

Artículo especial

Management of univentricular heart

Mark G. Hazekamp

Kinderhartcentrum D6-26, Leiden University Medical Center, Leiden, The Netherlands



ARTICLE INFO

Article history:

Received 6 November 2013

Accepted 5 March 2014

Available online 13 April 2014

Keywords:

Univentricular palliation

Norwood

Glenn

Fontan

ABSTRACT

Univentricular pathway is required in any congenital heart disease unsuitable for biventricular repair. In the next manuscript we will describe the three stages of univentricular palliation, i.e. neonatal palliation, Glenn and Fontan procedures, specially focusing on surgical timing and complications, as well as indications and contraindications.

© 2013 Sociedad Española de Cirugía Torácica-Cardiovascular. Published by Elsevier España, S.L. All rights reserved.

Gestión de la vía univentricular

RESUMEN

La vía univentricular es necesaria en cualquier anomalía congénita en la que no sea posible realizar una reparación biventricular con éxito. En el siguiente manuscrito describiremos los 3 estadios de paliación univentricular, es decir, la paliación neonatal, Glenn y Fontan y enfatizaremos especialmente sobre el momento de su realización y las complicaciones, así como sobre las indicaciones y las contraindicaciones.

© 2013 Sociedad Española de Cirugía Torácica-Cardiovascular. Publicado por Elsevier España, S.L. Todos los derechos reservados.

Palabras clave:

Paliación univentricular

Norwood

Glenn

Fontan

Introduction

Univentricular heart management is needed for all cardiac anomalies where successful biventricular repair is not possible.

A summary of management of patients with a univentricular heart will be provided. Surgical management will be described for the first and preparatory neonatal procedures, the bidirectional cavopulmonary anastomosis (BCPA), the total cavopulmonary connection (TCPC) and finally also for revision of older type of Fontan operations to extracardiac TCPC. A time frame is suggested for these steps as they are currently in use in the Leiden University Medical Center.

Indications and contra-indications for these operations will be discussed, as well as complications and when possible, prognosis.

Forms of univentricular heart

Univentricular heart means that only one cardiac ventricle is available to support the systemic circulation. Many cardiac conditions fall into this category: hypoplastic ventricles (hypoplastic left heart syndrome (HLHS), pulmonary atresia with intact ventricular septum or hypoplastic right heart syndrome as well as several other anomalies with hypoplasia of one ventricle (e.g. unbalanced atrioventricular septal defect, transposition of the great arteries with one hypoplastic ventricle, double inlet left ventricle, double

outlet right ventricle (DORV) with hypoplasia of the left ventricle, etc.), absent valve structures (e.g. tricuspid, mitral and sometimes aortic atresia), straddling atrioventricular valves (e.g. double outlet right ventricle with straddling tricuspid valve), abnormal valves (hypoplastic mitral or aortic valves, extreme forms of Ebstein's disease, etc.), many hearts with heterotaxy syndromes and also hearts that possess two well-developed ventricles but where biventricular repair is not obtainable (e.g. giant VSD, DORV with straddling tricuspid valve).

Management goals and contra-indications

The main goal of management of a univentricular heart or its equivalent is to have one ventricle (or sometimes two ventricles) that supports the systemic circulation while direct cavopulmonary connection is used for the pulmonary circulation. Thus, there will not be a cardiac chamber to assist the pulmonary circulation. There is only one exception here, namely in tricuspid atresia where in older types of Fontan operations (direct or indirect (valved or non-valved conduit) connection of right atrium to the (rudimentary) RV) small RV was used to provide some assistance for the pulmonary circulation.

As a consequence pulmonary blood flow must be as smooth as possible; an elevated pulmonary pressure or better, an elevated pulmonary vascular resistance, hypoplastic or distorted pulmonary arteries as well as obstructed pulmonary venous drainage are absolute contra-indications for any form of cavopulmonary connection. This also means that anything that increases pulmonary vascular resistance should be avoided. Hypertrophy of the systemic

E-mail address: m.g.hazekamp@lumc.nl

ventricle, stenotic or insufficient valves of the systemic ventricle will eventually elevate pulmonary vascular resistance and this will negatively influence the functioning of the cavopulmonary connection. Anomalies of the cardiac rhythm will also decrease the efficiency of the cavopulmonary connection.

All this has to be taken into account when entering the pathway towards total cavopulmonary connection. This means that in the neonatal phase pulmonary overflow must be corrected as this will lead to an increase in pulmonary vascular resistance that, however small this increase might be, will eventually result in an impaired functioning of the cavopulmonary connection. For the same reasons obstructive and regurgitant valves must be dealt with at the earliest stage possible as they all will lead to ventricular hypertrophy which in turn negatively affects pulmonary vascular resistance. Pulmonary artery distortions and obstructions should be avoided when constructing systemic to pulmonary shunts and repair of anomalous pulmonary venous drainage should be done as early as possible. Loss of sinus rhythm should be prevented, especially when constructing the bidirectional cavopulmonary anastomosis.

Time frame

Neonatal management is preferably performed in the first month of life and for some conditions like HLHS even within the first 2 weeks after birth.

The second step is BCPA and is typically performed at an age between 4 and 8 months. In some instances this can be done earlier (e.g. after Norwood/Sano procedure, when the Sano-shunt gets obstructed) but not earlier than 3 months as pulmonary vascular resistance has not decreased sufficiently as arborization of the pulmonary arteries has not been concluded at that point. In case of double vena cava superior it is usually wise to wait at least 8 months as here the superior caval veins will be subsequently smaller and more vulnerable to obstruction.

The final step is TCPC where the vena cava inferior is connected to the (right) pulmonary artery by means of a Gore-Tex vascular tube that is sufficiently wide for adult life. As this means a diameter of 18–20 mm of the Gore-Tex tube, patient weight will have to be at least 15 kg. This is normally at an age between 3 and 5 years.

When there is vena azygos continuation of the vena cava inferior to a vena cava superior, cavopulmonary connection will be almost complete when the superior vena cava (or two, if there is also a persistent left vena cava superior) is connected to the pulmonary arteries (Kawashima procedure). Only the hepatic and coronary veins will not drain to the pulmonary arteries. This implies that a Kawashima operation will be usually scheduled somewhere at an age of 8–18 months.

Connection of the caval veins to the pulmonary arteries is done in two steps: first end-to-side anastomosis of the superior vena cava to the (usually right) pulmonary artery (BCPA) and later TCPC connecting the vena cava inferior to the (usually right) pulmonary artery. Most high volume centers now avoid TCPC in one step as the postoperative course is usually much less complicated when cavopulmonary connection is performed in two steps. Advantages of connecting the vena cava superior to the pulmonary artery prior to TCPC are several. There is sufficient preload remaining for the systemic ventricle and unloading of the ventricle is more gradual. Furthermore, the BCPA may be used as a final palliation when TCPC is not possible. Finally, the BCPA can be used as an intermediary step in a one-and-half ventricle repair.

Neonatal management

Neonatal management is aimed to create a pulmonary flow that is balanced: if pulmonary flow (e.g. pulmonary atresia) is

insufficient a modified Blalock–Taussig shunt is made and if pulmonary flow is too high pulmonary artery banding is done. Both procedures are done through a median sternotomy. This has several advantages; a Blalock shunt can be placed more centrally both on the brachiocephalic artery and on the pulmonary artery and is easier to close at the next operation. A median sternotomy offers an opportunity to inspect the rest of the cardiac anatomy and as all further operations will be through the same approach there is evident cosmetic benefit. Finally, a median sternotomy offers full access for cardiopulmonary bypass and is much safer than a lateral thoracotomy. Typically, a 3.5–4.0 mm thin-walled Gore-Tex tube is used as a modified Blalock-shunt as this preserves vascularity to the arm and is easier to undo later. During surgery 1 mg/kg heparin is given intravenously and the patient is placed on low dose of oral Aspirin later on. For pulmonary artery banding a Teflon-band is used that is fixed into position by either a small hemoclip or a thin suture. It is of importance to fix the banding proximally to the pulmonary artery wall in order to avoid distal migration and obstruction of the PA-branches.

Another goal of neonatal management is to procure unobstructed flow to the aorta. In hypoplastic left heart syndrome or equivalents (e.g. unbalanced AVSD with LV hypoplasia) this is done by means of a Norwood procedure (with a modified Blalock-shunt or a RV to PA (Sano)-shunt) that uses the pulmonary valve as the main outlet valve. Some forms of univentricular heart (e.g. double inlet left ventricle with coarctation) can be treated neonatally by aortic arch repair and PA-banding; if the entrance to the aorta will obstruct at a later stage this can be corrected at the time of the BCPA with a Damus–Kaye–Stansel procedure.

Anomalous pulmonary venous drainage should be corrected as soon as possible, especially when pulmonary venous drainage is obstructed as this will result in elevation of pulmonary vascular resistance that will have a negative influence on the functioning of later cavopulmonary connections.

If possible, insufficient atrioventricular valves should be addressed and repaired in the neonatal phase although this can be postponed and done at the time of BCPA.

It is important to evaluate the atrial septal communication and to open this further either by interventional procedure (Rashkin balloon atrioseptostomy) or by surgical atrioseptectomy.

Hybrid procedures are of recent interest: bilateral pulmonary artery banding and maintaining the patency of the arterial duct with prostaglandin infusion or with a stent can be beneficial in selected subgroups (e.g. hybrid Norwood).^{1–3}

Management in infancy

The interstage interval between neonatal surgery and BCPA is well-known to have a risk of mortality. For this reason infants should be monitored very closely by a dedicated pediatric cardiologist. Many centers now have home monitoring programs directed towards this specific group and some HLHS programs keep their patients in hospital until they have received the BCPA.

Neonates and small infants are especially vulnerable after the Norwood procedure because of the risk of coronary steal (by the modified Blalock-shunt) that can rapidly result in cardiac ischemia and death. After the Norwood/Sano variant RV-function may be decreased because of the scar in the RV anterior wall or a rapid obstruction of the Sano-shunt. Some univentricular hearts carry a high risk of mortality after pulmonary artery banding and aortic arch repair when there is a combined obstruction towards the pulmonary and the systemic circulation.

In this phase between neonatal palliation and BCPA all infants should thus be monitored very closely and the threshold for admittance into the hospital should be very low.⁴

Management around bidirectional cavopulmonary anastomosis

Ideally BCPA should be performed between 4 and 8 months. When double vena cava superior is present BCPA must be bilateral (BCPA with only one vena cava superior will result in formation of veno-venous collaterals between the vena cava superior that is still connected to the heart and the vena cava inferior and results in less efficiency of the BCPA). If BCPA is bilateral it is wise to wait until the child has reached a weight of approximately 8–10 kg. Superior caval vein diameters are smaller when there are two superior caval veins and technically, the operation is more difficult.

As the sinus node resides in the zone between vena cava superior and right atrium there is always a risk to damage the sinus node when performing a BCPA. Therefore it is advised to always leave a (very) short segment of superior caval vein attached to the right atrium and to take only very shallow suture bites when closing the atrial opening.

When performing a BCPA the vena azygos (or vena hemi-azygos when left-sided) has to be closed and divided in all instances. Leaving a vena (hemi-) azygos open will result in flow from the superior vena cava system to the inferior caval vein system and consequently, a decreased performance of the BCPA.

At BCPA the vena cava superior is anastomosed end-to-side to the pulmonary artery. Normally, this can be done without any patch but in some instances an autologous pericardial patch (untreated) may be used to relieve any tension on the anastomosis or to augment the BCPA. Ideally, the flow of the vena cava superior should be directed more towards the right lung than to the left lung. This is because the distribution of blood flow to the right lung is higher than towards the left lung. At the time of the TCPC the flow from the vena cava inferior should preferably be directed towards the left pulmonary artery. This will result in the best possible flow profiles without any collision of superior and inferior flows.

Sometimes it may be reasonable to leave another source of additional flow to the pulmonary artery. This can only be done when flow is limited as too much flow will result in backward flow into the vena cava superior and competition with BCPA function. A tight banding of the pulmonary artery or a tight pulmonary stenosis may be left untouched as these sources of (additional) forward flow will help in postoperative recovery after BCPA with some increase in oxygen saturations.

When it is decided to disconnect the pulmonary artery from the heart it is important to close the pulmonary valve as failure to do so can result in later thrombus formation at that location (with a risk of embolization into the aorta).

When oxygen saturations after BCPA are insufficient it is advisable to add a (small) modified Blalock-shunt and thus provide some extra pulmonary flow. On the other hand and especially in young infants we know that it may take some days for complete BCPA function and adequate oxygen saturations and for that reason we sometimes can wait and in the meantime assist decrease of pulmonary vascular resistance with nitrous oxide ventilation.

At the time of TCPC all sources of additional pulmonary flow should be closed as they will negatively influence TCPC functioning. After TCPC there is no longer a need for additional oxygenation anyway.

At the time of BCPA all remaining cardiac defect should be dealt with effectively to prepare the patient as good as possible for later TCPC. Remaining aortic arch obstructions should be repaired (normally with stent placement), when the flow to the aorta is limited because of a subaortic restriction (restrictive VSD, small LV outflow tract, small aortic valve annulus or a valvular aortic stenosis) it must be treated. Combining pulmonary and aortic valves and outlets can be done by the Damus–Kaye–Stansel procedure where a double-barreled cardiac outlet is formed. Any obstruction or regurgitation

of atrioventricular valves should be corrected as they will negatively affect the functioning of the cavopulmonary connection. A restrictive atrial septal communication must be opened whenever necessary.

In the first week after the BCPA the patient should be nursed in a half-sitting position as this will decrease congestion of the upper body half.

Management around total cavopulmonary connection

TCPC is performed using an extracardiac conduit in almost all cases; only when a straight pathway from inferior vena cava to pulmonary artery cannot be constructed, a lateral tunnel type TCPC is done. As the extracardiac conduit needs to be sufficiently wide for adult life, an 18 or 20 mm Gore-Tex vascular prosthesis is minimally required. This implicates that the child needs to have a body weight that is at least 15 kg.⁵ In our situation this will typically be around 3–4 years of age. In all cases we use a fenestration of 4 mm diameter. The fenestration will either close spontaneously later or is closed 1–3 years after TCPC in the cath lab.

TCPC is carried out on moderately hypothermic (28–30 °C) cardiopulmonary bypass with bicaval cannulation and on beating heart. Only when intracardiac repairs are needed, the aorta is cross-clamped and the heart is arrested with cardioplegia. It is important that the vena cava inferior is sufficiently developed and that the cannula is placed at some distance from the atrium. First, the vena cava inferior is detached from the atrium and the atrial opening is temporarily clamped. The anastomosis between the Gore-Tex tube and the vena cava inferior is made using a continuous 6-0 Prolene suture. After this, a 4 mm fenestration is punched into the Gore-Tex tube and then the atrial opening (where the vena cava inferior was connected) is sewn to the Gore-Tex tube over this fenestration. The tube is clamped to prevent air entering into the heart. The right pulmonary artery is dissected at the underside and freed from the left atrium. Then the right pulmonary artery is clamped and longitudinally opened on its lower aspect. If clamping is difficult the CPB flow is lowered and using a small-tipped sucker is used to keep the pulmonary artery dry. The anastomosis between right PA and the Goretex tube is usually beveled so that there is some slight inclination towards the left pulmonary artery. When 6-0 Prolene sutures are used it is not necessary to use surgical glues. The Goretex tube is carefully de-aired and all clamps are released. All remaining sources of additional pulmonary flow (e.g. an obstructed pulmonary valve or important systemic to pulmonary collaterals) are closed as they may interfere with efficient functioning of the TCPC. As in BCPA, we feel it is important to repair all remaining valvular defects whenever possible. Then, the patient is rewarmed and with TEE the results are controlled.

Postoperative management is usually smooth when a two-step approach is used (first BCPA and later TCPC) and when patient selection has been properly done. It is not uncommon that in the first postoperative day filling pressures need to be maintained at higher levels than normal. When inotropic support is needed this will typically be a combination of milrinone and noradrenalin. It is important to have pulmonary vascular resistance as low as possible and for that reason early spontaneous ventilation and early extubation is encouraged. After TCPC pleural drain production can be prolonged for days and in some cases even for weeks. Fluid restriction, diuretics and patience will usually be sufficient and drain production will finally always stop.

The fenestration between the external IVC to PA conduit and the pulmonary venous atrium can result in some arterial desaturation, arterial saturations will return to 100% after the fenestration is closed.^{6–9}

Anticoagulation

There is no general consensus on all aspects of anticoagulation in the management of univentricular heart. We will discuss here our anticoagulation policy briefly. In the presence of a (Gore-Tex) central or (modified Blalock) aortopulmonary shunt, Aspirin is prescribed to prevent shunt thrombosis. In BCPA no anticoagulation is needed as there is tissue-to-tissue anastomosis, only in exceptional cases Aspirin may be given. In the presence of a stent (e.g. in the left pulmonary artery after the Norwood procedure) a combination of Aspirin and Clopidogrel is prescribed. All TCPC's will have oral anticoagulation with Cumadins and this should be continued at least until the fenestration is closed; we feel however that either Cumadin or Aspirin therapy should be prolonged lifelong in the presence of a TCPC.^{10,11}

In the direct postoperative phase subcutaneous administration of low molecular weight heparin is usually given for thrombosis prevention.

Long-term follow-up

Rhythm disturbances are very frequent in patients with a Fontan circulation. Rhythm disturbances are typically supraventricular and may lead to an important decrease in the hemodynamic efficiency of the Fontan circulation. Therefore it is important to address this problem aggressively and if needed with transcatheter electrophysiological ablation and wherever necessary with pacemaker therapy.

Older types of Fontan circulation (atriopulmonary connections) frequently suffer from late dilatation of the right atrium. This will result in supraventricular tachycardias and stasis of blood flow. Together with a diminished performance of the systemic ventricle this may finally end in multi-organ failure. Liver cirrhosis and kidney failure are well-known to occur in adult patients with a Fontan circulation.^{12–14}

Fontan conversion

Conversion of atriopulmonary Fontan circulations to extracardiac cavopulmonary connection can reverse dilatation of the right atrium, tachycardias and improve the hemodynamic efficiency of these patients. This should however be done before the patient has become inoperable because of systemic ventricle failure, renal insufficiency and hepatic cirrhosis. Patient selection for Fontan to TCPC conversion should therefore be early and selective. Fontan conversion is usually combined with (surgical) ablation therapy (mini Maze with radiofrequency ablation) and placement of permanent pacemaker leads and devices.

Limitations

Although in the majority of pediatric cardiac surgical units these steps and time frames are followed, there are differences in management that are not fully described here. Some centers prefer a hemi-Fontan procedure over a bidirectional cavopulmonary anastomosis, as well as some use the lateral tunnel technique instead of the extracardiac conduit technique to create a TCPC. Fenestrations are not uniformly used and anticoagulation management is also different around the globe.

Conflict of interests

The authors declare they have no conflict of interests.

References

1. Backer CL, Russell HM, Deal BJ. Optimal initial palliation for patients with functionally univentricular hearts. *World J Pediatr Congenit Heart Surg*. 2012;3:165–70.
2. Fraser Jr CD. Management of systemic outlet obstruction in patients undergoing single ventricle palliation. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu*. 2009;70–5.
3. Baba K, Kotani Y, Chetan D, Chaturvedi RR, Lee KJ, Benson LN, et al. Hybrid versus Norwood strategies for single-ventricle palliation. *Circulation*. 2012;126 11 Suppl. 1:S123–31.
4. Hansen JH, Furck AK, Petko C, Buchholz-Berda R, Voges I, Scheewe J, et al. Use of surveillance criteria reduces interstage mortality after the Norwood operation for hypoplastic left heart syndrome. *Eur J Cardiothorac Surg*. 2012;41:1013–8.
5. Madan P, Stout KK, Fitzpatrick AL. Age at Fontan procedure impacts exercise performance in adolescents: results from the Pediatric Heart Network Multicenter study. *Am Heart J*. 2013;166:365–72.
6. Kaulitz R, Hofbeck M. Current treatment and prognosis in children with functionally univentricular hearts. *Arch Dis Child*. 2005;90:757–62.
7. de Leval MR, Deanfield JE. Four decades of Fontan palliation. *Nat Rev Cardiol*. 2010;7:520–7.
8. Walker SG, Stuth EA. Single-ventricle physiology: perioperative implications. *Semin Pediatr Surg*. 2004;13:188–202.
9. Villa CR, Marino BS, Jacobs JP, Cooper DS. Intensive care and perioperative management of neonates with functionally univentricular hearts. *World J Pediatr Congenit Heart Surg*. 2012;3:359–63.
10. Potter BJ, Leong-Sit P, Fernandes SM, Feifer A, Mayer Jr JE, Triedman JK, et al. Effect of Aspirin and warfarin therapy on thromboembolic events in patients with univentricular hearts and Fontan palliation. *Int J Cardiol*. 2013;168:3940–3.
11. Marrone C, Galasso G, Piccolo R, de Leva F, Paladini R, Piscione F, et al. Antiplatelet versus anticoagulation therapy after extracardiac conduit Fontan: a systematic review and meta-analysis. *Pediatr Cardiol*. 2011;32:32–9.
12. Mondésert B, Marcotte F, Mongeon FP, Dore A, Mercier LA, Ibrahim R, et al. Fontan circulation: success or failure? *Can J Cardiol*. 2013;7:811–20.
13. Goldberg DJ, Shaddy RE, Ravishanker C, Rychik J. The failing Fontan: etiology, diagnosis and management. *Expert Rev Cardiovasc Ther*. 2011;9:785–93.
14. Collins KK. The spectrum of long-term electrophysiologic abnormalities in patients with univentricular hearts. *Congenit Heart Dis*. 2000;4:310–7.



BIOMED



unidix

Especialistas en cirugía cardiovascular

desde 1977 al cuidado de tu salud



91 803 28 02



info@biomed.es