation exposure especially in the pediatric population make MR of great interest for image-guided interventions in children.

Specific technical and diagnostic challenges applied to imaging children and adolescents with CHD. In pediatric patients the body size is small and heart rates are rapid. Imaging these patients requires operators trained to expedite image planning and optimize pulse sequences in this context and physicians with expertise in the anatomical and physiological changes of CHD. In adult patients, the technical challenges might be lower but the knowledge of anatomy and prior surgical interventions is crucial to perform an imaging focus for each individual patient.

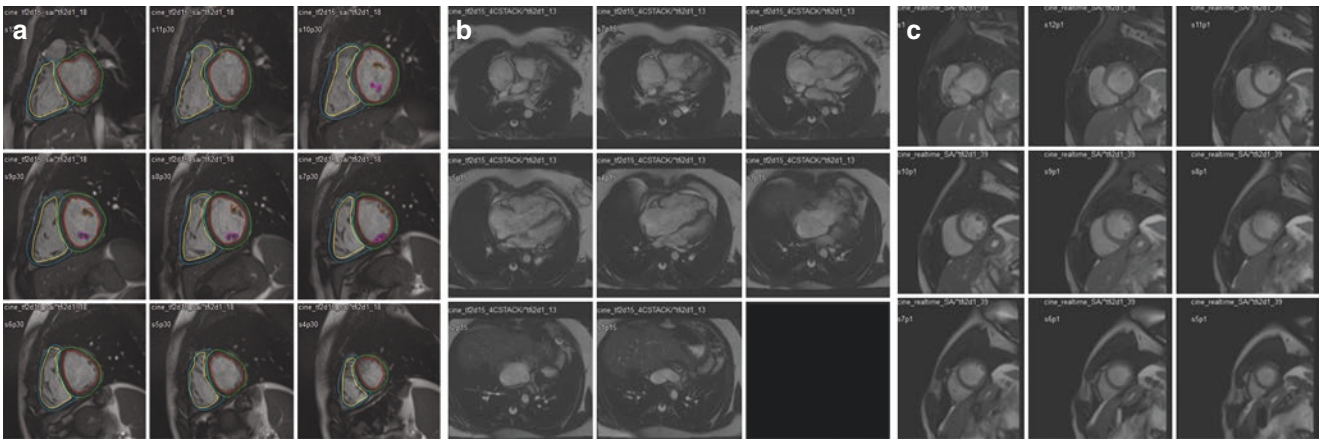



**Fig. 39.1** Ventricular volumes and function. 2D SSFP short axis (**a**), axial (**b**), and real-time (**c**) stack of the ventricles. Segmentation with commercial software of the epicardium and endocardium (**a**) gives estimation of ventricular volumes, mass, and function

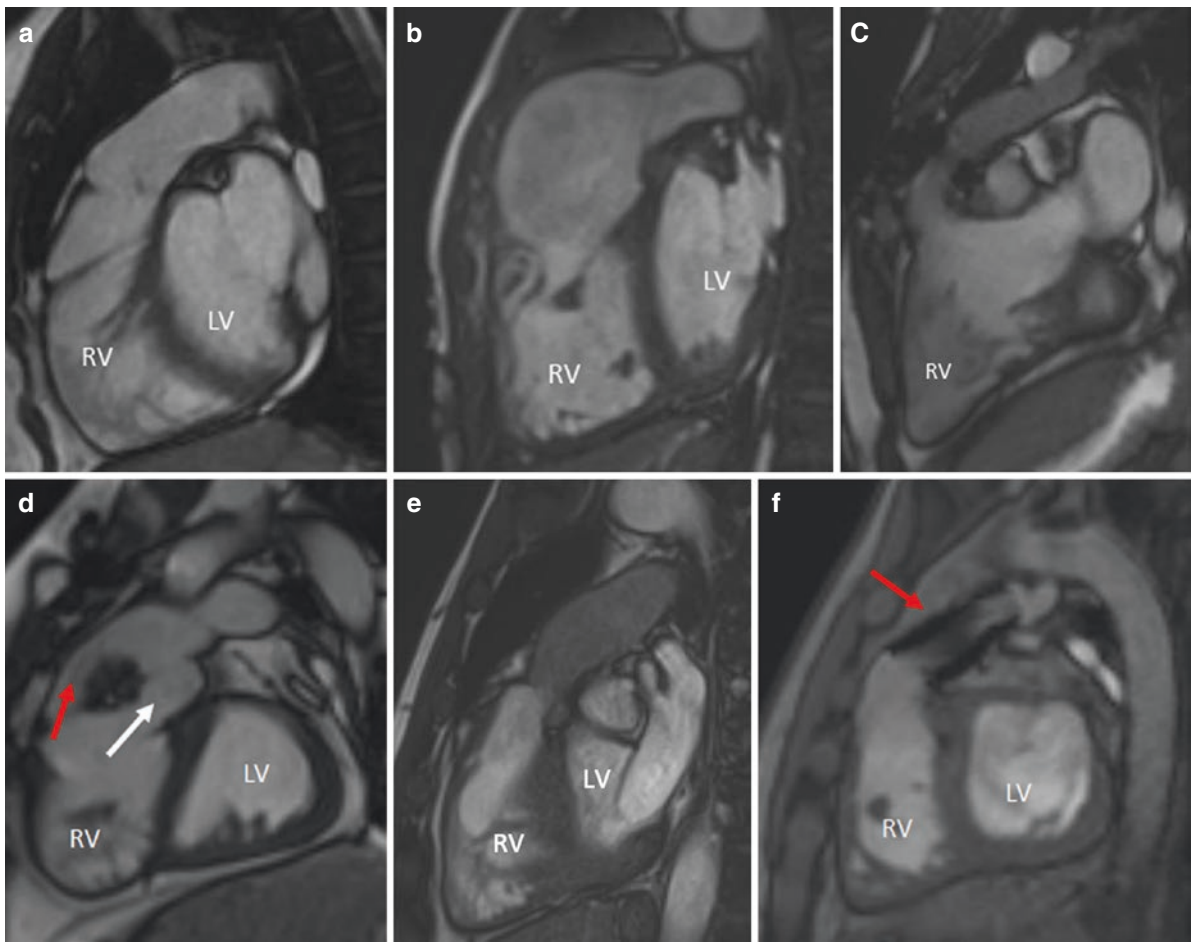



**Fig. 39.2** Right ventricular outflow tracts. 2D SSFP short axis view showing different right ventricular outflow tract. Note the variation in morphology, size, and narrowing. (**a**) Patient after repair of tetralogy of Fallot with ventricular septal defect closure and pulmonary commissurotomy. (**b**) Patient after repair of tetralogy of Fallot with ventricular septal defect closure and outflow patch enlargement. (**c**) Patient with transposition of the great arteries, ventricular septal defect, and pulmonary stenosis who underwent Rastelli operation and now presents with right ventricle to pulmonary artery conduit stenosis. (**d**) Patient after repair of tetralogy of Fallot with ventricular septal defect closure and right ventricle to pulmonary artery conduit (red arrow) for an anomalous course of the right coronary artery. Native right outflow tract is patent (white arrow). (**e**) Patient after Ross operation for bicuspid aortic valve who presents stenosis of the homograft in pulmonary position. (**f**) Patients with pulmonary atresia and ventricular septal defect who underwent surgical correction in infancy and placement of a Melody valve in the outflow tract later in life. The stent of the percutaneous pulmonary valve appears black (arrow). Legend: *LV* left ventricle, *RV* right ventricle

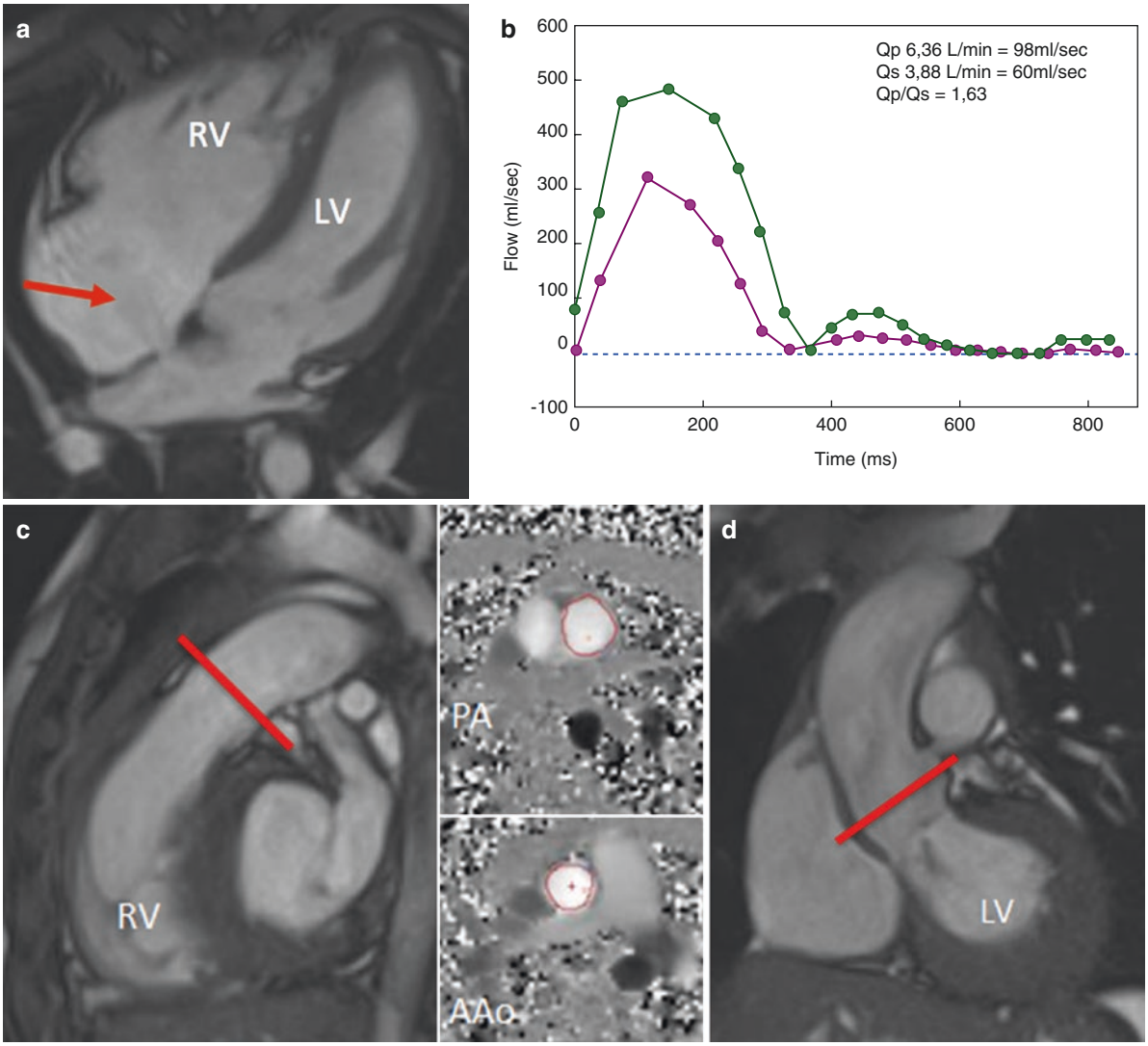



**Fig. 39.3** Atrial septal defect. Patient with unrepaired atrial septal defect, ostium secundum type. (**a**) 2D SSFP images showing the defect at the atrial level (arrow). In case of shunts, MR is useful to calculate the Qp/Qs ratio. Phase contrast sequence planned on the pulmonary valve (**c**) and on the aortic valve (**d**) gives accurate estimation of the antegrade flow through the valves. Flows are used to calculate the Qp/Qs ratio (**b**). Legend: *AAo* ascending aorta, *LV* left ventricle, *PA*, pulmonary artery, *RV*, right ventricle

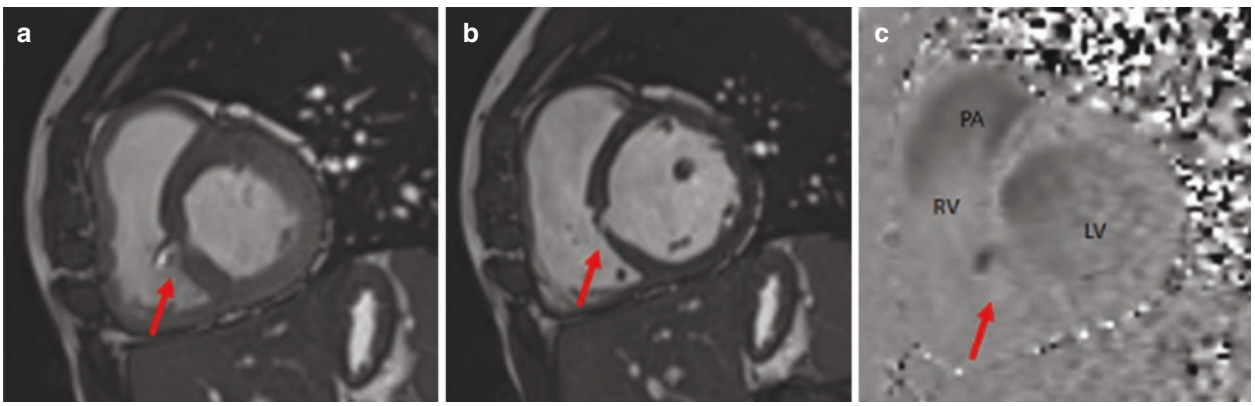



**Fig. 39.4** Interventricular septal defect. Patient with unrepaired interventricular septal defect, muscular type. (**a**) 2D SSFP short axis view in systole showing small mid-muscular interventricular septal defect (arrow). During systole the signal dephases secondary to turbulent flow through the defect. (**b**) 2D SSFP short axis view in diastole showing thinning of the muscular septum at the level of the defect. (**c**) Phase contrast MRI might be useful to show the flow through the defect during systole (arrow). Legend: *LV* left ventricle, *PA*, pulmonary artery, *RV* right ventricle

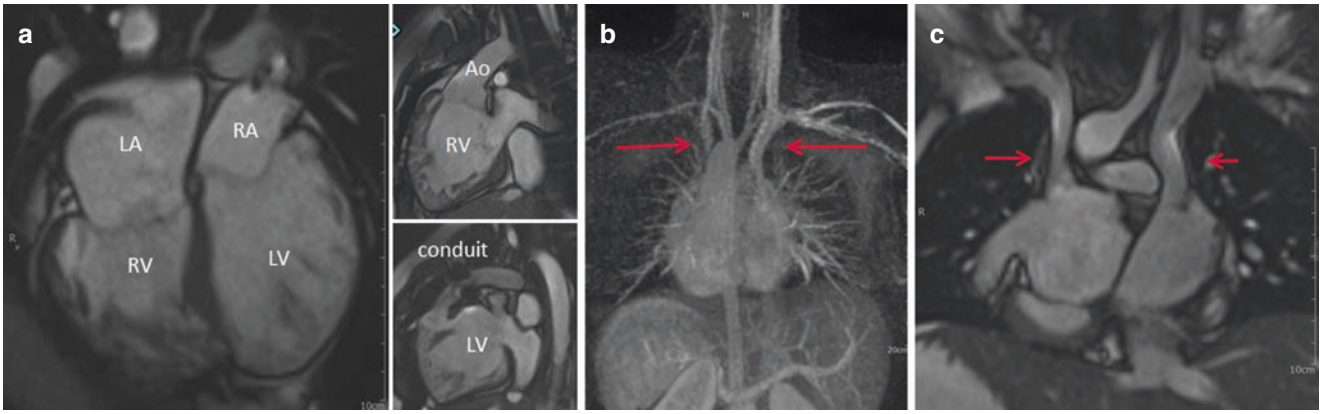



**Fig. 39.5** Anomalous systemic venous return. Patient with situs inversus, mesocardia, congenitally corrected TGA, ventricular septal defect, and pulmonary stenosis who underwent ventricular septal defect closure and position of a left ventricle to pulmonary artery conduit in infancy. She complains of increased fatigability and desaturation during exercise. (**a**) 2D SSFP long axis view showing the complex anatomy (the morphologically right atrium on the left side of the body connects to the left ventricle and to the conduit, while the morphologically left atrium on the right side of the body connects to the right ventricle and the aorta). (**b**) Time-resolved contrast-enhanced MR angiography showed two systemic venous return from the upper side of the body (arrows). (**c**) 2D SSFP coronal view showed presence of two symmetric superior venae cava (arrows) with no bridging vein. The right superior vena cava drains into the left atrium and to the pulmonary circulation. The left superior vena cava drains into the right atrium and the systemic circulation. The anomalous return of the left superior vena cava causes mix of arterial and venous blood in the morphologically right atrium and is responsible for the desaturation. Legend: *Ao* aorta, *LA* left atrium, *LV*, left ventricle, *RA* right atrium, *RV* right ventricle. Conduit: left ventricle to pulmonary artery conduit

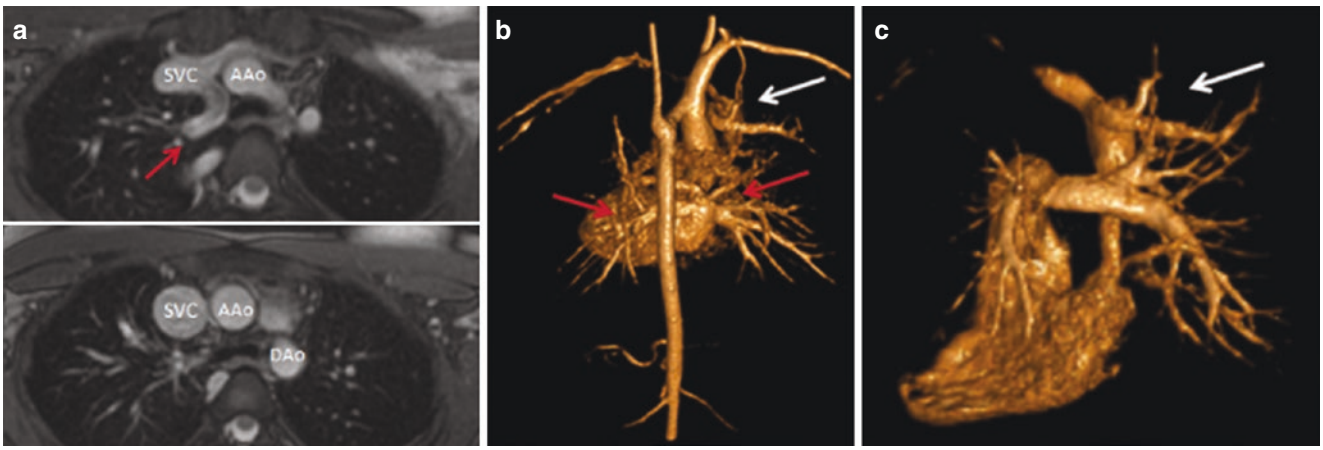



**Fig. 39.6** Anomalous pulmonary venous return. Eighteen-year-old patient operated for aortic coarctation at 6 months of age and treated with percutaneous angioplasty at 5 years of age. She complains of increased fatigability. Cardiac MR shows anomalous pulmonary venous return of a single upper right pulmonary vein to the superior vena cava, associated with mildly hypoplastic aortic arch and descending aorta. (**a**) 2D SSFP image shows the anomalous right upper pulmonary vein draining to the superior vena cava (arrow) which appears dilated (**b**). (**c** and **d**) 3D reconstruction from a contrast-enhanced MR angiography posterior view shows four pulmonary veins entering the left atrium (red arrows) and the superior right upper pulmonary vein draining to the superior vena cava (white arrow). Legend: *AAo* ascending aorta, *DAo* descending aorta, *SVC* superior vena cava

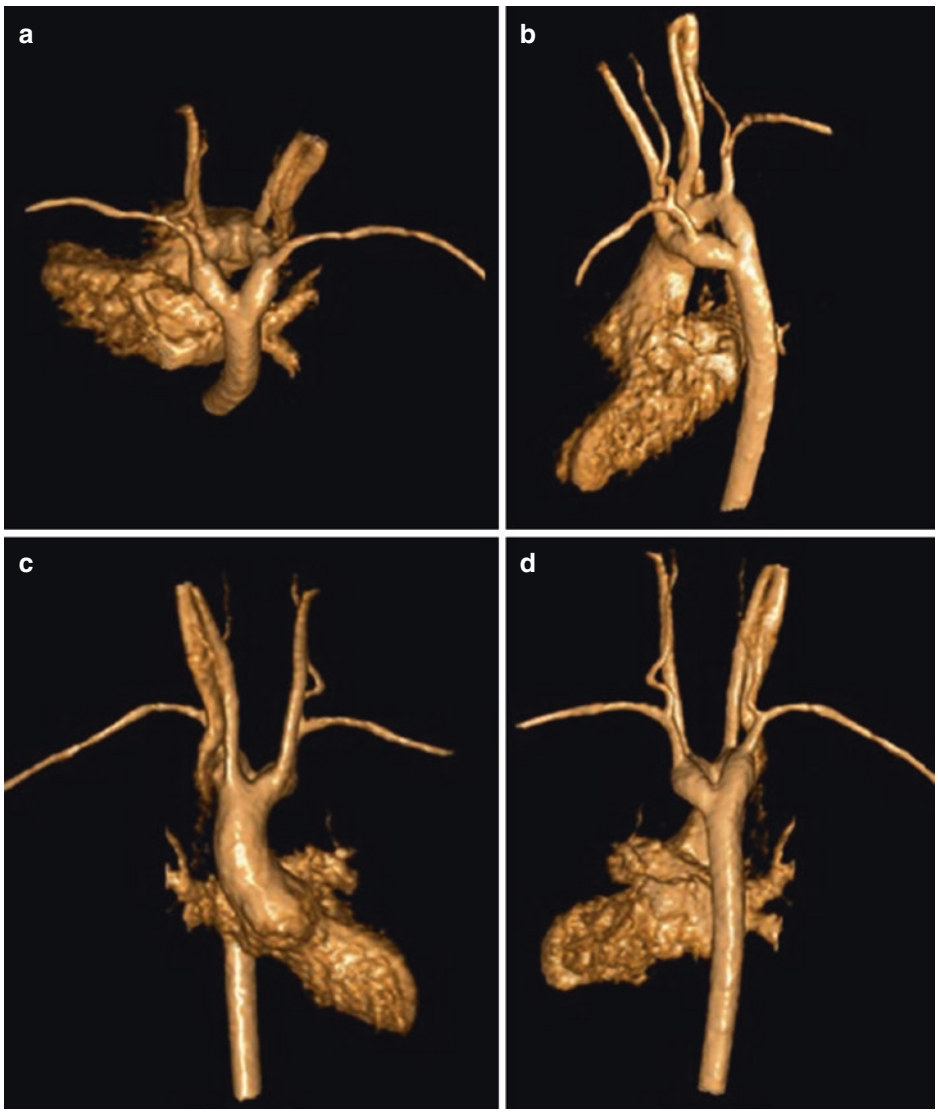



**Fig. 39.7** Double aortic arch. 3D reconstruction from a contrast-enhanced MR angiography in a patient with a double aortic arch. (**a**) Superior oblique view; (**b**) lateral oblique view; (**c**) anterior view; (**d**) posterior view

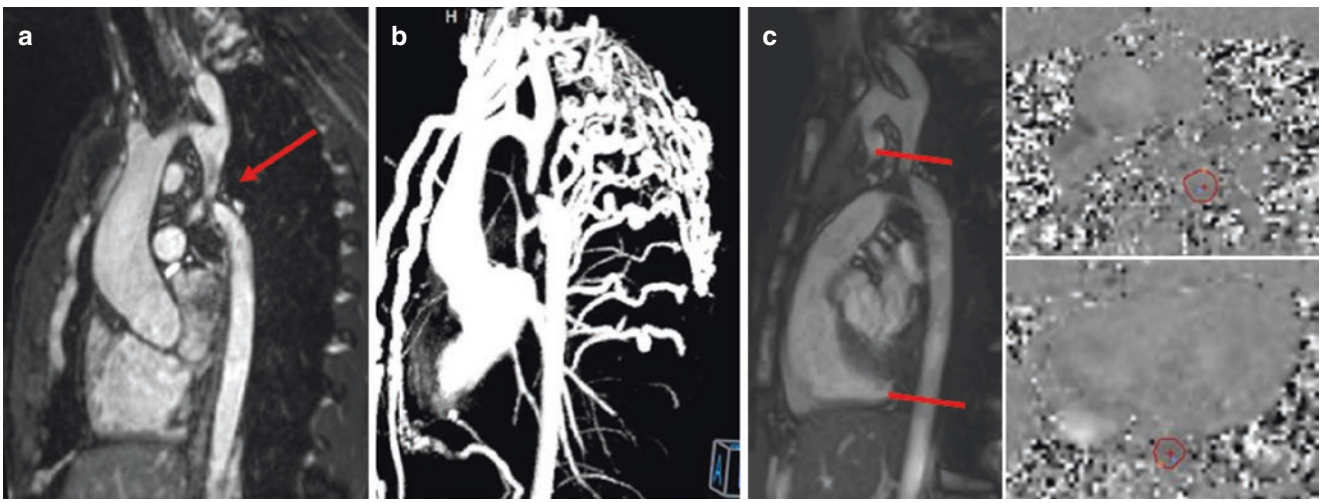



**Fig. 39.8** Patient with unrepaired aortic coarctation. (**a**) 3D SSFP oblique sagittal view showing tight coarctation at the aortic isthmus (arrow). (**b**) Maximum-intensity projection image from the contrast-enhanced MR angiography showing multiple collateral vessels that supply blood to the descending aorta. (**c**) In patients with aortic coarctation, phase contrast MRI is used to calculate the amount of collateral flow by measuring blood flow in the proximal descending aorta before the site of coarctation and in the diaphragmatic aorta. Collateral flow equals the difference between distal and proximal aortic flow

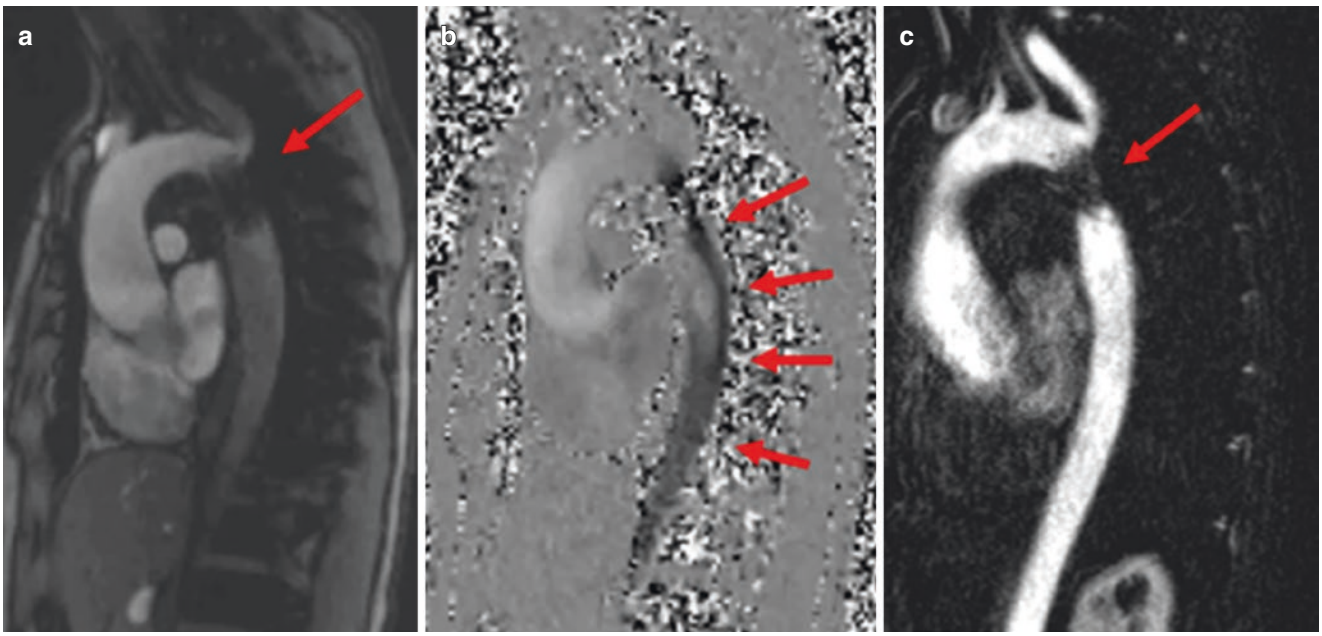



**Fig. 39.9** Patient with repaired aortic coarctation. (**a**) 2D SSFP oblique sagittal view showing the stent at the aortic isthmus (arrow). (**b**) Phase contrast MRI shows flow through the stent (black signal—arrow). (**c**) Maximum-intensity projection image from the contrast-enhanced MR angiography showing the adequate position of the stent in the aortic isthmus

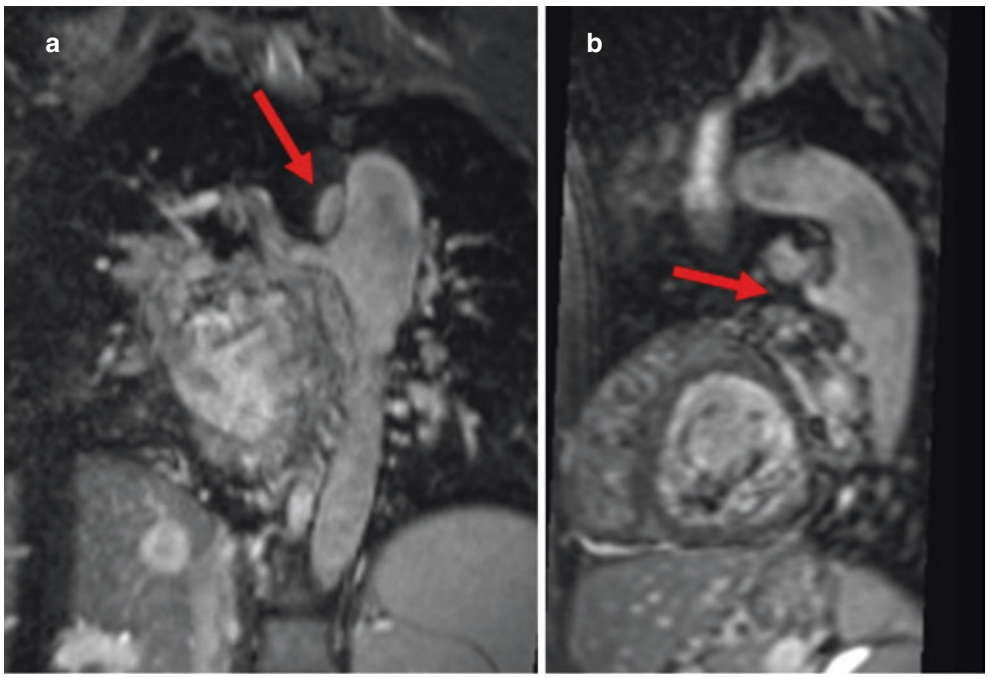
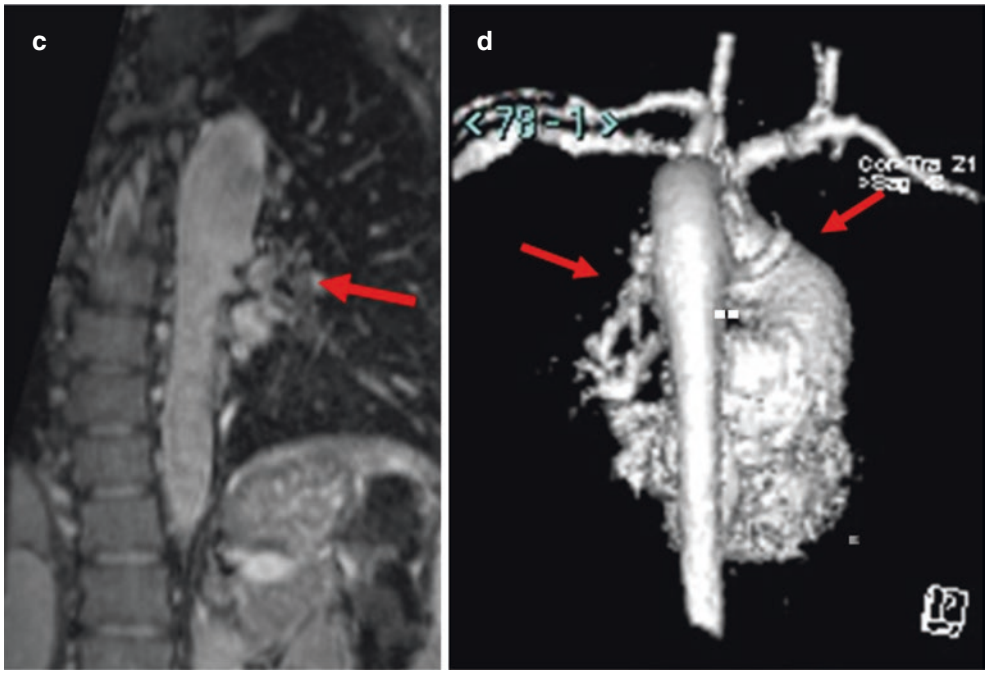



**Fig. 39.10** Major aortopulmonary collateral arteries (MAPCAs). (**a**) Anterior view; (**b**) lateral view; (**c**) posterior right view. All images taken from 3D SSFP data. (**d**) 3D CE-MRA viewed from posterior. The arrows show the MAPCAs

**Fig. 39.10** (continued)



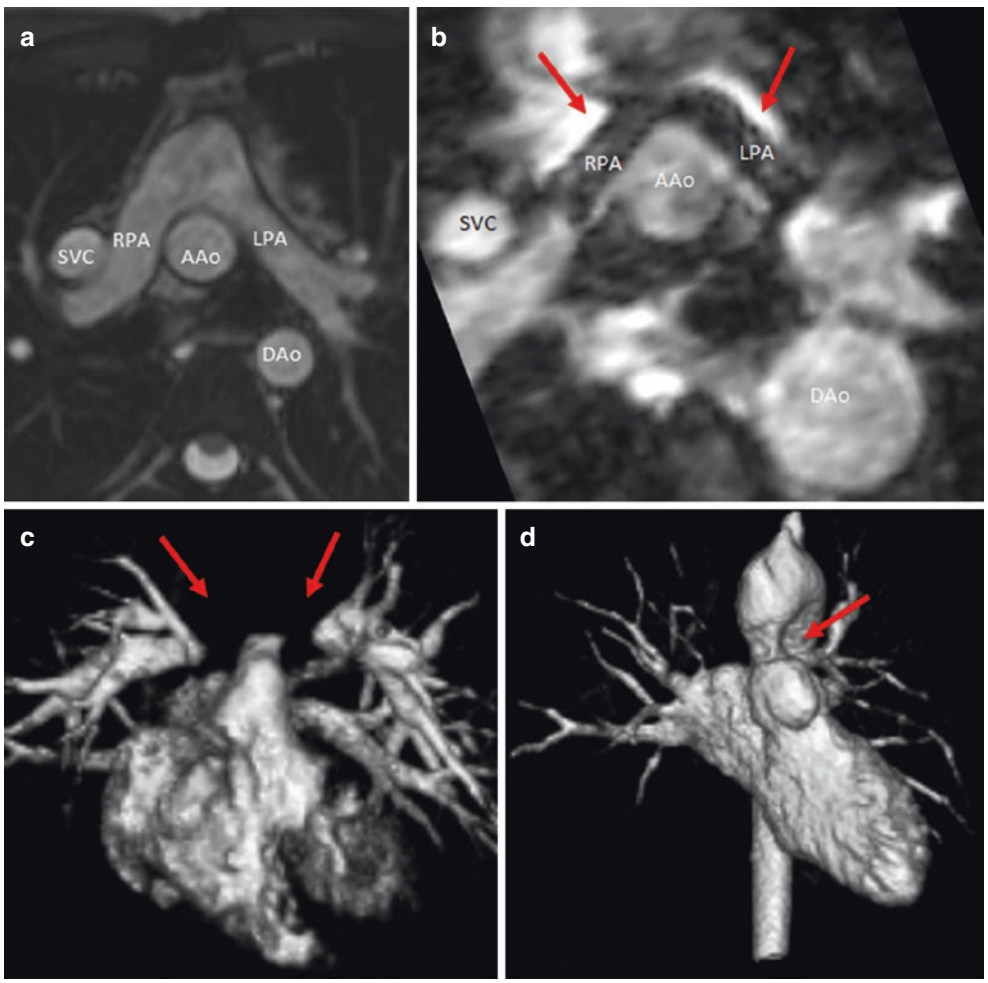




**Fig. 39.11** Transposition of the great arteries, arterial switch operation. (**a**) 2D SSFP axial view shows Lecompte maneuver with the pulmonary artery anterior to the descending aorta (Aao) with the right (RPA) and left (LPA) pulmonary arteries passing either side of the aorta. (**b**) 3D SSFP axial view showing stents in both pulmonary arteries (arrows) in patients who presented with severe pulmonary artery stenosis after arterial switch operation in infancy. (**c**) 3D contrast-enhanced MR angiography shows signal loss (black holes—arrows) inside the stents but well-represented peripheral pulmonary branches. (**d**) 3D contrast-enhanced MR angiography shows narrowing of the ascending aorta (arrows) where the stents passed in front. Legend: *SVC* superior vena cava, *Dao* descending aorta

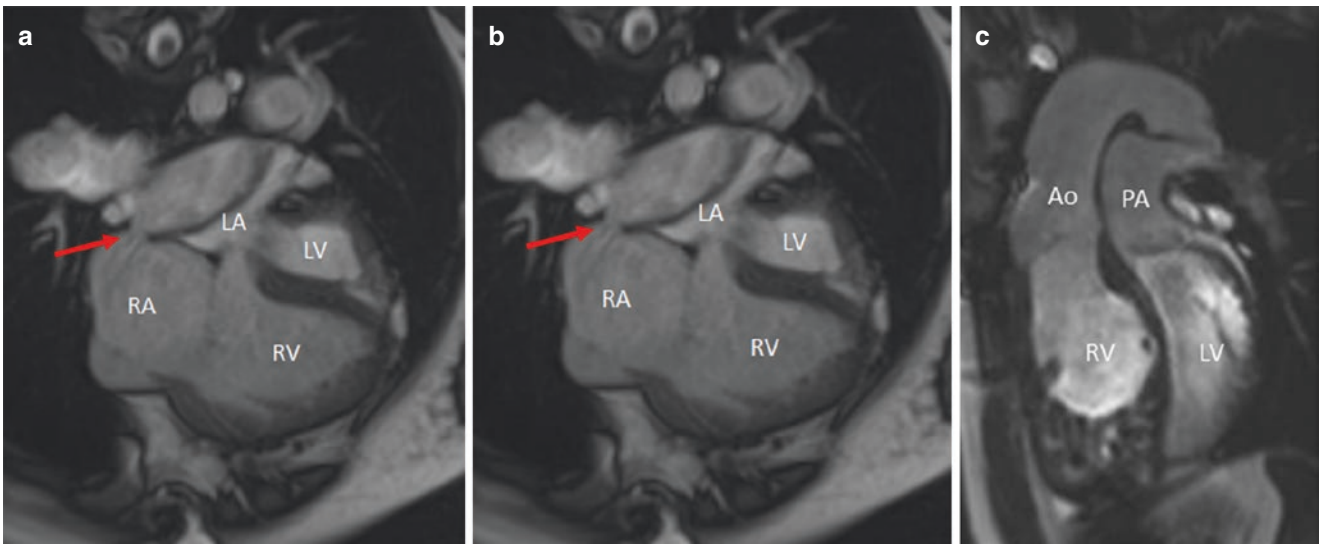



**Fig. 39.12** Transposition of the great arteries, atrial switch operation. (**a**) Oblique coronal view through the pulmonary venous baffle connecting the pulmonary veins to the right atrium and right ventricle; the arrow shows some degree of narrowing of the venous baffle. (**b**) Oblique coronal view through the systemic venous baffle with both the superior and inferior vena cava directed to the left atrium and then to the left ventricle. (**c**) Oblique sagittal view through the outflow tracts showing the aorta arising anteriorly from the right ventricle and the pulmonary trunk posteriorly from the left ventricle

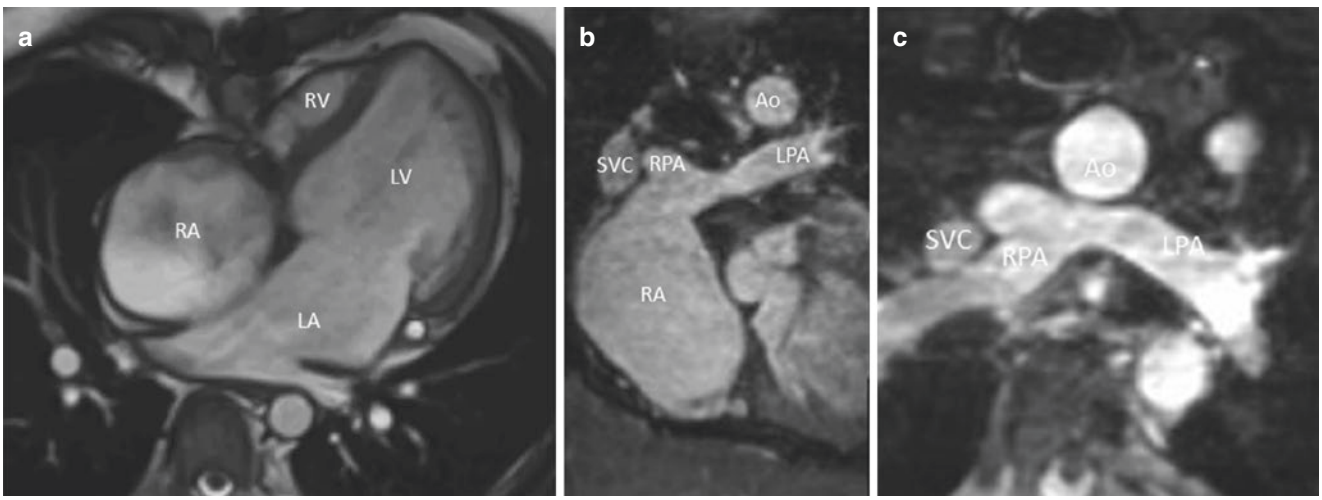



**Fig. 39.13** Atrio-pulmonary Fontan. Fifty-year-old man with tricuspid atresia, ventricular septal defect, and hypoplastic right ventricle who underwent atrio-pulmonary Fontan completion at 10 years of age. (**a**) 2D SSFP image showing the anatomy. Note the severely enlarged right atrium and the hypoplastic right ventricle. (**b**) 3D SSFP reformatted plane oriented to visualize the connection between the atrium and the pulmonary bifurcation. (**c**) 3D SSFP reformatted plane oriented to visualize the pulmonary arteries. Legend: *Ao* aorta, *LA* left atrium, *LPA* left pulmonary artery, *LV* left ventricle, *RPA* right pulmonary artery, *RV* right ventricle, *SVC* superior vena cava

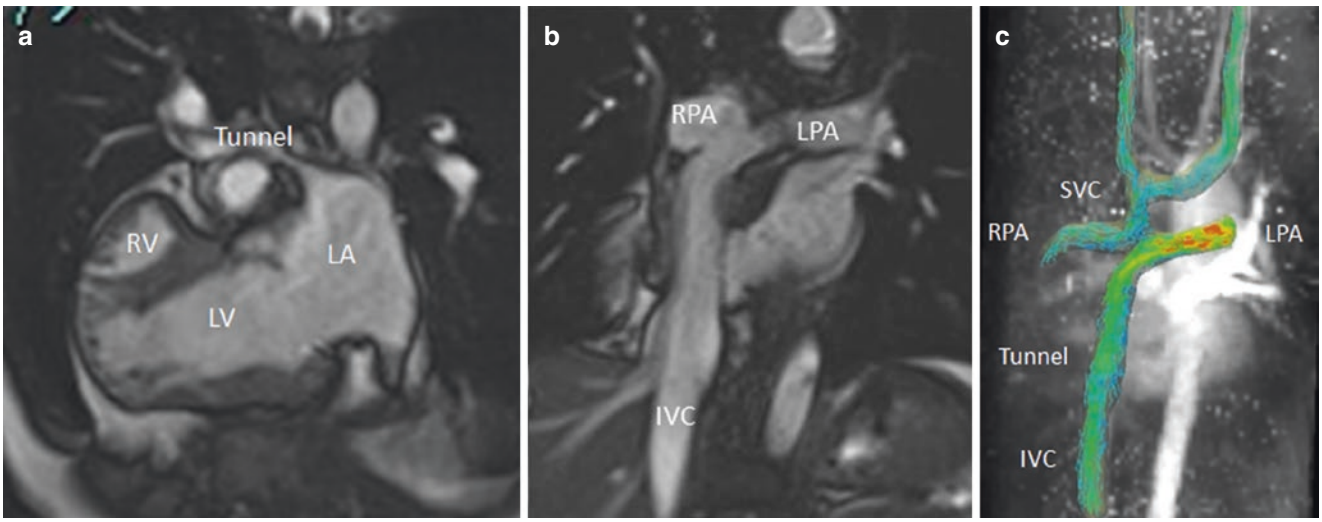



**Fig. 39.14** Atrial tunnel Fontan. Eleven-year-old boy with destrocardia, tricuspid atresia, ventricular septal defect, hypoplastic right ventricle, transposition of the great vessel, and subpulmonary and pulmonary valve stenosis. He underwent Fontan completion with an atrial tunnel at 6 years of age. (**a**) 2D SSFP image showing the position of the heart (destrocardia) and the anatomy. (**b**) 2D SSFP plane oriented to visualize the atrial tunnel from the inferior vena cava to the pulmonary bifurcation. (**c**) 4D flow imaging focused on the Fontan circulation. This innovative technique can accurately simultaneously visualize and quantify blood flow including great vessel flow and venous flow. Using 4D flow imaging, data is acquired as a 3D volume, which obviates the need for planning 2D imaging planes. 4D flow is also able to characterize multidirectional flow patterns furthering our understanding about the complex interaction of flow patterns in Fontan patients. Legend: *IVC* inferior vena cava, *LA* left atrium, *LPA* left pulmonary artery, *LV* left ventricle, *RPA* right pulmonary artery, *RV* right ventricle, *SVC* superior vena cava

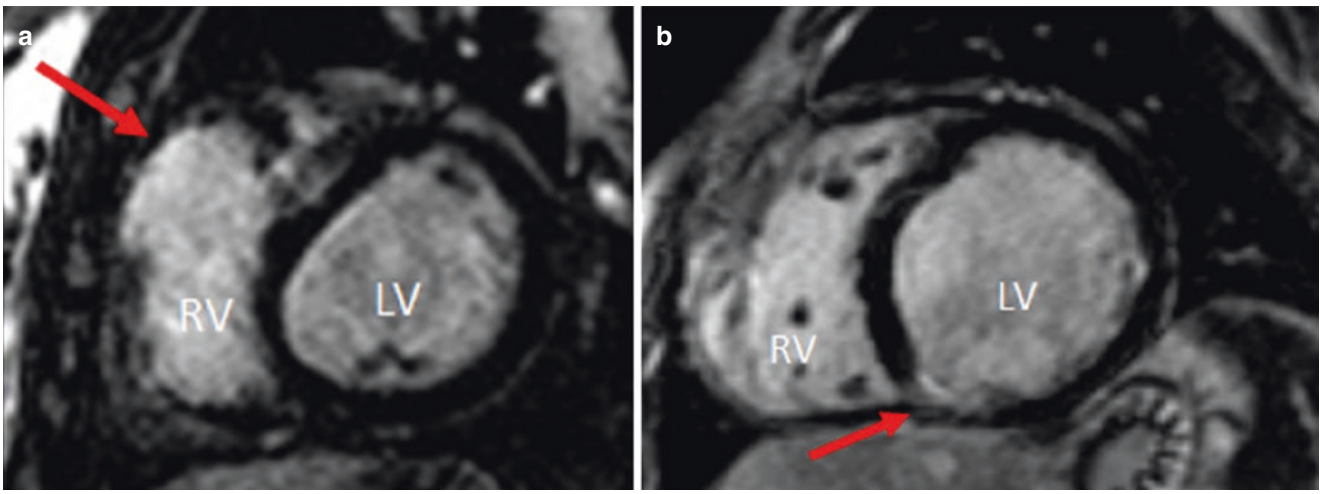



**Fig. 39.15** Myocardial scar. The extent of myocardial scar after surgery for congenital heart disease can be evaluated using the late gadolinium enhancement technique. (**a**) Patient with repair tetralogy of Fallot, late gadolinium enhancement is seen in the location of the right ventricular outflow tract patch (arrow). (**b**) Patient with transposition of the great arteries who underwent arterial switch operation and right coronary artery bypass surgery in infancy. Local transmural delayed enhancement is present in the inferior septal proximal wall (arrow)

**Video 1** Superior vena cava interruption—time-resolved contrast-enhanced MR angiography. Seventeen-year-old boy with transposition of the great arteries and pulmonary artery stenosis. He underwent arterial switch operation as a newborn and a cardiac catheterization to implant a stent in the right pulmonary artery at 9 years of age. Time-resolved contrast-enhanced MR angiography showed complete occlusion of the superior vena cava. Time-resolved contrast-enhanced MR angiography is a real-time dynamic contrast MR angiography to specifically assess contrast flow dynamics in arteries and veins. Rapid sequential images are acquired during the contrast bolus to obtain vascular anatomy and functional information. The contrast injected in a peripheral vein in the right upper arm does not enter the superior vena cava which is closed, but it takes an alternative path into the azygos vein. The azygos vein usually drains toward the superior vena cava. In this case it drains in a craniocaudal direction into the inferior vena cava and the right atrium. From the right atrium, contrast highlights right ventricle and pulmonary arteries. Pulmonary parenchymal perfusion can be evaluated. Contrast returns to the heart through the pulmonary veins, enters the left atrium and left ventricle, and highlights the aorta (AVI 637 kb)

**Video 2** Percutaneous pulmonary valve dislocation. Tetralogy of Fallot patient who underwent correction with transannular patch in infancy, stents in both pulmonary arteries to treat stenosis, and percutaneous pulmonary valve implantation (PPVI) for severe pulmonary regurgitation. Six months after PPVI, he has been evaluated with cardiac MR. On MR the stent of the percutaneous valve appears black. 2D SSFP cine images showed dislocation of the percutaneous pulmonary valve which was floating in the right ventricular outflow tract (MP4 2216 kb)

# CT in Congenital Heart Disease Diagnosis and Transcatheter Treatment

# 40

Andrew Taylor

## 40.1 Introduction

Cross-sectional imaging (cardiovascular MRI and CT) has become a crucial component in the diagnostic pathway in patients with congenital heart disease over the last decade. Their use compliments echocardiography and, for many cases, has superseded the use of diagnostic cardiac catheterisation. Though cardiovascular MRI is still the main form of cross-sectional imaging for CHD, increases in the speed of CT (examinations can be completed in seconds) and reductions in the radiation dose (sub-mSv radiation dose exposure) mean that cardiovascular CT is being increasingly used for the assessment of CHD, in particular in neonates (Figs. 40.1, 40.2, 40.3) and young children (Fig. 40.4) but also in adults (Figs. 40.5, 40.6).

Other areas where cardiovascular CT is predominantly used include investigation of:

- Coronary artery anatomy and structure (Fig. 40.7).
- Vascular rings, where it is important to visualise the airway anatomy (Figs. 40.1, 40.2, 40.3).
- Pulmonary venous anatomy (Figs. 40.8, 40.9). (in particular for the assessment of pulmonary venous stenosis (Fig. 40.10)).
- Pulmonary atresia with major aorta pulmonary collateral arteries (MAPCAs). Assessment prior to cardiac catheterisation can identify the number of large aorta pulmonary collaterals and the presence of any central pulmonary arteries, and this information can be used to guide cardiac catheterisation.
- Patients with stents and metallic implants (Figs. 40.11–40.14, 40.15).
- Patients in whom there is a contraindication to MRI (e.g. older permanent pacemaker).

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_40) contains supplementary material, which is available to authorized users.

A. Taylor (✉)
Cardiovascular Imaging, Great Ormond Street Hospital for Children & UCL Institute of Cardiovascular Science, London, UK
e-mail: a.taylor76@ucl.ac.uk


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_40

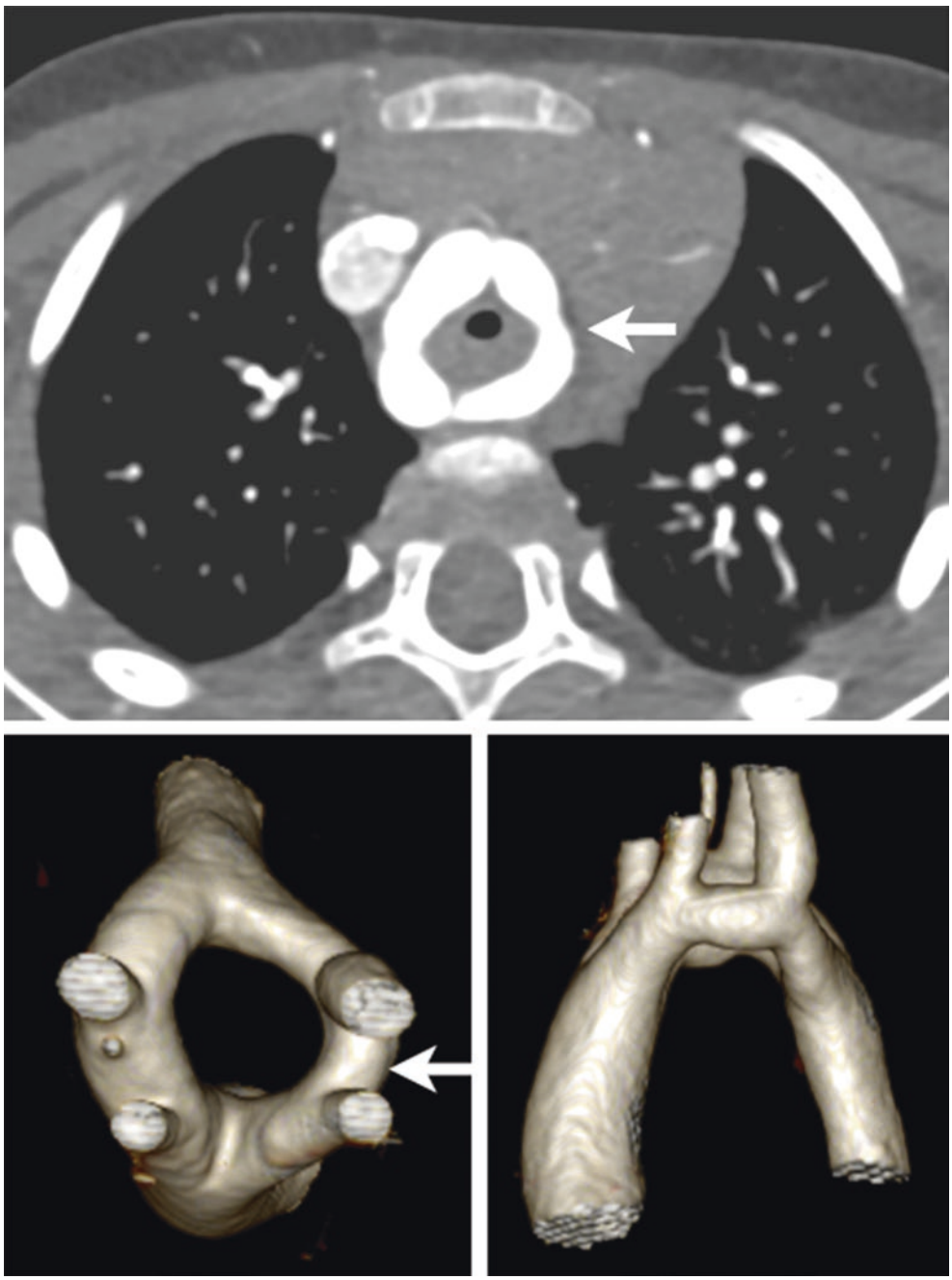


**Fig. 40.1** Double aortic arch in a neonate, contrast-enhanced CT. Top image: axial image through the upper thorax—arrow shows the left arch, trachea (black) within the complete ring. Bottom images: 3D volume-rendered images; left, viewed from above; right, viewed from the left

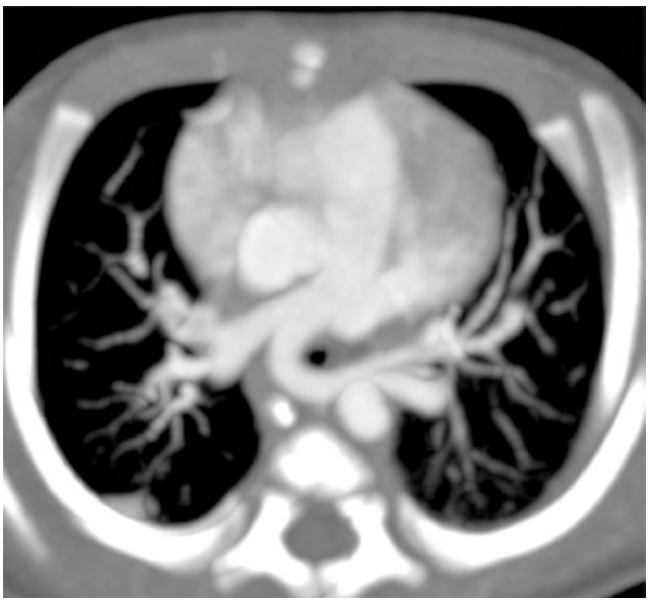


**Fig. 40.2** Left pulmonary artery sling in a neonate, contrast-enhanced CT. The left pulmonary artery passes behind the trachea and anterior to the oesophagus (Note contrast in the nasogastric tube)

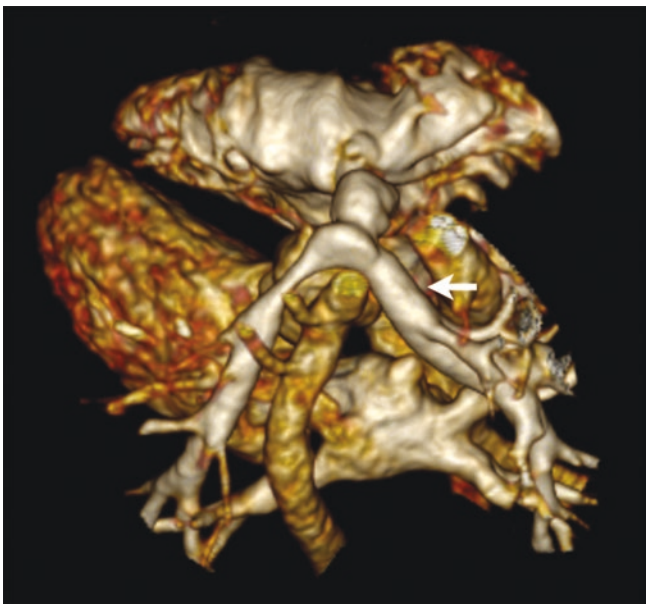


**Fig. 40.4** Transposition of the great arteries following an arterial switch operation in a 6-month-old, contrast-enhanced CT. Oblique axial, 3D volume-rendered image viewed from above, showing the branch pulmonary arteries (arrow shows the left pulmonary artery) passing anterior to the aorta (central structure). Note the proximal right pulmonary artery narrowing

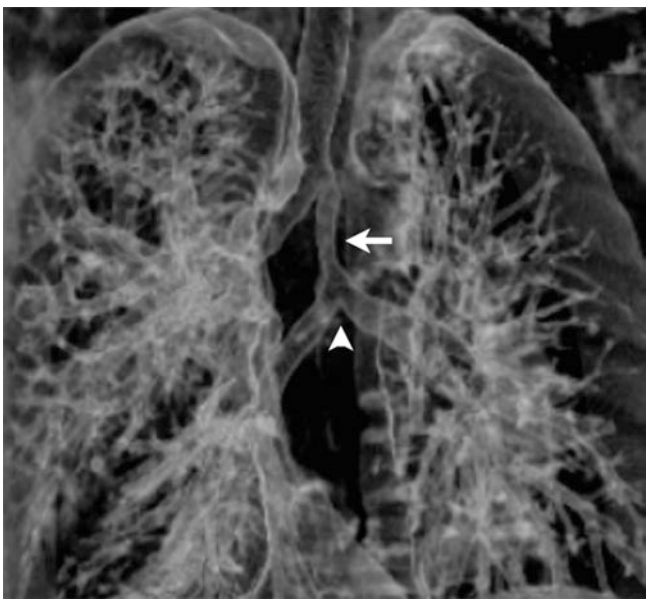


**Fig. 40.3** Left pulmonary artery sling in a neonate, CT. 3D volume-rendered reconstruction of the airways. There is a high right bronchus, a narrowed, 'stove-pipe' trachea (arrow) leading to the true bifurcation (arrowhead)

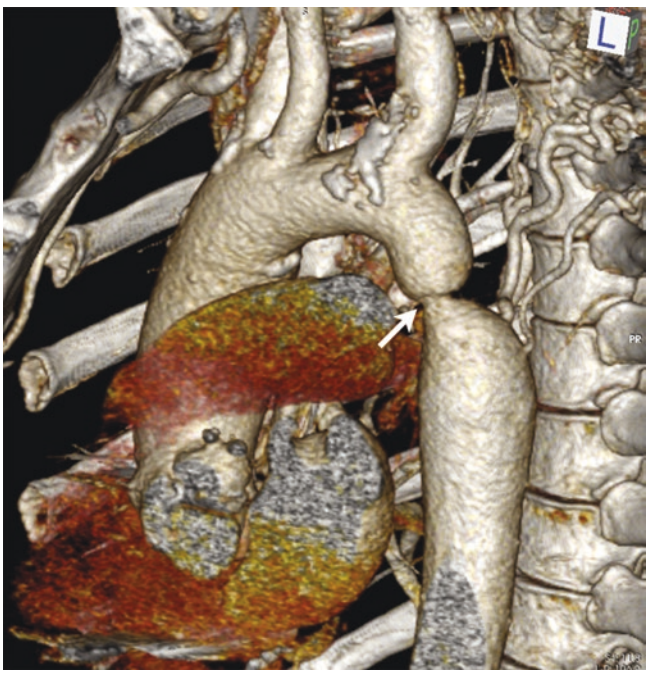



**Fig. 40.5** Coarctation of the aorta in a 17-year-old, first presentation, contrast-enhanced CT. 3D volume-rendered image, viewed from the left lateral aspect showing a tight native coarctation (arrow). Note multiple large collateral vessels around the site of the coarctation. CT imaging can define the optimal treatment option, stent diameter and length and risks associated with potential collateral damage

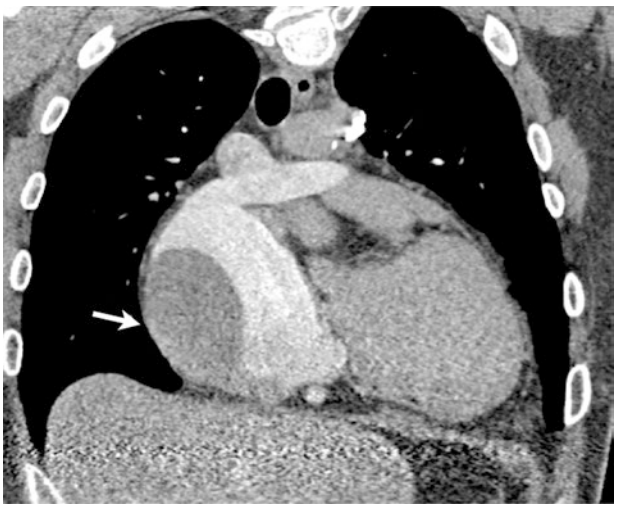


**Fig. 40.6** Classical Fontan procedure in a 43-year-old, contrast-enhanced CT. Coronal view showing large, chronic right atrial thrombus (arrow)

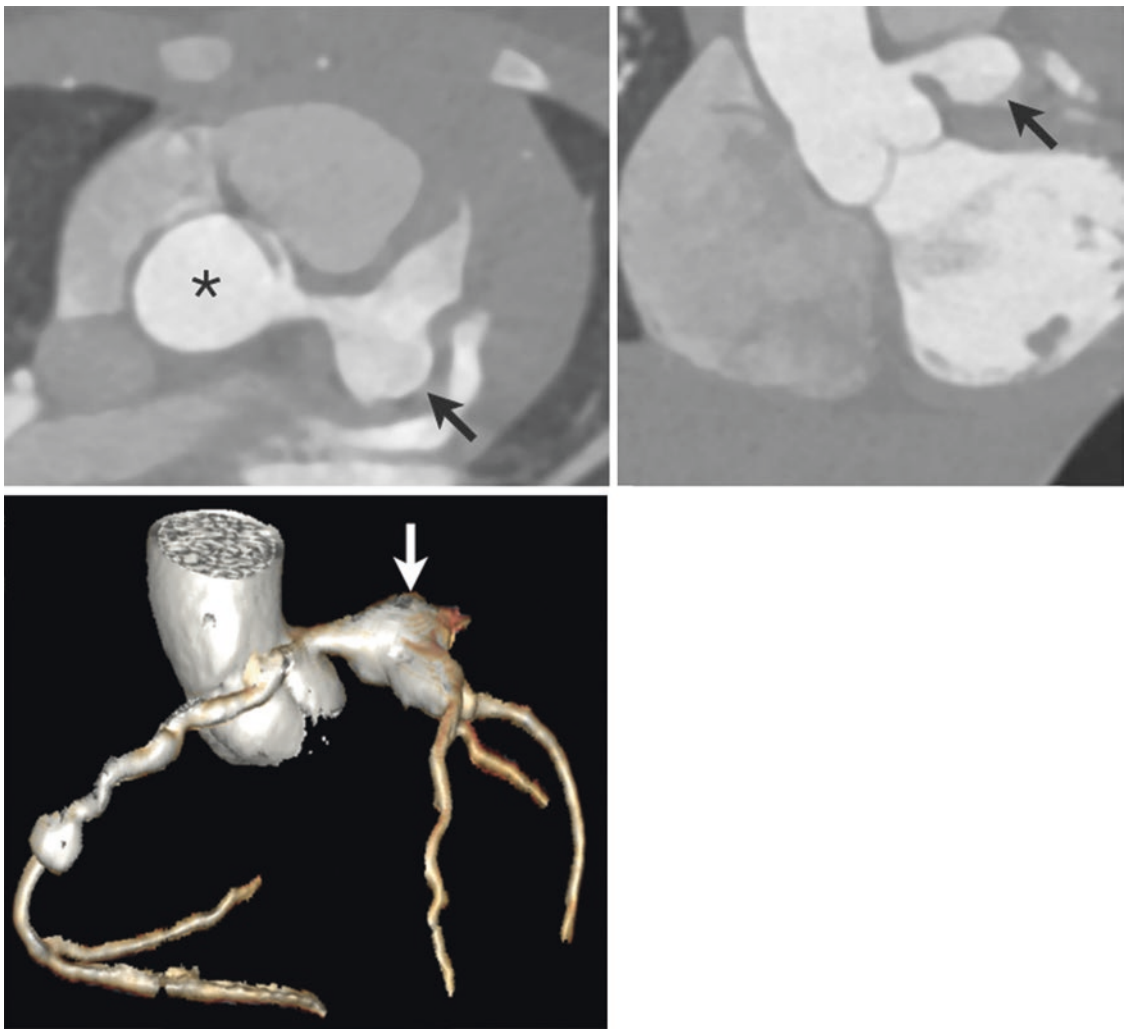



**Fig. 40.7** Kawasaki disease with giant left coronary artery aneurysm in a 7-year-old, contrast-enhanced CT. Upper right image (axial view) shows a patent, giant aneurysm of the left main stem (LMS) coronary artery (arrow), aortic root (*). Upper left image shows the LMS in an oblique coronal view. Lower left image shows a 3D volume-rendered image of the aortic root and proximal coronary arteries. Note the giant LMS aneurysm (arrow) and smaller aneurysmal dilatations in the RCA

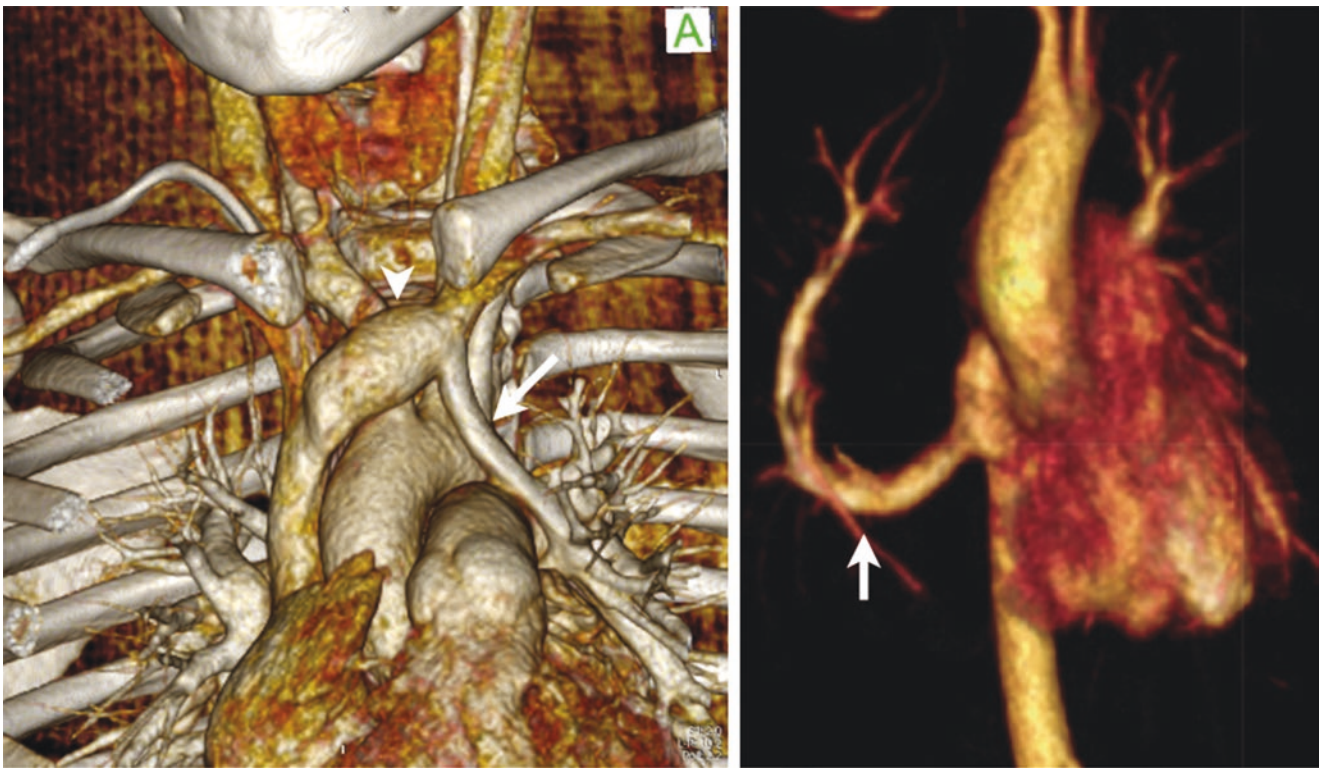



**Fig. 40.8** Partial anomalous pulmonary venous drainage. Left image, 3D volume-rendered image, viewed from anterior, of a left upper pulmonary vein (arrow) draining into the brachiocephalic vein (arrowhead) in a 32-year-old, contrast-enhanced CT. Right image, 3D volume-rendered image, viewed from anterior, of a right lower pulmonary vein (arrow) draining into the inferior caval vein in a 10-year-old, contrast-enhanced CT

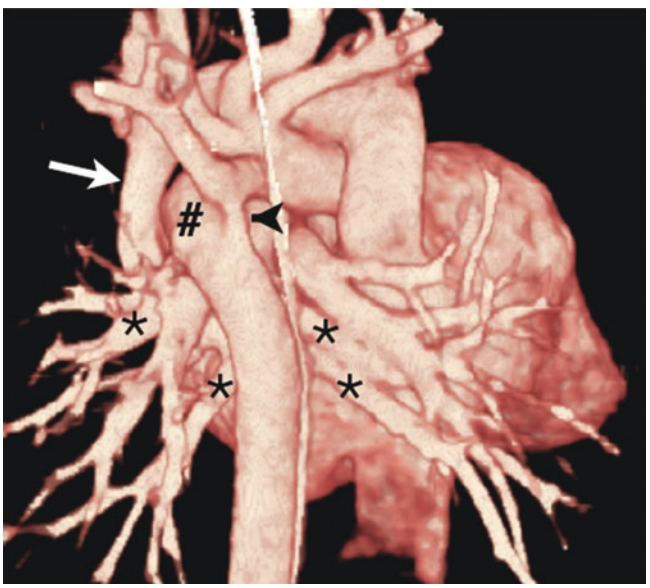


**Fig. 40.9** Supra-cardiac total anomalous pulmonary venous drainage in a neonate, contrast-enhanced CT. 3D volume-rendered image, viewed from posterior, shows the four pulmonary veins (*) coming together behind to form a single ascending vein on the left that drains into the brachiocephalic vein. Note also the dextrocardia, the aortic coarctation (arrowhead), the large patent ductus arteriosus (#) that continues to the descending aorta and the contrast-filled nasogastric tube

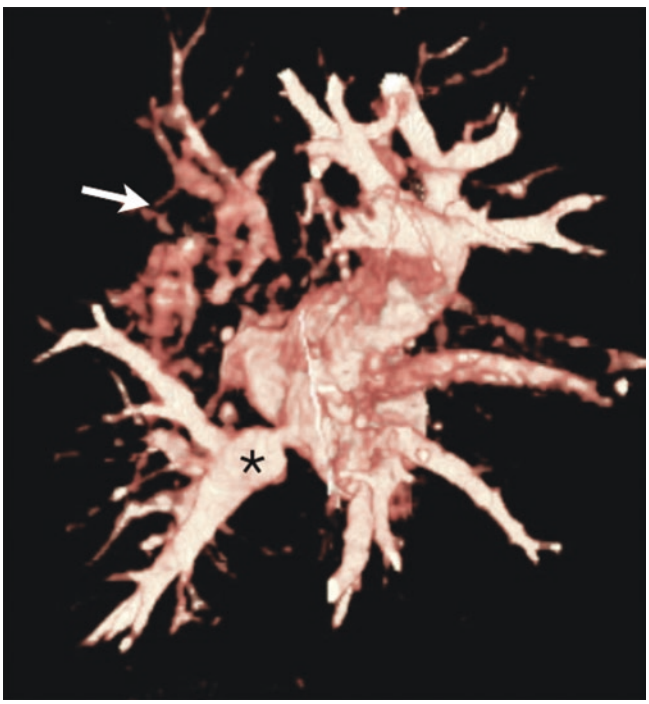


**Fig. 40.10** Pulmonary vein stenosis in a 9-month-old, contrast-enhanced CT. 3D volume-rendered image, viewed from posterior, of the pulmonary veins showing a discrete left lower pulmonary vein stenosis (*). The arrow shows the faint outline of the peripheral left upper pulmonary veins due to complete occlusion of the left upper pulmonary vein

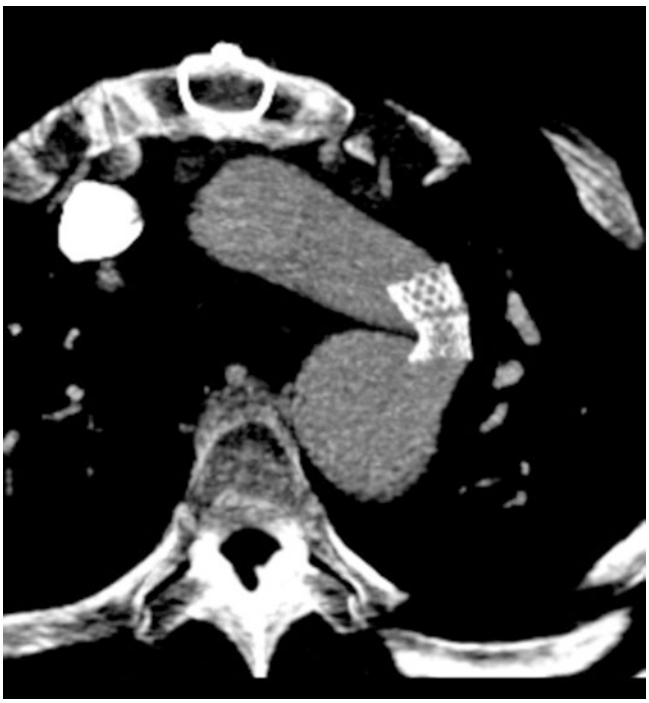


**Fig. 40.11** Stented aortic coarctation in a 25-year-old, contrast-enhanced CT. Axial image in the upper thorax shows a stent fracture with a degree of restenosis at the site of previously treated aortic coarctation

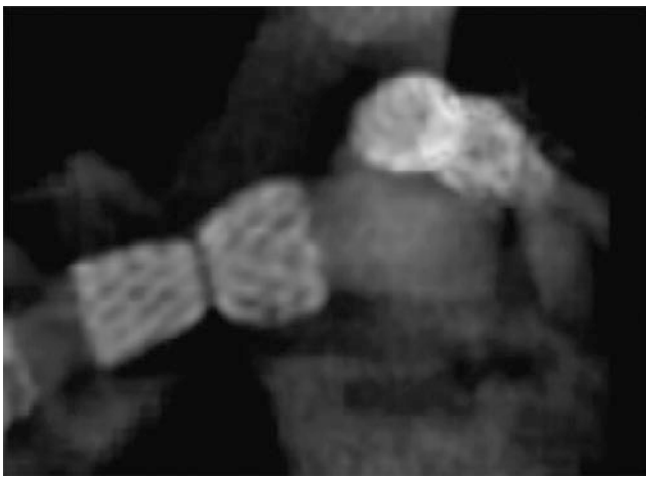


**Fig. 40.12** Bilateral pulmonary artery stenting for branch pulmonary artery stenosis in a 4-year-old, contrast-enhanced CT. Coronal image viewed from the front of a right pulmonary artery fractured stent with pulmonary artery restenosis. The left pulmonary artery stent is seen end on and is seen to be narrowed

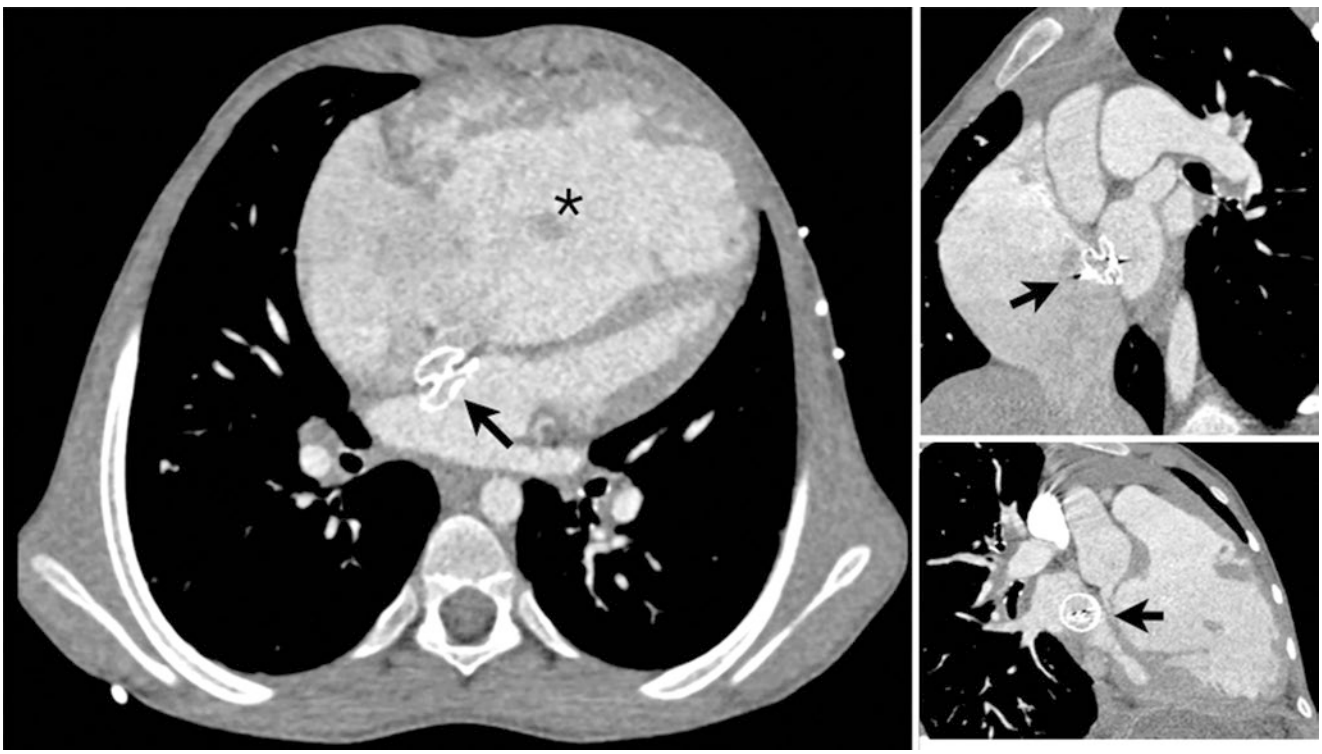


**Fig. 40.13** Atrial septal defect device in a 30-year-old, contrast-enhanced CT. Left image shows an axial four-chamber view. ASD device seen in atrial septum (arrow). Note the marked dilatation of the right ventricle (*) and the right atrium. Right upper image shows an oblique sagittal view, ASD device (arrow) and a dilated right atrium. Right lower image shows an oblique coronal view of the ASD device en face

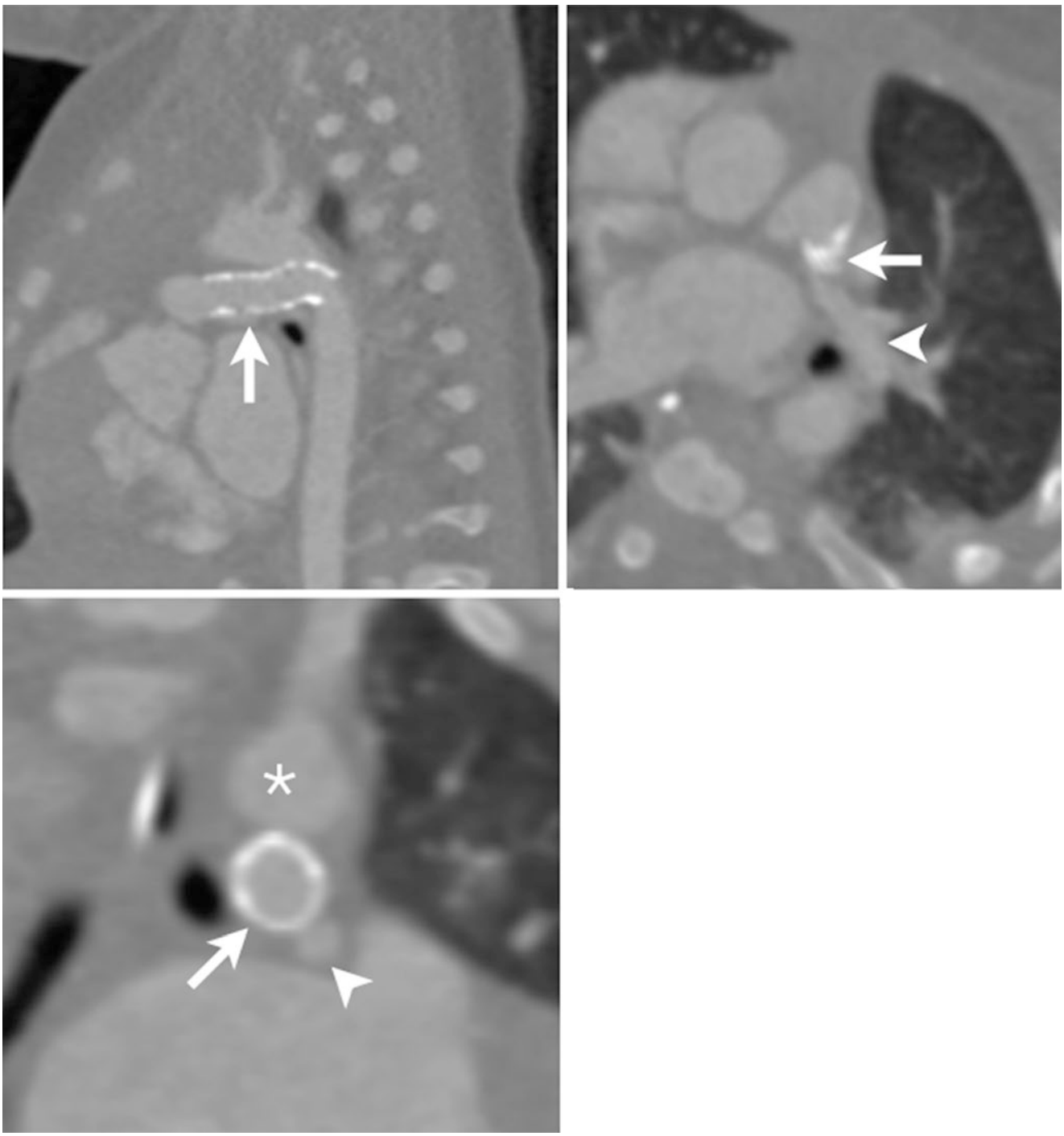


**Fig. 40.14** Proximal left pulmonary artery compression following patent ductus arteriosus (PDA) stenting in a neonate, contrast-enhanced CT. Upper right image (sagittal view) shows the widely patent stent in the PDA (arrow). Upper left image (axial view) shows the left pulmonary artery (LPA—arrowhead). Note the proximal narrowing and stent artefact (arrow). Lower left image (coronal view) shows the significantly narrowed LPA (arrowhead) inferior to the PDA stent (arrow), which lies inferior to the aortic arch (*)

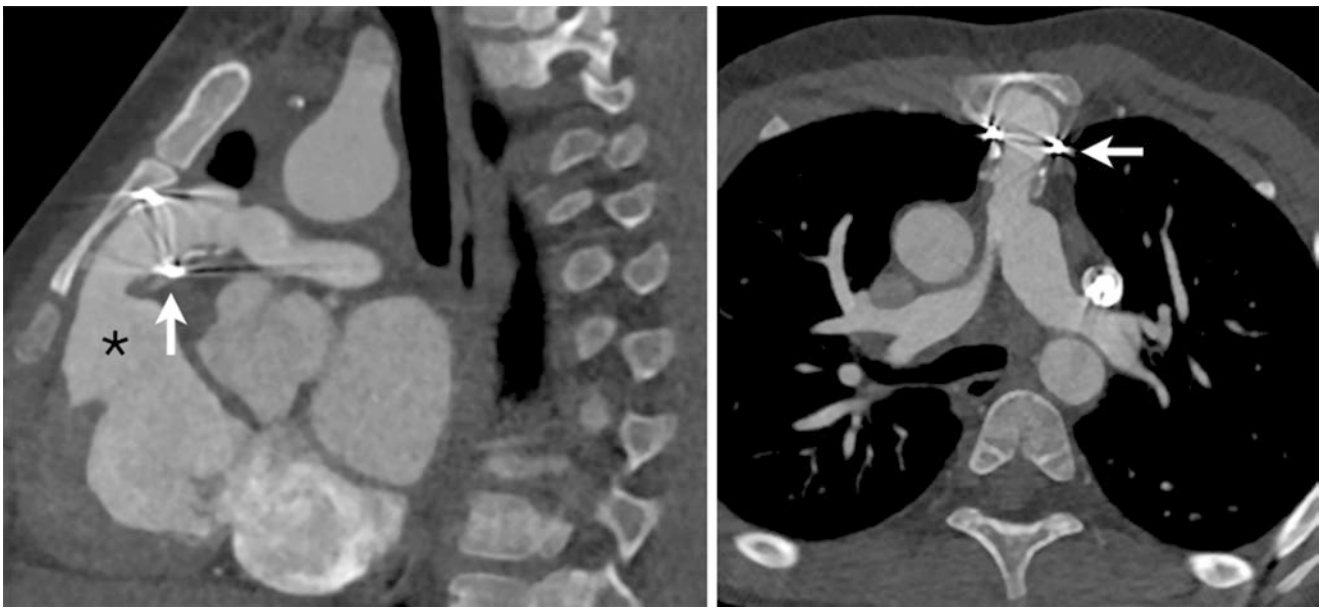


**Fig. 40.15** Erosion of a 'Hancock' pulmonary conduit through the sternum in a 25-year-old, contrast-enhanced CT. Right image (coronal view); left image (axial view)—'Hancock' pulmonary valve metal ring artefact (arrow), right ventricular outflow tract (*). Note the proximal right pulmonary artery narrowing in the right image

**Video 1** Unrepaired tetralogy of Fallot in a neonate, contrast-enhanced CT. Axial slices through the thorax and upper abdomen, starting in the neck. (1) Left-sided aortic arch with normal branching pattern; (2) tortuous patent ductus arteriosus (PDA) from the left, inferior surface of the aortic arch passing to the mid-left pulmonary artery (LPA); (3) proximal LPA stenosis; (4) small pulmonary valve; (5) clockwise rotation of the origins of the coronary arteries; (6) right ventricular outflow tract narrowing; (7) ventricular septal defect (VSD); (8) atrial septal defect (ASD) (MOV 29372 kb)

**Video 2** Dynamic 3D volume-rendered reconstructions of the right ventricular outflow tract, pulmonary trunk and proximal branch pulmonary arteries, all viewed from anterior. Images from 12 patients all with an original diagnosis of tetralogy of Fallot, treated with complete repair in infancy and then imaged 15 to 20 years later. Note the wide range of anatomies and dynamic motion. Imaging useful for pulmonary valve replacement, in particular for selecting if patients are suitable for percutaneous pulmonary valve implantation (MOV 18043 kb)

**Video 3** Cine contrast-enhanced CT, acquired during prospective gating. 35-year-old with tetralogy of Fallot, assessment for severe pulmonary regurgitation. Right upper image—four-chamber view of the heart; left upper image, basal short-axis view; right lower image, right ventricular outflow tract view; left lower image, mid-short-axis view. Note patient not suitable for cardiac MRI as permanent pacemaker in situ—pacing wire artefact seen on all images (MOV 14060 kb)

**Video 4** Anomalous left coronary artery from the pulmonary artery (ALCAPA) in a 17-year-old, contrast-enhanced CT. Rotation of 3D volume-rendered image. Note large right coronary artery, with large collateral vessels that cross the right ventricular outflow tract. The left coronary artery arises from the posterior left side of the pulmonary artery (MOV 26559 kb)

**Video 5** Transposition of the great arteries with an atrial switch operation in a 38-year-old, contrast-enhanced CT. The right upper image (axial view) shows the origin of the left coronary artery (LCA) from the posterior aortic sinus. The LCA passes directly between the aorta and the pulmonary trunk. Right lower image shows the normal course of the right coronary artery, which arises from the anterior aortic sinus (not shown). Left image, sagittal cine of the outflow tracts (aorta anterior, pulmonary trunk posterior). Note the dynamic compression of the LCA between the two great vessels (MOV 14635 kb)

**Video 6** Cine sagittal image of the right ventricular outflow tract (RVOT) in a 12-year-old, contrast-enhanced CT. The patient had been previously treated with a pulmonary artery stent and percutaneous pulmonary valve implantation. Encroachment of the stents into the dynamic RVOT had caused stent fracture and dynamic RVOT obstruction (MOV 8373 kb)

**Video 7** Dynamic 3D volume-rendered reconstruction of the right ventricular outflow tract, pulmonary trunk and proximal branch pulmonary arteries, viewed from anterior. The cine image shows the first-in-man implantation of the Medtronic Harmony(TM) Transcatheter Pulmonary Valve (MOV 11953 kb)

**Video 8** Stented aortic coarctation in a 25-year-old, contrast-enhanced CT. 3D volume-rendered image that has then been rotated to show the narrowing of the coarctation stent, the relationship of the stent to the origin of the left subclavian artery and a small pseudoaneurysm at the distal end of the stent. These images were used to plan for the insertion of a covered stent to cover the mouth of the pseudoaneurysm and relieve the re-coarctation whilst ensuring that the origin of the left subclavian artery was not covered (MOV 20004 kb)

# Intracardiac Echocardiography

# 41

Jason H. Anderson and Allison K. Cabalka

Intracardiac echocardiography (ICE) serves as an attractive adjunct for the structural and congenital interventionalist. The experience to date has been primarily obtained utilizing the AcuNav system (Boston Scientific, San Jose, California) consisting of a 64-element phased array transducer with steerable handle and locking knob utilizing a frequency range of 5–10 MHz with a tissue penetration of around 15 cm. The catheter is available in 8 and 10 French single-use designs. As with all intraprocedural imaging modalities, there are several advantages and disadvantages to the utilization of ICE imaging for procedural guidance.

Advantages of ICE:

- Excellent near-field imaging with high resolution (Figs. 41.1–41.8)
- Catheter manipulation performed by the interventionalist
- Freely torquable system for image acquisition through multiple different planes of the heart
- Minimal invasiveness obviating the need for general anesthesia in the older patient
- Reduction in procedural and fluoroscopy time
- Ease (over TEE) of imaging the inferior atrial septum near the IVC with visualization of the posterior/inferior atrial septal rim (Fig. 41.9)
- Imaging of anterior cardiac structures when prosthetic valve interference limits TEE assessment (Figs. 41.10–41.12)

Disadvantages of ICE:

- Limited depth of tissue penetration.
- Lack of adequate resolution with 3D imaging.
- Single-use design increasing the cost associated with use (albeit this may be offset by the reduced personnel required or, in some centers, by resterilization and reuse of the probe).
- Limited Doppler capability.
- Specific training is essential to become confident with ICE imaging.

The utilization of intraprocedural ICE has led to improved safety and reduction in fluoroscopic time during complex procedures. This system increases the accuracy of placing percutaneous closure devices, performing transatrial punctures, and evaluation of transcatheter valves post-deployment. Furthermore, it provides a real-time assessment of complications making it a useful adjunct for interventional procedures.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_41) contains supplementary material, which is available to authorized users.

J. H. Anderson · A. K. Cabalka (✉)
Division of Pediatric Cardiology, Mayo Clinic,
Rochester, MN, USA
e-mail: cabalka.allison@mayo.edu


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_41

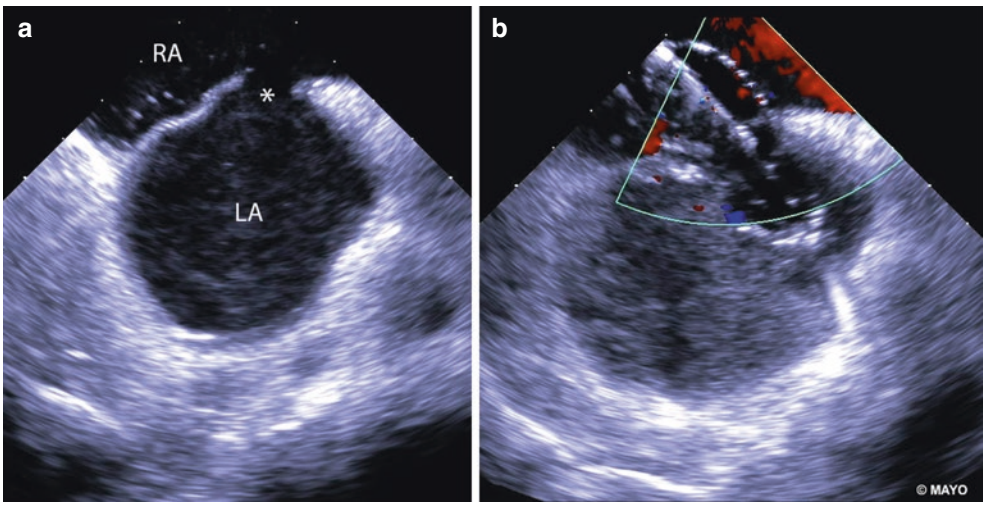



**Fig. 41.1** Patient 1: 65-year-old male (75 kg) with TOF repaired in 1957; now with a multi-fenestrated atrial septum. There are two secundum atrial septal defects; an anterior/superior defect and posterior/inferior defect with stretched diameters of 18 mm and 12 mm, respectively. (**a**) Static, unstretched anterior/superior defect [asterisk]. (**b**) Balloon sizing with color flow Doppler and stop flow technique of the anterior/superior secundum atrial septal defect. The stretched diameter of the defect is 18 mm with ICE measurements correlating with fluoroscopic measurements

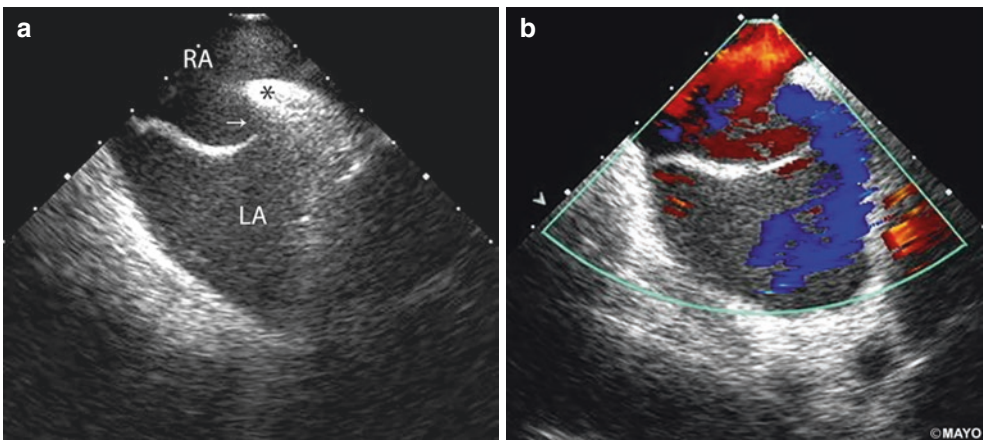
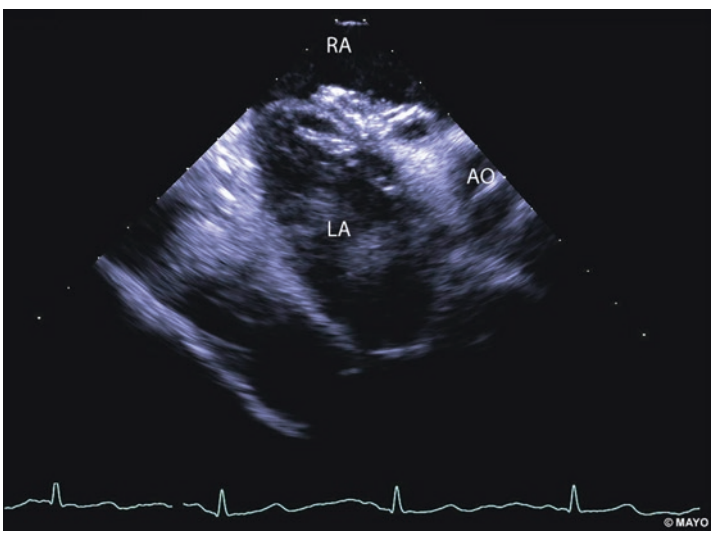



**Fig. 41.2** Patient 2: 65-year-old male (88 kg) with history of thromboembolic stroke and PFO. (**a**) The atrial septum is aneurysmal with the thin, redundant septum primum bowing toward the left atrium. There is a large PFO [arrow] (10 mm static, unstretched diameter) with a thick superior limbus [asterisk] of the septum secundum. (**b**) Color flow Doppler evaluation demonstrates a right-to-left shunt [blue flow] at the atrial level across the PFO

**Fig. 41.3** Patient 3: 55-year-old male (90 kg) with history of cryptogenic stroke with an atrial septal aneurysm and PFO with bidirectional shunt. Balloon sizing demonstrated a stretched PFO diameter of 5 mm with a 5 mm limbus. A 35 mm GORE® HELEX® Septal Occluder (W.L. Gore & Associates Inc., Newark, DE), the precursor to the CARDIOFORM® device, was placed with good apposition of the right atrial disc and left atrial disc. There was no distortion of the aortic root and no residual shunt on color flow Doppler evaluation



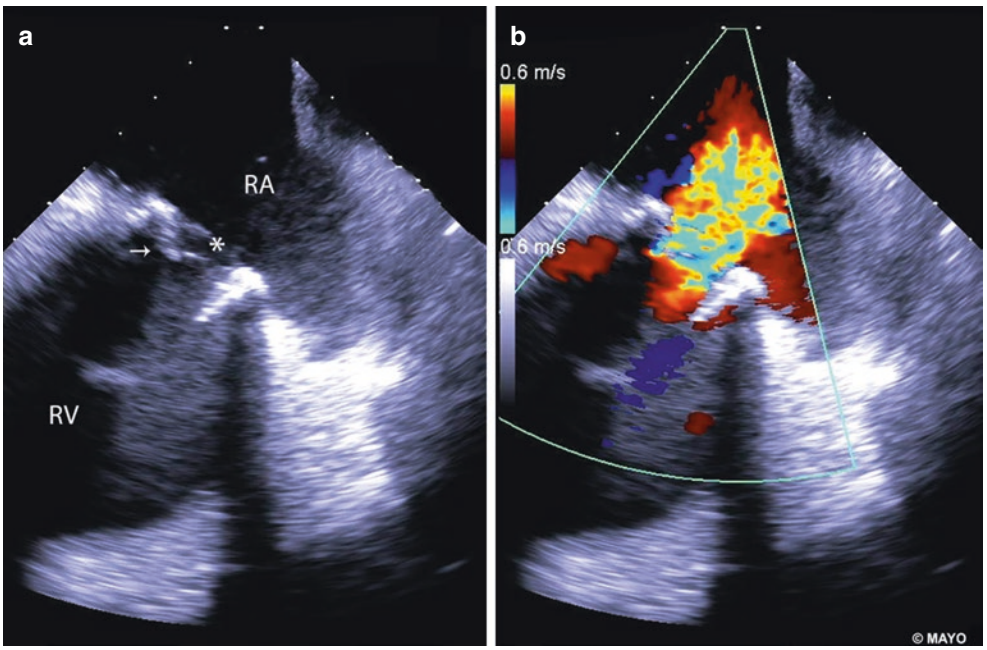



**Fig. 41.4** Patient 6: 17-year-old female (51 kg) with Ebstein's anomaly and tricuspid valve replacement in 2010; S/P tricuspid valve re-replacement with a 23 mm Medtronic Hancock II bioprosthetic valve [asterisk] (Medtronic, Minneapolis, MN) and bidirectional cavopulmonary anastomosis. (**a**) The bioprosthetic valve leaflets appear thickened [arrow] with mild-moderate stenosis (mean diastolic inflow gradient = 9 mmHg). (**b**) There is severe valvular regurgitation demonstrated via color flow Doppler with no paravalvular/intravalvular leak

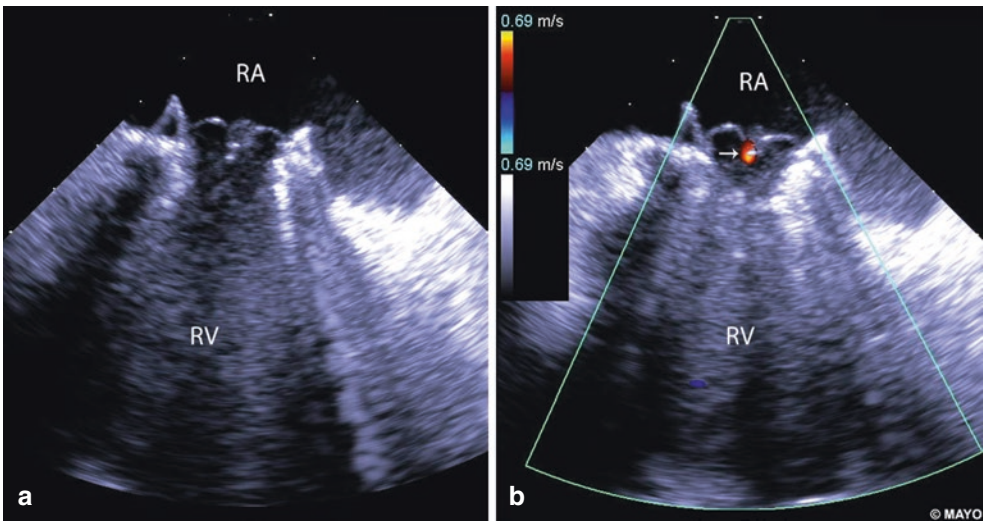



**Fig. 41.5** Patient 6: A 22 mm Melody® transcatheter heart valve (Medtronic, Minneapolis, MN) has been placed within the existing bioprosthesis in the tricuspid valve position. (**a**) The Melody® valve stent is well positioned. The three leaflets of the Melody® valve are easily visualized with normal mobility and normal coaptation. Due to deep coaptation of the bovine jugular vein, the leaflets appear relatively proximal in the valve stent. (**b**) Trivial regurgitation [arrow] is present with no stenosis (mean diastolic Doppler inflow gradient = 5 mmHg) or paravalvular/intravalvular leak

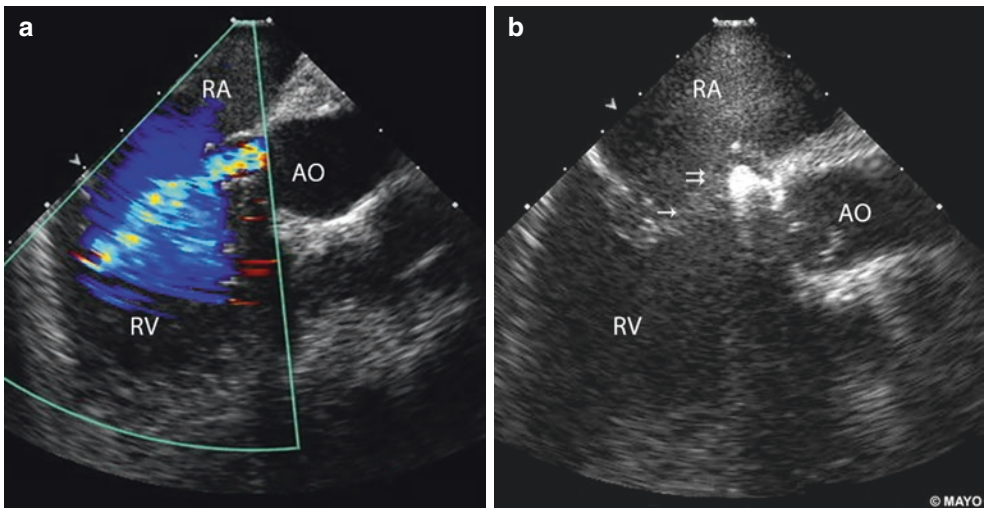



**Fig. 41.6** Patient 7: 27-year-old male (37 kg) with rupture of a sinus of Valsalva aneurysm and fistula formation from the posterior, noncoronary cusp to the right atrium. (**a**) A large tissue membrane is present at the fistula site prolapsing into the right atrium. (**b**) This defect was closed utilizing a 6/4 AMPLATZER™ Duct Occluder device [double arrow] (St. Jude Medical, Minneapolis, MN) with no impingement of the tricuspid valve septal leaflet [arrow]

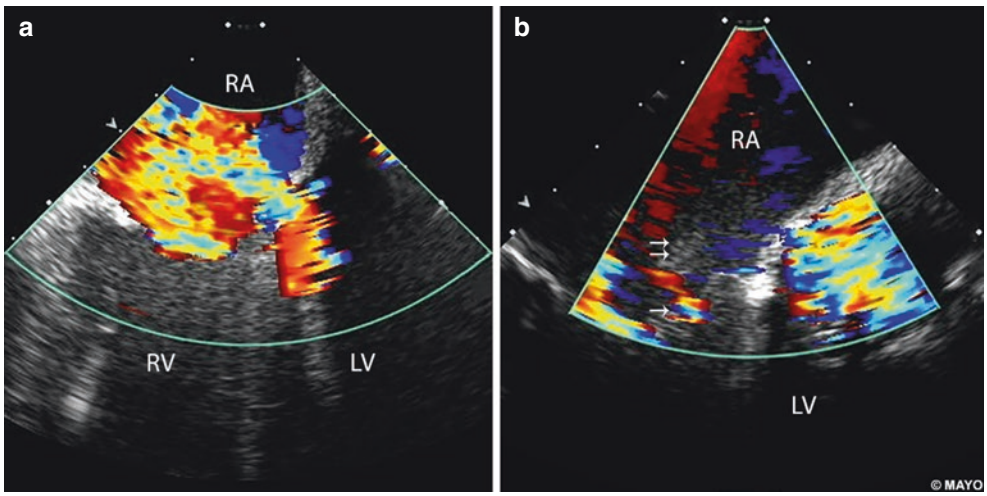



**Fig. 41.7** Patient 8: 70-year-old male (82 kg) with a LV to RA ventriculoatrial defect [Gerbode defect]; S/P AVR and root reconstruction in 2003 with a residual, iatrogenic ventriculoatrial shunt. ICE imaging is obtained from the right atrium. (**a**) There is holosystolic shunting through a 7 mm orifice. (**b**) This defect was successfully occluded in the cath lab utilizing a 12/10 mm AMPLATZER™ Duct Occluder device [double arrow] (St. Jude Medical, Minneapolis, MN) with a trivial residual shunt [arrow]

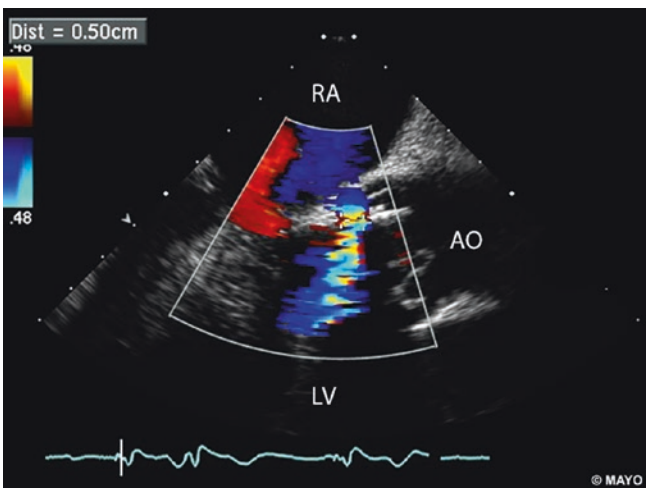



**Fig. 41.8** Patient 9: 67-year-old male (51 kg) with history of aortic valve replacement with a 21 mm Medtronic Mosaic tissue valve (Medtronic, Minneapolis, MN) in 2012, now with paravalvular regurgitation. ICE imaging is obtained from the right atrium demonstrating a posterior paravalvular defect measuring 5 mm in size. This defect was successfully occluded utilizing an 8 mm AMPLATZER™ Vascular Plug II device (St. Jude Medical, Minneapolis, MN)

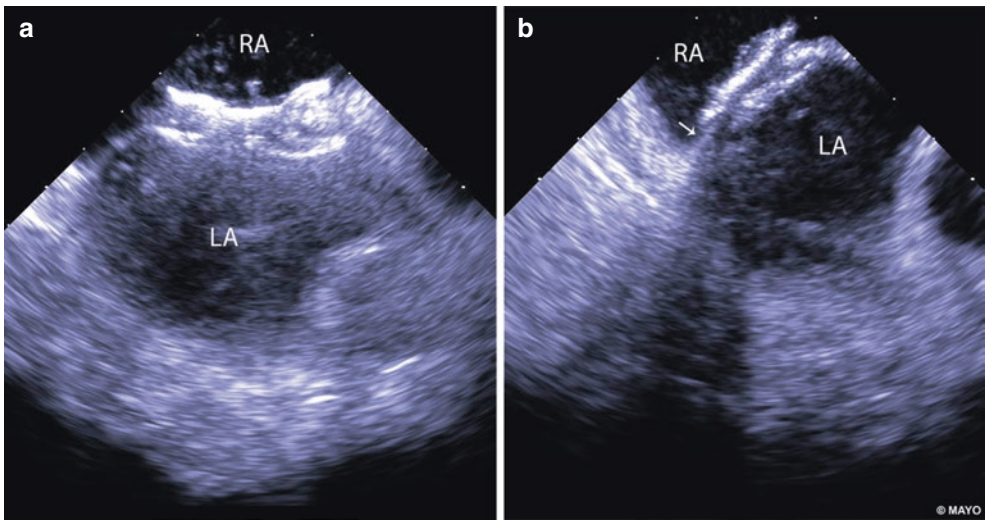



**Fig. 41.9** Patient 1: Successful device closure performed for both secundum ASDs. The two defects are visualized in separate imaging planes demonstrating the need for a complete evaluation following device ASD closure to evaluate for additional/residual defects. (**a**) The anterior/superior secundum ASD with stretched diameter of 18 mm was closed utilizing a 22 mm AMPLATZER™ Septal Occluder device (St. Jude Medical, Minneapolis, MN). There is secure apposition on the retroaortic rim and no interference with the aortic root. (**b**) The posterior/inferior secundum ASD with stretched diameter of 12 mm was closed utilizing a 25 mm GORE® CARDIOFORM® Septal Occluder (W.L. Gore & Associates Inc., Newark, DE). The left atrial disc and right atrial disc demonstrate good apposition with the posterior/inferior rim securely captured [arrow]

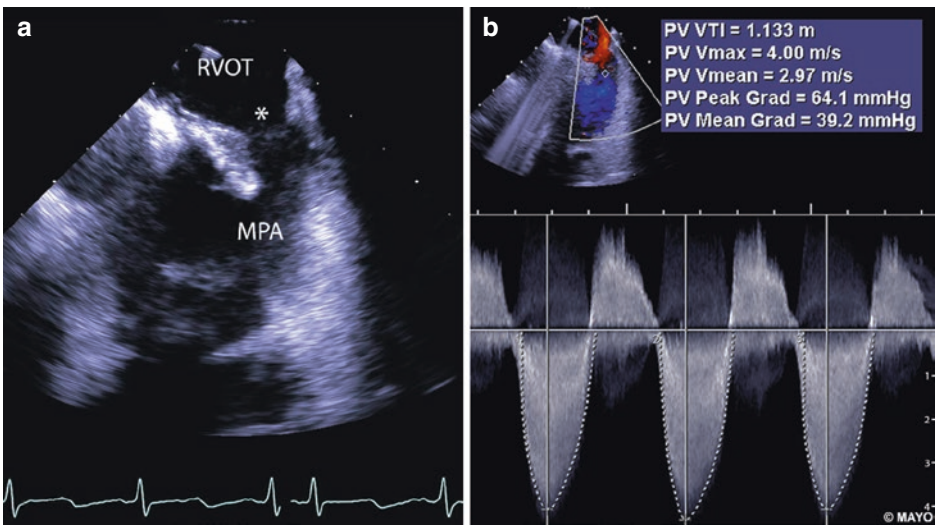



**Fig. 41.10** Patient 4: 23-year-old female (67 kg) with a history of congenital valvar pulmonic stenosis and prior valvuloplasty; S/P PVR with a 27 mm Medtronic Hancock II bioprosthetic valve (Medtronic, Minneapolis, MN) in 2011; now with mixed bioprosthetic valve dysfunction. The ICE catheter has been advanced into the right ventricular outflow tract by retroflexing from the right atrium, directing the catheter through the tricuspid valve with the imaging plane angled superiorly; with relaxation of the retroflexion the probe will advance into the RVOT. (**a**) There are thickened valve leaflets with fibrous ingrowth and a coaptation defect [asterisk]. (**b**) Continuous wave Doppler evaluation of the bioprosthetic tricuspid valve replacement. The mean systolic Doppler gradient is 39 mmHg with a holosystolic regurgitant envelope with a signal density equal to the inflow jet consistent with severe stenosis and regurgitation

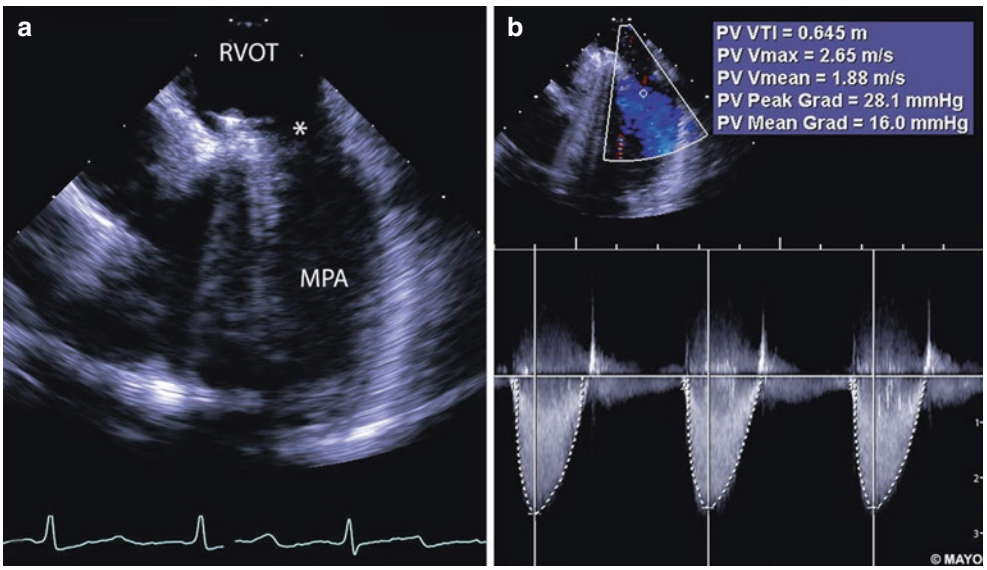



**Fig. 41.11** Patient 4: A 22 mm Melody® transcatheter heart valve (Medtronic, Minneapolis, MN) has been placed within the existing bioprosthesis. (**a**) The valve stent is well visualized and well positioned [asterisk]. The valve leaflets are thin with complete coaptation. (**b**) Continuous wave Doppler evaluation across the Melody® valve demonstrates a mean systolic Doppler gradient of 16 mmHg. There is a trivial regurgitant signal immediately following valve closure

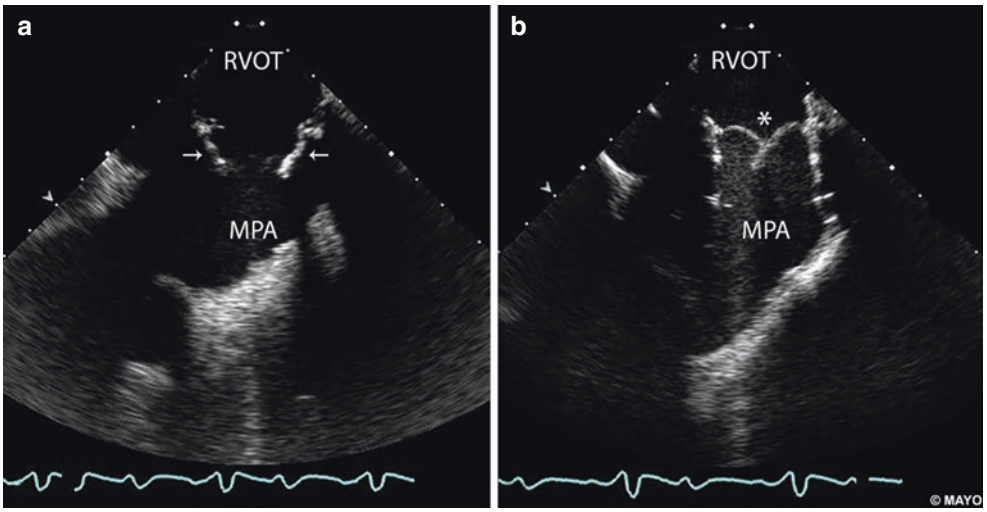



**Fig. 41.12** Patient 5: 34-year-old male (68 kg) with remote history of native pulmonary valve endocarditis; S/P PVR with a 29 mm Carpentier-Edwards Perimount Bioprosthesis (Edwards Lifesciences Corp., Irvine, CA) in 2009, now with severe bioprosthetic valve dysfunction. (**a**) The ICE probe has been advanced just beneath the bioprosthesis in the RVOT where immobilized leaflets [arrows] and turbulent flow can be demonstrated in addition to coaptation failure resulting in severe regurgitation. (**b**) A 29 mm SAPIEN 3 transcatheter heart valve [asterisk] (Edwards Lifesciences Corp., Irvine, CA) has been placed within the existing bioprosthesis. The SAPIEN valve is well visualized and well positioned. The valve leaflets are thin with no stenosis and complete coaptation. ICE imaging documented trivial pulmonary valve regurgitation and no paravalvular/intravalvular leak [Reference Video 6]

**Video 1** Patient A: 22-year-old female (53 kg) with a secundum ASD. ICE imaging is performed from the right atrium. Prior to ASD evaluation, mild tricuspid valve regurgitation, moderate right ventricular enlargement, and normal pulmonary venous return were documented. The defect demonstrates a 5 mm retroaortic rim and 23 mm posterior/inferior rim. The static, unstretched ASD diameter is 12 mm (AVI 9913 kb)

**Video 2** Patient A: Color flow Doppler echocardiography demonstrates primarily left-to-right shunting at the atrial level across the secundum ASD (AVI 7629 kb)

**Video 3** Patient A: Successful device closure of the secundum ASD was performed with a 30 mm GORE® CARDIOFORM® Septal Occluder device (W.L. Gore & Associates Inc., Newark, DE). The left atrial disc and right atrial disc demonstrate good apposition with all rims adequately captured (AVI 10387 kb)

**Video 4** Patient B: 23-year-old female (52 kg) with Ebstein's anomaly; S/p tricuspid valve replacement with a 25 mm St. Jude Biocor Bioprosthesis (St. Jude Medical, Minneapolis, MN), now with mixed bioprosthetic valve dysfunction. The tricuspid valve prosthesis is well visualized with one leaflet completely tethered and doming in diastole and incomplete opening of the remaining leaflets causing severe stenosis and regurgitation (AVI 4689 kb)

**Video 5** Patient B: A 22 mm Melody® transcatheter heart valve (Medtronic, Minneapolis, MN) has been placed within the existing bioprosthesis. The valve stent is well visualized and well positioned. The valve leaflets are thin with complete coaptation (AVI 4729 kb)

**Video 6** Patient C: 34-year-old male (68 kg) with remote history of native pulmonary valve endocarditis; S/P PVR with a 29 mm Carpentier-Edwards Perimount Bioprosthesis (Edwards Lifesciences Corp., Irvine, CA) in 2009. A 29 mm SAPIEN 3 transcatheter heart valve (Edwards Lifesciences Corp., Irvine, CA) has been placed within the existing bioprosthesis. The SAPIEN valve is well visualized and well positioned. The valve leaflets are thin with complete coaptation (AVI 4625 kb)

# 3D Echocardiography in Congenital Heart Disease Diagnosis and Transcatheter Treatment

# 42

Carmelo Arcidiacono

## 42.1 Introduction

Transesophageal echocardiography (TEE) is widely established in most centers as a mandatory imaging modality for guidance of percutaneous closure of septal defects: coupled with fluoroscopy, it provides detailed and reliable information in real time to the operator, enabling measurement of the defects and of their rims, visualization of devices during their deployment and evaluation of the results after release. Conventional two-dimensional TEE has however some intrinsic limitations. Obtaining detailed information such as number, size, shape, and spatial relationships of multiple, multifenestrated, or complex-shaped defects can often be challenging with two-dimensional imaging. Two-dimensional TEE guidance is also significantly limited in locating guidewires and catheters and assessing device position, particularly when multiple devices are used.

Three-dimensional transesophageal echocardiography (3DTEE) is a rather recent innovation in echocardiographic imaging, based on miniaturized matrix-array transducers coupled with dedicated software that, in modern versions, allows three-dimensional imaging in real time without the need for multiple-beat acquisition. This imaging modality is therefore particularly useful for guidance of percutaneous procedures. As reported in many studies, 3DTEE enables accurate identification of guidewires and catheters when crossing the defects and monitoring of device deployment and release (Figs. 42.4, 42.5, 42.6, 42.8, 42.11, 42.14, 42.15). In cases where multiple devices are used, 3DTEE provides information on their arrangement and relationship with surrounding structures (Figs. 42.2, 42.3 and 42.7).

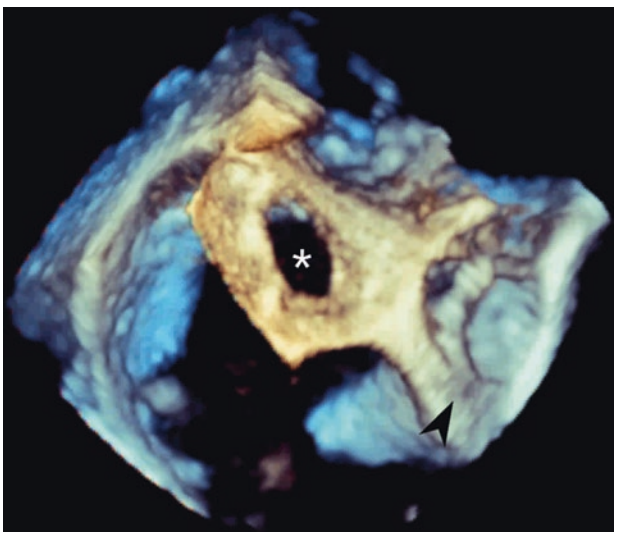


**Fig. 42.1** 3DTEE imaging in a case of an ostium secundum atrial septal defect. The defect is centrally located (asterisk), and the imaging mode can clearly show its relationship to the tricuspid and mitral valve annuli (lower left and upper right of the image, respectively) and with the aortic root (black arrowhead)

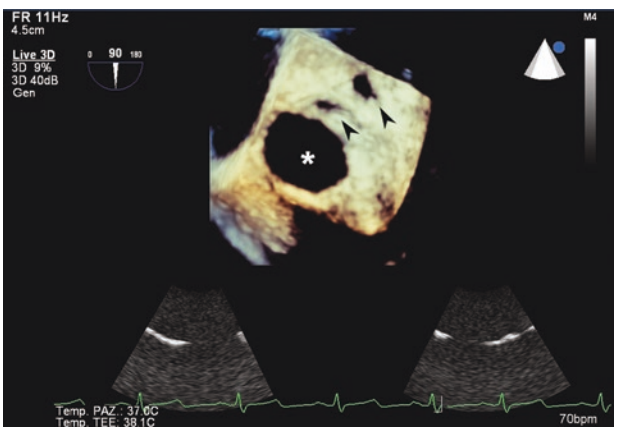



**Fig. 42.2** 3DTEE image of a case of multiple atrial septal defects. The view shows the left atrial side of the atrial septum. The main defect, indicated by an asterisk, is located in the middle portion of the oval fossa, while two small accessory defects are detectable on the postero-inferior portion of the septum (arrowheads)

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_42) contains supplementary material, which is available to authorized users.

C. Arcidiacono (✉)
Department of Pediatric Cardiology, Policlinico San Donato IRCCS, San Donato Milanese, Italy
e-mail: Carmelo.arcidiacono@grupposandonato.it



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_42

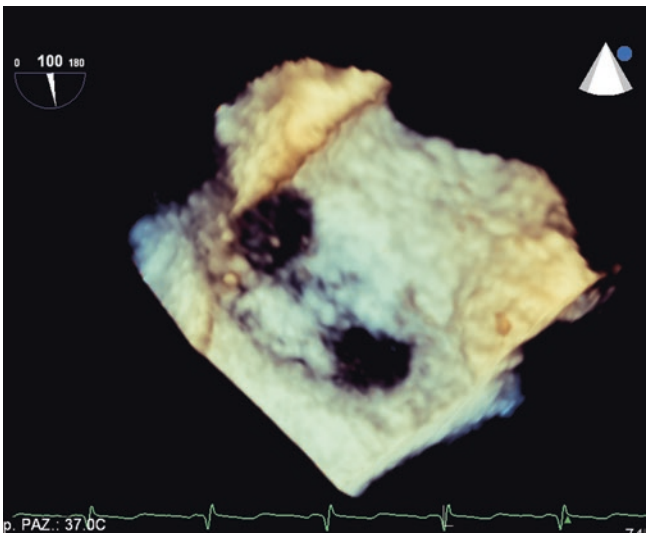



**Fig. 42.3** 3DTEE image of a case of multiple atrial septal defects. The view shows the left atrial side of the atrial septum. Two secundum-type defects of similar size are shown, both round shaped, separated by a large band of septal tissue

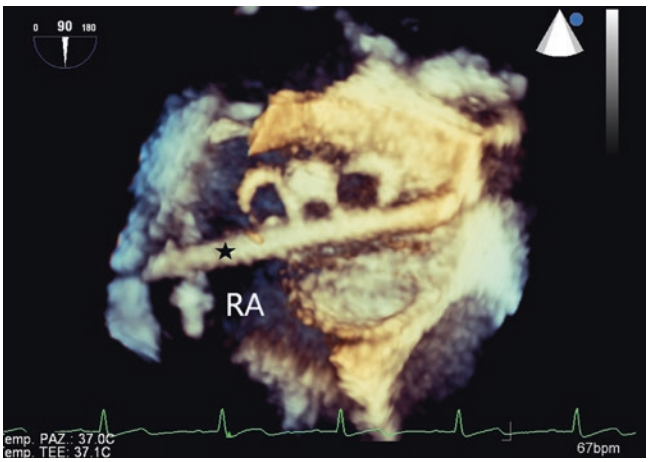



**Fig. 42.5** A case of multifenestrated atrial septal defect. The catheter, indicated by a black star, is crossing the central fenestration from the right atrium

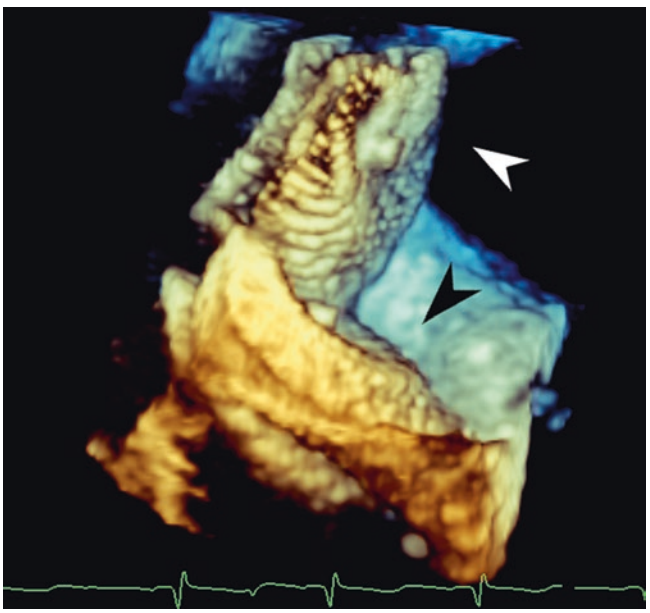


**Fig. 42.4** The same case as in Fig. 42.3, after placement of two Amplatzer ASO devices (black and white arrowheads). Of note, the two devices are arranged at an almost orthogonal angle, reflecting the curved surface of the atrial septum

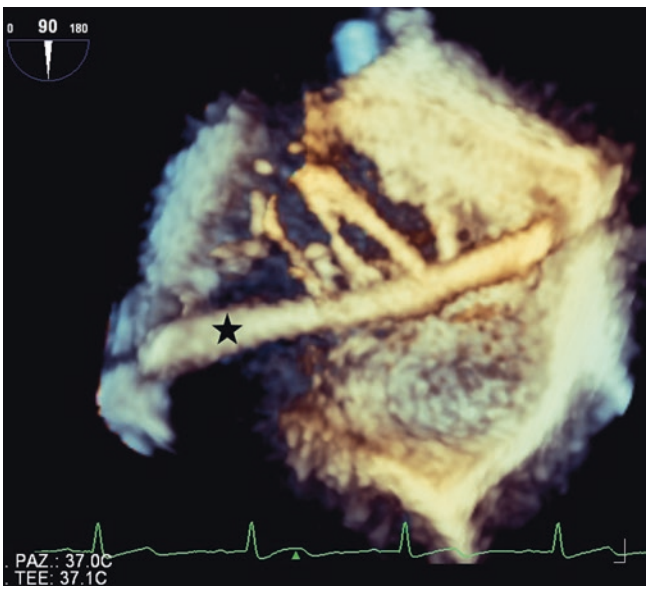



**Fig. 42.6** The same case as in Fig. 42.5. When some tension is applied to the catheter, this stretches the strands of tissue separating the fenestrations, as clearly shown by the 3DTEE image

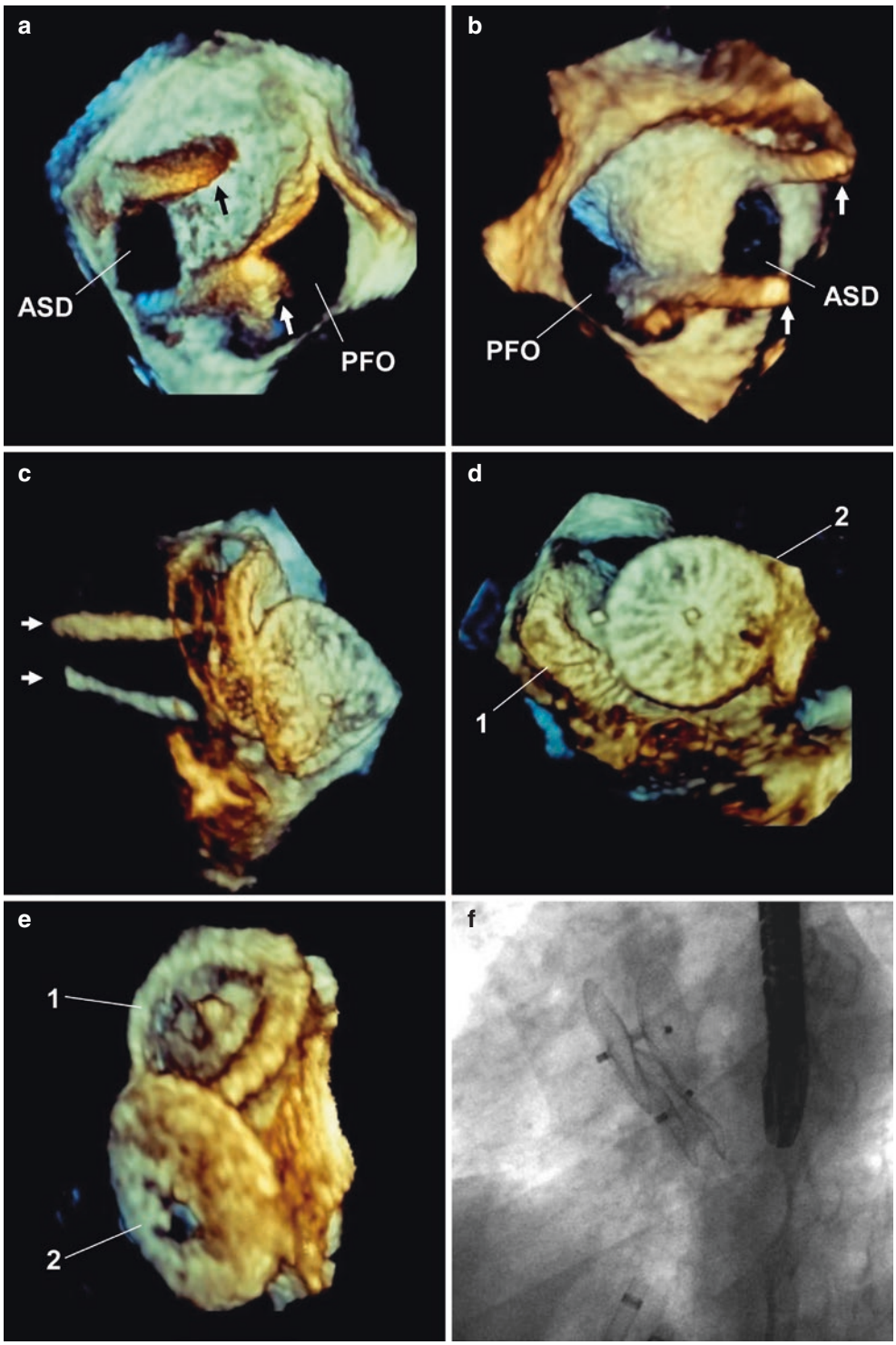



**Fig. 42.7** Panels (**a**, **b**) show 3DTEE images of the atrial septum, seen from the left and right side: a posterior secundum atrial septal defect is associated to an anterosuperior patent foramen ovale with a very compliant and aneurysmal flap valve. The defects are both crossed by catheters (arrows). On panel (**c**) the two defects are closed with occluding devices (angiogram on Panel **f**). The devices are still attached to the delivery wires (arrows). On Panel (**d**, **e**) the two devices have been released and are seen from the left and from the right atrial side, respectively. The different shape of the two devices is evident (PFO device, number 1; ASD device, number 2)

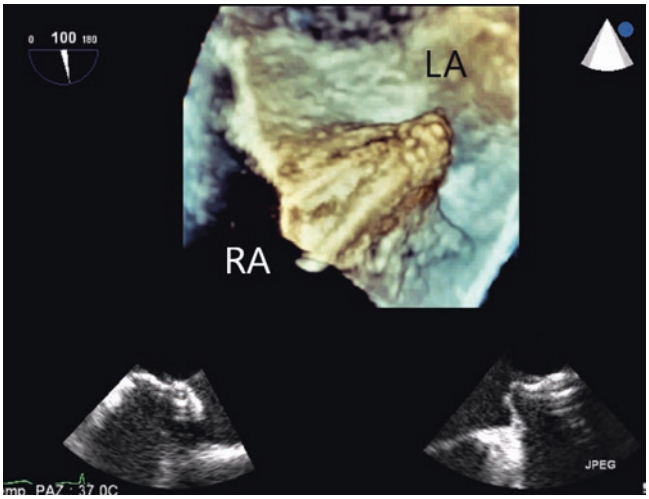



**Fig. 42.8** 3DTEE imaging during an attempt at crossing a multifenestrated and floppy atrial septum with a catheter: the tenting of the septum is obvious. *RA* right atrium, *LA* left atrium

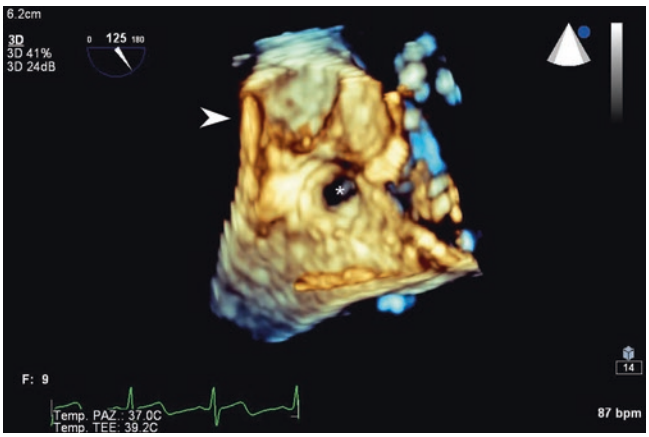



**Fig. 42.9** 3DTEE image of a case of perimembranous ventricular septal defect. The viewpoint is from the left ventricular outflow tract, and the defect, indicated by an asterisk, is clearly detected in close proximity to the aortic valve (white arrow)

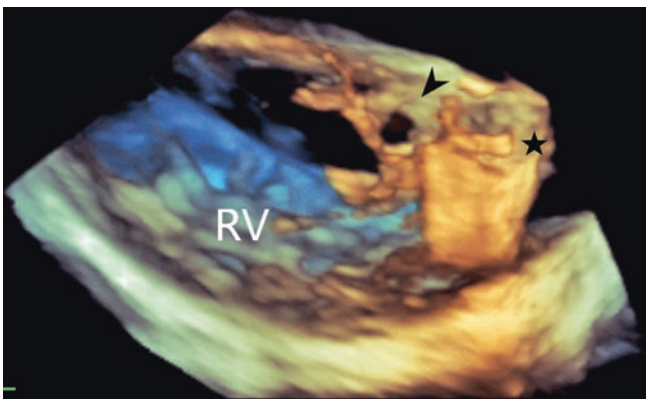



**Fig. 42.10** A case of perimembranous ventricular septal defect, where the defect, oval in shape (black arrowhead), is at some distance from the plane of the aortic valve (black star). Accessory fibrous tissue from the tricuspid valve apparatus is also evident

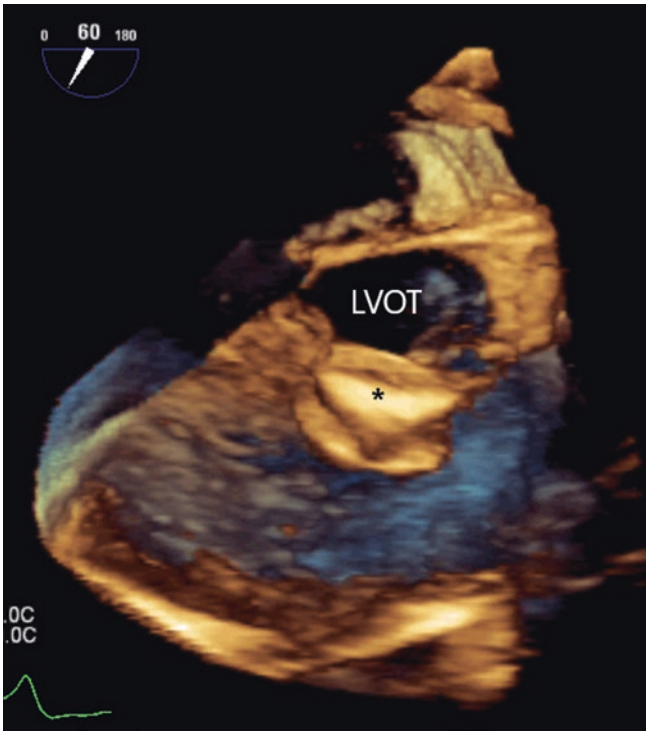



**Fig. 42.11** Final check of the position of an Amplatzer muscular VSD device after release. In this transverse 3DTEE view across the left ventricular outflow tract (LVOT), we can check the profile of the device (asterisk) and its relationship to the ventricular septum

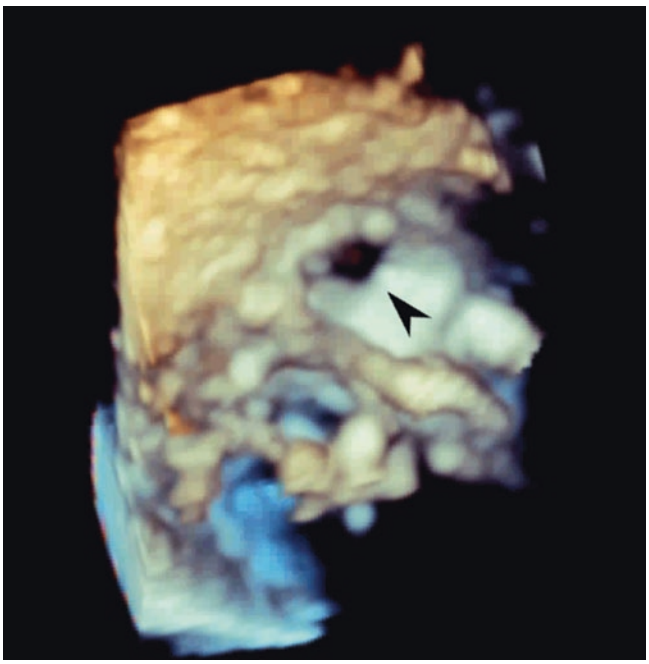


**Fig. 42.12** 3DTEE view of the left ventricular cavity in an unusual case of muscular VSD in a hypertrabeculated left ventricle. The round-shaped defect (black arrowhead) is seen above a very prominent trabeculation, resembling the moderator band

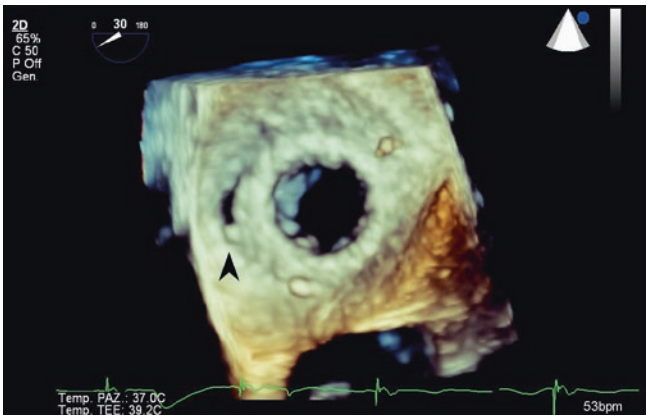



**Fig. 42.13** Left atrial view of a case of paravalvular leak on a prosthetic mitral valve. The leak, pointed by a black arrowhead, has an elongated shape and is located toward the atrial septum

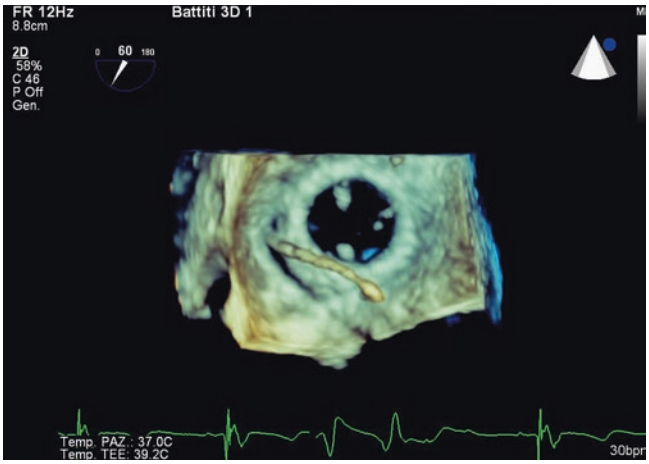



**Fig. 42.14** The same case as in Fig. 42.13. A guide wire, as clearly shown by 3DTEE imaging, has crossed the leak

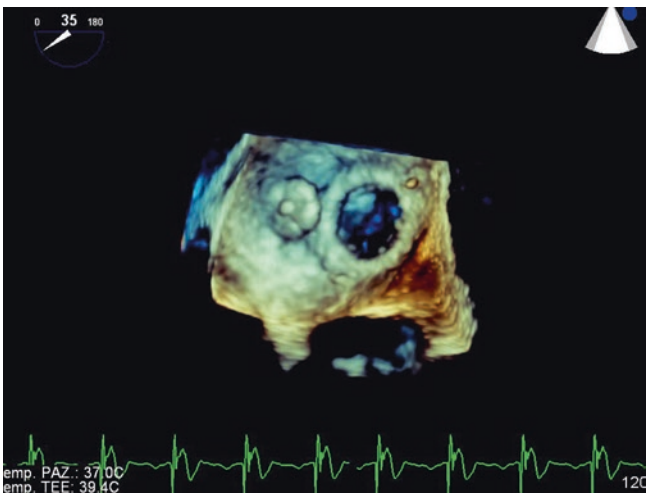



**Fig. 42.15** Final check after placement of an occluding device, obtaining occlusion of the paravalvular leak

**Video 1** 3DTEE video showing a secundum-type atrial septal defect from the left atrial aspect. The defect is located toward the septum secundum, close to the orifice of the superior vena cava. The floppiness of the thin-walled septum of the oval fossa is suggested by its "trembling" movement in the video (AVI 3046 kb)

**Video 2** As in Fig. 42.7, this 3DTEE video shows a case of secundum atrial septal defect associated to a very compliant patent foramen ovale. Both defects, seen from the right atrial side, are crossed by catheters (AVI 1570 kb)

**Video 3** As in Fig. 42.4, this 3DTEE video was obtained after placement of two occluding devices in two secundum-type atrial septal defects. The two devices are arranged at an angle, reflecting the normal curvature of the atrial septum (AVI 3684 kb)

**Video 4** 3DTEE video showing, as in Fig. 42.10, a case of perimembranous ventricular septal defect, where the defect, oval in shape, is at some distance from the plane of the aortic valve. Accessory fibrous tissue from the tricuspid valve apparatus is also evident (AVI 4387 kb)

**Video 5** 3DTEE video with color flow map of a case of paravalvular leak from a prosthetic mitral valve. The leak, clearly indicated by the jet of color, is crossed by a guide wire, which is seen oscillating with the cardiac cycle. This imaging technique proved to be very useful to locate the leak and to confirm its crossing by wires and catheters during the procedure (AVI 1914 kb)

# 3D Mapping: Live Integration and Overlay of 3D Data from MRI and CT for Improved Guidance of Interventional Cardiac Therapy

43

Stephan Schubert and Felix Berger

## 43.1 Introduction

Multimodal picture integration has been developed for improved visualization of interventional procedure in congenital and structural heart disease. 3D rotational angiography (3DRA) was used for that purpose in the past, and a 3D dataset was than imposed as an overlay for fluoroscopy. But 3DRA includes a 180° turn of the X-ray arm in order to generate a 3D dataset, including additional contrast application of 4–5 seconds, may need rapid pacing and preparation of the cath lab to allow X-ray arm movement.

VesselNavigator (Philips Healthcare) allows reuse of 3D vascular anatomical information from existing CT and MRI datasets as a 3D road map overlay on a live X-ray image (Fig. 43.1). Quality and success of interventional therapy may be improved by this 3D dataset, and generation of this data can be done prior to the catheterization. Additionally with the use of MRI data, X-ray and contrast exposure can be reduced significantly in comparison to 3DRA.

In this chapter, we want to demonstrate the use of 3D overlay in different interventional procedures.

## 43.2 CASE: PPVI with Melody

Percutaneous pulmonary valve implantation (PPVI) was performed in a 13-year-old male patient after correction of Ebstein anomaly with tricuspid reconstruction, PCPC, and pulmonary valve replacement with an 18 mm Contegra (Medtronic) 2 years before. Now a high-grade Contegra stenosis was intended to be treated by PPVI (see Video 1).

After segmentation (MRI dataset) of the pulmonary artery and aorta, two rings were blue placed for marking the landing zone for stenting and pulmonary revalvulation (Fig. 43.1 and Video 1 and 3). The left and right coronary artery were also marked by green points (Fig. 43.2). Percutaneous valve implantation was performed with the use of Melody TPV on a 18mm Ensemble (Fig. 43.3 and Video 2) and guided by the 3D overlay.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_43) contains supplementary material, which is available to authorized users.

S. Schubert (✉) · F. Berger
Department of Congenital Heart Disease/Pediatric Cardiology, German Heart Center Berlin, Berlin, Germany
e-mail: sschubert@dhzb.de


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_43


**Fig. 43.1** Work Flow for the use of 3D overlay with the use of VesselNavigator (Philips Healthcare): *A*. SEGMENTATION is done by importing a 3D dataset (DICOM) into the workstation (CT of a complete thoracic scan of the heart and vessels and thorax and 3D MRI of the whole heart sequence are optimal). The targeted vessels are selected with the mouse nearly automatically (aorta, pulmonary arteries, coronaries). *B*. PLANNING is then performed by using ring markers and small points for marking stenotic regions (landing zone) or landmarks (branches, coronaries). Additionally the 3D dataset can be rotated in order to identify the best projection and visualization, which can be saved for at the workstation. *C*. REGISTRATION is performed, if the patient is lying on the table with the use of fluoroscopy or angiography and in the AP plane only. A difference of >30° is demanded in order to do the registration. This can be done from inside at the patients table or outside at the workstation. *D*. LIVE GUIDANCE will start after steps *A*–*C*, where also re-calibration of the accuracy of the overlay is possible during the whole examination

## 43.3 CASE: RPA Stenting

Right pulmonary artery (RPA) stenting was performed in an 8-year-old patient with pulmonary atresia with shunt palliation and right ventricular to PA conduit placement. Several surgical and catheter interventions had been performed, but RPA stenosis was reluctant to balloon dilatation or surgical reconstruction. Actual MRI showed a residual RPA stenosis with flow and size mismatch of left > right pulmonary arteries including supravalvular stenosis at distal conduit anastomosis (see Fig. 43.4 and Videos 4, 5 and 6).

## 43.4 CASE: CoA Stenting

A 35-year-old patient was treated for native and subatretic coarctation of the aorta (CoA). Vessel navigator was used after import of an external CT scan (see Figs. 43.5, 43.6, 43.7 and 43.8.

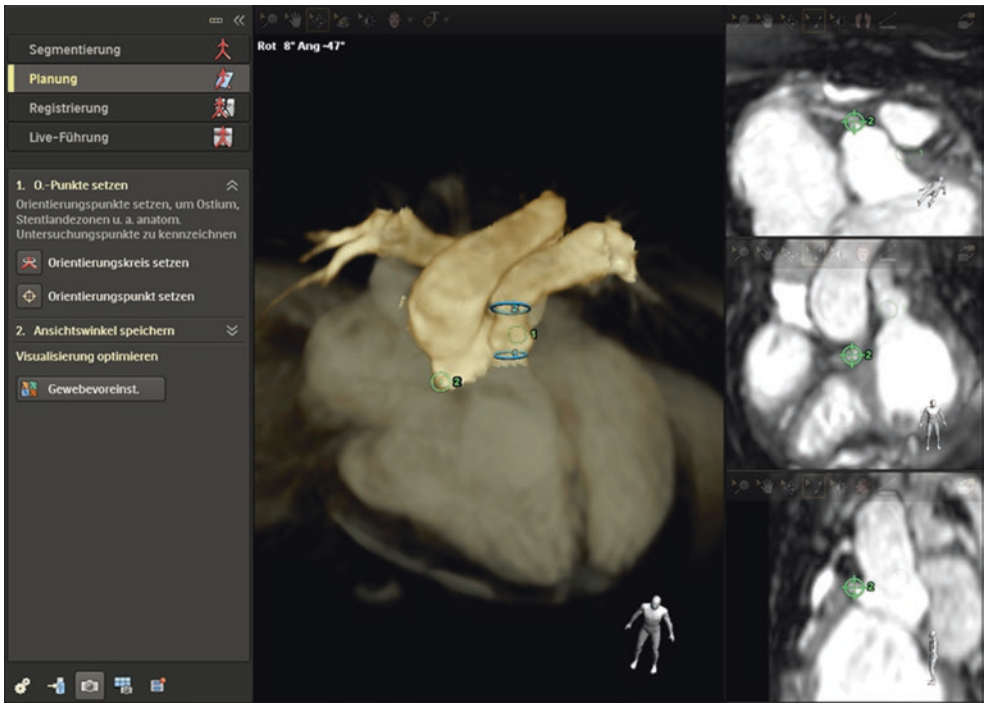



**Fig. 43.2** Segmented vessel for PPVI in a 13-year-old patient with Contegra stenosis in RAO 8° and cranial 47°. Two rings are marking the proximal and distal landing zone for stent placement. 1 and 2 are marking LCA and RCA (see Video 2)

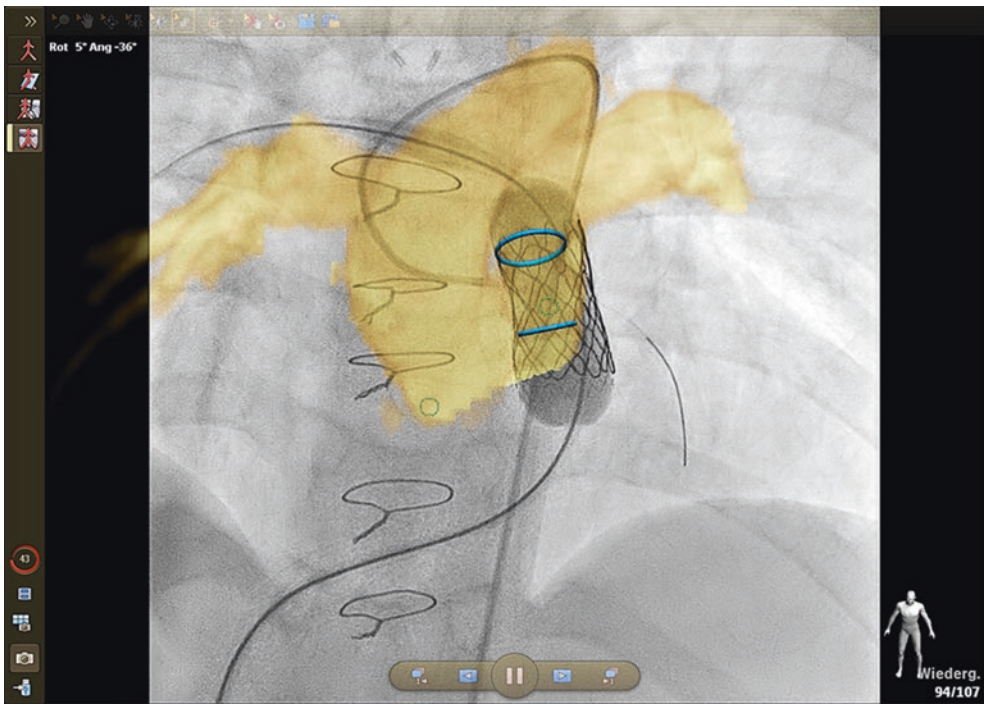



**Fig. 43.3** Percutaneous Pulmonary valve (Melody) implantation in a pre-stented stenotic contegra with live guidance by MRI based image fusion (PA and Ao). Blue rings act as marker for the valve region. Left coronary are marked by 0.014 inch wire

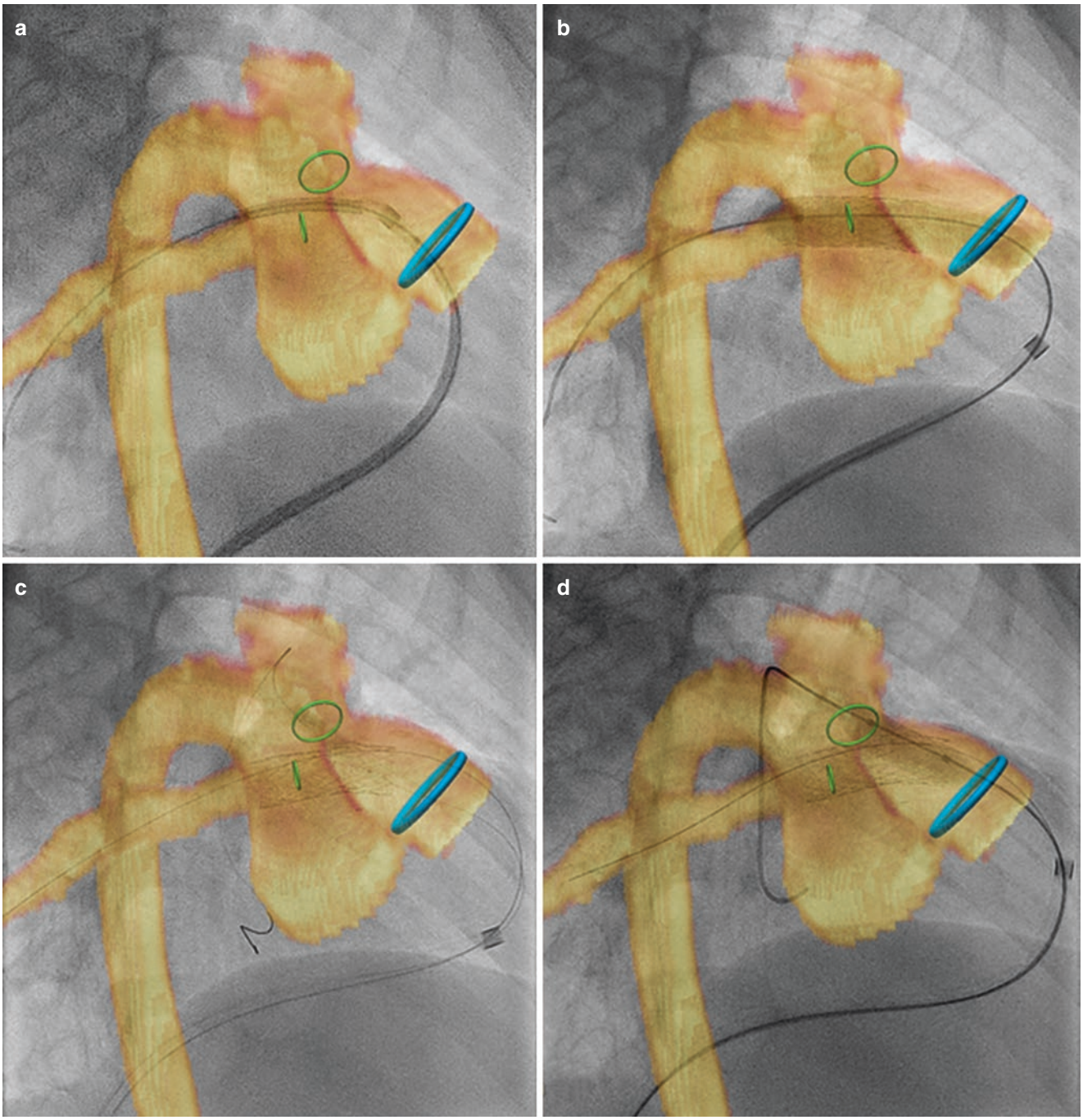



**Fig. 43.4** 3D overlay with pictures generated from MRI with segmented vessels (PA and Ao = yellow) during treatment of pulmonary branch stenosis. Green rings are marking the stenotic part in RPA and LPA ostium. (**a**, **b**) Stent implantation of a Mega LD (Ev3, 26 mm) on a 10 mm Powerflex balloon with a use of a 9 Fr long sheath (Cook Flexor). (**c**, **d**) Re-opening of the LPA ostium after stent placement with the use of a 4 mm coronary balloon and 8 mm Powerflex afterward

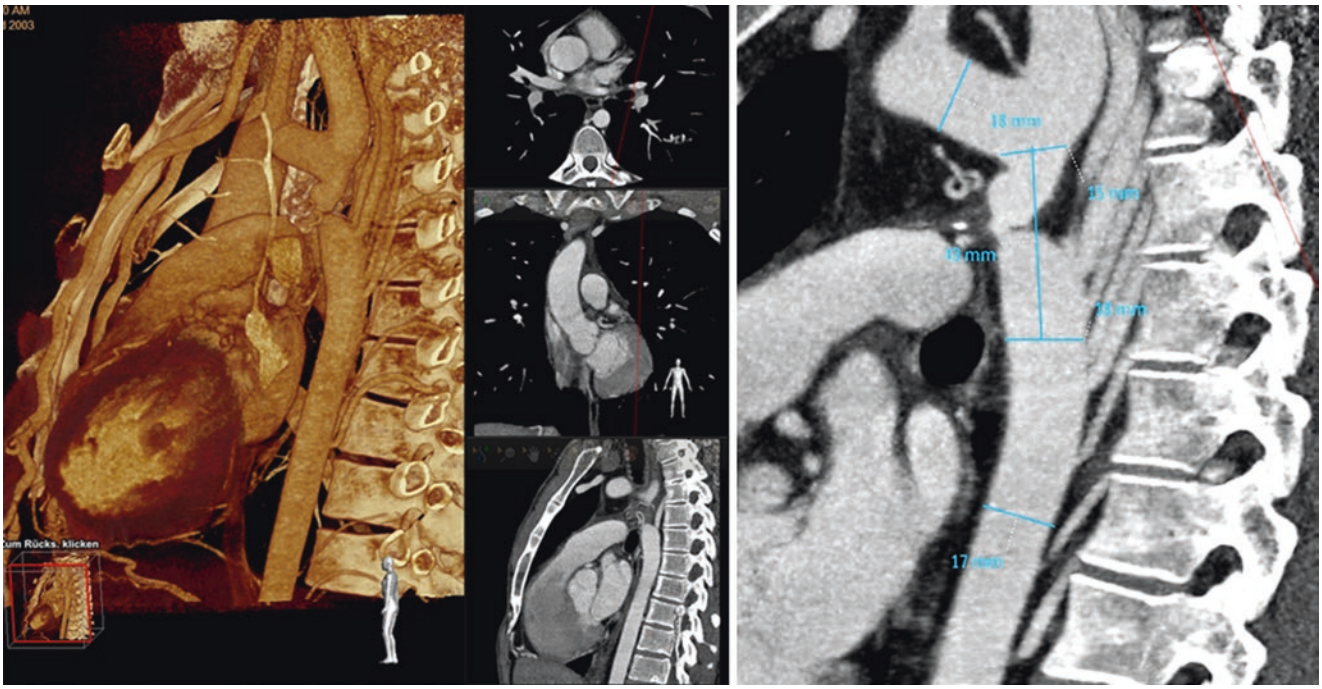



**Fig. 43.5** CT data as used for planning of the procedure in this patient with native CoA imported into VesselNavigator (left). Numerous collateral vessels can be seen connecting from dorsally into the descending aorta. Measurements (right) were performed, and landing zone length (42 mm) and diameter (18 mm) of the stent were defined

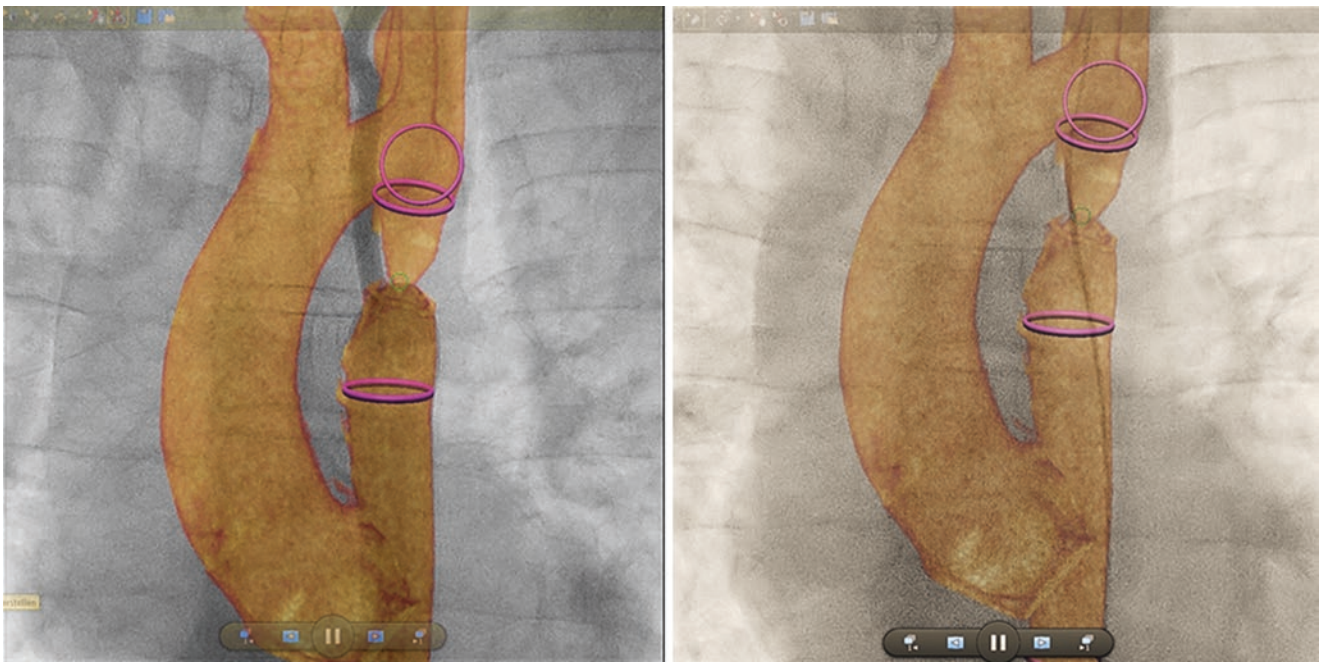


**Fig. 43.6** Live guidance with the use of CT data was used. For calibration, two angiographies in the descending aorta were used (left) and in a second 40° RAO projection. Passage over the CoA into the left subclavian artery was then performed (right), and rings were placed in order to define the landing zone for stent placement

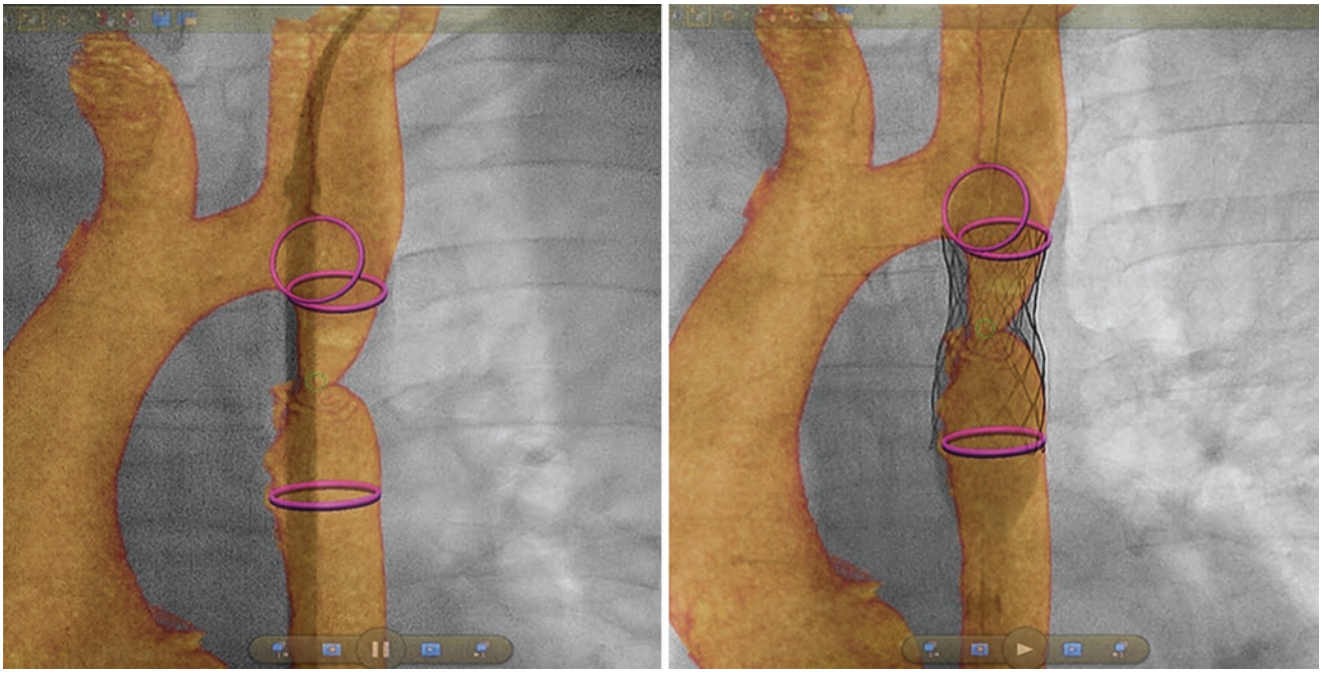


**Fig. 43.7** Live guidance for 16 Fr-long sheath (Mullins, Cook) placement and implantation of a custom-made covered 10 zig CP Stent on a 22 mm Balloon-in-Balloon (BIB®) Catheter and with a length of 50 mm (NuMed)

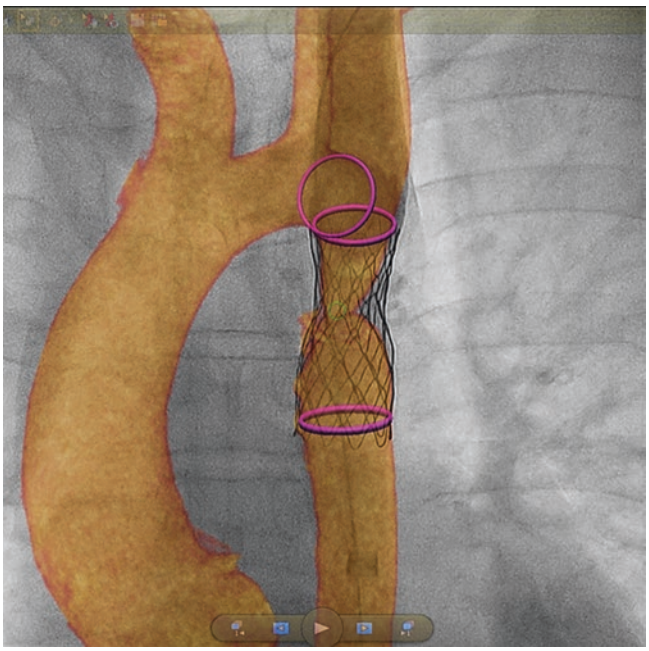


**Fig. 43.8** Angiographic result after covered CP stent implantation into a native CoA with the use of overlay by VesselNavigator from CT data. No residual gradient was measured

**Video 1** SEGEMENTATION is performed with the use of a 3D dataset from MRI (3D of the whole heart), and the pulmonary artery is selected by semiautomatic registration. On the right side, the MRI data is still visible (MP4 8312 kb)

**Video 2** Live guidance of PPVI with Melody into a pre-stenting (Max LD (EV3) stent) valve region in RAO 5° with cranial 35°. A floppy 0.014 in. wire is marking the LCA. Overlay was implicated from MRI data; see Figs. 43.2 and 43.3 (MP4 50133 kb)

**Video 3** Segmentation of target vessels (blue) prior to intervention with data achieved by MRI (3D of the whole heart). Right and left pulmonary artery, conduit with distal stenosis, and aorta are selected and visualized (blue). Coronary arteries are also selected, as they are far away from the region of intervention. Movement of the dataset can be used for achieving optimal angulation (MP4 4766 kb)

**Video 4** LIVE Guidance during sheath placement (9 Fr Flexor sheath, Cook) after segmentation and calibration. The three rings are marking each of the pulmonary valve, RPA ostium, and LPA ostium (MP4 20692 kb)

**Video 5** LIVE Guidance during balloon dilatation of the LPA ostium through the stent meshes of the Mega LD stent (26 mm) with the use of an Atlas (Bard) 12 mm (MP4 25744 kb)

**Video 6** LIVE Guidance during balloon dilatation of the RPA ostium through the stent meshes of the Mega LD stent (26 mm) with the use of an Atlas (Bard) 12 mm (MP4 13558 kb)

# Part XI

# Future Directions

# Holography in Congenital Heart Disease: Diagnosis and Transcatheter Treatment

44

Elchanan Bruckheimer and Carmel Rotschild

## 44.1 Introduction

The heart is comprised of three-dimensional components whose size, shape, and spatial interrelationships constantly change in an organized rhythmic fashion. The field of congenital heart disease encompasses the result of a disruption of the normal, but complex, three-dimensional internal anatomy and functioning of the heart. The diagnosis and treatment of congenital heart defects demand a precise and profound understanding of this dynamic anatomy and are therefore heavily reliant on real-time imaging. The open-heart surgeon has the benefit of comprehension and interaction with the real 3D anatomy; however this is usually static during open-heart surgery. The interventionalist wishing to employ minimally invasive transcatheter techniques in the beating heart has to build a mental concept of the anatomy usually derived from 2D echocardiographic and fluoroscopic data displayed on a screen. In general, the surgeon's understanding of the anatomy is more complete and intuitive than that of the interventionalist even though they may be performing similar procedures, e.g., closure of an ASD secundum. In an attempt to bridge that gap, high-quality 3D volumetric data acquisition systems such as 3D transthoracic and transesophageal echocardiography, 3D rotational angiography, CT, and MRI have been developed. However this volumetric data is typically displayed on 2D screens which restrict the image to a single plane of view, preclude interaction directly with the image, and hamper the perception of depth. The development of a simple, accurate, and accessible 3D display for these data is a challenge, especially when attempting to display true depth.

Depth perception defines our ability to see an object, comprehend its position in space, and interact precisely with that object. The object's three dimensionality and distance are perceived owing to the capacity of the visual cortex to simultaneously combine multiple depth cues including binocular stereopsis, convergence, and accommodation. Each eye, since it has a different position in the head, views an object from a slightly different angle, and this disparity, stereopsis, is employed by the brain as one of the cues for depth perception. Stereoscopy is a method which mimics stereopsis by presenting each eye with two slightly different versions of an image which, when combined in the brain, produce an illusion of depth. It is an illusion since all the points of light in the stereoscopic image are focused in a singular focal plane, whereas in a real object, these points arise from multiple different planes, and true depth perception has infinite focal planes (Fig. 44.1).

Even the highest-quality stereoscopic image is confined to one focal plane, and therefore interactions such as placing a device in the image, or manipulating the image with the observer's hand inside it, are not possible since the device or hand occupies multiple focal planes. In addition, the source of stereoscopic images is at a distance from the observer; however the overlapping of the images to create the 3D illusion occurs up close. When attempting to interact with the up close 3D image, the eyes converge to focus on the hand or tool and lose their focus on the distant source, and the image becomes blurred. This discrepancy between the convergence depth cue and the distant focus creates a conflict in the brain often resulting in nausea. Recent attempts at enhancing the "image-anatomy" interaction and the operator's experience with stereoscopic displays include virtual reality (VR) and augmented reality (AR). Although these methodologies may provide a better understanding of the patient's anatomy and spatial relationship to the operator, they lack *true* depth perception and cannot be used for accurate navigation within the image.

E. Bruckheimer (✉)
Section of Pediatric Cardiology, Schneider Children's Medical Center Israel, Petach Tikva, Israel
e-mail: elchananb@bezeqint.net

C. Rotschild
Department of Mechanical Engineering, Technion, Haifa, Israel



G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_44

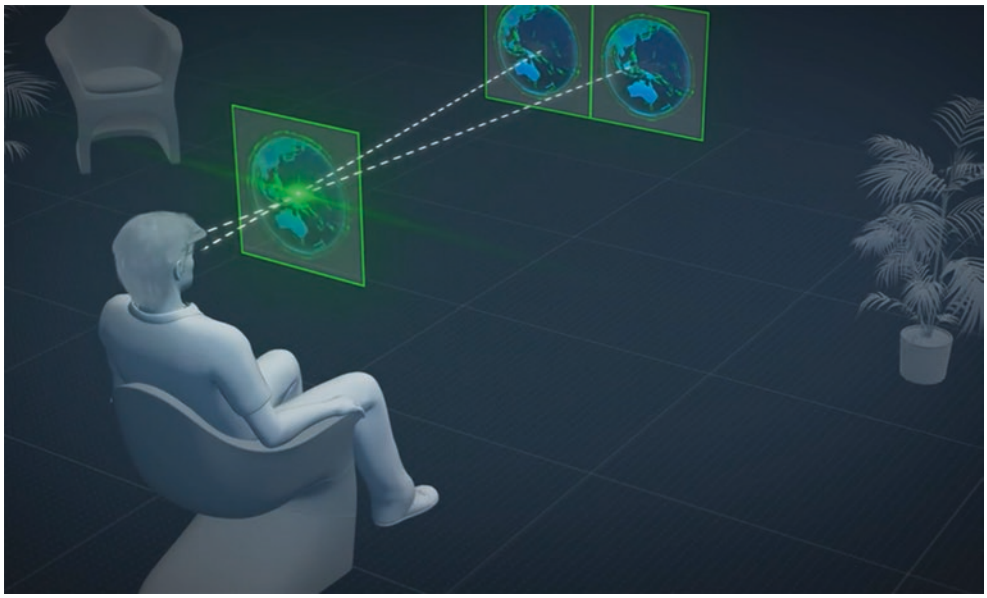


**Fig. 44.1** Stereoscopic images of an object creating a 3D illusion of that object. The observer is presented with two slightly different images of the same object which overlap. The 3D illusion is confined to one focal plane

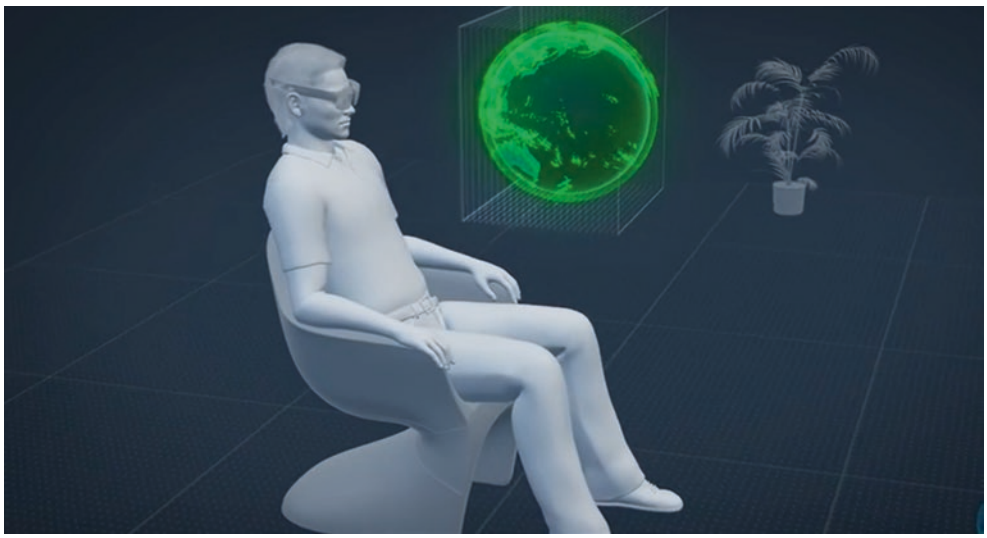


**Fig. 44.2** The same object in Fig. 44.1 is projected as a hologram that occupies 3D space, as opposed to one single focal plane. The fixed coordinates and multiple focal planes afford interaction with and within the image at arm's reach of the observer

Holography, from the Greek for "whole" and "drawing," is a methodology which creates an exact visual replication of an object in three physical dimensions including all the depth cues, focal planes, and specific coordinates. The holographic image occupies an actual space and can be likened to an object created by a 3D printer where the material used to print is light. A high-quality hologram should be indistinguishable from the true object it represents (Fig. 44.2).

Computer-generated holograms (CGH) of patients' 3D data could be useful in the clinical setting by providing the physician with a 3D image which is a true and spatially accurate representation of the patient's anatomy, having all the visual depth cues. Since each point of light in the generated hologram is a distinct physical entity in real 3D space, it represents a precise anatomical coordinate in the patient, and interaction with and within the holographic image will simulate such interaction with and within the patients' particular anatomy. Initial attempts at creating holograms in real time in an actual clinical setting have been recently reported using the RealView 3D dynamic holographic display and interface system [RealView Imaging, Yokneam, Israel]. The system projects high-resolution 3D live holographic images "floating in mid-air" without the need for any human-mounted device, goggles, or a conventional 2D screen. The projected 3D images appear in free space, allowing the user limitless accessibility to the image affording direct and precise interaction with and within the dynamic image. Therefore, a true volumetric display based on current 3D acquisition modalities maintains the interventionalist's advantage of a minimally invasive approach and provides "real" 3D dynamic

anatomy similar or even better than that of the surgeon, since the surgeon sees static 3D surfaces, while the hologram can display the organ at different levels of transparency to the viewer and can be easily cropped and manipulated as required.

The following figures are examples of holographic images created using the RealView system and demonstrate some of the interactions that can be performed and 3D acquisition modalities that can be used to create holograms in the clinical setting. Please note that holograms cannot be fully appreciated on a 2D page, but need to be observed directly. [Videos of holograms shown here in the figures can be viewed at www.realviewimaging.com]

**Acknowledgement** *Conflict of interest*: EB and CR are stockholders and employees of RealView Imaging.

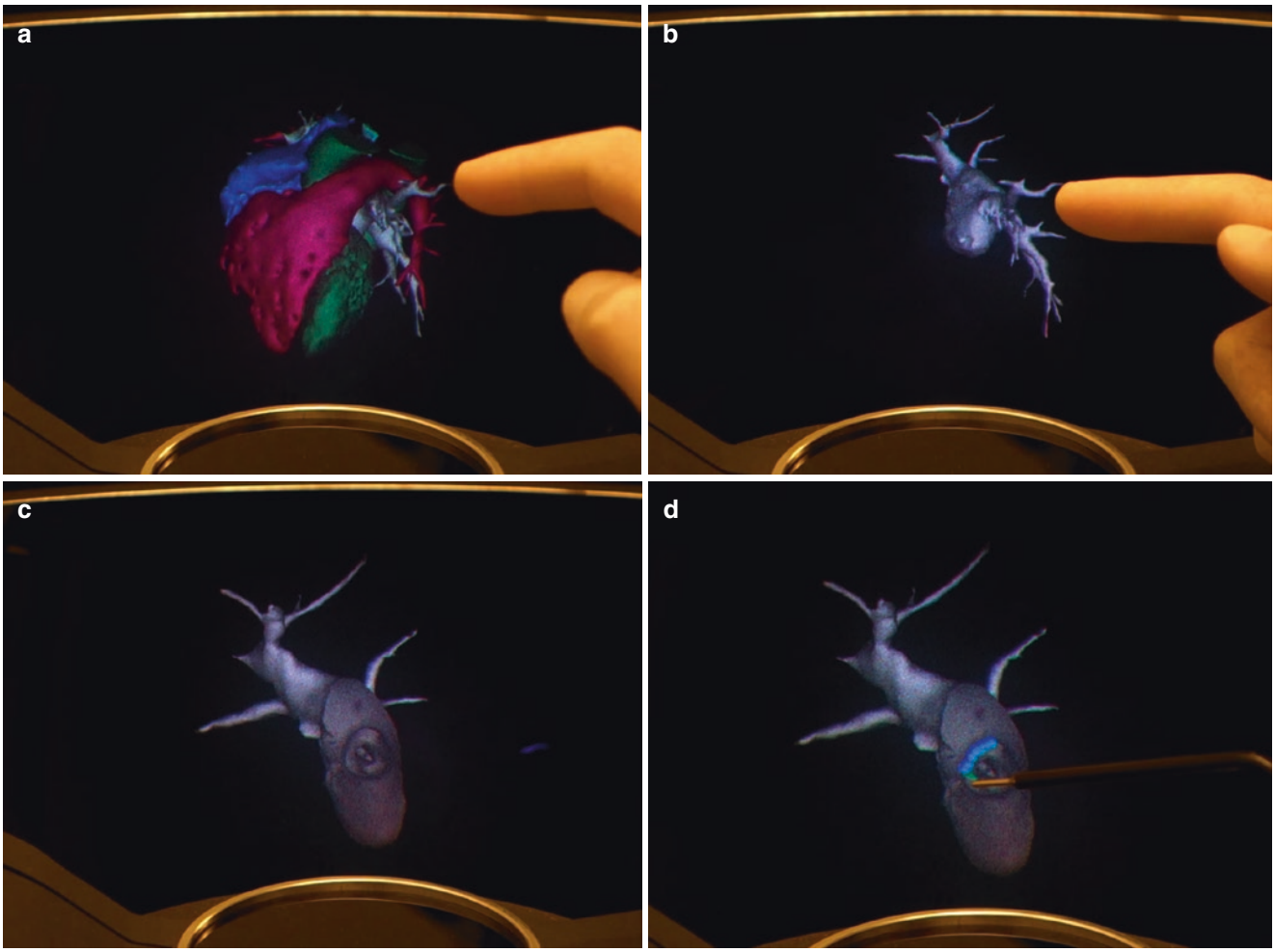



**Fig. 44.3** (**a**) Hologram created from a segmented cardiac CT showing the anterior right ventricle [red] and the posterior left atrium [gray]. The finger is touching the left upper pulmonary vein and rotates the image to the desired viewing angle. (**b**) The volumetric data of the left atrium is isolated, and the atrium is rotated anteriorly prior to slicing as seen in Fig. 44.3c. (**c**) The hologram of the left atrial volume has been sliced open and rotated so that the pulmonary venous orifice of the left upper pulmonary vein is viewed en face. (**d**) Ablation points are simulated directly on the image at the junction of pulmonary vein orifice to the left atrial wall using a tool. These points were intuitively positioned at the exact anatomical position and depth creating a continuous and contiguous line in a similar manner to painting directly on a 3D object

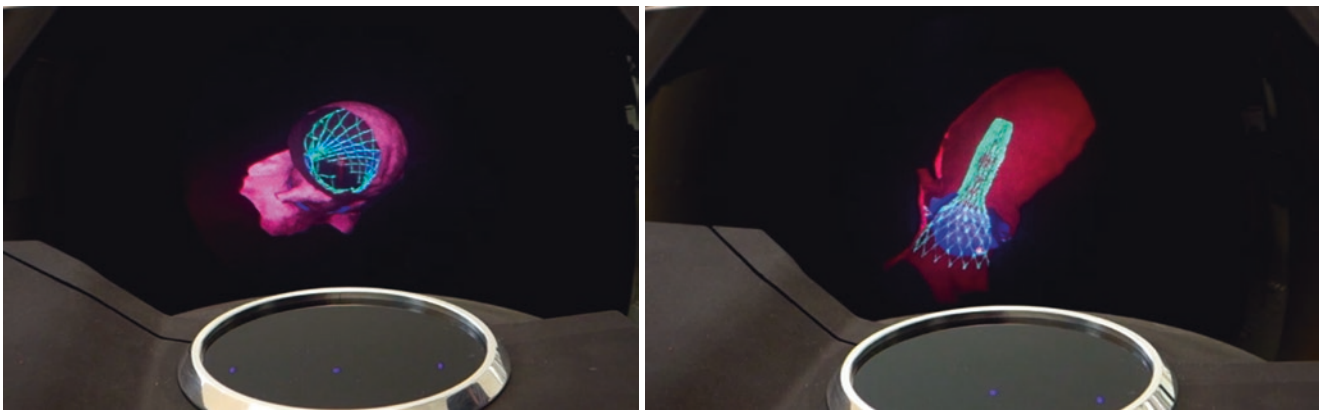


**Fig. 44.4** Dynamic color holograms created from volumetric data simulated with the TAVIguide™ technology (FEops, Ghent, Belgium) of a virtually deployed CoreValve® wire frame (Medtronic, Minneapolis, MN, USA). The TAVIguide technology predicts how a certain TAVI device will interact with a specific patient. Upper plane: The aorta and fully deployed CoreValve seen from the ascending aorta. Lower pane: The partially deployed CoreValve is seen by slicing into the ascending aorta, and the interaction with the native aortic leaflets, in blue, can be seen from any angle

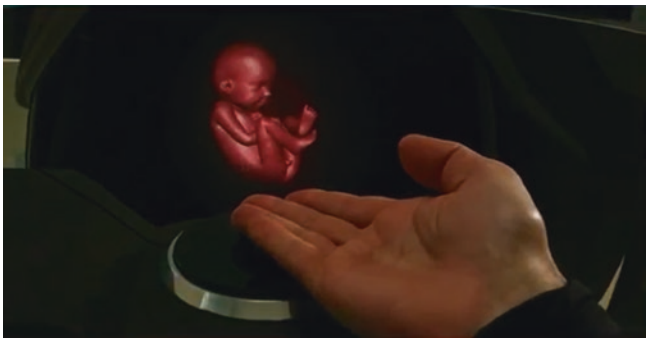


**Fig. 44.5** Static hologram created from volumetric data of 3D model of a fetus. The hologram is "floating" in mid-air above the palm of the observer

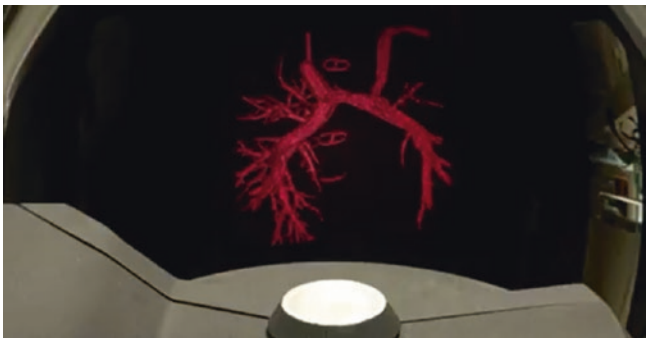


**Fig. 44.6** Bicaval bidirectional Glenn palliation in a 2-year-old patient: Static hologram created in real time from an intra-procedure 3D rotational angiogram using a Philips Allura Cath Lab System [Philips Medical, Eindhoven, Netherlands]. Contrast was injected into the right SVC and left arm [hand injection] simultaneously

**Fig. 44.7** ASD secundum in a 16-year-old girl. Dynamic color holograms created in real time from intra-procedure 3D transesophageal electrocardiography using a Philips ie33 echocardiography system [Philips Medical, Eindhoven, Netherlands]. Upper plane: The ASD secundum is demonstrated from the left atrial aspect in the center of the septum. Lower pane: The ASD is closed by a septal occlude, and the mitral valve is seen en face, while the observer holds the heart in his hand

# Integrating Imaging Modalities

# 45

Tilak K. R. Pasala, Vladimir Jelnin, and Carlos E. Ruiz

## 45.1 Introduction

Transcatheter interventions for congenital heart disease can be challenging and require not only the understanding of anatomy but a working knowledge of modern imaging modalities. Fluoroscopy has poor characterization of non-radiopaque structures and has limitations in providing three-dimensional (3D) spatial information. Similarly, echocardiography by itself has limitations in detecting the position of the catheters and wires. Additionally, the orientation of the images from various imaging modalities can be different which poses an added challenge to the operator. A real-time integration of the imaging modalities with 3D information and live fluoroscopy provides a rapid recognition and orientation of the cardiac structures during percutaneous interventions. It can also improve the communication between various members for the team while performing the procedure.

**Electronic Supplementary Material** The online version of this chapter (https://doi.org/10.1007/978-3-319-72443-0_45) contains supplementary material, which is available to authorized users.

T. K. R. Pasala · V. Jelnin · C. E. Ruiz (✉)
Structural and Congenital Heart Center, Hackensack University Medical Center and the Joseph M. Sanzari Children's Hospital, Hackensack, NJ, USA
e-mail: CRuiz@StructuralHeartCenter.org; Carlos.Ruiz@HackensackMeridian.org


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_45

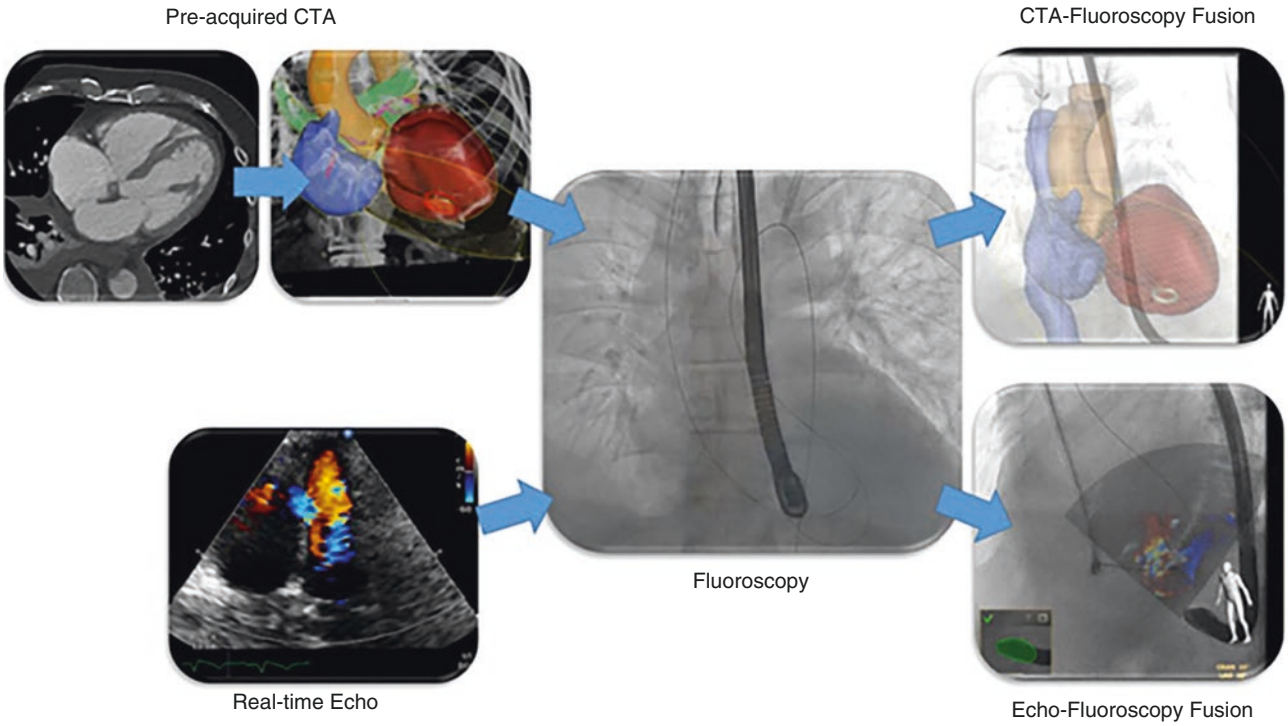



**Fig. 45.1** Integration of various imaging modalities. The pre-acquired computer tomography angiography (CTA) data is post-processed with advanced software tools and is overlaid on live fluoroscopy in a comparable scale for CTA-fluoroscopy fusion. Similarly, real-time integration of echo with fluoroscopy (echo-fluoroscopy fusion) is used during procedures at various steps

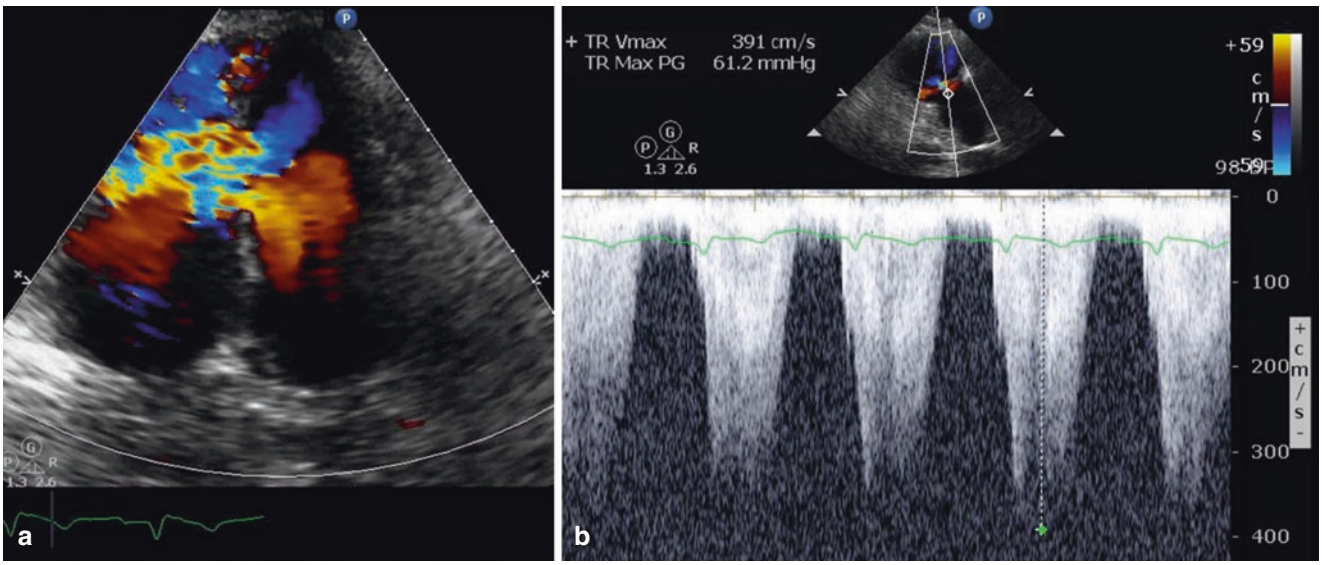



**Fig. 45.2** Case presentation. We present a case of a large muscular VSD to demonstrate the benefits of integrated imaging. (**a**) The color Doppler on the transthoracic echo (TTE) shows a large muscular VSD with predominantly left-to-right flow. (**b**) The continuous Doppler shows a significantly elevated pulmonary pressure with a right ventricular systolic pressure of 61 mmHg

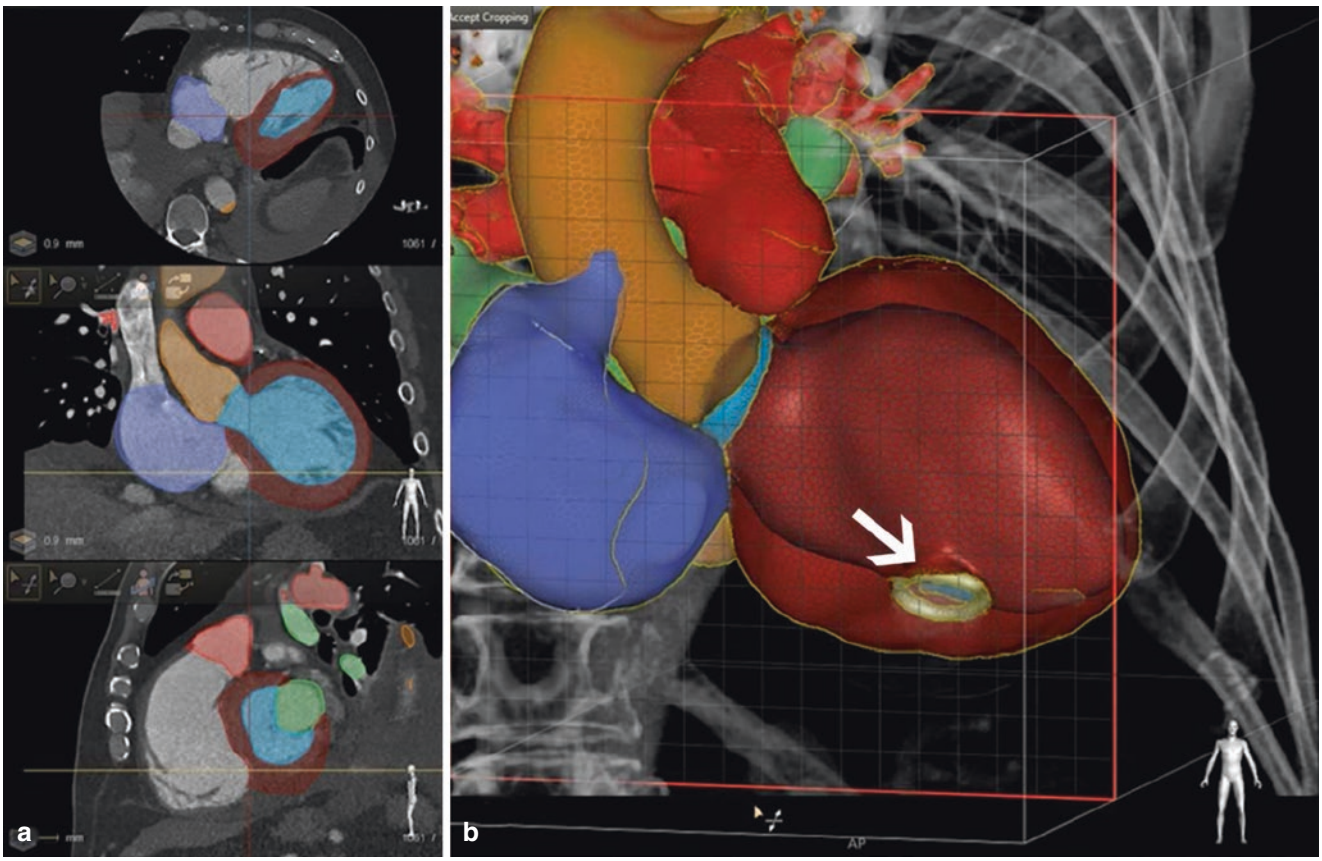



**Fig. 45.3** Cardiac computer tomography angiography (CTA)—post-processing. The acquisition of CT requires expertise and can be obtained by using helical CT, multi-row detectors (MDCT), and ECG gating. (**a**) The standard CT images with anatomical planes (axial, sagittal, and coronal) are shown. Various cardiac structures are coded with specific colors by using "model-based" segmentation method. (**b**) Specialized software (Heart Navigator, Philips Healthcare, Inc.) are used to build a 3D model of the heart. The white arrow is pointing at the ventricular septal defect reconstructed from the CT images with its exact size, shape, and precise location

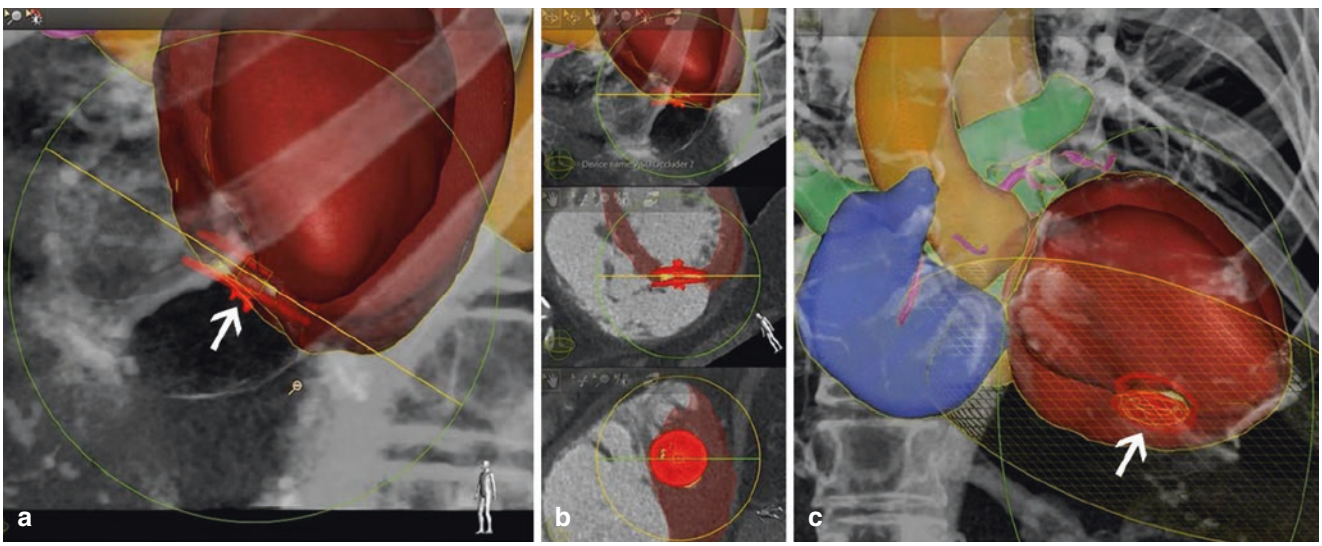



**Fig. 45.4** Cardiac computer tomography angiography—virtual planning. Virtual devices (white arrows) are placed at the position of the ventricular septal defect to make decisions on the size and shape. Additionally, the interaction with surrounding structures can be examined

**Video 1** Cardiac computer tomography angiography—virtual planning. Virtual devices (white arrows) are placed at the position of the ventricular septal defect to make decisions on the size and shape. Additionally, the interaction with surrounding structures can be examined (MP4 59345 kb)

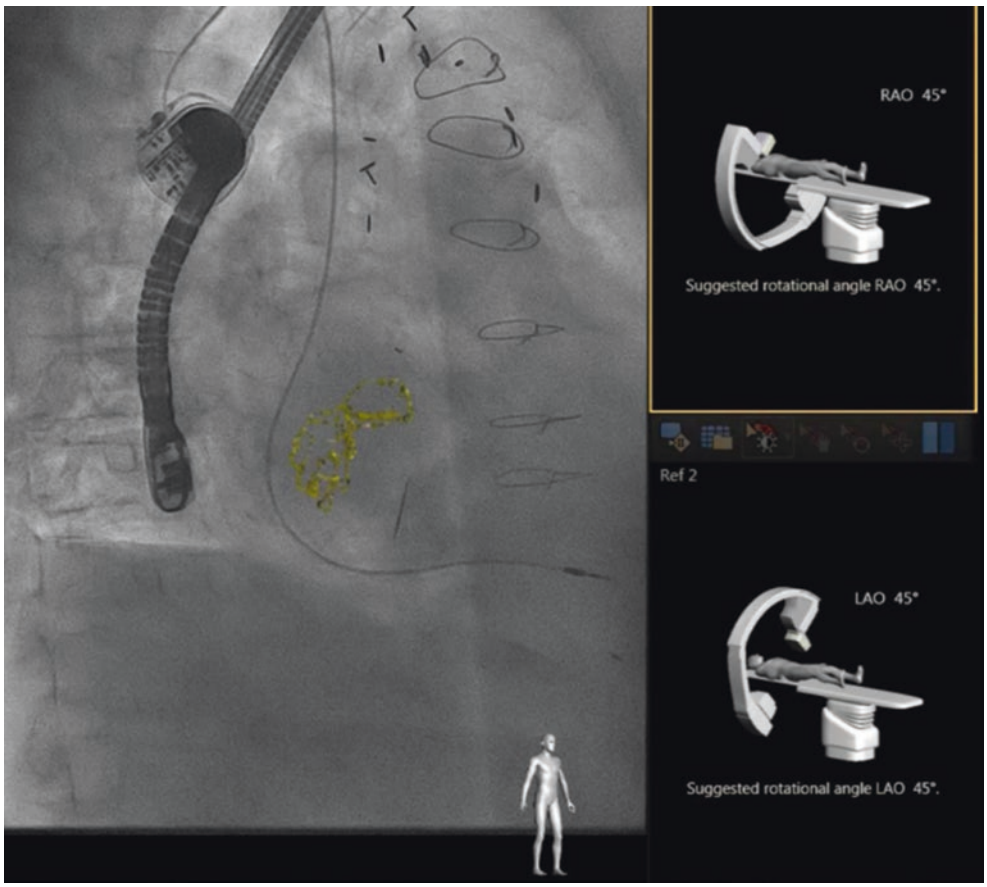



**Fig. 45.5** Registration of processed CT data over fluoroscopy. Registration is the process of orienting post-processed CTA images on live fluoroscopy. It is achieved by manually adjusting any segmented radiopaque structures, in the above example prosthetic valves (in yellow), to the same structures on fluoroscopy. Two orthogonal planes (RAO 45° and LAO 45°) are used adjust manually. After the desired position is achieved, the 3D model of the cardiac structures is overlaid which will follow the C-arm. In the case of lack of usable radiopaque structures, aortogram is performed to aid in the registration process

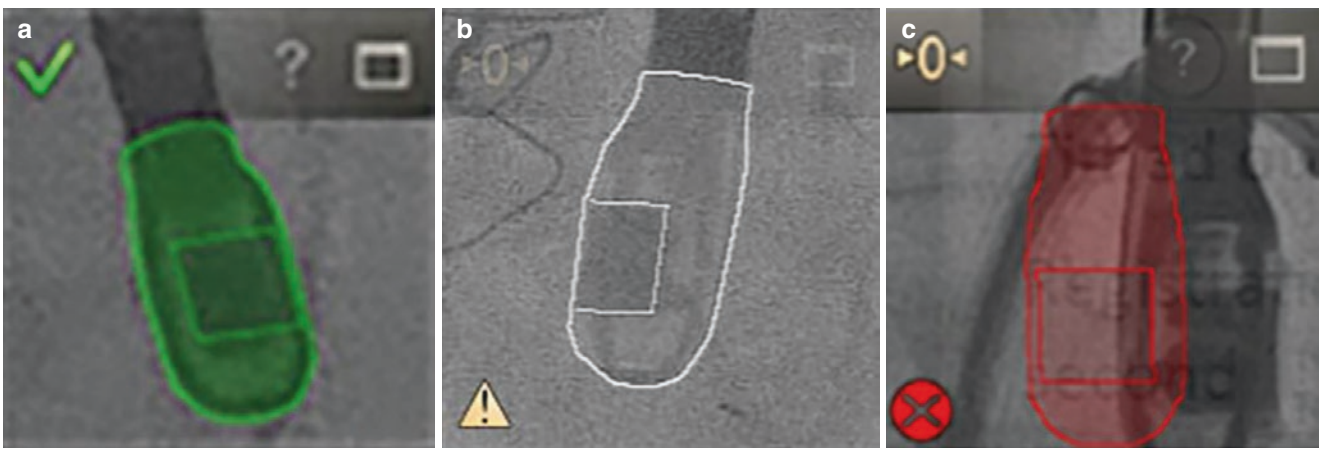



**Fig. 45.6** Registration using TEE probe for echo-fluoroscopy fusion. A small window with the TEE probe is displayed on the bottom left of the echo-fluoroscopy fusion display. When the TEE probe is seen on the *X*-ray field, specialized software (Echo Navigator, Philips Healthcare, Inc.) can perform registration automatically. (**a**) A check mark is seen, and the probe is displayed in green color confirming successful registration. (**b**) An exclamation mark is seen, and the probe is displayed in white/transparent color suggesting that the registration is timed out. (**c**) An X mark is seen with the probe displayed in red color suggesting an unsuccessful registration

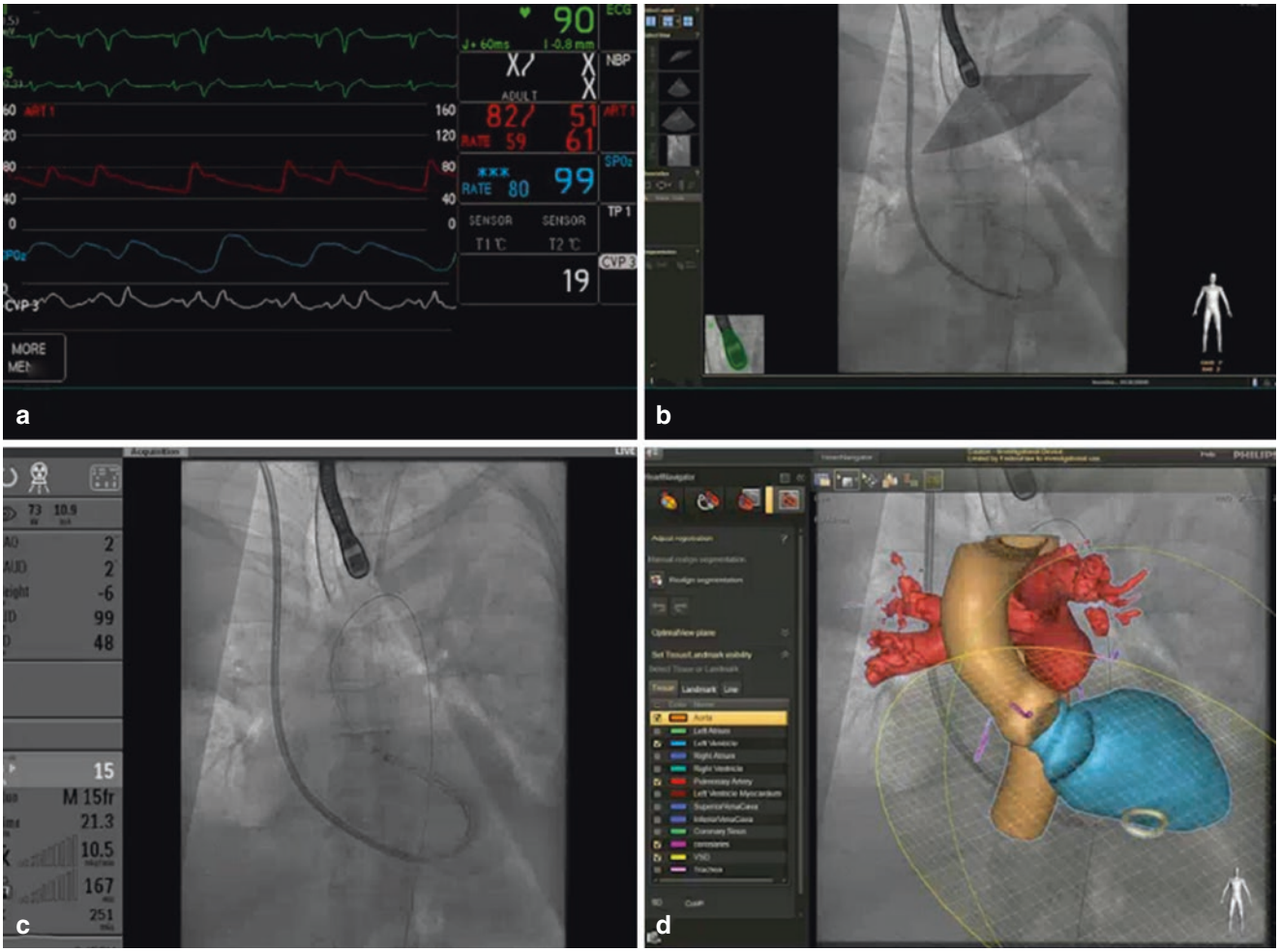



**Fig. 45.7** Display of hemodynamic and integrated imaging during procedures. Operators can have comprehensive information displayed on the screen. (**a**) Live hemodynamic data is displayed on the upper left. (**b**) Echo-fluoroscopy fusion is shown where relevant information from transesophageal echocardiography (TEE) are fused to fluoroscopy. (**c**) Live fluoroscopy is shown with catheters and wires. (**d**) CTA-fluoroscopy fusion is shown. A three-dimensional model of the heart with relevant cardiac structures is fused to fluoroscopy. The windows can be changed per operator preference

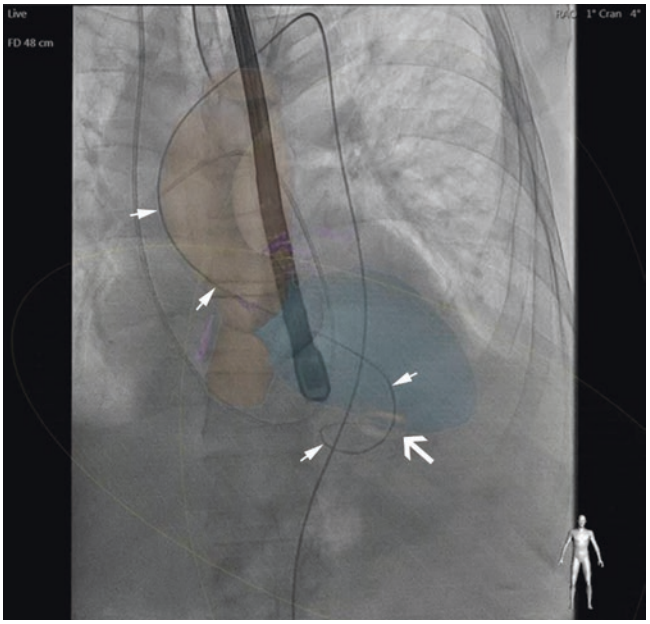



**Fig. 45.8** CTA-fluoroscopy fusion—crossing ventricular septal defect. Fusion of the CTA with live fluoroscopy shows a 3D model of the heart overlaid over the fluoroscopy during the procedure for guidance. Segmentation of the heart is visible, but it is dimmed to avoid blocking the view of the wires. A circular marker at the position of the ventricular septal defect (VSD) helps the operator in steering the guidewire. The guidewire (small white arrows) is seen crossing the ventricular septal defect (VSD) retrograde from the aorta. The guidewire is inserted into the right ventricle through the VSD (large white arrow)

**Video 2** CTA-fluoroscopy fusion—crossing ventricular septal defect. Fusion of the CTA with live fluoroscopy shows a 3D model of the heart overlaid over the fluoroscopy during the procedure for guidance. Segmentation of the heart is visible, but it is dimmed to avoid blocking the view of the wires. A circular marker at the position of the ventricular septal defect (VSD) helps the operator in steering the guidewire. The guidewire (small white arrows) is seen crossing the ventricular septal defect (VSD) retrograde from the aorta. The guidewire is inserted into the right ventricle through the VSD (large white arrow) (MP4 59628 kb)

**Video 3** CTA-fluoroscopy fusion—creating arterial-venous (AV) rail. An AV rail is created to support the delivery of device. The left ventricle is seen in blue, aorta in orange, and pulmonary arterial system in red color. The retrograde guidewire (white arrows) crossing the VSD is advanced and positioned in the left pulmonary artery. Then, a 6F JR4 guide catheter with an 18 × 30 mm Ensnare system (black arrows) is advanced through the right internal jugular vein. The retrograde guidewire is snared and externalized creating an AV rail (MP4 153400 kb)

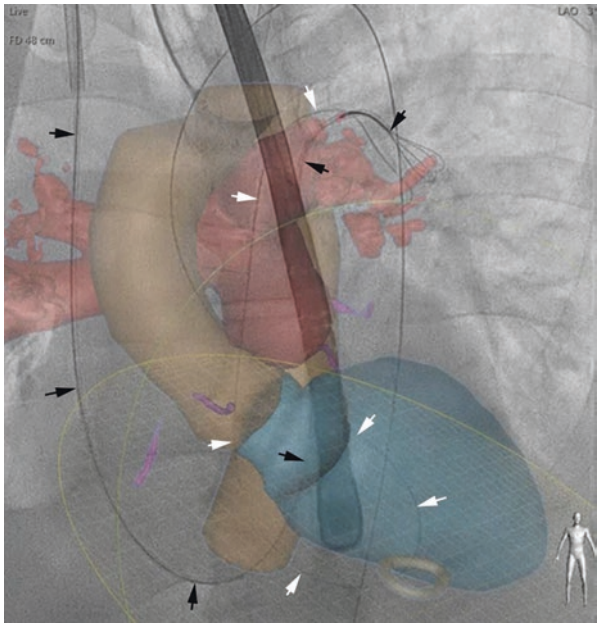



**Fig. 45.9** CTA-fluoroscopy fusion—creating arterial-venous (AV) rail. An AV rail is created to support the delivery of device. The left ventricle is seen in blue, aorta in orange, and pulmonary arterial system in red color. The retrograde guidewire (white arrows) crossing the VSD is advanced and positioned in the left pulmonary artery. Then, a 6F JR4 guide catheter with an 18 × 30 mm Ensnare system (black arrows) is advanced through the right internal jugular vein. The retrograde guidewire is snared and externalized creating an AV rail

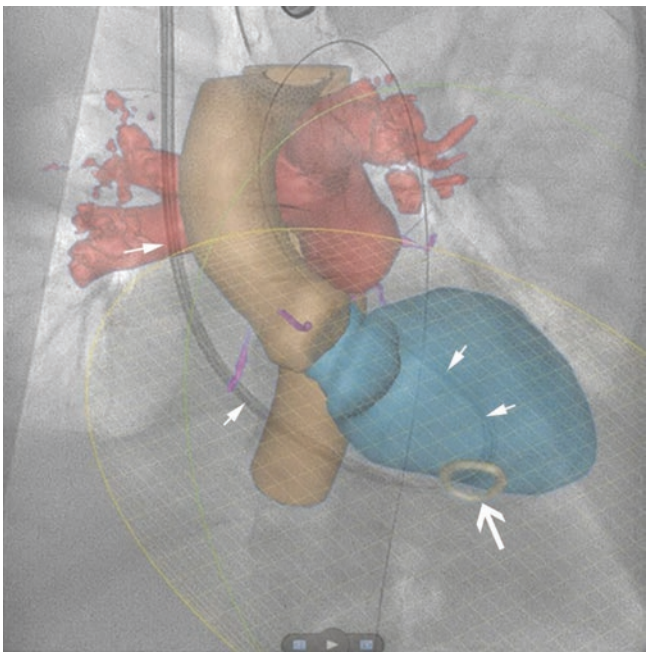


**Fig. 45.10** CTA-fluoroscopy fusion—advancing delivery sheath. Delivery sheath is introduced through the VSD (large white arrow) into the left ventricle (small white arrows) using the AV rail and is advanced into the aorta (Video 3)

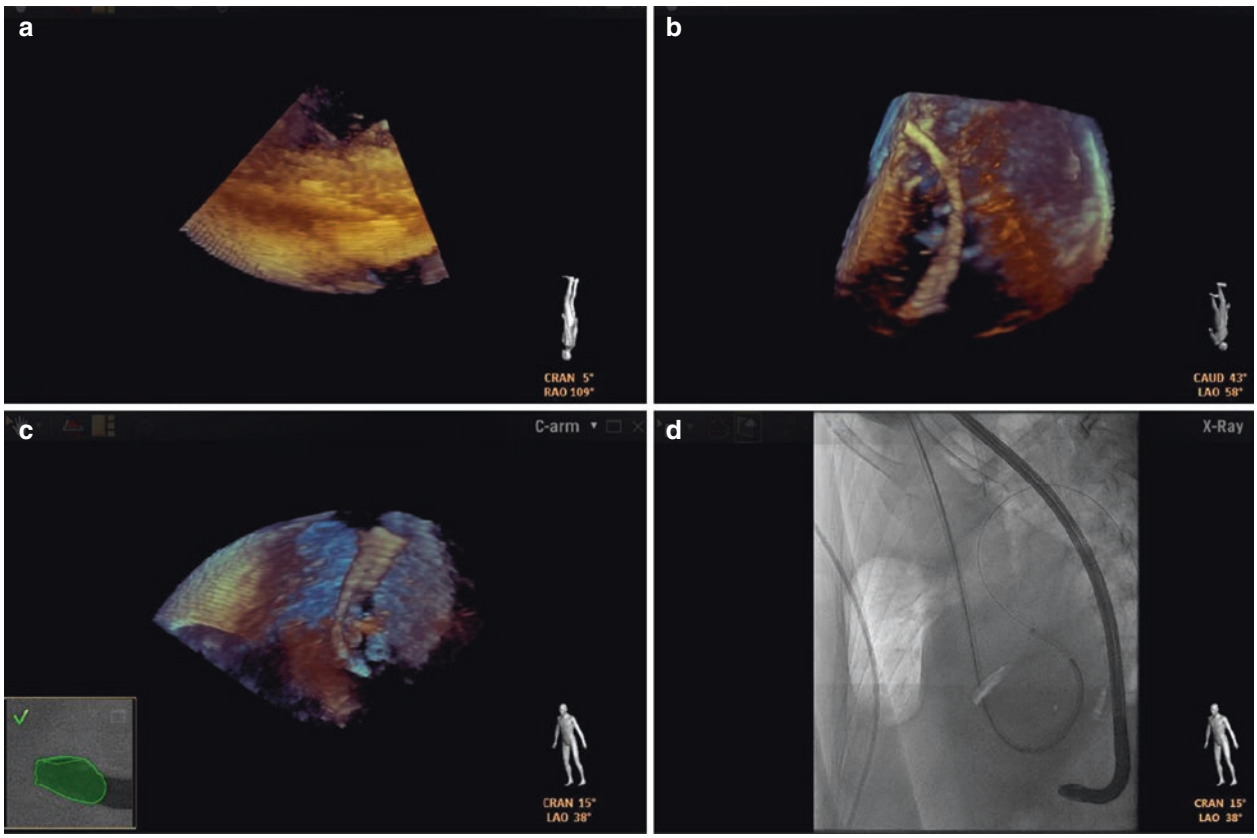



**Fig. 45.11** . Echo-fluoroscopy fusion—advancing VSD closure device. Different views of the 3D TEE are shown. (**a**) The "free view" allows the operator to cut the 3D reconstructed volume and perform 360° rotation. (**b**) This view allows the echocardiographer to perform additional review. (**c**) This view coincides with the position of TEE probe. (**d**) The C-arm view where relevant TEE images are fused with live fluoroscopy. Echo-fluoroscopy fusion helps in controlling the position of the delivery system into LV through the VSD. The VSD closure device is seen inside delivery sheath along with the security wire going from the sheath to the aortic arch (Videos 2 and 3)

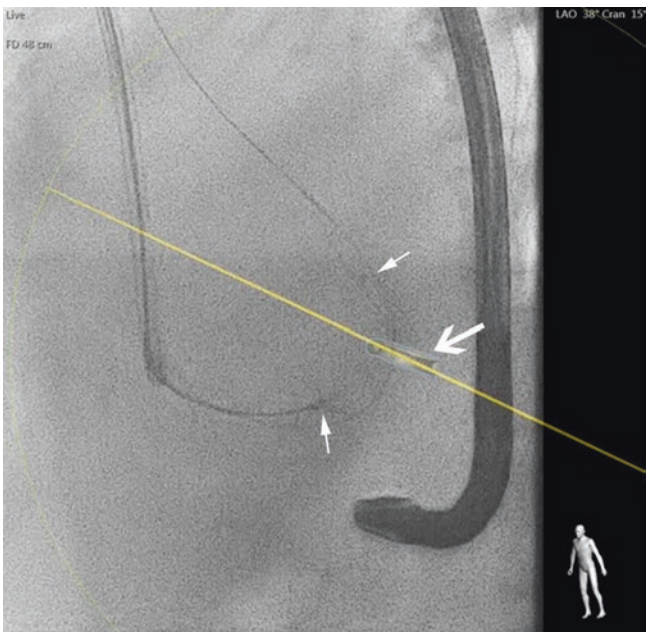



**Fig. 45.12** CTA-fluoroscopy fusion—VSD closure device delivery. The opened Amplatzer VSD closure device which is positioned at the VSD (large white arrow) and still connected to the delivery wire. Having the segmented VSD overlaid on fluoroscopy along with echo-fluoroscopy fusion aids the operator in positioning the VSD device

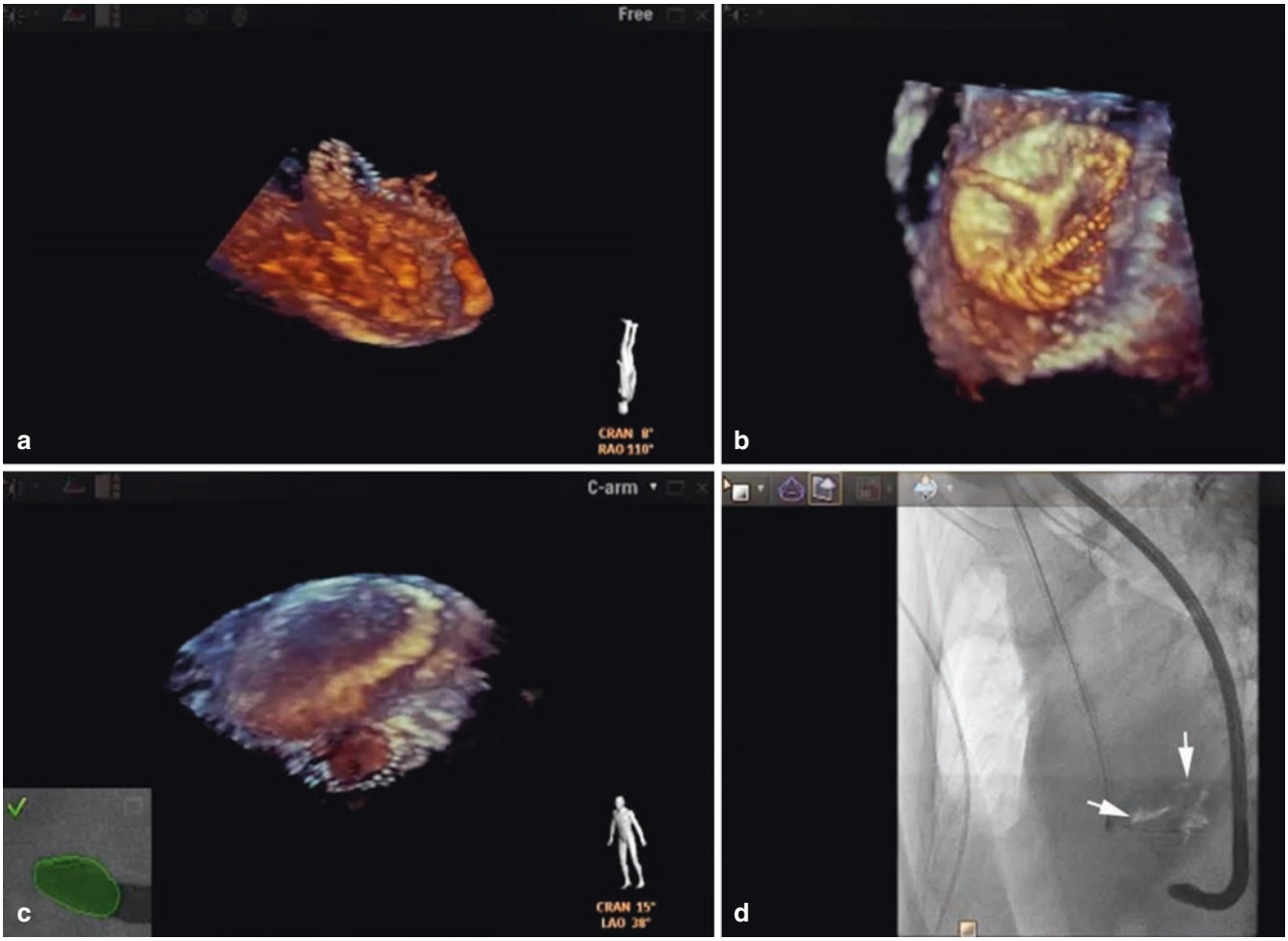



**Fig. 45.13** Echo-fluoroscopy fusion—confirmation of device position**.** The most important functions of echo-fluoroscopy fusion are positioning the VSD closure device and confirming the device position. (**a**, **b**, **c**) The 3D TEE images of the device are shown. These images aid in the positioning of the device with the help of echo-fluoroscopy fusion (**d**). The left disc of the Amplatzer device (**d**, white arrows) is anchored on the LV side, and the right disc is attached to the delivery wire. Echo-fluoroscopy fusion adds anatomical relevance to the fluoroscopic image

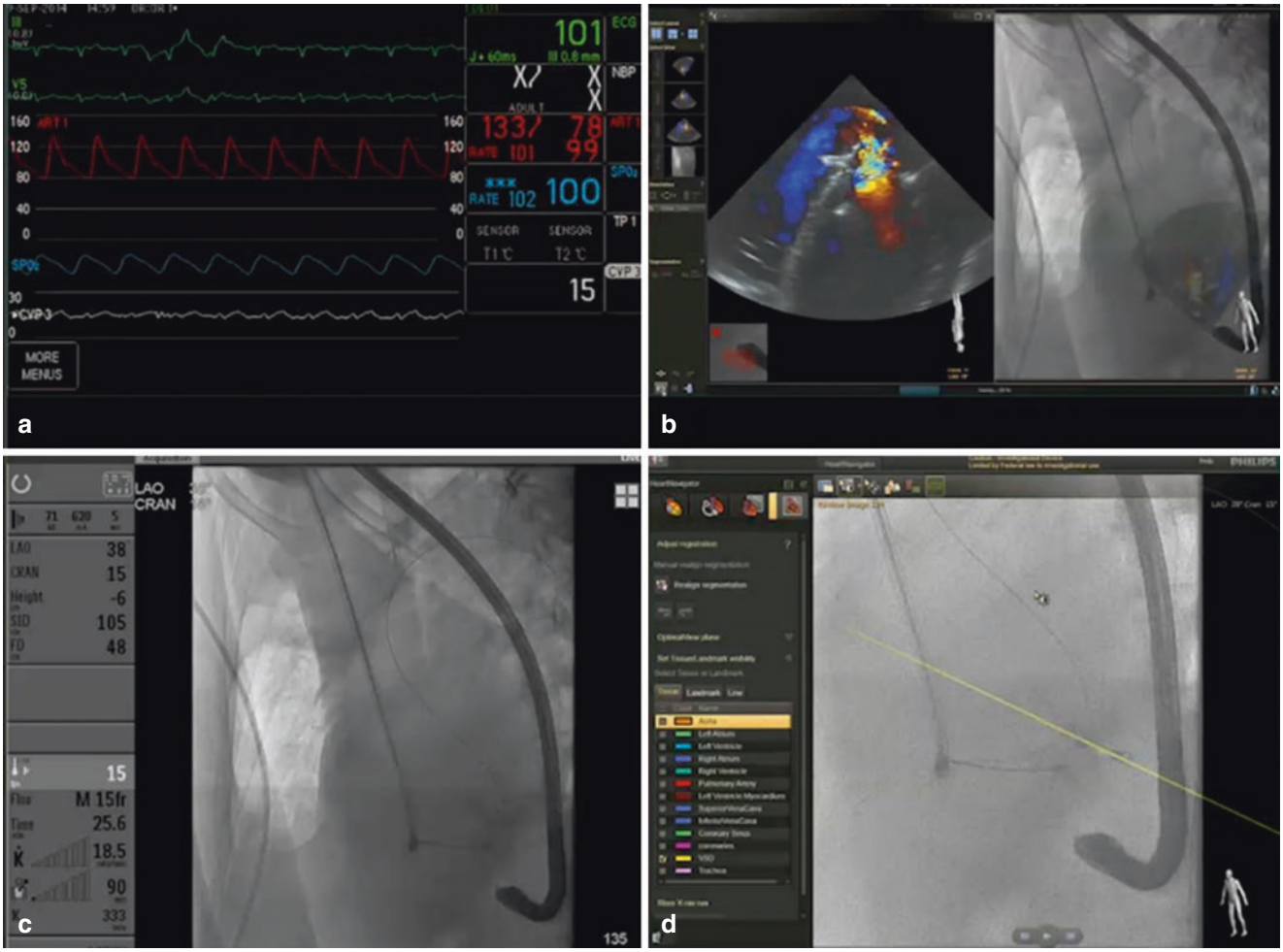



**Fig. 45.14** Integrated imaging—final stages of VSD closure. The final stage of the VSD closure procedure using integration of the different imaging modalities is shown. (**a**) Vitals of the patient. (**b**) Echo-fluoroscopy fusion showing the color Doppler overlay on fluoroscopy. The color Doppler demonstrates a mild residual flow around the Amplatzer VSD closure. (**c**) Live fluoroscopy showing the VSD device. (**d**) CTA-fluoroscopy fusion showing the device that is still attached to the delivery wire. After a thorough assessment, the device is deployed, and its position and stability are checked (Videos 2 and 3)

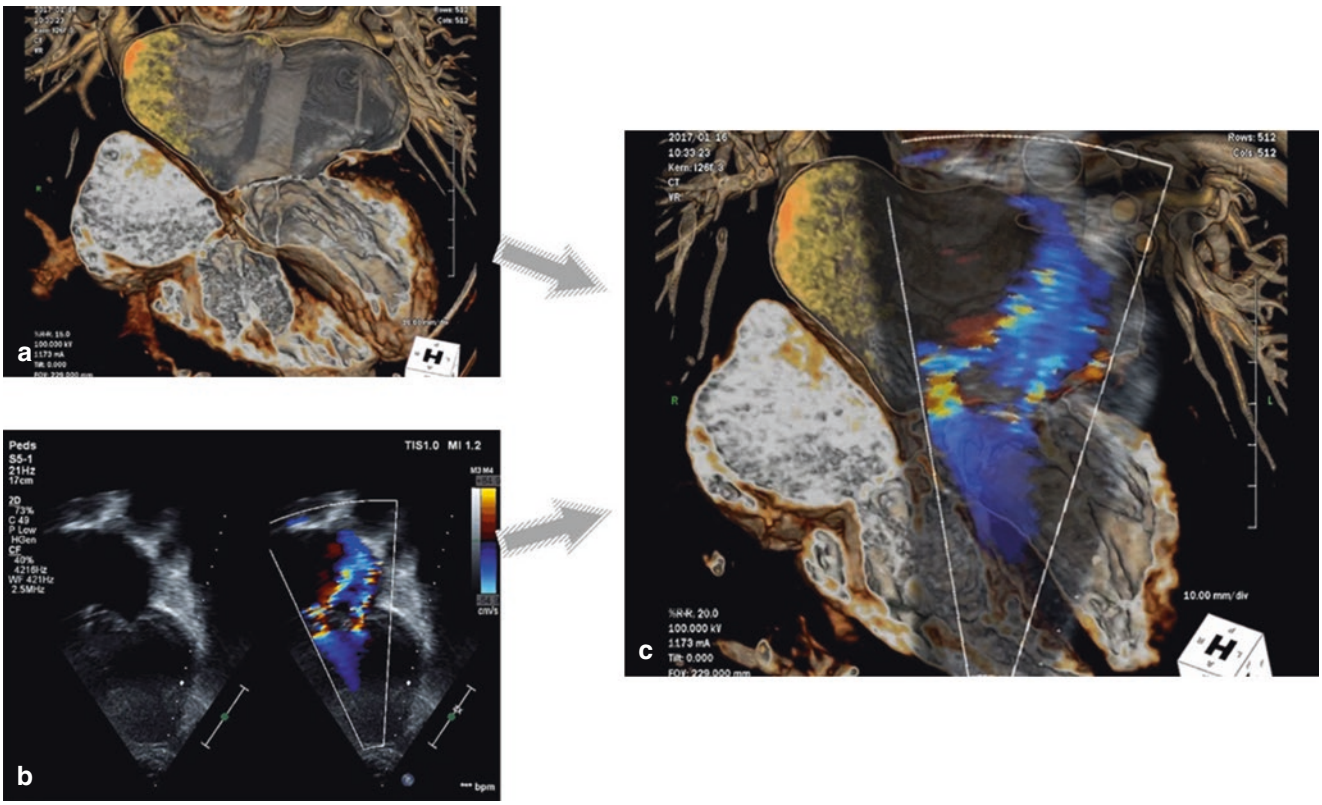



**Fig. 45.15** Future direction—echo-CTA fusion. In the future, the integration of processed CTA (**a**) and live echocardiography (**b**) has the potential to decrease the overall radiation dose and contrast use during procedures. In the above image, two separate jets of mitral regurgitation are seen in a patient with repaired endocardial cushion defect. The origin of the two jets is seen on color Doppler (**b**, **c**). One of the jets is originating from the lateral cleft of left-sided valve and the other from the base of the left-sided valve due to dehiscence

# Correction to: Atlas of Cardiac Catheterization for Congenital Heart Disease

Gianfranco Butera, Massimo Chessa, Andreas Eicken, and John D. Thomson

**Correction to:**
**G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease,***
**https://doi.org/10.1007/978-3-319-72443-0**

This book was inadvertently published with patients' names in figures and videos for the following chapters

a. Chapter 1, Fig. 1.9
b. Chapter 20, Figs. 20.8, 20.12, 20.13, and all videos
c. Chapter 28, Fig. 28.12
d. Chapter 30, Video 1

Patient's names have been removed from the book.

The updated online version of these chapters can be found at
https://doi.org/10.1007/978-3-319-72443-0_1
https://doi.org/10.1007/978-3-319-72443-0_20
https://doi.org/10.1007/978-3-319-72443-0_28
https://doi.org/10.1007/978-3-319-72443-0_30


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_46

# Correction to: Reopening of Peripheral and Central Arteries and Veins

Henri Justino

**Correction to:**
**G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease,***
**https://doi.org/10.1007/978-3-319-72443-0**

This chapter was originally published without the author "Athar M. Qureshi". The author name has been included in the corrected version of the chapter.

The updated online version of this chapter can be found at
https://doi.org/10.1007/978-3-319-72443-0_15


G. Butera et al. (eds.), *Atlas of Cardiac Catheterization for Congenital Heart Disease*,
https://doi.org/10.1007/978-3-319-72443-0_47