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EDITORIAL COMMENT

Transcatheter Superior Cavopulmonary Anastomosis

Interesting Technique, Limited Applicability*

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The superior vena cava to pulmonary artery anastomosis (Glenn shunt) was a fundamental advance in the treatment of patients with complex heart disease (1). For patients who are not candidates for a 2-ventricle septated circulation, the Glenn shunt is usually the second surgical procedure that patients undergo on their way to a Fontan circulation, which completes the passive redirection of all venous return to the pulmonary arteries. Although not a "cure," the Fontan palliation has allowed many thousands of children with cyanotic congenital heart disease to survive to adulthood with a good quality of life. Sadly, the burdens of a Fontan circulation are becoming increasingly recognized as these patients begin to encounter morbidity and mortality, often in the second decade and beyond (2). In the absence of a better alternative, however, the infant born with a single-ventricle type of congenital heart disease is looking at these surgical palliations in his or her future.

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In this issue of the *Journal*, Ratnayaka et al. (3) describe a transcatheter method of performing a superior vena cava to pulmonary artery anastomosis—equivalent to a Glenn shunt. They performed this procedure on a previously unpalliated adult with severe cyanotic congenital heart disease and describe improvement in the patient's symptoms. Although this represents an interesting technique, it appears destined to be limited to a very rare set of adult congenital patients.

The usual course for an infant born with a single-ventricle type of circulation is to secure a pulsatile source of pulmonary blood flow that allows for sufficient pulmonary blood flow to achieve decent arterial saturation while avoiding too much pulmonary flow, which would "flood" the lungs causing heart failure and would prohibit somatic growth and the normal fall in pulmonary vascular resistance that is critical over the first months of age. If the infant is too cyanotic, this is accomplished by means of a systemic-to-pulmonary artery shunt (1 example of which is the Blalock-Taussig shunt). More recently, in select cases, ductal stenting has become another option to provide this temporary circulation (4). If there is too much pulmonary flow, pulmonary artery stenosis is created surgically by means of a pulmonary artery band to limit pulmonary flow and pressure. The infant with single ventricle and a banded or shunted circulation usually has an arterial oxygen saturation of around 80%, which empirically seems to provide the best balance between systemic oxygen delivery, while limiting the adverse effects of over-circulation (**Figure 1A**). Because the blood flow to the lungs in the shunted or banded circulation is already partially oxygenated, the circulation is inefficient. At an arterial saturation of 80%, the systemic to pulmonary artery flow ratio (Qp/Qs) is about 2:1, meaning that there is twice as much blood flow going to the lungs as is going to the body. Because there is only 1 ventricle doing the job, that ventricle is seeing the entire volume (3×).

At about 4 to 6 months of age, the infant with a systemic-to-pulmonary artery shunt or pulmonary

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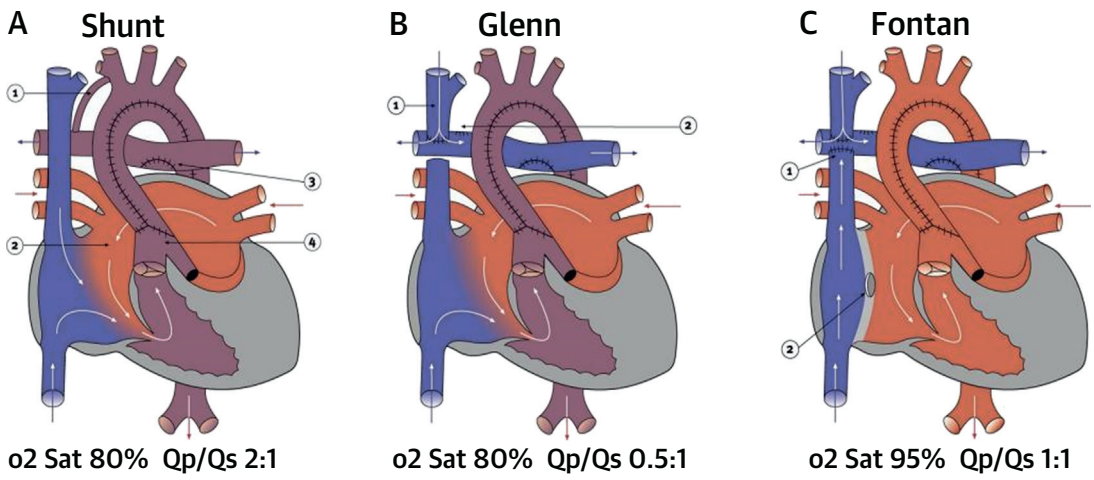
FIGURE 1 3 Staged Operations for Single Ventricle Physiology



(A) Shunted circulation after first stage repair for hypoplastic left heart. **(1)** Modified Blalock-Taussig shunt between the right subclavian artery and the pulmonary artery provides pulmonary blood flow. **(2)** The atrial septum has been removed to allow for mixing of systemic and pulmonary venous blood. The arterial saturation in a shunted circulation is ~80% with a Qp/Qs of 2:1. **(3)** Oversewn pulmonary artery stump. **(4)** Main pulmonary artery anastomosed to diminutive ascending aorta to create an unobstructed egress from the single ventricle. **(B)** After the Glenn operation at ~6 months of age, **(1)** the superior vena cava is disconnected from the right atrium and anastomosed to the cranial end of the right pulmonary artery. **(2)** The arterial shunt is removed. After the Glenn, the arterial saturation is still ~80%, but the Qp/Qs is now ~0.5:1. **(C)** At 2 to 5 years of age, the Fontan operation is performed. **(1)** The inferior vena cava is connected to the caudal aspect of the pulmonary artery. All of the venous return is now directed passively to the pulmonary arteries. A small communication is often left in the baffle to allow some residual right-to-left mixing, which improves perioperative hemodynamics. This communication (fenestration) often closes spontaneously or can be closed percutaneously some time later, increasing the systemic oxygen saturation to >95%.

artery band typically undergoes his next surgery—the Glenn operation (Figure 1B). With a Glenn, the superior vena cava (SVC) is disconnected from the right atrium and anastomosed in an end-to-side fashion to the cranial aspect of the right pulmonary artery. The blood from the SVC now passes to both lungs in a passive manner. The prior source of pulmonary blood flow is removed or severely limited. The inferior vena cava flow continues to return and mix with the pulmonary venous flow and is pumped to the body. The "magic" of the Glenn operation is that afterwards, the infant's systemic saturation is still about 80%; however, because only desaturated blood is going to the lungs, it is much more efficient. The Qp/Qs in a Glenn circulation is about 0.5:1. Because the pulmonary flow to the lungs is passive, the ventricle needs to only pump the systemic component (1×). The Glenn cannot be performed much before 3 to 4 months of age because the pulmonary vascular resistance is too high before that point to allow passive flow at an acceptable pressure. After the Glenn operation, infants tend to grow much better than with a shunted circulation having removed considerable cardiac and pulmonary work. In addition, by removing the volume-load on the single ventricle, the diastolic compliance of the ventricle is preserved—an important consideration for the future where a compliant single ventricle is critical for passive venous blood flow.

The completion of the single ventricle palliation called the Fontan is done between 2 and 5 years of age. It consists of directing the inferior vena cava to the pulmonary arteries (usually with a synthetic tube) completing the passive circulation to the lungs (Figure 1C). The patient is now fully saturated without a volume load on the single ventricle, albeit at the expense of elevated central venous pressure. Although generally well-tolerated in childhood, the chronic effects of elevated central venous pressure are being appreciated in an aging group of adult patients with Fontan circulation. These effects include arrhythmias, liver fibrosis and cirrhosis, lymphatic problems, thrombotic complications, protein losing enteropathy, and right heart failure symptoms (2).

Some patients with single ventricle physiology are born "well-balanced" with limited pulmonary blood flow so as to prevent the development of pulmonary vascular disease ("Eisenmeinger's") but enough pulmonary blood flow to allow an adequate arterial saturation typically in the 80% range. Even so, the

approach to that patient today would usually be to recommend the Glenn at age 6 months and the Fontan thereafter. That being said, decades ago or in a location without access to congenital cardiac services, or for reasons of patient/parent preference, the rare well-balanced single ventricle patient could escape surgery and grow to adulthood. With growth, however, this circulation that was "well-balanced" in childhood becomes inadequate, with a decrease in the systemic arterial saturation, severe polycythemia, and severe exercise limitation. Ratnayaka et al. (3) describe such a patient. Limited by severe cyanosis and secondary polycythemia, the patient they report was in a poor functional state. As the authors correctly point out, cardiac surgery in an adult patient such as this would have a high predicted morbidity and mortality. They describe a novel method to create a Glenn percutaneously using covered and uncovered stents. This method appears to have palliated the patient, significantly improving her oxygen saturation and functional status.

Although not diminishing the authors work, this technique is unlikely to have applicability much outside the rare, unoperated adult single-ventricle patient. Even if this procedure could be scaled down and done on much younger patients, a 6-month-old patient is a long way from an adult. There would also need to be some fundamental advances in stent technology to allow this type of an approach to be used in infants, as somatic growth would quickly outgrow the implanted size of any stent. Although the authors rightly point out that the surgical Glenn is not without complications, many of these complications are due to unfavorable hemodynamics. It is difficult to accept that a young patient in need of a Glenn would be better served with a percutaneous rather than surgical shunt.

Over the past 2 decades, there have been attempts to supplant the surgical Fontan step of the palliation with a transcatheter approach (5). The general idea has been to prepare for a transcatheter Fontan completion at the time of a surgical Glenn procedure such that the inferior vena cava to pulmonary artery connection could then be accomplished with a single covered stent from a "cuff" at the inferior vena cava to a "cuff" at the caudal aspect of the pulmonary artery. This is a much simpler prospect than a transcatheter Glenn procedure done in a much older patient (age 4 to 5 years compared with 6 months). Even this approach, however, has been mired in difficulties that have limited its clinically applicability, and except for some small series, this approach has not gained much traction.

Could the percutaneous Glenn approach be used outside of congenital heart disease in the adult patient? Right heart failure symptoms due to severe tricuspid regurgitation or right ventricle myocardial infarction could theoretically be ameliorated by unloading the right ventricle and routing part of the venous circulation directly to the lungs. Despite a few successful case reports (6), the surgical experience has been poor in this setting likely due to the noncompliance of the left heart in the older adult and prohibitively high pulmonary artery and SVC pressure that results after a Glenn in this setting. Although this is an interesting report of a technique, the transcatheter Glenn procedure is not yet poised to have a large effect in the field of congenital heart disease.

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