Significance of systemic to pulmonary artery collaterals in single ventricle physiology: new insights from CMR imaging

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INTRODUCTION

Systemic to pulmonary artery collaterals (SPCs) are ubiquitous in single ventricle physiology, being found in nearly two-thirds of patients after a bidirectional Glenn operation and in approximately half of patients after a Fontan operation. They arise most commonly from the subclavian artery or one of its branches and supply systemic arterial blood to the distal pulmonary vasculature. The risk factors and pathogenic mechanisms that lead to the formation of SPCs remain poorly characterized, but chronic hypoxaemia, diminished pulmonary blood flow, surgical scarring, branch pulmonary artery stenosis, inflammation and abnormal flow patterns in the pulmonary arteries have all been hypothesised to contribute to their formation, although these hypotheses remain largely untested.

In theory, SPCs may have beneficial effects in single ventricle physiology. By providing additional pulmonary blood flow, SPCs may promote pulmonary artery growth and improve systemic oxygen saturation. They may also prevent the formation of pulmonary arteriovenous malformations by supplying ‘hepatic factor’. However, more attention is usually focused on the potential harmful consequences of SPC flow including ventricular dilatation, recirculation of oxygenated blood to the lungs resulting in ineffective pulmonary blood flow, energy losses related to competing sources of pulmonary blood flow and, rarely, erosion into a bronchus resulting in life-threatening haemoptysis. These physiological abnormalities have been hypothesised to contribute to adverse clinical outcomes.

Excessive SPC flow is thought to contribute to increased pleural drainage after a Fontan operation, which can prolong recovery in the postoperative period. In the longer term, persistent SPC flow has been implicated in the pathogenesis of ventricular dysfunction, heart failure and protein-losing enteropathy. However, most of these hypotheses have not been tested and the few data that exist are contradictory. For example, two similar studies that sought to investigate if increased SPC flow was associated with immediate postoperative outcomes after a Fontan operation arrived at conflicting conclusions. Spicer et al graded SPC on pre-Fontan x-ray angiography and found that higher angiographic SPC grade was associated with longer duration of pleural drainage after the Fontan operation. In contrast, Bradley et al quantified SPC flow intraoperatively by measuring the flow rate of pulmonary venous effluent while on cardiopulmonary bypass and found no association between this estimate of SPC flow and a number of postoperative outcomes including the duration of pleural drainage, pulmonary artery pressure or resource utilisation.

A lack of clarity regarding the clinical implications of SPCs has hindered the development of evidence-based management guidelines including indications for coil embolisation. This has contributed to wide practice variations between institutions. At one end of the spectrum are institutions that follow a practice of aggressive detection and coil embolisation of all SPCs, and at the other are institutions that do not embolise SPCs in any patients. It is not clear if a particular strategy leads to more favourable outcomes. Retrospective data from the Paediatric Heart Network did not find differences in early postoperative outcomes among participating centres despite widely varying rates of embolisation. Research in this area has been hindered by the lack of a reliable technique to quantify flow through SPCs in order to assess the haemodynamic burden they pose. x-Ray angiography is commonly used to identify SPCs, but grading SPC flow is confounded by technical and reader-specific factors. Other methods for quantification such as thermodilution techniques and nuclear scintigraphy have been proposed but have significant drawbacks that limit practical use.

CARDIAC MAGNETIC RESONANCE QUANTIFICATION OF SPC FLOW

The recent development of cardiac magnetic resonance (CMR) techniques for the quantification of SPC flow has led to a renewed interest in this field. Two independent but related methods have been described. The ‘systemic flow method’ calculates SPC flow as the difference between ascending aorta and total branch pulmonary artery blood flows. However, this method is invalid in the presence of a Blalock—Taussig shunt and neither method can differentiate between SPCs and systemic to pulmonary vein collaterals. Although no reference technique exists for comparative validation, indirect evidence suggests that the methods are valid. These data include good correlation between SPC flow and measures of ventricular dilatation, estimates of SPC flow in normal controls that agree with prior estimates of normal bronchial flow, good correlation and agreement between the two CMR techniques and modest correlation with x-ray angiographic SPC grade.

SPC FLOW AND FONTAN OUTCOMES

In the paper by Odenwald et al the results of a single-centre prospective study are reported in which SPC flow was estimated by CMR prior to the Fontan operation in 65 patients and data on early postoperative outcomes after the Fontan operation were collected in 41 patients. Their estimates of SPC flow in this cohort were consistent with those obtained by other investigators prior to a Fontan operation. In accordance with their institutional practice, no patient underwent cardiac catheterisation or embolisation of SPCs prior to the Fontan operation. Higher SPC flow on CMR was associated with adverse outcomes in the immediate postoperative period, including higher volume and...
duration of pleural drainage and longer duration of intensive care unit and hospital stay. Importantly, the effect of SPC flow on these outcomes was independent of other known predictors of outcomes such as age at operation, left or right ventricular morphology, fenestration, atrioventricular valve regurgitation, ejection fraction and transmural gradient. The relationships between SPC flow and measured outcomes were statistically significant but relatively weak (correlation coefficients 0.51–0.51 and area under the curve 0.74 for ROC analysis with postoperative chest drain volume >5 l/m²). Relationships with postoperative outcomes were stronger with the pulmonary flow method than with the systemic flow method. The relationship of SPC flow with mortality could not be assessed owing to the rarity of events.

These results are similar to those of Glatz et al.11 who also measured SPC flow by CMR prior to a Fontan operation and demonstrated that, after adjustment for Fontan type and fenestration, higher SPC flow was associated with dichotomous outcome variables including hospital duration ≥7 days (OR 9.2) and pleural drainage duration ≥10 days (OR 22.7). Linear relationships with outcome variables were either absent or less robust. Notably, similar to Odenwald et al., no patient underwent coil embolisation of SPCs prior to the Fontan operation, again reflecting institutional practice.

Both these studies—although limited by small numbers of patients—convincingly and consistently demonstrate a relationship between SPC flow and increased resource utilisation immediately after the Fontan operation. Although the association between SPCs and Fontan outcomes has been examined previously,7–12 those attempts were hampered by the lack of an objective method of estimating SPC burden. The introduction of CMR techniques brings more objectivity to the assessment of SPC burden, although it is important to note that the techniques are new and remain incompletely validated due to the absence of a comparable reference technique. Furthermore, while it is interesting to note the relationship between SPC flow and resource utilisation after a Fontan operation, it remains unclear if this relationship has implications for longer term and ‘harder’ outcomes such as death, transplantation, heart failure and protein-losing enteropathy. There is evidence to suggest that SPC flow is most prominent between the bidirectional Glenn and Fontan operations and declines progressively after completion of the Fontan operation, especially relative to body size. Late after the Fontan operation, SPC flow is not associated with adverse clinical outcomes apart from mild ventricular dilation.9

Longer-term follow-up of the cohort studied by Odenwald et al. will help improve our understanding of the ‘natural history’ of SPCs.

**PREDICTORS OF SPC FLOW**

In addition to assessing the relationship of SPC flow with outcomes, Odenwald et al. also examined relationships with possible predictors. In multivariable linear regression analysis, higher SPC flow was associated with younger age at bidirectional Glenn shunt, lower branch pulmonary artery cross-sectional area and higher systemic oxygen saturation. Separate analysis of a subset of patients showed that the presence of pulsatile antegrade pulmonary artery flow was associated with lower SPC flow. These findings are consistent with theoretical considerations and build upon previous work in this field.9 Risk factors and pathogenetic mechanisms for the formation of SPCs remain largely uncharacterised. Objective assessment of SPC flow by CMR should allow for further exploration of these and other predictors in the future.

**IMPLICATIONS FOR CLINICAL MANAGEMENT**

There is an increasing body of literature to suggest the validity and consistency of CMR estimates of SPC flow. In independent but nearly identical studies, Glatz et al.11 and Odenwald et al.10 have taken the next logical step by demonstrating a relationship between these estimates of SPC flow and immediate postoperative outcomes. The impact of these results on the clinical management of patients with single ventricle physiology remains unclear. An objective measure of SPC flow could prove useful in a variety of ways. CMR estimates of SPC flow could be used to guide the management of SPCs by providing a threshold for intervention and to monitor the short- and long-term efficacy of these interventions. It is important to note that most institutions continue to rely on cardiac catheterisation for diagnostic evaluation before staged surgical palliations in single ventricle physiology, although there is mounting evidence that, in a majority of patients, this could be accomplished at lower risk and cost by using a combination of echocardiography and CMR.13–15 The ability to quantify SPC flow adds further value to CMR evaluation, which can also provide additional information about vascular flow patterns that may be relevant to surgical planning.16 However, CMR in young children requires general anaesthesia or deep sedation with associated costs and risks. Identifying cost-effective diagnostic algorithms that help achieve the best outcomes while minimising risk should be the focus of further investigation.

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