Interventions in Adult Congenital Heart Disease

A Case-Based Approach

Vaikom S. Mahadevan
Editor

MOREMEDIA

Springer

Interventions in Adult Congenital Heart Disease

Vaikom S. Mahadevan
Editor

Interventions in Adult Congenital Heart Disease

A Case-Based Approach

Springer

Editor
Vaikom S. Mahadevan
University of Massachusetts
Worcester, MA, USA

ISBN 978-3-030-85407-2 ISBN 978-3-030-85408-9 (eBook)
https://doi.org/10.1007/978-3-030-85408-9

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

Foreword

When my colleague Vaikom Mahadevan asked me to write a Foreword for his new textbook *Interventions in Adult Congenital Heart Disease*, I agreed without hesitation. The very next day, however, I wondered: "Is there a difference between a Foreword and a Preface, and if so, what is it?" I am embarrassed to say that although I had a fair amount of experience writing Prefaces, I had not previously been asked to write a Foreword. And so, I went to Google where I l learned that a **Foreword is written by someone other than the book's author** and tells potential readers why they should read the book. In contrast, a Preface is written by the author, and tells readers why he or she undertook to write the book in the first place. This was very good news for me, because in the 8 years that I have had the pleasure of working with Dr. Mahadevan here at UCSF, it has become abundantly clear why he is one of the world's leaders in catheter-based interventions for treatment of adult congenital heart disease, as well as structural heart disease in general. He knows his stuff!

As is appropriate and proper, this comprehensive textbook starts out with the basics. The first chapter, by Drs. David Teitel and Elena Amin in Pediatric Cardiology here at UCSF, reviews the fundamentals of vascular access, hemodynamic measurements, and shunt calculations. Written by two experts with many years of experience, this chapter provides an essential starting point for the physician intent on mastering diagnosis and treatment of the patient with congenital heart disease, or indeed any form of structural problem within the heart leading to altered hemodynamics. This beginning section of the book is followed in sequence by sections on venous interventions, interatrial and ventricular septal interventions, valvular interventions including prosthetic valve implantations, and also interventions to attenuate or eliminate valvular leaks. There are also sections on innovative approaches to Eisenmenger physiology, as well as interventions in univentricular physiology, coronary interventions in congenital heart disease, interventions in pregnancy, and an intriguing chapter on 3D modeling in congenital cardiac interventions. All the chapter authors are renowned experts in the field of adult congenital cardiac intervention.

It is not an exaggeration to say that Dr. Vaikom Mahadevan is truly a world-renowned expert in interventional techniques for treating adult congenital heart disease (ACHD) and structural heart disease, including replacing all four valves of the heart. He has brought his expertise to the organization and editing of this outstanding textbook. It is easy to read and well illustrated,

both for the beginner and the experienced interventionist. Accordingly, it is with great enthusiasm that I recommend this book to cardiologists and cardiology trainees embarking on a career in interventional cardiology. They will learn much from it and will not be disappointed.

William Grossman
University of California
San Francisco, USA

Preface

The increasing survival of children born with congenital heart disease to adulthood due to advances, initially in congenital cardiac surgery, led to the development of the specialty of Adult Congenital Heart Disease (ACHD). This development started in the late 1990s and now has become a well-established area of its own.

The developments in interventional cardiology evidently have made a huge impact in the treatment of these patients. It would be worthwhile to note that many of the structural cardiac interventions being done today with large bore access have their origins in the treatment of patients with congenital heart disease such as stenting of aortic coarctation and septal defect closures. In fact, the first transcatheter valve implant in a human was done in the year 2000 in the pulmonic position, as treatment for a patient with congenital heart disease, long before transcatheter aortic and other valve replacements were performed.

While there are many guides for adult cardiac interventional procedures, ACHD intervention by its nature of being done in specialist, mostly academic centers, with a relatively smaller pool of skilled operators, does not have a large amount of such guidance for these procedures. In the era of such advanced ACHD interventions, the aim of this book is to bring together the expertise of skilled adult congenital interventional cardiologists from the United States, Europe, and other parts of the world to provide an easy guide to these complex interventions using a simple case-based approach. While the emphasis is largely on the interventional techniques, the indications for such interventions are equally important to understand for successful outcomes, and these are also addressed in each of the chapters. Congenital cardiology was one of the early adopters of the "heart team" approach, long before it becomes routine in acquired heart disease cardiac care, and the interventions described in this book are based on such consensus by the respective authors' teams.

I would like to sincerely thank all the chapter authors for kindly sharing their time and expertise, all of whom are established experts in this field, and Dr. Sarah Blissett for providing her valuable input on chapter format and titles, with her background of being an ACHD cardiologist and an educator.

Finally, but not least, this project would not have been possible without the support of my family, my parents, Mrs. and Mr. Mani, my wife Kritika, and my children Manish and Harishnarayanan who, with all their love and affection, make all my efforts worthwhile.

I hope all the readers will find this book a very useful guide to treat and help their ACHD patients, whose well-being is the ultimate goal for all the physicians involved in the care of this population.

Worcester, MA, USA Vaikom S. Mahadevan

Contents

Part I

Fundamentals of Catheterisation in Patients with ACHD

Principles of Vascular Access, Hemodynamic Measurements, and Shunt Calculations

David Teitel and Elena Amin

Abstract

Knowledge of the fundamental principles of vascular access, hemodynamic measurements, and shunt calculations is essential prior to undertaking any cardiac catheterization procedure. Vascular access site selection in adults with congenital heart disease is frequently complicated by multiple prior catheterizations, the congenital or post-surgical anatomy, and the delivery systems required for deployment of an ever-growing array of stents, devices, and valves. Likewise, hemodynamic measurements and shunt calculations are far more complex in adults with congenital heart disease compared to those with structurally normal hearts. Performing cardiac catheterization in a way that obtains accurate data with careful intra-procedural interpretation is essential to ensure the best contemporaneous and post-intervention care.

Keywords

Vascular access · Hemodynamics · Cardiac index · Shunt calculation

Introduction

Adult congenital heart disease encompasses a wide variety of anatomy and presentation. Some patients with congenital heart defects either were identified in adulthood or did not require an intervention until later in life. Others have congenital heart defects that were repaired or palliated in infancy or childhood, with hemodynamic sequelae of the defect itself or of the intervention. Most adult patients with significant congenital heart disease, such as tetralogy of Fallot, fall into this latter category. Those requiring palliation, often with multiple staged interventions, such as patients with single ventricle physiology, likely underwent many cardiac catheterizations and surgical procedures during their lifetime. Catheter interventions on adults with congenital heart disease may also be directed toward acquired cardiovascular problems, such as iatrogenic pulmonary vein stenosis following electrophysiology ablation procedures. The approach to vascular access, hemodynamic measurements, and shunt calculations is universally applied to all of these patients, the complexity of which is dependent on the underlying defects, previous interventions, and the current clinical status.

D. Teitel · E. Amin (✉)
UCSF School of Medicine, Benioff Children's Hospital, San Francisco, CA, USA
e-mail: David.Teitel@ucsf.edu; Elena.Amin@ucsf.edu

V. S. Mahadevan (ed.), *Interventions in Adult Congenital Heart Disease*,
https://doi.org/10.1007/978-3-030-85408-9_1

Case 1: Complex Congenital Heart Disease with Limited Vascular Access

Prior History, Current Symptoms, and Outcome

A young woman with hypoplastic left heart syndrome underwent staged surgical single ventricle palliation, consisting of a modified Norwood procedure with a right ventricle to pulmonary artery conduit in the neonatal period, followed by a cavopulmonary anastomosis with conduit takedown in early childhood. She underwent multiple diagnostic cardiac catheterizations for hemodynamic assessment and angiography for surgical planning, and interventional catheterization procedures to address recurrent coarctation of the aorta. Consequently, she developed multiple vessel occlusions including the right internal jugular vein and the bilateral femoral veins and arteries. She never underwent the third and final single ventricle palliation, the modified Fontan procedure, because she had poor right ventricular function. She required a cardiac transplantation for right ventricular failure at 10 years of age.

Physical Examination

Occluded femoral arteries were evident by absence of femoral arterial pulses with normal capillary refill time indicating adequate arterial supply to the lower extremities via collateral arterial vessels.

Investigations

Ultrasound evaluation of vessels was performed in the cardiac catheterization laboratory prior to access attempts to determine vessel patency. When vessel occlusion was suspected by ultrasound or failure to obtain access, angiography through distal vessels was performed to confirm occlusion. When distal vessels were not accessed, formal doppler ultrasound evaluations of the upper and lower extremity vasculature were performed to document vessel occlusions.

Management Options, Procedural Management, and Outcome

For catheterization procedures after vessel occlusions or surgical isolation of vessels, access to her cavopulmonary system for angiography was achieved via sheath placement in the left internal jugular vein as shown in Fig. 1a. Access to her cardiac chambers, pulmonary veins, and aorta was achieved with transhepatic access (Fig. 1c, d), which was performed on several occasions, as

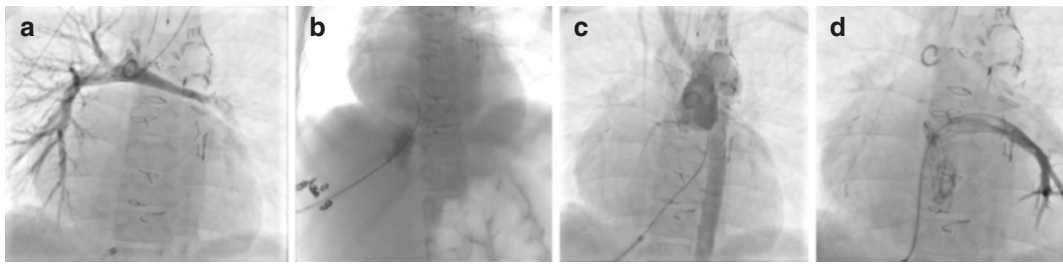

Fig. 1 Patient with hypoplastic left heart syndrome s/p single ventricle palliation with a bidirectional cavopulmonary anastomosis, and with a history of multiple vascular occlusions. (**a**) Bidirectional cavopulmonary anastomosis angiogram performed via a pigtail catheter advanced antegrade from the left internal jugular vein to the superior vena cava to right pulmonary artery anastomosis; (**b**) Wire through the transhepatic access tract with tip in the right atrium and injection of contrast confirming the right atrium to hepatic vein anatomy and course prior to sheath advance. Multiple embolization coils are visible in the lateral right subcostal region after multiple previous transhepatic access tract closures; (**c**) Ascending aortogram performed via a pigtail catheter advanced via the transhepatic access sheath to the right atrium to the single right ventricle and antegrade to the aorta; (**d**) Left lower pulmonary vein angiogram performed via an end hole catheter advanced via the transhepatic access sheath to the right atrium, across the atrial septectomy to the left atrium and pulmonary vein

evidenced by multiple radio-opaque coils in the subcostal space from transhepatic tract closures (Fig. 1b).

Following her orthotopic heart transplant, routine post heart transplant hemodynamic assessments with right heart catheterization and endomyocardial biopsy were performed via the left internal jugular vein. For annual cardiac catheterizations, when hemodynamic measurements from the left heart and coronary angiography were desired, arterial access was secured via the axillary or brachial artery as her bilateral radial arteries were occluded and the ulnar arteries were diminutive.

This case illustrates the creative solutions required to obtain vascular access in patients with complex congenital heart disease who require multiple cardiac catheterization procedures. Alternative management options and post-procedure management of patients with vessel occlusions are discussed below in the principles of vascular access section.

Discussion and Brief Literature Review

Principles of Vascular Access

Vascular access to the arterial and/or venous systems is essential to perform any type of diagnostic or interventional catheterization procedure. Site selection may be complicated in adults with congenital heart disease, particularly those who have undergone cardiac surgery, isolating portions of their vasculature from intracardiac structures, or have multiple vessel occlusions caused by repeated catheterization procedures. The potential vascular sites detailed below are by no means comprehensive, and creative access routes are frequently required in patients with adult congenital heart disease. Depending on the etiology and timing of the occlusion, some vessels may be amenable to recanalization; however, careful consideration is required prior to performing an intervention via a freshly recanalized vessel, given the inherent increased risks of hemorrhage and further vessel damage associated with these procedures [1, 2]. With the array of new devices available for complex transcatheter interventions in recent years, the choice of the optimal access site for device delivery in terms of catheter course, vessel size, and subsequent vascular integrity has become increasingly complicated. It is reasonable to start with the largest venous sheath likely to be required for the diagnostic portion of the case; for example, a sizing balloon may be required prior to device delivery. This limits the number of sheath exchanges, each of which may theoretically contribute to vessel injury. Selecting the smallest sheath possible to complete the diagnostic portion of the case is more prudent when accessing the arterial vessels, given their less compliant and more calcific profile, and the serious consequences of vessel disruption or occlusion in the adult population. In that regard, whenever possible, it is best to avoid arterial access; for example, left heart hemodynamics can be obtained via venous access when there is an interatrial communication, or via transseptal puncture. For those who require only arterial monitoring but not full sided hemodynamics or angiography, a peripheral arterial line can be considered as an alternative, to reduce the risk of vessel damage and occlusion. Reported complications of vascular access include hemorrhage, hematoma, pseudoaneurysm, acute or chronic vessel occlusion, arteriovenous fistula, and vessel dissection. Most of these can be avoided with a stepwise procedural approach using ultrasound guidance and fluoroscopy confirmation of wire position, and with attention to post-procedure hemostasis with the minimum of pressure, and with close monitoring of the access site.

Site Selection The following points are considered when selecting the most appropriate site for vascular access:

- Prior access
  - For example, significant right femoral venous narrowing may be present after repeated cardiac catheterizations via that vessel, which may make the left femoral vein more favorable for the large sheaths

required for a percutaneous valve placement, despite the slight increase in tortuosity of the sheath course.

- Known vessel occlusions
  - Prior difficulty accessing a vessel with or without documented vessel occlusions, by ultrasound or angiography, may require creative alternative access points such as transhepatic access (Case 1).
- Congenital vascular anatomy
  - For example, patients with heterotaxy (left atrial isomerism) may have a congenitally interrupted inferior vena cava with azygos continuation. Selection of an alternate route to the intracardiac structures via access to the internal jugular vein is likely to facilitate successful interventions (Case 3).
- Congenital and post-surgical intracardiac anatomy
  - Patients post cavopulmonary anastomosis (bidirectional Glenn circulation) will require separate access to the pulmonary arteries, usually via the right internal jugular vein, than the intracardiac chambers, usually via the femoral veins, and occasionally, the femoral arteries.
- Location of defects likely to require intervention
  - For example, delivery of a device for a muscular ventricular septal defect is often more likely to succeed with a sheath coursing from the right internal jugular vein than from a femoral vein.

General Vascular Access Technique One of the most significant changes to percutaneous vascular access in the catheterization laboratory in recent years has been adoption of the standard use of ultrasound to guide access. While it is possible to access the vessels using bony landmarks, location of pulses and acquired skill, ultrasound is a useful adjunct. 2D and color imaging of the bilateral femoral vessels, and, if relevant, the neck vessels, prior to vascular access can lessen access times and avoid attempts at accessing occluded vessels. It is also helpful to image the vessels while administering local anesthesia and accessing the vessel, to confirm needle trajectory and vessel entry. In adult congenital heart disease patients who have developed significant groin or neck scar tissue after multiple cardiac catheterization procedures, a stepwise introduction of a stiff Micropuncture (Cook) introducer over the initial access wire followed by exchange for a stiff wire and then serial incremental dilators is required prior to successful placement of the desired sheath.

Femoral Artery and Vein In patients who require both arterial and venous access, or for those who are likely to require a structural intervention, the femoral vessels are most often selected. It is critical to make sure that the vessel entry site is inferior to the inguinal ligament, which runs between the anterior superior iliac spine and the pubic tubercle, so that the entry site can be adequately compressed after removal of the sheath. Entry to the vessel above the inguinal ligament risks a retroperitoneal hemorrhage that may not be immediately identified in the catheterization laboratory and cannot be managed with manual compression. Ultrasound visualization of the bilateral femoral vessels with confirmation of patency, bifurcation sites, and assessment of degree of calcification is performed to guide site selection, vessel entry point, and the use of vascular closure devices at the conclusion of the procedure. The femoral vessels are entered at a 45° angle. A heparinized saline filled syringe for gentle aspiration to confirm vascular entry is helpful in most cases. In cases with high venous pressure or systemic desaturation, wire confirmation in the inferior vena cava or aorta can be performed with fluoroscopy or by connecting a cannula or Micropuncture introducer to pressure or sampling saturation prior to exchanging the introducer for a larger sheath. In some patients, particularly in obese patients, a doppler needle can be useful to access vessels and to distinguish the pulsatile signal of the artery compared to the phasic venous flow signal.

Internal Jugular and Subclavian Veins Right internal jugular vein access is the first-line alternative in cases of bilateral femoral vein occlusion. It is also often the selected site of venous

access in patients undergoing right heart catheterization only, because the post-procedure flat time is much shorter than with femoral venous access. This allows for earlier discharge. The right internal jugular vein is preferred over the left because it affords a more direct route and straight trajectory to the superior vena cava, and the proceduralist can more easily work to the right of the patient, which minimizes radiation in a biplane laboratory. If the right internal jugular vein is occluded, the left internal jugular vein is often used; using a longer sheath with the tip in the superior vena cava may simplify catheter manipulation. Ultrasound-guided access is particularly useful for the internal jugular veins, to avoid inadvertent entry into the adjacent structures including the carotid artery, trachea, and lung. The internal jugular vein is located deep to the sternocleidomastoid muscle, slightly lateral and anterior to the carotid artery. In cases where the identity of the vessels is unclear, which may be the case if the central venous pressure is high and both vessels appear pulsatile, the internal jugular vein can be distinguished from the carotid artery by its compressibility when pressure is applied via the ultrasound probe. A doppler needle can also be useful in this circumstance to confirm entry to the internal jugular vein rather than the carotid artery. Fluoroscopic confirmation of wire position in the right atrium (or the pulmonary artery in patients with cavopulmonary anastomosis) is useful prior to advancing the sheath. The subclavian vein, located beneath the clavicle and caudal to the subclavian artery, is an alternative access site in instances of multiple venous occlusions.

Transhepatic Access Transhepatic access is considered in cases with multiple vessel occlusions for whom there is no alternative route to the intracardiac chambers. Patients with multiple vessel occlusions likely require many cardiac catheterizations, and can therefore have multiple transhepatic access procedures and tract closures (Case 1). They tend to be well tolerated and do not appear to result in overt long-term hepatic dysfunction. There are, however, significant risks to transhepatic access. The most common is bleeding, although there is potential for damage to the bile ducts, portal vein thrombosis, gallbladder perforation, and other intra-abdominal complications [3]. Ultrasound guidance can also be helpful for transhepatic access, by identifying large hepatic veins and planning a trajectory that is likely to be successful. With ultrasound and fluoroscopic guidance, the needle enters the right subcostal skin usually between the mid-axillary and mid-clavicular line aiming posterior, leftward and superiorly toward the junction of the inferior vena cava and right atrium. Gentle suction on a contrast-filled syringe is applied during needle advancement. When the needle tip has been advanced close to the inferior vena cava a small amount of contrast is injected to confirm entry to a hepatic vein. A long needle may be required for transhepatic access. Some centers routinely use a 22-gauge Chiba needle (Inrad, Kentwood, MI), which is 15 or 22 cm long; the obturator is left in place until the needle tip is close to the inferior vena cava. Once position is confirmed by injection of contrast into a hepatic vein, an 0.018″ floppy guidewire is advanced through the needle into the inferior vena cava and right atrium. The needle is then exchanged for a long sheath and serial exchanges of dilators over a stiff wire are then feasible, to secure a large sheath if necessary. One advantage of the transhepatic access route is for interventions which are directed at the atrial septum; the course to the septum is short and fairly straight. On the other hand, manipulation of catheters to the right ventricle and pulmonary arteries, and the superior vena cava, can be more challenging than via other venous access sites. At the conclusion of the procedure, manual pressure to the subcostal region is applied and, in most cases, the transhepatic access tract is closed to prevent hemorrhage. Closure of the transhepatic tract at the liver capsule can be effectively achieved using various methods, including coils (Case 1), vascular plug type devices, or absorbable gelatin compressed sponge such as Gelfoam (Pfizer, New York, NY).

Alternative Arterial Access Access to the aorta has historically been obtained via a femoral arterial sheath. In keeping with the adult standard

cardiac catheterization for coronary artery disease, it is possible to complete the diagnostic portion of a left-sided cardiac catheterization via the radial, ulnar artery, or brachial artery. This can be particularly useful for adult congenital heart disease patients, who may have bilateral femoral artery occlusion (Case 1), but these arteries may not accommodate the large sheaths required for structural interventions (for example, stent dilation of a coarctation of the aorta). Common carotid artery or axillary artery access is then considered. In infants the use of percutaneous common carotid artery access has increased, with great success over recent years [4]. It offers a straight course to the aortic valve and to the more vertical patent ductus arteriosus seen in some patients with pulmonary atresia. In adults, despite the straightforward delivery sheath course, the carotid arteries have been avoided due to carotid calcification and stenosis, with the attendant risk of neurologic insult. Data from transcatheter aortic valve replacement studies in recent years confirms transcarotid access, via a surgical cutdown, is a viable alternative for large-bore sheaths in cases of bilateral femoral artery occlusion [5, 6]. There is also growing experience, also largely gained through transcatheter aortic valve replacement cases, with transcaval access and transapical access as alternate venous and arterial routes [6–8]. In older patients, with likelihood of significant vessel calcification, pre-procedural CT angiography of the aorta and iliofemoral vessels in addition to ultrasound aids in planning access sites and sheath selection.

Vascular Closure Devices In most diagnostic cardiac catheterization cases for adults with congenital heart disease the vascular access sites can be closed with manual pressure alone. For interventional cases, with the frequent need for large-bore sheaths for delivery of transcatheter valves and for complex stent angioplasty cases, there has been a concomitant use of vascular access closure devices. The most commonly used devices employ a suture-based closure such as ProGlide Perclose (Abbott, Abbott Park, IL) to achieve vessel closure and hemostasis. Other devices which are either commercially available or in a preclinical testing stage achieve vessel closure using a collagen plug, a clip mechanism, or a membrane closure of the vessel [9]. These vascular closure devices are generally intended for the femoral arteries but have also been used successfully for femoral vein closure following transcatheter pulmonary valve placement. In cases where a vascular closure device might be employed, extra care is taken to puncture the vessel at the optimal site. For the femoral artery, the optimal site is in the common femoral artery, distal to the epigastric artery origin and proximal to the femoral artery bifurcation. Fluoroscopy of the femoral head in addition to ultrasound guidance is helpful to increase the likelihood of optimal vessel entry site. Angiography of the femoral artery is necessary to confirm adequate vessel size and position of the entry site prior to use of a closure device. In cases where there is a high likelihood of requiring a large-bore femoral venous sheath, one or two suture-based closure devices can be placed using a pre-closure method after the skin incision is performed but prior to insertion of the short sheath, or prior to exchange for the long interventional sheath. Suture closure is then performed following removal of the long sheath at the conclusion of the case. A temporary subcutaneous figure-of-eight suture can help with compression for large-bore venous access as an alternative to a vascular closure device, when the vascular closure device fails or if the vessel site is not suitable for the use of a closure device [10].

Case 2: Hemodynamic Assessment of Long-Term Congenital Heart Disease Sequelae

Prior History

A 20-year-old female was referred to the cardiac catheterization laboratory for hemodynamic assessment with pulmonary vasodilator testing. Her history was significant for coarctation of the

aorta that was identified and surgically repaired as a newborn. At 3 years of age, she was found to have a large aortopulmonary window, which had not been appreciated earlier, and underwent surgical repair at that time. At 15 years of age, she was diagnosed with pulmonary hypertension, likely secondary to late repair of the aortopulmonary window. She underwent a cardiac catheterization at that time and was found to have a pulmonary arterial pressure of 70/30 mmHg with a mean of 49 mmHg, and a pulmonary vascular resistance of 10.7 indexed Wood units. Triple pulmonary hypertension therapy was initiated consisting of bosentan, tadalafil, and subcutaneous Remodulin. There was subsequent improvement in pulmonary artery pressures, to 63/19 mmHg with a mean of 39 mmHg, and pulmonary vascular resistance, to 5.8 indexed Wood units. Unfortunately, she was intolerant of both subcutaneous and intravenous Remodulin, so she was transitioned to oral selexipag prior to repeat cardiac catheterization.

Current Symptoms

She was asymptomatic.

Examination Findings

Cardiac examination was significant for a loud S2 but was otherwise unremarkable.

Investigations

Echocardiography was significant for greater than 50% systemic right ventricular pressure estimate based on tricuspid regurgitation doppler. The right ventricle was mildly dilated and hypertrophy with normal biventricular function. There was no residual arch obstruction. Cardiac catheterization with hemodynamic assessment remains the gold standard for evaluation of pulmonary hypertension with vasodilator testing to guide long-term pharmacologic therapy.

Procedural Management

To maintain conditions as close as possible the patient's baseline condition, cardiac catheterization was performed under moderate sedation with the patient spontaneously breathing in room air. As per routine for cardiac catheterization of patients with pulmonary hypertension, the sedation was performed by anesthesiologists with additional training in cardiac anesthesia. Following ultrasound inspection of the femoral vessels, vascular entry was by the modified percutaneous technique using a Doppler needle (Fig. 2). A 6-French sheath was placed in the right femoral vein to facilitate right heart catheterization. A 4-French arterial cannula was placed in the right femoral artery for continuous pressure and intermittent blood gas monitoring, to obtain rapid and accurate systemic arterial data acquisition during the pulmonary hypertension study. The patient was heparinized after access. Monitoring during the procedure included continuous surface electrocardiogram, continuous pulse oximetry, and cycled cuff blood pressure in addition to intravascular pressures. Flows were calculated by the thermodilution technique because the patient had no shunts (Fig. 3), and oxygen consumption was back-calculated from the thermodilution cardiac output and oxygen contents derived from hemoglobin oxygen satu-

Fig. 2 Femoral vessel access guided by anatomic landmarks, palpation of the arterial pulse, and doppler needle

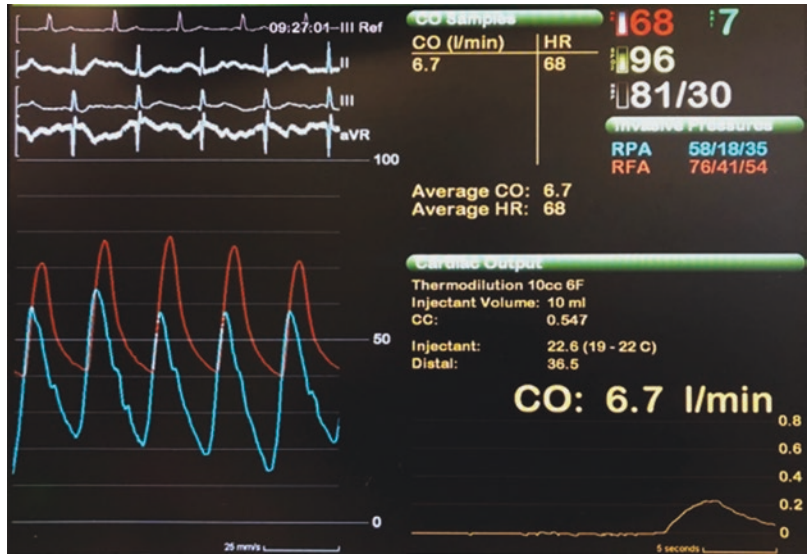

Fig. 3 Hemodynamic monitoring system displaying continuous monitoring of the 3-lead electrocardiogram, SpO_2 monitor, cycled cuff blood pressure, pressure tracing (blue) from the distal port of the thermodilution catheter in the right pulmonary artery, pressure tracing (red) from the red femoral arterial cannula, and the first of three 10 ml injections of hypothermic saline solution for thermodilution calculation of cardiac output

rations in the pulmonary and femoral arteries and hemoglobin capacity, for confirmation of the accuracy of the thermodilution data.

Oxygen consumption based on the patient's age of 20 years, female sex, and baseline heart rate of 70 beats/min using oxygen consumption charts per Body Surface Area was estimated to be 116 mL oxygen/min/m^2 [11]. Back calculation of oxygen consumption in this patient showed it to be nearly identical to expected, at 117 mL of oxygen/min/m^2. Saturation and pressure data are detailed in the cardiac catheterization diagram (Fig. 4). Pulmonary and femoral arterial saturations were normal. Femoral arterial blood gas results, detailed in Fig. 4, were within the normal range. Thermodilution cardiac index was normal, at 3.9 L/min/m^2.

Right atrial phasic and mean pressures were normal (Fig. 5a) with a dominant *a* wave (resulting from atrial contraction) that was similar to the right ventricular end diastolic pressure, excluding significant tricuspid valve stenosis and demonstrating normal compliance of the right ventricle (Fig. 5b). The right atrial *v* wave was normal (Fig. 5a), excluding significant tricuspid regurgitation. Right ventricular peak systolic pressure was moderately elevated (Fig. 5b) and similar to that in the bilateral branch pulmo-

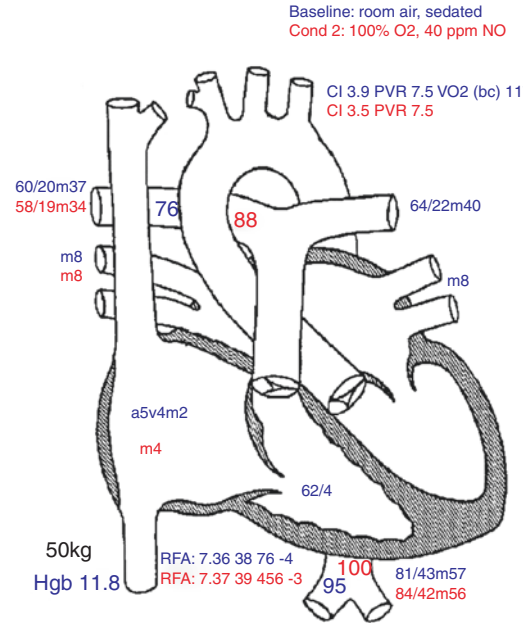

Fig. 4 Case 2, cardiac catheterization diagram displaying saturations (large font) and pressure data in mmHg with indexed calculations and blood gas results. The baseline condition performed without supplemental oxygen is illustrated with blue font and the second condition with administration of FiO_2 100% and 40 ppm inhaled nitric oxide is illustrated with red font. *Cond* condition, *ppm* parts per million, *NO* nitric oxide, *CI* cardiac index, *PVR* indexed pulmonary vascular resistance in Wood units, *bc* back-calculated, *m* mean

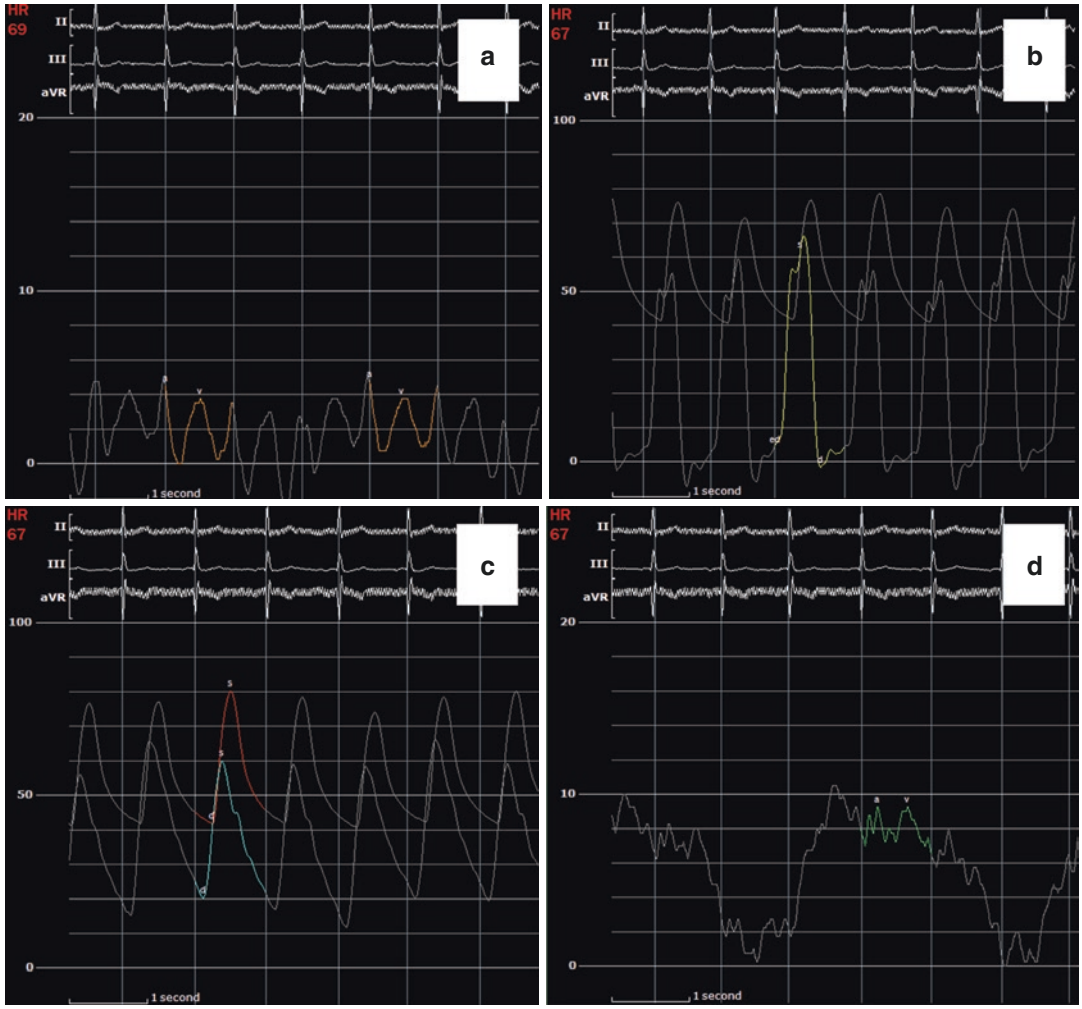

Fig. 5 (**a**) Right atrial pressure tracing with scale set at 20 mmHg showing respiratory variation. (**b**) Right ventricular pressure tracing with selected beat highlighted in yellow, end diastole is notated as "ed" with scale set at 100 mmHg and displayed with the femoral arterial tracing. (**c**) Right pulmonary artery pressure tracing with selected beat highlighted in blue shown with the femoral arterial pressure tracing with selected beat highlighted in red with scale set at 100 mmHg. (**d**) Left pulmonary capillary wedge pressure demonstrating respiratory variation with selected beat highlighted in green with scale set at 20 mmHg

nary arteries, excluding pulmonary subvalvar, valvar, or supravalvar stenosis. Bilateral pulmonary arterial phasic and mean pressures were moderately elevated and nearly identical to those found at last catheterization, at 60/20 mmHg with a mean of 37 mmHg in the right pulmonary artery (Fig. 5c). The 16 mmHg difference in pressure between the pulmonary arterial diastolic and right ventricular end diastolic pressure excluded significant pulmonary valve insufficiency. Wedge pressures were normal bilaterally (Fig. 5d) with mean pressure of 8 mmHg, indicating a normal left atrial pressure. The pulmonary capillary wedge tracing had a normal dominant *v* wave and a normal *a* wave (Fig. 5d), excluding significant mitral valve stenosis or regurgitation, and demonstrating a normally compliant left ventricle. The femoral artery pressure tracing (Fig. 5c) was normal, with a normal pulse pressure and dicrotic notch (Fig. 5c), excluding significant aortic valve insufficiency and demonstrating normal aortic compliance. The systolic blood pressure of 81 mmHg was slightly lower than would be expected for age

and likely reflects the effects of moderate sedation; her pre-procedural cuff systolic blood pressure when the patient was awake was 119 mmHg. She had undergone multiple previous cardiac catheterizations and echocardiograms, none of which showed pressure gradients between the left ventricle and the femoral arteries, so that further evaluation of left-sided hemodynamics was unnecessary. The peak systolic pressure in the left ventricle can be assumed to be approximately 10 mmHg lower than that obtained at the femoral artery due to pulse-wave amplification.

The elevated mean pulmonary artery pressure of 37 mmHg and normal wedge pressure of 8 mmHg gave a significantly elevated transpulmonary gradient (mean PA pressure [37 mmHg]—mean pulmonary capillary wedge pressure [8 mmHg]) of 29 mmHg. Calculated pulmonary vascular resistance (transpulmonary gradient [29 mmHg]/Qp [3.9 L/min/m^2]) was moderately elevated, at 7.5 indexed Wood units.

After baseline measurements, hemodynamics were repeated during pulmonary vasodilator testing, with the patient breathing 100% oxygen using a nonrebreather mask with 40 ppm of inhaled nitric oxide. There were the expected increases in saturations in the pulmonary artery and aorta. There were no significant changes in the right atrial, wedge, or systemic arterial pressure, but there was a modest decrease in pulmonary arterial pressures, which were 58/19 mmHg with a mean of 34 mmHg. The decrease in pulmonary arterial pressure was due to the expected decrease in cardiac index with supplemental oxygen, to 3.5 L/min/m^2, and pulmonary vascular resistance was unchanged. Nitric oxide and oxygen were weaned while continuing invasive monitoring of the pulmonary and systemic arterial pressures prior to removal of catheters and sheaths.

Post-procedure Management

Given the patient's significant pulmonary hypertension, post anesthesia recovery care was provided in the cardiac intensive care unit setting where close monitoring and inhaled nitric oxide are available at bedside to treat any potential pulmonary hypertensive crisis episodes.

Outcome

The patient recovered without incident and was discharged to home with continued triple oral pulmonary hypertension therapy.

Discussion and Brief Literature Review

Principles of Hemodynamic Measurements

The cardiac catheterization laboratory remains a true laboratory, where a rigorous approach to data collection occurs. This includes blood sampling for saturation and oxygen content data, and pressure measurements, when the patient is in an environment of an unchanging cardiorespiratory state. Baseline measurements are obtained, so that analysis of effects of non-invasive and transcatheter interventions can be compared to these data. A thorough understanding of the principles of hemodynamic measurements and calculations, with the ability to interpret the data throughout the case, is required, to determine whether to perform interventions and if so, whether the intended outcome of the intervention has been achieved.

Cardiac anesthesiologists experienced in the management of patients with congenital heart disease are essential. Anemia should be identified and evaluated prior to cardiac catheterization. Red blood cell transfusion should be considered in cases of severe anemia prior to the catheterization procedure, if necessary to safely perform the procedure and to obtain optimal baseline hemodynamics, particularly in anticipation of potential bleeding during sheath exchanges and interventions. The pH, base excess, and pCO_2 on the initial arterial blood gas should be noted, and any changes in ventilatory strategy or correction of metabolic derangements should be made at the start of the case, prior to obtaining baseline data. In cases where accuracy of the pulmonary vascular resistance calculation is essential to patient management, pH and pCO_2 are particularly important, because abnormalities in either greatly impact measurements of pulmonary vascular resistance. Intravenous fluids should be adminis-

tered to maintain adequate hydration, which is particularly important for patients with pulmonary hypertension, who require adequate preload, or cyanotic patients who are polycythemic and are at high risk for thrombosis.

The optimal sedation and airway management conditions are as close as possible to the patient's baseline awake state, particularly in patients for whom hemodynamic evaluation is most important. If tolerated, the patient can be conscious and communicating with the team during the procedure, though in most cases some degree of sedation is administered for patient comfort. Every effort is made to make the patient comfortable during the procedure with local anesthesia and sedation, but spontaneous breathing is preferable to intubation and mechanical ventilation, when possible. The mechanism of ventilation will determine which pressure measurements to select. In general, expiratory pressures are closer to atmospheric than inspiratory pressures. Thus, in a patient breathing spontaneously (Case 2, Fig. 5a–d) the higher pressure beats are selected (inspiration leads to negative intrathoracic pressures, lowering intravascular pressures), while during mechanical ventilation the lower pressure beats are selected (the positive inspiratory pressures lead to higher intravascular pressures). In cases where there is significant airway obstruction, there is a large variation in pressures between inspiration and expiration, and end-expiration is associated with high intrathoracic pressures. Thus, near-atmospheric pressures cannot be obtained. A jaw thrust or an oral airway can be applied by the anesthesiology team. Occasionally an LMA can be used, which would require greater sedation. If these maneuvers do not mitigate the wide variation in pressures, the average pressures over a full recorded respiratory cycle can be used to best approximate near-atmospheric pressures. If this is necessary, this technique must be applied for each pressure tracing obtained. Ventilation type is a particularly important consideration for patients with pulmonary hypertension, in whom positive end expiratory pressure will elevate the pulmonary capillary wedge pressure, and in patients with single ventricle palliation, who are reliant on passive pulmonary blood flow, which is in part driven by negative intrathoracic pressure.

The patient's level of sedation should be considered when determining whether valve gradients are significant enough to require intervention, because deeper sedation or anesthesia will result in a lower valve gradient. Valve area measurements do not have the same problem since the stroke volume across the valve is included in its calculation, along with the pressure gradient, and are discussed in the chapter on valve intervention. In order to more accurately reflect a state of exertion, inotropic agents as dobutamine can be administered during the case, to increase cardiac output and allow for an assessment more akin to exercise conditions. Valve gradients should be considered in comparison to the pre-procedure echocardiographic gradients, with the catheterization derived peak to peak valve gradient usually corresponding more closely to the echo mean gradient rather than the peak instantaneous gradient. Comparative values of the systemic and pulmonary hemodynamics, such as when pulmonary arterial pressures are reported as being equal to systemic pressures, should be interpreted with great caution, given that the major impact of sedation state and anesthetic agents is on the systemic vascular resistance. For example, an increase in the pulmonary-to-systemic pressure or vascular resistance ratio caused by systemic vasodilation in the catheterization laboratory is not evidence of worsening in the state of the pulmonary vascular bed.

Saturations

For a first right heart catheterization performed in a patient (i.e., prior to being certain of the presence or absence of shunts), hemoglobin oxygen saturations should be obtained in both the high and low superior vena cava. A high saturation in the upper SVC suggests anomalous connection of the left upper pulmonary vein to the innominate vein, while a high saturation only in the lower SVC suggests anomalous connection of the right upper pulmonary vein. Both of these defects can be missed on transthoracic echocardiography. Saturations are then obtained in the right atrium, right ventricle, and bilateral pulmonary arteries, in consideration of shunts at each

level. For patients returning for repeat cardiac catheterization in whom the absence of any shunts has been confirmed, a single saturation in a branch pulmonary artery will suffice for the purpose of blood flow calculations. Where feasible, one left-sided saturation should be obtained in the absence of a right-to-left shunt, usually in a systemic artery. If a shunt is present, saturations proximal to and as distal as possible from the shunt should be obtained. For example, if a patient may have a right-to-left atrial shunt, pulmonary venous saturations should be obtained proximal to it, and aortic saturations distal, as a reflection of the complete mixing of the systemic and pulmonary venous blood. If the presence of pulmonary arteriovenous malformations is being evaluated, saturations should be obtained in all of the pulmonary veins.

Flow Calculations

Normal cardiac output is 5–8 l of blood flow per minute and is determined by stroke volume multiplied by heart rate. Cardiac index is cardiac output normalized to body surface area with normal values of 2.4–4 L/min/m^2. Cardiac output is measured in the cardiac catheterization laboratory predominantly by one of two methods. Thermodilution is the most accurate of these methods. Thermodilution measures the extent of change in temperature at a thermistor at the tip of a catheter in the pulmonary arterial blood after injection of a fixed volume, usually 10 mL in an adult patient, of relatively cold saline (approximately 20 °C) fluid, injected via a proximal port in the same catheter into the right atrium. The resulting temperature-time curve is used to determine cardiac output. This technique is inaccurate in the presence of shunts or tricuspid valve regurgitation, both of which are frequently encountered in adults with congenital heart disease.

When thermodilution is not appropriate, the Fick principle is commonly used. It is based upon the amount of oxygen consumed by the body (which is equal to that taken in by the lungs) and the difference in oxygen content across the circulation of interest. Thus, pulmonary blood flow (Qp) is calculated by dividing oxygen consumption by the difference between the oxygen content of the pulmonary venous and pulmonary arterial blood. Systemic blood flow (Qs) is calculated by dividing oxygen consumption by the difference between the systemic arterial oxygen content and the mixed systemic venous oxygen content. Oxygen consumption (VO_2) can be measured in the cardiac catheterization laboratory, although the commercially available equipment cannot be used in the ventilated patient or in patients who are breathing supplemental oxygen. In most cases oxygen consumption is assumed, as is based on sex, age, and heart rate such as the LaFarge values [11]. Although such assumptions are not perfect, they are necessary when thermodilution cannot be used and oxygen consumption cannot be measured.

Oxygen content is usually calculated only from hemoglobin oxygen saturation and hemoglobin capacity (13.6 × Hgb in g/dL × saturation) because the amount of oxygen dissolved in blood is relatively low compared to that attached to hemoglobin. There are 3 mL of oxygen per 100 mmHg of pO_2 per dL of blood. Using Case 2 as an example, in the first condition while breathing room air, her systemic arterial saturation was 95%, her hemoglobin concentration was 11.8 g/dL, and her pO_2 was 76 mmHg. Thus, there were 161 mL of oxygen attached to hemoglobin in 1 dL of her systemic arterial blood, but only 2 mL of oxygen dissolved in that same amount of blood. When a patient is in a significant amount of supplemental oxygen and has a pO_2 of greater than about 200 mmHg in any chamber measured, dissolved oxygen should be added when calculating oxygen content in all of the sites in which saturations are measured. This occurred in the second condition of Case 2, when she was breathing 100% oxygen via a nonrebreather mask and had a systemic arterial pO_2 of 456 mmHg.

For Qp measurements, the values to use for pulmonary arterial and pulmonary venous bloods are usually obvious. However, the value to use for mixed systemic venous blood flow in Qs measurements is not so obvious. The best approach is to use the most DISTAL sample in a well-mixed chamber or vessel PROXIMAL to any left-to-right shunt. Thus, in the absence of shunts, pulmonary arterial saturation is the best estimate of mixed systemic venous saturation. In the pres-

ence of a ductus arteriosus, right ventricular saturation should be used. In the presence of either a VSD or ASD, superior vena caval saturation should be used. This is because the right atrium is not a well-mixed chamber; the SVC, coronary sinus, and IVC all have very different saturations, so the location in the right atrium where the sample is taken significantly affects the value obtained. Because the hepatic veins deliver highly saturated blood into the IVC very close to the right atrium, the IVC is not a well-mixed location either. It is important to note that using SVC saturation as an estimate of mixed venous saturation usually underestimates Qs somewhat. This is because IVC blood is usually more saturated than SVC blood and is demonstrated by the fact that pulmonary arterial saturation is usually about 2–3% more saturated than SVC blood in a patient with normal hemodynamics who does not have a shunt. However, it is the best site to use in a patient with either a VSD or ASD. One just needs to appreciate the small underestimate in Qs that is associated with its use.

In addition to calculation of Qp and Qs when a shunt is present, effective pulmonary blood flow, or Qep, is often calculated. It is an estimate of the amount of mixed venous blood that goes to the lung for oxygenation, and thus is calculated as oxygen consumption divided by the difference in oxygen content between the pulmonary venous and mixed systemic venous blood. Because pulmonary venous blood is always the most saturated blood and mixed venous the least saturated blood used in calculations, it can never be greater than either Qp or Qs and is lower than both in the presence of bidirectional shunting.

While straightforward calculation of the Qp, Qep, and Qs will in most cases completely describe a patient's hemodynamics, there are some patients for whom it is not possible to accurately calculate shunt magnitude in the cardiac catheterization laboratory. That occurs when it is not possible to obtain samples representing either mixed systemic venous, pulmonary arterial, or pulmonary venous blood. In such circumstances cardiac magnetic resonance velocity encoded flow calculations obtained concurrently can be a useful adjunct to determining blood flows. Case 3 is an example of a patient with complex congenital cardiac and post-surgical anatomy and physiology for whom simple shunt calculations cannot be reliably calculated.

Pressures and Resistances

Commercially available recording systems allow for continuous display of pressure tracings, a short period of storage of the complete record of the pressure tracings during the case, saving of specified tracings during the case, and built-in analysis tools for valve areas. Detailed review of the recorded pressures can be performed, with analysis in relation to the contemporaneous electrocardiogram to accurately determine the timing of the pressure acquired for wave analysis.

Many factors affect pressure tracings, all of which must be considered prior to concluding that the waveform displayed is, in fact, accurate enough to form the basis for clinical decisions. The pressure transducers should be as close to the level of the patient's mid left atrium as possible prior to zeroing to ambient air pressure. Care is taken to exclude air in the transducer tubing and within the catheter. Excess blood in the catheter will dampen the tracing though in some circumstances, generally in the left ventricle, a small volume of blood in the catheter will reduce catheter "fling" caused by the kinetic energy of the catheter hitting the ventricular myocardium, thereby making estimations of pressure gradients more accurate. Ensuring the catheter tip is free in the vessel or chamber rather than against a wall will provide a more accurate pressure tracing. It is important to remember that measurement of intravascular pressures caused by the potential energy within the fluid is the goal, that contact forces and kinetic energy are also converted to voltage by the pizoelectric transducers, and that any resistor within the system dampens the pressure wave before it is seen by the transducer. Thus, large-bore catheters produce a more accurate pressure tracing than microcatheters. In locations only accessible with a microcatheter, such as an area proximal to the stenotic lesion of a pulmonary vein, wave analysis may not be accurate; however, the mean pressure obtained will provide useful data. If the initial pressures obtained are

higher, or lower, or more dampened than excepted, taking additional time is prudent, to confirm correct transducer height, eliminate all air from the transducer system, and re-flush the catheters prior to recording the pressure. If the patient has been NPO for considerable time prior to the procedure there may indeed be a lower-than-normal filling pressures, which should be addressed with administration of intravenous fluids. If feasible, simultaneous measurement of pressures of interest is prudent to determine accurate gradients. If not, measurements should be made as contemporaneously as possible while ensuring that the patient is in a hemodynamically stable state.

Pressure waves and interpretation are detailed in Case 2 above. Normal pressures vary with age and congenital cardiac anatomy. For example, "normal" pulmonary arterial phasic pressures in a patient with a biventricular heart will be pulsatile and considerably different from those with a Fontan circulation, where pulmonary blood flow is passive. However, mean pressures should be similar, as long as pulmonary capillary wedge pressures are normal. Normal pressure ranges in adults with a structurally normal heart and normal systemic blood pressure are generally in the following range: right atrial mean and right ventricular end diastolic pressure are 1–8 mmHg, right ventricular and pulmonary artery peak systolic pressures are 15–30 mmHg, pulmonary artery diastolic, pulmonary capillary wedge, left atrial and left ventricular end diastolic pressures are 4–12 mmHg.

Because most adults with congenital heart disease have been followed throughout childhood, their blood flows and vascular resistance measurements in the catheterization laboratory against which they are being compared are usually indexed to body surface area. Indexed systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR) are expressed using measurements of pressure in mmHg and of flow in L/min, and are referred to as indexed Wood units. They are calculated by using the simple equations below, derived from a variation on Ohm's law, in which resistance is equal to the change in pressure divided by flow. If only right heart data is obtained, pulmonary capillary wedge pressure is used as an estimate of left atrial pressure.

$$\mathrm{SVR} = \left(\text{mean aortic pressure} - \text{mean right atrial pressure}\right) / \text{indexed Qs}$$

$$\mathrm{PVR} = \left(\text{mean pulmonary artery pressure} - \text{mean left atrial pressure}\right) / \text{indexed Qp}$$

Alternative resistance units and pulmonary vascular reactivity testing will be covered in detail in chapter "Pulmonary Hypertension".

Case 3: Vascular Access, Shunt Calculation, and Transcatheter Interventions for Sequelae of Complex Congenital and Post-surgical Cardiac Anatomy

Prior History and Current Symptoms

A 24-year-old woman was referred for cardiac catheterization to evaluate progressive cyanosis and exertional dyspnea. Her congenital cardiac history was significant for heterotaxy with dextrocardia, left atrial isomerism, interrupted inferior vena cava with azygos continuation, an unbalanced complete atrioventricular canal with double outlet right ventricle and d-malposed great vessels and severe pulmonary stenosis. She underwent staged single ventricle palliation, with a left modified Blalock-Taussig shunt as a newborn followed by a Kawashima procedure at 8 months of age without disruption of her limited antegrade pulmonary flow. She developed progressive cyanosis secondary to pulmonary arteriovenous malformations. She then underwent surgical redirection of hepatic venous drainage to

the left pulmonary artery at 11 years of age. At 19 years of age a second conduit was placed between the hepatic veins and the right innominate vein because of persistence of right-sided pulmonary arteriovenous malformations. At the same time an epicardial dual chamber pacemaker was placed because of sinus node dysfunction. Her only routine medication was daily aspirin.

Physical Examination

On physical examination she was noted to be cyanotic at rest, with an SpO_2 of 80% and with prominent clubbing. The cardiac examination was significant for jugular venous distention, a well-healed median sternotomy scar, apical impulse palpated in the 5th intercostal space just medial to the right mid-axillary line in keeping with dextrocardia, a regular heart rate of 92/min, a normal S1 and single S2 without clicks or gallop. There was a 2/6 soft holosystolic murmur at the right lower sternal border. The pulses were full and equal in well-perfused extremities, and her blood pressure was 119/74 mmHg. The liver was palpable in the midline, in keeping with patient's known heterotaxy, polysplenia, and previously CT scan confirmed transverse liver.

Investigations

Pre-catheterization investigations included an electrocardiogram, which showed an atrial paced rhythm with prolonged AV conduction. Transthoracic echocardiography showed the post-surgical anatomy as detailed above, with adequate biventricular function, mild atrioventricular valve regurgitation, and limited imaging of the pulmonary arteries. She was referred to the cardiac catheterization laboratory for hemodynamic and angiographic assessment of the Kawashima, the hepatic inclusion conduits, and the pulmonary arteries to determine the etiology of her symptoms.

Management Options, Procedural Management, and Discussion

With a known interrupted inferior vena cava and single ventricle palliation, access to the intracardiac structures was approached from the arterial system, via with a 5-French sheath in the right femoral artery. Although access to the pulmonary arterial circulation could have been obtained via a femoral vein, via the azygos continuation of the interrupted inferior vena cava, internal jugular venous access was obtained instead, to facilitate the most straightforward access to her pulmonary arteries and to each hepatic inclusion conduit, for likely intervention. The modified percutaneous Seldinger technique was used, with ultrasound guidance. The hemodynamic portion of the procedure was performed with moderate sedation, with conversion to general anesthesia and endotracheal intubation for the interventional portion.

Patients who have undergone a bidirectional cavopulmonary anastomosis with hepatic inclusion should have no shunts except for the small amount of coronary sinus blood which goes to the atrium and then out the ventricle and aorta. In this patient's case, given the systemic desaturation, there is likely a much more significant right-to-left shunt from a different source.

As shown in Fig. 6, there was no increase in saturation between the systemic venous return (73%) from the lower body (azygos) and upper body (superior vena cava) saturations (72%) to the right pulmonary artery saturation (72%). There was an increase in saturation from the hepatic veins (53%) to the left pulmonary artery (62%) likely explained by admixture with right pulmonary arterial blood, although there might also be left-sided aortopulmonary collateral vessels that would be investigated by systemic arterial angiography. Systemic venous angiography confirmed multiple systemic venous to pulmonary venous collateral vessels. They were successfully closed with coils. Because of the differential pulmonary arterial saturations and likely different pulmonary venous saturations

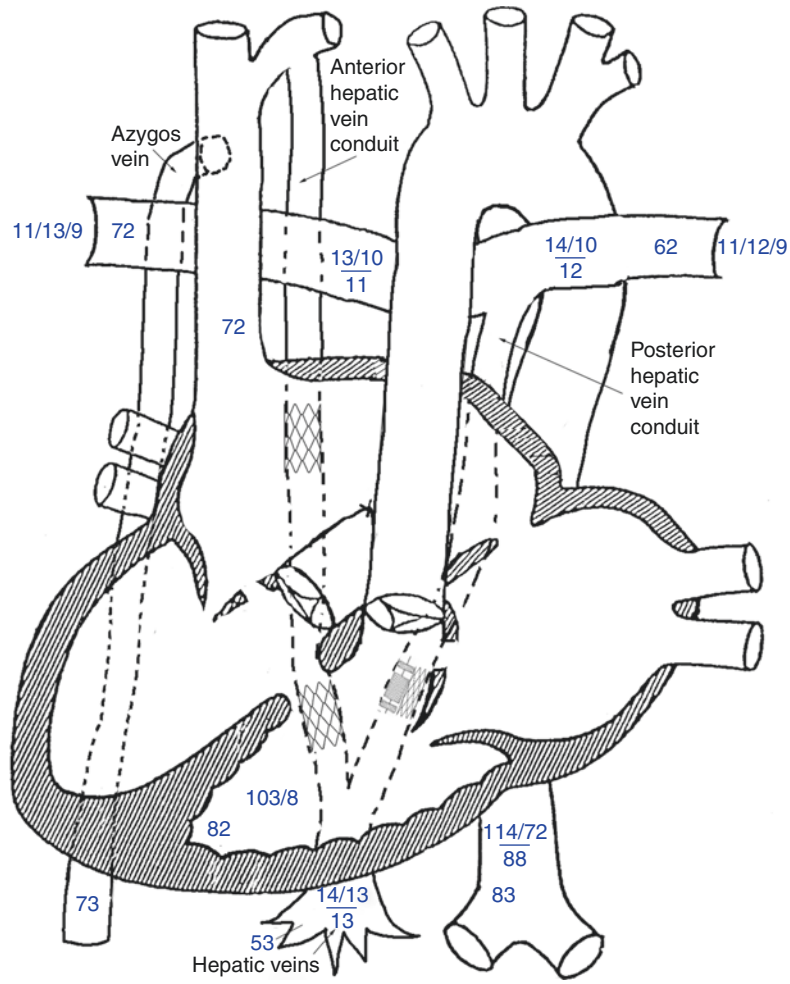

Fig. 6 Case 3, cardiac catheterization diagram displaying saturations (blue large font) and pressure data in mmHg (blue small font). Hemodynamic data was obtained prior to the interventions

due to the venous collaterals, pulmonary blood flow could not accurately be calculated. Qs could be calculated using SVC saturation as the estimate of mixed systemic venous saturation, knowing its inherent small inaccuracy.

The systemic arterial desaturation (83%) indicated the presence of a significant right-to-left shunt, lung disease, or a combination of both. Because the right-to-left shunt in this patient went directly to the pulmonary veins, it would be impossible to use catheterization data to distinguish between the two. Her history of pulmonary arteriovenous malformations and the absence of a history of lung disease or elevated ventricular filling pressures (which could lead to pulmonary edema) indicate that parenchymal lung disease is not a significant component of the desaturation. When in doubt, one can perform transthoracic echocardiography while injecting a mixture of agitated blood and saline into each branch pulmonary artery. If this "saline contrast" is seen in the atrium via the pulmonary veins, then pulmonary arteriovenous malformations are present.

Venous angiography also demonstrated that the anterior hepatic to right innominate vein conduit was stenotic; it was successfully stented. The bilateral internal jugular vein access sites helped to facilitate angiography and intervention on the hepatic venous conduits to encourage flow of hepatic venous blood to the right lung; simultaneous placement of large vascular plug and stent within the hepatic vein to left pulmonary artery conduit was performed to reduce flow through this conduit. This procedure required bilateral internal jugular vein access, as shown in Fig. 7, due to requirement of two large venous sheaths

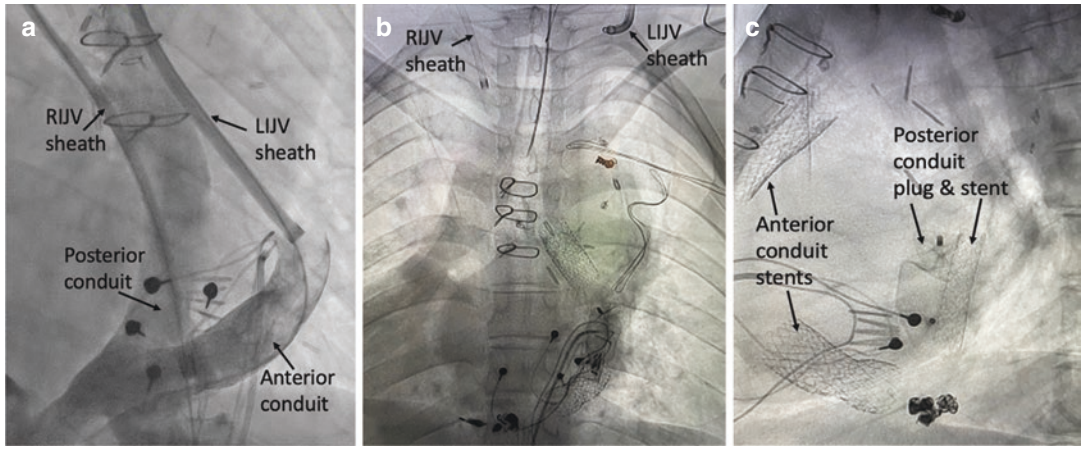

Fig. 7 (**a**) Simultaneous hepatic venous conduit angiography via the right internal jugular vein (RIJV) sheath in the posterior conduit and the left internal jugular vein (LIJV) sheath in the anterior conduit in the RAO projection. (**b**) Right internal jugular vein and left internal jugular vein sheaths post stent angioplasty of the anterior conduit and posterior conduit narrowing with simultaneous stent and vascular plug placement in the straight AP projection and (**c**) straight lateral projection

for simultaneous vascular plug and stent deployment within the hepatic vein to left pulmonary artery conduit. All of these interventions would have been much more challenging via the interrupted inferior vena cava with azygos continuation to the pulmonary artery anastomosis.

Post-procedure Management

Post-procedure hemostasis was achieved by simple manual pressure at the groin and neck entry sites. She was discharged the day following the procedure on warfarin to maintain stent patency.

Outcome

Her oxygen saturations have been in the 85–93% range with no change in symptoms after 3 years of follow-up.

References

1. Frazer JR, Ing FF. Stenting of stenotic or occluded iliofemoral veins, superior and inferior vena cavae in children with congenital heart disease: acute results and intermediate follow up. Catheter Cardiovasc Interv. 2009;73(2):181–8. https://doi.org/10.1002/ccd.21790. PMID: 19156885.
2. Ligon RA, Petit CJ. Not that atretic: use of an atretic femoral vein for transcatheter pulmonary valve implantation. Catheter Cardiovasc Interv. 2017;89(2):321–3. https://doi.org/10.1002/ccd.26687. Epub 2016 Aug 12. PMID: 27516338.
3. Qureshi AM, Prieto LR, Bradley-Skelton S, Latson LA. Complications related to transhepatic venous access in the catheterization laboratory–a single center 12-year experience of 124 procedures. Catheter Cardiovasc Interv. 2014;84(1):94–100. https://doi.org/10.1002/ccd.25278. Epub 2014 Mar 21. PMID: 24989344.
4. Justino H, Petit CJ. Percutaneous common carotid artery access for pediatric interventional cardiac catheterization. Circ Cardiovasc Interv. 2016;9(4):e003003. https://doi.org/10.1161/CIRCINTERVENTIONS.115.003003. PMID: 26984925.
5. Kirker E, Korngold E, Hodson RW, Jones BM, McKay R, Cheema M, Heimansohn D, Moainie S, Hermiller J, Chatriwalla A, Saxon J, Allen KB. Transcarotid versus subclavian/axillary access for transcatheter aortic valve replacement with SAPIEN 3. Ann Thorac Surg. 2020;110(6):1892–7. https://doi.org/10.1016/j.athoracsur.2020.05.141. Epub 2020 Jul 24. PMID: 32712098.
6. Overtchouk P, Modine T. A comparison of alternative access routes for transcatheter aortic valve implantation. Expert Rev Cardiovasc Ther. 2018;16(10):749–56. https://doi.org/10.1080/14779072.2018.1524295. Epub 2018 Sep 28. PMID: 30232918.
7. Heyden CM, Moore JW, Ryan JR, Lederman RJ, El-Said HG, Ratnayaka K. Alternative access in congenital heart disease. JACC Case Rep. 2020;2(11):1734–5. Epub 2020 Sep 15. PMID: 33000000; PMCID: PMC7520567.
8. Lederman RJ, Babaliaros VC, Greenbaum AB. How to perform transcaval access and closure for transcath-

eter aortic valve implantation. Catheter Cardiovasc Interv. 2015;86(7):1242–54. https://doi.org/10.1002/ccd.26141. Epub 2015 Sep 10. PMID: 26356244; PMCID: PMC4740457.

9. Cilingiroglu M, Salinger M, Zhao D, Feldman T. Technique of temporary subcutaneous "figure-of-eight" sutures to achieve hemostasis after removal of large-caliber femoral venous sheaths. Catheter Cardiovasc Interv. 2011;78(1):155–60. https://doi.org/10.1002/ccd.22946. PMID: 21681904.
10. van Wiechen MP, Ligthart JM, Van Mieghem NM. Large-bore vascular closure: new devices and techniques. Interv Cardiol. 2019;14(1):17–21. https://doi.org/10.15420/icr.2018.36.1. PMID: 30858887; PMCID: PMC6406132.
11. LaFarge CG, Miettinen OS. The estimation of oxygen consumption. Cardiovasc Res. 1970;4:23.