Catheter hemodynamic assessment of the univentricular circulation

Oliver Stumper, Gemma Penford

Department of Pediatric Cardiology, Birmingham Children's Hospital, Birmingham, United Kingdom

ABSTRACT

Children with very complex congenital heart disease not amenable to biventricular repair are increasingly being considered for a palliative univentricular care pathway. This involves a staged surgical approach culminating in the Fontan circulation with passive pulmonary blood flow and added resistances. The catheter based hemodynamic assessment at all three stages of this palliation is described in detail. Frequent pitfalls, inherent limitations and potential errors are discussed and clinical examples are illustrated.

Keywords: Cardiac output, cavopulmonary shunt, Fontan, hemodynamics, univentricular heart

INTRODUCTION

Children born with highly complex congenital heart disease may not survive to diagnosis. For those who are diagnosed, there are now treatment options available, all of which are rather high risk and ultimately palliative in nature. The surgical Fontan pathway[1,2] offers reasonable quality of life for the majority of survivors for a number of decades although there is the need for close ongoing follow-up, long-term medication, and repeated surgical and catheter interventions. Families have to receive detailed counseling about the palliative nature of this approach and how it will affect the whole family's life.

Univentricular hearts are all those congenital cardiac lesions, which cannot be corrected or repaired to a circulation with two pumping chambers. This is either because of one of the atrioventricular (AV) valves being atretic (tricuspid or mitral atresia), or both AV valves emptying into the same ventricle (double inlet ventricles), or either of the ventricles being severely hypoplastic (hypoplastic left or right heart syndrome). In other cases, associated lesions such as straddling AV valves, multiple ventricular septal defects, multilevel

outflow obstructions, or a combination of these may make the univentricular treatment pathway a less risky and more predictable choice for parents who are aware of the palliative nature of these treatment avenues.[3]

PRINCIPLES OF UNIVENTRICULAR PALLIATION

On (ultrasound) diagnosis of the complex lesion, an attempt is made to protect the lungs from increased pulmonary blood flow and increased perfusion pressure. In cases with increased pulmonary blood flow and a normal pulmonary valve, the application of a pulmonary artery band may be required to limit pulmonary blood flow. Alternatively in patients with decreased pulmonary blood flow, this can be achieved by establishing pulmonary blood flow through small diameter systemic to pulmonary artery shunts or right ventricle to pulmonary artery (RV-PA) conduits (Blalock-Taussig (BT) shunt, Norwood/Sano procedure). Significant aortic arch obstruction also has to be addressed at this stage. In both case scenarios, the single ventricle will have to

Access this article online

Quick Response Code:

Website:
www.annalspc.com

DOI:
10.4103/apc.APC_160_16

Address for correspondence: Dr. Oliver Stumper, Birmingham Children's Hospital, Steelhouse Lane, Birmingham B4 6NH, United Kingdom.
E-mail: oliver.stumper@bch.nhs.uk

pump the entire pulmonary and systemic blood flow, often reaching 300% of normal output and resulting in ventricular dilatation and hypertrophy.[4]

At 4–6 months of age (when pulmonary vascular resistance has dropped to its lowest level), the mode of pulmonary blood supply is then changed over to a bidirectional cavopulmonary (CP) shunt (end to side anastomosis of the superior vena cava [SVC] to the [right] pulmonary artery). This leads to volume unloading of the (hypertrophied) systemic ventricle and results in nonpulsatile low flow perfusion of the pulmonary arteries. The systemic venous return from the inferior vena cava (IVC) drains back to the heart and mixes with pulmonary venous blood. Despite these dramatic changes, the superior bidirectional CP shunt normally provides infants and young children with sufficient pulmonary blood flow to maintain saturations of around 85%. During exercise (running around), the saturations will drop significantly, the child will get breathless and will have to rest between short bursts of (largely anaerobic) exercise.

At around 4–5 years of age, the Fontan circulation is then completed by redirecting the IVC return to the pulmonary artery.

This establishes a circulation in series with only one ventricular pump.

The conversion to a Fontan (total CP anastomosis) results in persistently elevated systemic venous pressures affecting liver and gut function and reduced cardiac output both at rest and during exercise. After the Fontan completion, cardiac output is governed by the added resistances of the pulmonary and systemic circulation and by the gradually declining function of the single ventricle.[4]

MANAGEMENT CONCEPTS AND PHYSIOLOGIC CONSEQUENCES

At initial presentation

On presentation of children with complex congenital heart disease not amenable to biventricular repair, clinical assessment, pulse oximetry, chest X-ray, and detailed cardiac ultrasound are all that are needed to determine initial counseling and potential future management.

Invasive diagnostic cardiac catheter evaluation is rarely needed except for cases of late presentation with increased pulmonary blood flow to determine flows and vascular resistances. Interventional catheterization to enlarge the atrial septum (Rashkind balloon septostomy) and stent the arterial duct or a severely stenosed subpulmonary outflow tract may be considered as an

alternative to surgical systemic-to-pulmonary artery shunting or open septectomy.

Consideration for creation of cavopulmonary shunt

At 4–6 months of age, infants will have had a significant somatic growth and will have largely outgrown their initial palliative procedure. Saturations will typically be in the high 70 s or low 80 s. Pulmonary vascular resistance should have dropped to its lowest level. At this stage, it is important to get detailed insight into the pulmonary vasculature and pressures and overall morphology.

At Birmingham Children's Hospital, which has one of the largest univentricular treatment programs in Europe, we routinely perform cardiac catheter assessment before CP shunt. The pattern of systemic venous return is confirmed and the pulmonary vascular bed is imaged in great detail, documenting the pulmonary arteries, the capillary phase, and the pulmonary venous return in real time. In patients who initially started out with reduced pulmonary blood flow, there is no need to get direct pulmonary artery pressures by cannulation of the pulmonary artery system if the angiographic appearance of the pulmonary vascular bed (supply, arborization and venous return) is satisfactory. Advancing a four French catheter down a 3.5 mm BT shunt will reduce the lumen of the shunt by 15%–20%. Saturations will drop, and pulmonary arterial pressure measurements will be compromised. Several pulmonary vein wedge pressure measurements are more reliable, in our opinion.

If there is doubt about the size and adequacy of the pulmonary vascular bed (on angiography) to accommodate a low-pressure superior CP shunt circulation, we would consider catheter intervention to augment the size of the shunt or a limiting RV-PA conduit or balloon dilatation of any peripheral stenosis. Pulmonary arteries are distensible and their overall diameter/cross-sectional area is dependent on flow – the Nakata et al. index[5] is not load independent.[6] As long as peripheral arborization and pulmonary venous return are normal, together with satisfactory pulmonary vein wedge pressures, we would consider conversion to a CP shunt at 4–6 months and a minimum weight of 5–6 kg.

In univentricular pathway patients who have undergone initial pulmonary artery banding, we undertake detailed cardiac catheter hemodynamic evaluation when resting saturations are normally 75%–80% or below. Sampling for oxygen saturations is routinely performed in the SVC, the distal pulmonary arteries, the aorta, and one of the pulmonary veins to calculate Qp/Qs along the Fick principle.[7] Saturation sampling within the atria or the inferior caval vein is notoriously unreliable[8] with variation in measurement of up to 5%–8%, and thus, has been largely abandoned, other than in patients with a superior CP shunt (see below).

It is also important to accept the fact that the Fick principle to calculate relative systemic and pulmonary arterial blood flow in complete mixing/single inlet or outlet lesions is generally very good (subject to sampling variations in different locations within the circulation), but that it may be false or contaminated by streaming effects of other lesions. These streaming effects are sometimes more significant in patients with two outlets and a pulmonary arterial band, rather than a single outlet and a shunt. The clinical findings and the chest X-ray appearances of pulmonary vasculature markings, pulmonary venous congestion, and cardiomegaly should always be taken into account. If the table-side calculations are not in keeping with the clinical picture (simply do not make sense), it is important to repeat the entire saturation run. Shunt calculations by oxygen sampling are subject to very numerous potential errors with variations of some 10%–20% for each measurement run.

Consideration for Fontan completion

In infants with a CP shunt as the sole source of pulmonary blood supply, the saturations are initially 85% or above as the SVC return constitutes around 60% of total systemic venous return. As children grow up and get more active (and start running around), the relative proportion of systemic venous return from the SVC decreases to around 40% and resting saturations drop with age. Most centers consider completion of the Fontan circulation at about 4 years of age and 15 kg in bodyweight. Of late, the creation of an extracardiac fenestrated Fontan conduit from the IVC to the pulmonary artery using 18–20 mm GoreTex tubes has gained widespread acceptance.[9]

Invasive cardiac catheter evaluation is performed at BCH in all patients before Fontan completion. Typically, five French sheaths are inserted into both the right internal jugular vein and the femoral vein. If the femoral veins are blocked (some 15%) after staged surgery and intensive care therapy in the neonatal period, the femoral arteries are used and the heart and the atria are cannulated retrogradely. Pressure measurements are obtained at all sites.

Fontan hemodynamics

With Fontan completion, there is further unloading of the hypertrophied and enlarged ventricle. Whereas the ventricle had to initially pump both the systemic and pulmonary blood flow (some 300% of normal) during the first-stage palliation, the conversion to a CP shunt (Stage 2) implies a very significant volume reduction of the systemic ventricle. Blood flow from the SVC now constitutes the sole source of pulmonary blood supply. Ventricular load (Q_{vent}) changes dramatically during the course of the palliation [Table 1].

After the CP shunt, ventricular volume equals SVC return (= pulmonary blood flow) and the systemic venous return from the IVC.[10] After Fontan completion, the ventricle has to pump SVC and IVC return. Any right-to-left shunt through a fenestration improves ventricular preload by reducing total effective systemic venous vascular resistance and thereby improving overall cardiac output.

CATHETER HEMODYNAMIC AND ANGIOGRAPHIC ASSESSMENT

Invasive pressure measurements

The first pressure measurement to be taken in all children with univentricular heart circulation should be the ventricular pressure! The zero on the transducer should be adjusted to the level that the early diastolic pressure in the ventricle touches the baseline [Figure 1].

Most cardiac catheter hemodynamic studies nowadays are being undertaken under general anesthesia with intubation and positive pressure ventilation.

This practice of obtaining invasive hemodynamics under general anesthesia in these patients may be wrong as one of the major driving forces of the CP/Fontan circulation is self-ventilation, i.e., the generation of negative intrathoracic pressure during inspiration to increase systemic venous return from the liver and the body.

If studies are performed under general anesthesia and ventilation, it is important to use minimal positive

Table I: Ventricular volume, pulmonary and systemic flow ratios, and measurements during staged univentricular/Fontan palliation

	BT shunt/Sano	CP shunt	Fontan
Q_P/Q_S	$\dfrac{Ao_{Sat} - SVC_{Sat}}{PV_{Sat} - Ao_{Sat}}$	$\dfrac{Ao_{Sat} - IVC_{Sat}}{PV_{Sat} - SVC_{Sat}}$	$\dfrac{Ao_{Sat} - SVC_{Sat}}{PV_{Sat} - PA_{Sat}}$
Q_{Vent}	$Q_P + Q_S$	$Q_P + Q_{IVC}$	$Q_P + Q_{Fenestration}$
Q_S	$\dfrac{VO_2}{(Ao_{Sat} - SVC_{Sat}) \times Hb \times 0.134}$	$\dfrac{VO_2 \times (PV_{Sat} - Ao_{Sat})}{(PV_{Sat} - PA_{Sat}) \times (Ao_{Sat} - IVC_{Sat}) \times Hb \times 0.134}$	$\dfrac{VO_2}{(Ao_{Sat} - SVC_{Sat}) \times Hb \times 0.134}$
Q_P	$\dfrac{VO_2}{(PV_{Sat} - Ao_{Sat}) \times Hb \times 0.134}$	$\dfrac{VO_2}{(PV_{Sat} - Ao_{Sat}) \times Hb \times 0.134}$	$\dfrac{VO_2}{(PV_{Sat} - PA_{Sat}) \times Hb \times 0.134}$

Ao: Aortic, IVC: Inferior vena cava, Hb: Hemoglobin in g/dl, PA: Pulmonary artery, PV: Pulmonary vein, Qp: Pulmonary blood flow, Qs: Systemic blood flow, Sat: Saturations

Figure 1: Examples of incorrect zeroing of the pressure transducer in Fontan patients, which will contaminate/invalidate all subsequent measurements in this low pressure circulation in series. Top: Even during mechanical expiration, the early diastolic pressure reads 2–3 mmHg; thus, the transducer level has to be rezeroed at a level 3 cm higher. Bottom: The traces under-read by at least 5 mmHg! When readjusted, this patient will be documented to have very high ventricular end-diastolic pressures and may not be any longer a Fontan candidate, probably due to pump failure. Transducer level has to be rezeroed at 4 cm lower to get meaningful measurements
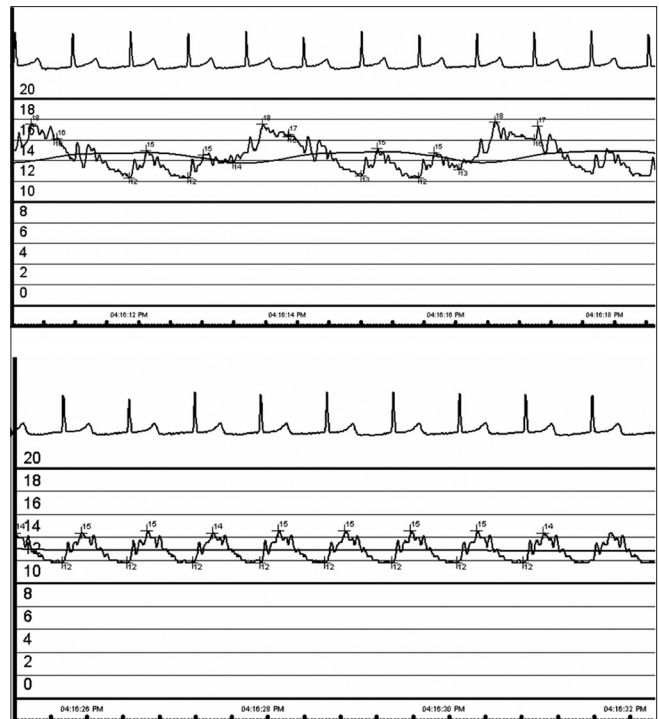

Figure 2: Effect of mechanical positive pressure ventilation on left pulmonary artery pressure recordings after Fontan completion. Top: During mechanical inspiration, there is a significant rise in pulmonary artery pressure to some 15 mmHg. During mechanical expiration, the pulmonary artery pressure drops to a mean of 13 mmHg. Note the time lag for mean pressure curve – rising during expiration and falling during early mechanical inspiration (averaged mean pulmonary artery pressure would be recorded as 14.5 mmHg), whereas when using apnea (held expiration), it is only 13 mm Hg (bottom). Pulsations on the left pulmonary artery tracing are due to transmitted compression by the reconstructed aortic arch in a post-Norwood patient

end-expiratory pressure so as not to reduce cardiac output. During mechanical inspiration, the intrathoracic pressure will increase and will decrease during mechanical expiration. Personal preference is to take all pressure measurements during mechanical expiration or while stopping ventilation for 30 s intervals. It is important to take/document all measurements during the same physiological state/phase of mechanical ventilation [Figure 2].

Most hemodynamic recording equipments calculate min/max/mean pressures over 2–5 cardiac cycles. It is advisable to go through the traces at the end of the case rather than taking the computer-generated numbers! This is essential data interpretation and should be done by experienced staff cognizant of the potential pitfalls and limitations of these traces.[8] Equally, the intermittent conversion to junctional rhythm will largely affect atrial and pulmonary arterial pressure recordings [Figure 3]. Care should be taken to record all tracings during the same rhythm, to make the data meaningful. A 24-h electrocardiogram tape is performed in all patients with intermittent junctional rhythm so as to inform

the decision as to whether sequential epicardial pacing lead implantation should be considered at the time of Fontan surgery.

One mmHg equates to 1.36 cm of water column. In a passive circulation in series (CP shunt and Fontan [total CP connection]), blood behaves like water – it flows along the path of least resistance!

It is extremely rare to record pressure gradients of more than 1 mmHg (during held expiration) anywhere in the CP shunt/Fontan pathways. These minor gradients represent very important obstructions to the circulation in series and will largely affect pulmonary afterload and with that overall cardiac output [Figure 4]. Such lesions require addressing by interventional stenting procedures of the branch pulmonary arteries.[11]

The most important aspect of pressure measurements is in the calculation of the transpulmonary gradient, which equals the difference between CP shunt/Fontan pressure and the pulmonary venous atrial mean pressure. That is, the figure that counts! That is the figure that overrides all zeroing errors and all the above limitations encountered
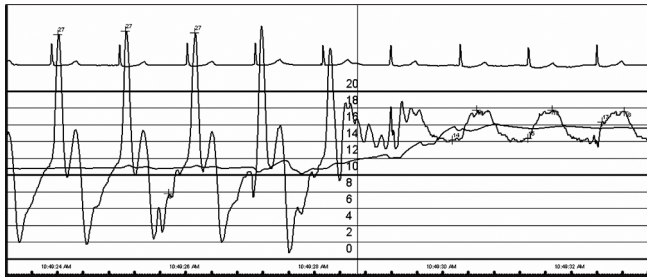
Figure 3: Pull-back trace from the pulmonary venous atrium (right atrium) to the Fontan circuit in a patient with fenestrated Fontan circulation and in junctional rhythm. Transpulmonary gradient is 5.5 mmHg. Note the huge a-waves with a peak of 27 mmHg within the pulmonary venous atrium. All pressure recordings should be taken during the same rhythm to be meaningful. If patients in junctional rhythm are symptomatic/have post-Fontan complications, then repeat measurements should be obtained with atrial pacing so as to inform the decision as to whether permanent (epicardial) pacing may improve hemodynamics/be of benefit

during mechanical ventilation. Again, it should be calculated during the same respiratory phase, the same rhythm, and ideally, during apnea [Figure 5].

A transpulmonary gradient of 3 mm Hg is acceptable for CP shunt patients. A transpulmonary gradient of more than 5 mmHg is very concerning in Fontan patients. A Fontan pressure of more than 20 mmHg is dramatically associated with poor outcome.[12] It will have to trigger (catheter) intervention where possible– or surgical takedown– to improve survival [Figure 6]. Thus, it is of crucial importance to be meticulous and absolutely confident about the baseline zero.

Pressure measurements are taken within the ventricle, the pulmonary venous atrium, the SVC, branch pulmonary arteries, the Fontan conduit, and also the IVC. If these are not completely uniform, allowing for the respiratory cycle, and there is concern about any potential narrowing on angiography pull-back gradients should be obtained during apnea [Figure 7]. Finally, it is important to obtain a pull-back gradient across the reconstructed aortic arch. Any arch obstruction with a gradient of more than 10 mmHg systolic, and with appropriate morphology on angiography, should be addressed by transcatheter balloon angioplasty. Cardiac output in the Fontan circulation is governed by the added resistances of the various components of the Fontan circuit, the resistance of the pulmonary vascular bed, and the pulmonary venous return back to the ventricle. Any added resistance or stenosis will further reduce overall cardiac output.

Calculations of flows and resistances by catheter

Oxygen saturations are being taken at most sites of the CP shunt or Fontan circulation. There is a significant error in sampling for oxygen saturations up to 3%–8% for repeat measurements in the systemic veins. Nonetheless, a step up of saturations from the SVC to the distal branch pulmonary arteries in a patient with a CP shunt should
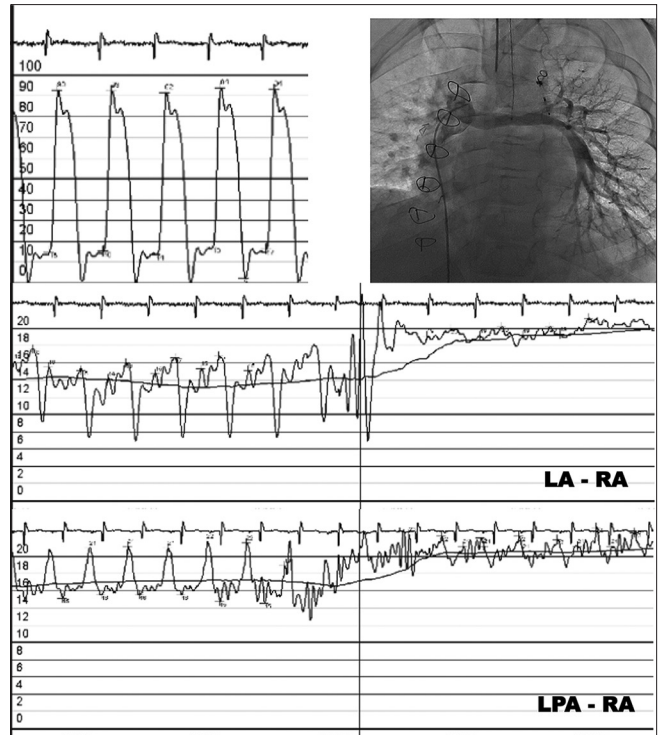

Figure 4: Hemodynamic study in a 13-year-old boy after staged fenestrated Fontan palliation with marked exercised induced cyanosis and in New York Heart Association II. The left ventricular trace shows an end-diastolic pressure of some 15 mmHg. Mean left atrial pressure is some 14 mmHg and Fontan pressure is approaching 20 mmHg mean even during apnea (middle). Pull-back gradient from the left pulmonary artery to the Fontan tube is approximately 3 mmHg mean during apnea, confirming very significant long segment left pulmonary artery stenosis as documented in the angiogram. This was addressed by stent implantation (relieving the gradient) and increasing cardiac output. In view of high Fontan pressures, fenestration closure was not contemplated

alert to the likelihood of significant systemic arterial collaterals to the lung. Equally, noticeably low saturations within the pulmonary veins should alert to the potential presence of pulmonary arteriovenous fistulae after superior or total CP shunt.[Figure 8].

Most centers perform cardiac catheterization under general anesthesia with an inspired oxygen concentration of 25%–30%. Assumed oxygen consumption in an anesthetized child after CP shunt procedure and reduced cardiac output who has been starved for some 6 h is low. It is generally overestimated using the original Lafarge tables.[13] The accurate measurement of oxygen consumption is only performed for research purposes.[14-17] Thermodilution techniques to determine cardiac output and flows have been largely abandoned. For clinical practice, the assumed values of 150 ml/min/m^2 in infants and 120 ml/min/m^2 in post-Fontan children are appropriate and accurate enough. At BCH, we do not measure dissolved oxygen in catheter studies undertaken in 30% oxygen or less.
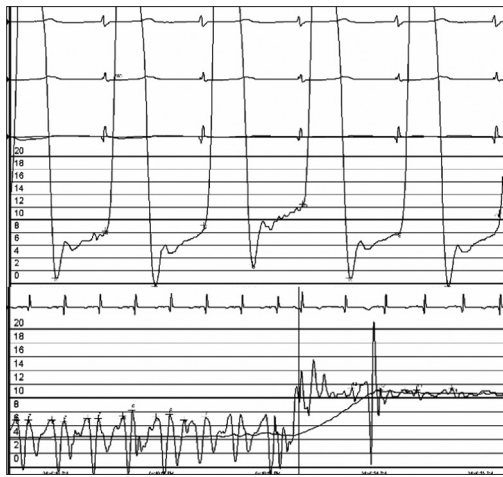
Figure 5: Catheter traces in an 8-year-old boy after fenestrated Fontan procedure for tricuspid atresia. There is good left ventricular function with low-end-diastolic pressure and low left atrial mean pressure of some 4 mmHg. Nonetheless, transpulmonary gradient is elevated with mean Fontan pressure approaching 11 mmHg. In the absence of Fontan pathway obstructions, the use of pulmonary vasodilators should be considered
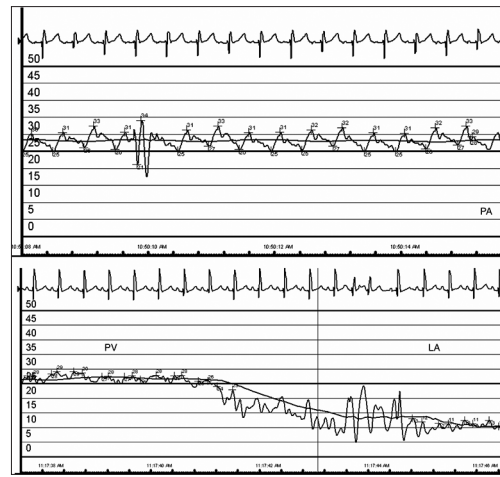

Figure 6: Early postoperative tracings after superior cavopulmonary shunt in a child with right atrial isomerism and anomalous pulmonary venous return. The mean pulmonary artery pressure (top) is 27 mmHg. Pull-back traces from the pulmonary venous confluence to the pulmonary venous atrium (left atrium) revealed a pressure gradient of some 16 mmHg requiring reoperation
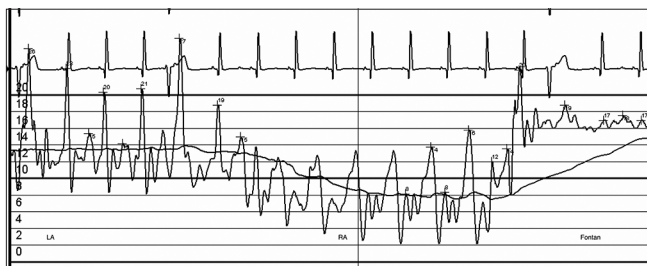

Figure 7: Pressure pull-back trace from the pulmonary veins to the morphologic right atrium and then to the Fontan circuit in a patient after fenestrated Fontan for double inlet left ventricle with a hypoplastic mitral valve. The trace reveals a major pressure drop at the level of the atrial septum of some 6 mm Hg (during general anesthesia), resulting in Fontan pressures of some 17 mmHg. The obstruction could be relieved by stent implantation
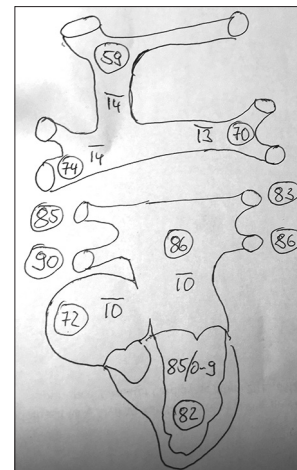

Figure 8: Schematic diagram of catheter hemodynamic study in a 2-year-old child after cavopulmonary shunt for pulmonary atresia with intact septum. Saturation recordings are listed at all sites as are mean pressures. Review of the data suggests systemic arterial collaterals (step-up of saturations from superior vena cava to branch pulmonary arteries) and significant pulmonary arteriovenous fistulae (low pulmonary venous saturations). At the same time, the transpulmonary gradient remains relatively high at 4 mmHg despite the fistulae!

Considering the significant sample variations of oxygen concentrations in systemic venous blood (some 3%–8%), the catheter calculations of flows and resistances remain simply a rough estimate [Table 1]. As a sense check, it is always useful to calculate systemic vascular resistance as well. This should typically calculate to be between 12 and 20 Wood Units. If unrealistically high or low values are obtained, repeat calculations should be made using lower or higher assumed values for oxygen consumptions. Indexed systemic cardiac output under general anesthesia, even in a good Fontan patient, rarely exceeds 3l/min/m^2.

As the catheter evaluation of estimated flows using the Fick principle and assumed oxygen consumption is limited and fraught with problems in a CP or Fontan circulation, all further calculations of vascular resistances are potentially contaminated at least they have to be interpreted against

the potential margin of error! As an adjunct to catheter evaluation of hemodynamics, a number of centers now utilize electrical bioimpedance cardiography which has been shown to give good estimates of systemic blood flow in a number of series.[18-20]

The gold standard in cardiac output measurement and relative flows to the pulmonary arteries, systemic collaterals, or fenestration flow at present is magnetic resonance imaging.[21-23] Nonetheless, even that technique relies on several assumptions (such that venous vessels

do not change caliber during the respiratory or cardiac cycle) and the inherent needs to measure small vessels by a technique which has limited spatial resolution and the need to trace flow curves. At present, only a few centers have the ability to combine invasive catheter hemodynamics and magnetic resonance imaging (MRI) evaluation simultaneously.[24-26] However, ultimately, this should be the best clinical practice, especially in patients who are considered for transplantation, for a failing Fontan circulation.

CONCLUSION

Cardiac catheter hemodynamic evaluation remains standard practice during the staged surgical approach in the management of univentricular hearts. Studies have to be conducted paying particular detail to the basics of catheter hemodynamic evaluation, and being aware of the many pitfalls and limitations, to best inform patient management. Newer techniques, such as MRI and impedance cardiography, should be added when possible so as to obtain the maximal information during catheter intervention or to inform future surgical management.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax 1971;26:240-8.

2. Gewillig M. The Fontan circulation. Heart 2005;91:839-46.

3. Gentles TL, Mayer JE Jr., Gauvreau K, Newburger JW, Lock JE, Kupferschmid JP, *et al.* Fontan operation in five hundred consecutive patients: Factors influencing early and late outcome. J Thorac Cardiovasc Surg 1997;114:376-91.

4. Gewillig M, Brown SC. The Fontan circulation after 45 years: Update in physiology. Heart 2016;102:1081-6.

5. Nakata S, Imai Y, Takanashi Y, Kurosawa H, Tezuka K, Nakazawa M, *et al.* A new method for the quantitative standardization of cross-sectional areas of the pulmonary arteries in congenital heart diseases with decreased pulmonary blood flow. J Thorac Cardiovasc Surg 1984;88:610-9.

6. Senzaki H, Isoda T, Ishizawa A, Hishi T. Reconsideration of criteria for the Fontan operation. Influence of pulmonary artery size on postoperative hemodynamics of the Fontan operation. Circulation 1994;89:1196-202.

7. Fick A. Überdie Messung des Blutquantums in den Herzventrikeln. Stahelschen Universitats-Buch Kunsthandlung; Sitzungsberichte der physikalisch-medicinischen Gesellschaftzu Würzburg; 1870. p. XVI-XVII.

8. Rudolph AM, Cayler GG. Cardiac catheterization in infants and children. Pediatr Clin North Am 1958;5:907-43.

9. Schreiber C, Hörer J, Vogt M, Cleuziou J, Prodan Z, Lange R. Nonfenestrated extracardiac total cavopulmonary connection in 132 consecutive patients. Ann Thorac Surg 2007;84:894-9.

10. Salim MA, Case CL, Sade RM, Watson DC, Alpert BS, DiSessa TG. Pulmonary/systemic flow ratio in children after cavopulmonary anastomosis. J Am Coll Cardiol 1995;25:735-8.

11. Noonan P, Kudumula V, Anderson B, Ramchandani B, Miller P, Dhillon R, *et al.* Stenting of the left pulmonary artery after palliation of hypoplastic left heart syndrome. Catheter Cardiovasc Interv 2016;88:225-32.

12. Kirklin JW, Fernandez G, Fontan F, Naftel DC, Ebner A, Blackstone EH. Therapeutic use of right atrial pressures early after the Fontan operation. Eur J Cardiothorac Surg 1990;4:2-7.

13. LaFarge CG, Miettinen OS. The estimation of oxygen consumption. Cardiovasc Res 1970;4:23-30.

14. Wolf A, Pollman MJ, Trindade PT, Fowler MB, Alderman EL. Use of assumed versus measured oxygen consumption for the determination of cardiac output using the Fick principle. Cathet Cardiovasc Diagn 1998;43:372-80.

15. Fakler U, Pauli C, Hennig M, Sebening W, Hess J. Assumed oxygen consumption frequently results in large errors in the determination of cardiac output. J Thorac Cardiovasc Surg 2005;130:272-6.

16. Li J, Bush A, Schulze-Neick I, Penny DJ, Redington AN, Shekerdemian LS. Measured versus estimated oxygen consumption in ventilated patients with congenital heart disease: The validity of predictive equations. Crit Care Med 2003;31:1235-40.

17. Rutledge J, Bush A, Shekerdemian L, Schulze-Neick I, Penny D, Cai S, *et al.* Validity of the LaFarge equation for estimation of oxygen consumption in ventilated children with congenital heart disease younger than 3 years – A revisit. Am Heart J 2010;160:109-14.

18. Zoremba N, Bickenbach J, Krauss B, Rossaint R, Kuhlen R, Schälte G. Comparison of electrical velocimetry and thermodilution techniques for the measurement of cardiac output. Acta Anaesthesiol Scand 2007;51:1314-9.

19. Norozi K, Beck C, Osthaus WA, Wille I, Wessel A, Bertram H. Electrical velocimetry for measuring cardiac output in children with congenital heart disease. Br J Anaesth 2008;100:88-94.

20. Noonan PM, Viswanathan S, Chambers A, Stumper O. Non-invasive cardiac output monitoring during catheter interventions in patients with cavopulmonary circulations. Cardiol Young 2014;24:417-21.

21. Grosse-Wortmann L, Al-Otay A, Yoo SJ. Aortopulmonary collaterals after bidirectional cavopulmonary connection or Fontan completion: Quantification with MRI. Circ Cardiovasc Imaging 2009;2:219-25.

22. Bossers SS, Cibis M, Kapusta L, Potters WV, Snoeren MM, Wentzel JJ, *et al.* Long-term serial follow-up of pulmonary artery size and wall shear stress in Fontan patients. Pediatr Cardiol 2016;37:637-45.

23. Kutty S, Rathod RH, Danford DA, Celermajer DS. Role of imaging in the evaluation of single ventricle with the Fontan palliation. Heart 2016;102:174-83.

24. Razavi R, Hill DL, Keevil SF, Miquel ME, Muthurangu V, Hegde S, *et al.* Cardiac catheterisation guided by MRI in children and adults with congenital heart disease. Lancet 2003;362:1877-82.

25. Kuehne T, Yilmaz S, Schulze-Neick I, Wellnhofer E, Ewert P, Nagel E, *et al.* Magnetic resonance imaging guided catheterisation for assessment of pulmonary vascular resistance: *In vivo* validation and clinical application in patients with pulmonary hypertension. Heart 2005;91:1064-9.

26. Fogel MA, Pawlowski TW, Whitehead KK, Harris MA, Keller MS, Glatz AC, *et al.* Cardiac magnetic resonance and the need for routine cardiac catheterization in single ventricle patients prior to Fontan: A comparison of 3 groups: Pre-Fontan CMR versus cath evaluation. J Am Coll Cardiol 2012;60:1094-102.