Interactive CardioVascular and Thoracic Surgery

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Interact CardioVasc Thorac Surg 2010;10:262-265; originally published online Nov 27, 2009;
DOI: 10.1510/icvts.2009.218586

doi:10.1510/icvts.2009.218586

INTERACTIVE CARDIOVASCULAR AND THORACIC SURGERY

Interactive CardioVascular and Thoracic Surgery 10 (2010) 262–265

www.icvts.org

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Volume load paradox while preparing for the Fontan: not too much for the ventricle, not too little for the lungs

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Received 11 August 2009; received in revised form 29 October 2009; accepted 2 November 2009

Summary

Ventricular dysfunction is frequently encountered in Fontan patients. Cardiologists and cardiac surgeons have, therefore, mainly focused on preservation of cardiac function, limiting the early volume overload as much as possible both in magnitude and duration. This resulted in improved cardiac function but, in some patients, also in poor pulmonary artery (PA) growth which in turn resulted in a poor final Fontan circuit. The volume requirements for optimal growth and development of the ventricle and the lungs are different and divergent. Avoiding overload of the ventricle is important, but excessive protection from volume overload may not be necessary and may result in PA hypoplasia, which in turn will severely affect the Fontan circuit.

Keywords: Univentricular heart; Fontan circulation; Cavo pulmonary connection; Palliation; Growth and resistance pulmonary arteries

1. Introduction

Over the past 50 years, palliation for the functionally univentricular heart (UVH) has vastly improved. In the 1950–1970s, large systemic to pulmonary artery (PA) shunts for adequate long-term relief of cyanosis were the treatment of choice. The dictum was: as pink as possible for as long as possible, typically many years. The enormous volume overload of the lungs resulted in satisfactory relief of cyanosis, but also in significant ventricular dysfunction, congestive cardiac failure and pulmonary hypertension, with few survivors beyond the 4th decade [1, 2].

Since the introduction of the Fontan operation [3, 4] almost 40 years ago, emphasis has shifted towards reducing the volume load of the ventricle; long-term overshunting proved to be detrimental, frequently resulting in dysfunction of the single ventricle as well as in some degree of pulmonary hypertension [5–7] (Fig. 1). Subsequently, neonatal shunts became smaller, designed to prepare the patient for Fontan repair. The staged approach in the beginning of the 1990s with early placement of a partial cavopulmonary Glenn shunt (usually performed at 4–7 months) further shifted the emphasis to limitation of volume load as early as possible. This strategy allowed the neonatal shunt to become smaller, typically lasting only a couple of months. Results improved even further, paralleled by an improvement of ventricular function.

Currently, at the new frontiers of UVH (hypoplastic left heart syndrome, unbalanced atrioventricular septal defect with right ventricular dominance), a very small neonatal systemic to pulmonary shunt is almost mandatory to maximally preserve the precious ventricular function [8]. The dictum has become: as blue as possible in order to keep the ventricle maximally unloaded.

However, many centres are now being confronted with new limitations associated with this latest strategy: patients have good ventricular function, but suffer from poor function of the Glenn shunt (significant cyanosis, congestion in upper body), and have an even worse Fontan circulation with low output and significant congestion [9].

The aim of this review is to reappraise the strategy of volume load to the cardiopulmonary system from birth to beyond Fontan.

1.1. Volume load and the single ventricle

At birth the single ventricle of a foetus with a UVH-complex is large and overgrown when compared to a left ventricle normal for body surface area (BSA). A normal left ventricle carries 40–45% of foetal cardiac volume output. If related to the size of this ventricle, relational mathematics dictate that a single ventricle carries 220–250% of the expected volume output [10]. This load leads to adapted overgrowth.

After birth a parallel circulation needs to be maintained until the pulmonary vascular resistance (PVR) has declined significantly, and the superior caval vein(s) and PAs have

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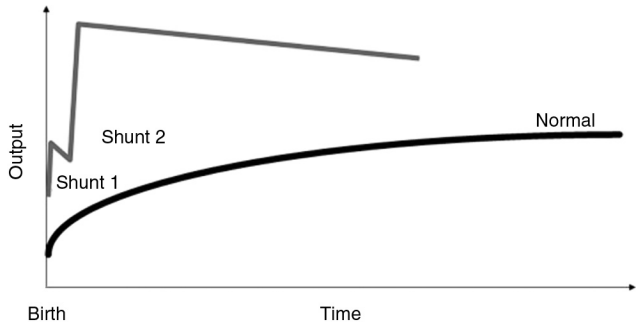

Fig. 1. Volume load on ventricle in management with two systemic to PA shunts prior to Fontan operation: UVH vs. normal (<1990s). The high volume load due to lesion and shunt demonstrated in this diagram represents management prior to 1990 (two or more shunts) for survivors to their 4th decade. Note that volume load on ventricle is at least double normal (thick line represents normal). PA, pulmonary artery; UVH, univentricular heart.

grown adequately. The pulmonary circulation is manipulated through banding of the PA (which decreases volume and flow in the PA) or placement of a systemic to PA shunt (which increases volume and flow in the PA). Volume load of the single ventricle will thus be around 250–350% when related to normal for BSA, but obviously less when related to expected ventricular size (Figs. 2 and 3). Such a significant volume load has several effects on the ventricle [11–15]:

- dilation of atrium and ventricle, partially reversible
- eccentric hypertrophy, similar to that seen in an athlete's heart and in patients with aortic regurgitation
- accelerated growth of heart and valves
- spherical remodelling, with re-orientation of myocardial fibres, partially irreversible
- increased wall stress, potentially leading to subendocardial ischaemia
- annular dilatation causing the atrioventricular valve to leak (more).

Prior to the 1990s, a Fontan circuit was typically created after two preceding aorta-pulmonary shunt procedures. The shunts caused significant dilation and overgrowth of the ventricle, which at the time of the Fontan operation was suddenly depleted of its customary volume load (Fig. 2). At the time of a Glenn or Fontan operation, the ventricle is unloaded to levels well below normal for BSA. The interposition of the PVR between superior caval flow (Glenn shunt) or the combined superior and inferior caval flows (Fontan circuit) and the systemic heart, limits venous return to the ventricle (Fig. 3). The Fontan operation unloads the ventricle to 50–70% of normal for BSA, but to <50% if significant overgrowth had occurred [16, 17]. Critical unloading in some patients resulted in 'collapse' of the ventricle as it suddenly had to perform at its closing volume [18]. Leaving a fenestration at the time of Fontan repair augmented the ventricular preload, however, at the expense of arterial desaturation [19].

It was concluded in those days that a large chronic volume overload is bad for the ventricle. Recommendations were: band early, do not be fooled by mild pulmonary valve stenosis, avoid a generous neonatal shunt, avoid a second arterial high-pressure shunt.

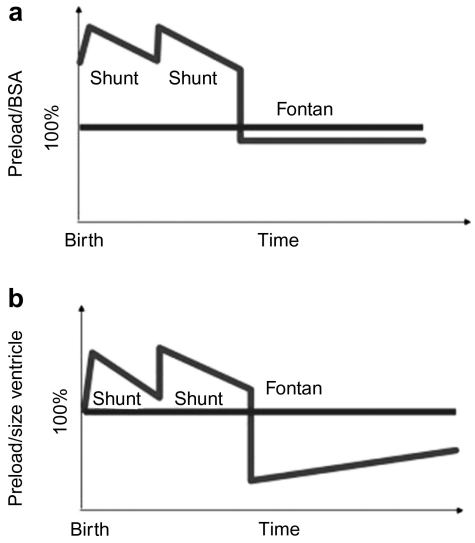

Fig. 2. Volume load to ventricle related to (a) body surface area (BSA) and (b) size of ventricle <1990s. Management with two classical systemic to PA shunts (typical <1990s). When relating all to BSA, one assumes a large ventricle due to pure stretch and dilation without any overgrowth. When related to ventricular size, one assumes a large ventricle due to adapted overgrowth without any stretch or dilation. Most patients will be positioned between both extremes with some dilation and some overgrowth. Unloading at the time of Fontan operation in model (a) down to 70% of 'normal' looks acceptable, but in model (b) when related to ventricular size makes the ventricle work at its closing volume. PA, pulmonary artery.

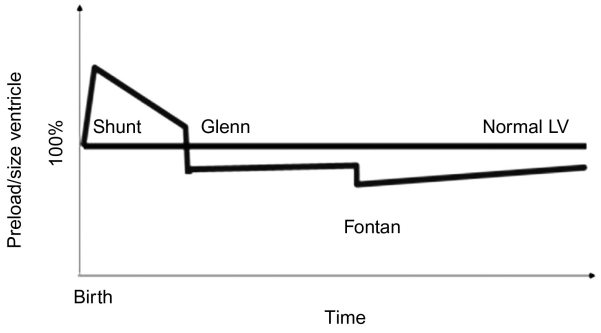

Fig. 3. Volume load to ventricle, current management since 1990. Since the 1990s, the ventricle is unloaded early at the time of a Glenn operation. By avoiding significant overload, excessive overgrowth and unloading is also avoided.

It became equally clear that excessive unloading also needed to be avoided. Recommendations were: rather perform the Fontan procedure for a blue than a pink (overshunted) patient, beware of atrioventricular valve regurgitation and concomitant repair or – if in doubt – perform stepwise repair and fenestrate the Fontan circuit.

1.2. Pulmonary vascular growth

A pulmonary vascular bed of high quality is essential for a good outcome after the Fontan circulation since, in most patients, PVR will control cardiac output. The importance of PA size on outcomes has been reported in several studies [20–31]. The logic is simple: in order to have a good Fontan, a well developed PA with a large diameter pulmonary vascular bed and low PVR are necessary [32].

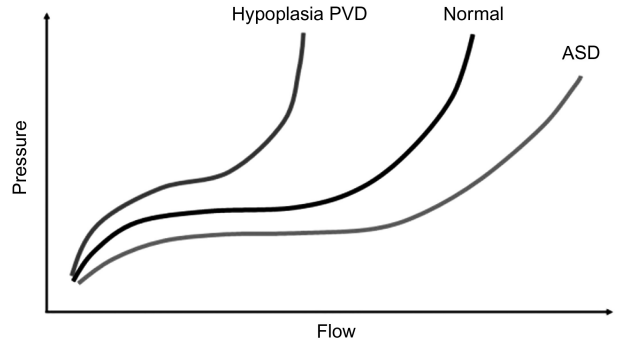

Fig. 4. Effect of flow on pulmonary arterial development and pressure. Patients with mild to moderate overflow due to intracardiac left to right shunts at low pressure for a limited time (e.g. ASDs) will have a large PA with low PVR. Patients with either excessive overflow or limited flow (e.g. hypoplastic PA) will have high resistance PA. PVD, pulmonary vascular disease; ASD, atrial septal defect; PA, pulmonary artery; PVR, pulmonary vascular resistance.

Sufficient flow at an adequate pressure is required to ensure continuing lung growth. This effect is demonstrated by studies assessing the haemodynamic effects of atrial septal defects (ASDs) on the PAs – significant PA growth associated with low PVR correlated with the magnitude of the shunt [14] (Fig. 4).

However, with the current staged strategy for UVH, only a limited period of controlled pulmonary 'overflow' is allowed to stimulate PA growth. The only phase of significant pulmonary flow is immediately after birth when a shunt or band is placed (Fig. 5). In many situations the systemic to pulmonary shunt is made as small as possible to avoid volume overload of the ventricle. If flow to the lungs is too low for too short a period, it may lead to inadequate PA growth, high PVR, and eventually a poor Fontan circulation (Fig. 5).

When creating a Glenn shunt as the sole pulmonary flow, the PA flow is reduced to $<50\%$ of normal for BSA. Potential for growth is thereby significantly restricted [33]. In many patients this is sufficient, but PA hypoplasia persists in some. In these, decreased capacitance and increased PVR is the inevitable result. Such high resistance will lead to poor performance of the final Fontan circuit.

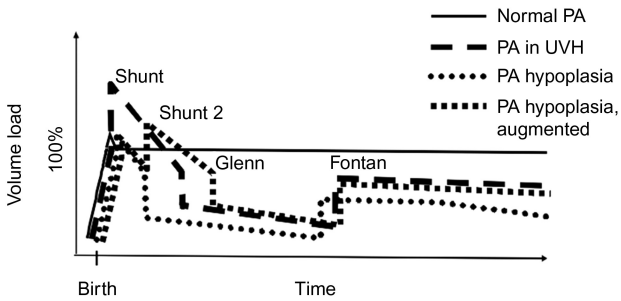

Fig. 5. Effect of pulmonary volume load on outcome of Fontan. Solid line, normal; broken line, ideal patient with UVH; dotted line, poor candidate with hypoplastic PA; square dots, poor candidate with hypoplastic PA, but augmented PA flow using a second or augmented systemic to PA shunt. PA, pulmonary artery; UVA, functionally univentricular heart.

Performing a Fontan operation will again increase PA flow, but flow will always be lower than normal, making catch-up growth after Fontan unlikely.

2. Clinical implications

In patients with significant hypoplasia of PAs, it might be advantageous if flow to the lungs could be augmented during the pre-Glenn period. The increase in pulmonary flow may again place a burden on the ventricle with additional overgrowth and possible dysfunction, but will enhance pulmonary vascular development. Ultimately, in patients with UVH evolving to Fontan circulations, it will be of more importance for long-term outcome to have good PAs with low PVR as opposed to perfect ventricular function. Many reports have demonstrated that mild impairment of ventricular function in Fontan patients results in acceptable clinical outcomes [34]. Therefore, if one has to choose between a Fontan circuit with a perfect ventricle and mildly increased PVR and a Fontan circuit with excellent lungs but mild ventricular dysfunction, clearly the choice will be biased towards the latter: such a circuit will have a better cardiac output!

In patients with mild to moderate PA hypoplasia, going for a classic bidirectional Glenn at the usual recommended time may thus not be the best option for good long-term results. Alternatives are: Glenn with additional pulmonary anterograde flow, creation of a second shunt, or increasing flow through the existing pathway (dilation of pulmonary stenosis, dilation of a dilatable band [35], stent expansion of stented duct [36] or expandable shunts [37]). This strategy could stimulate PA growth and lead to a better Fontan circuit with improved flow and even better ventricular function due to enhanced preload. This is demonstrated in Fig. 5. In a normal individual (solid line), there is a significant increase in pulmonary flow to 100% of normal at birth. In the ideal patient with a UVH (broken line), neonatal shunt/band allows for temporary pulmonary overflow and adequate PA growth; the patient slowly outgrows his palliation, at which time a Glenn shunt is performed and later followed by the Fontan operation. PA flow in this scenario equals cardiac output, and is only mildly decreased from normal. In the worst patients, the neonatal shunt provides only a limited period of overflow, quickly resulting in significant cyanosis. The dotted line shows the effect of a Glenn shunt in these: limited PA flow, finally resulting in a poor Fontan circuit with low and diminishing output. If however, flow is augmented after the first shunt (square dots), the augmentation of pulmonary flow may potentially result in additional growth; the Glenn shunt and Fontan are delayed, but with a better late hemodynamic result (more output).

3. Conclusions

It is better to have a patient after the Fontan operation with a well-developed pulmonary vasculature bed and mild ventricular dysfunction than one with normal ventricular function and poor cardiac output as a result of increased PVR. Avoiding significant volume overload of the ventricle is important, but aggressive protection from volume over-

load may not be essential. It may even be detrimental in the presence of PA hypoplasia, which could severely affect the Fontan circuit. Current strategies place too much emphasis on preserving cardiac function, which has short-term benefits. In order to allow good long-term outcomes, adequate growth and development of the lungs are more important.

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