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Physiology of single ventricle, birth and beyond

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SINGLE VENTRICLE PHYSIOLOGY

This term is used to describe the situation wherein **complete mixing of pulmonary venous and systemic venous blood** occurs at the atrial or ventricular level and the ventricle(s) then distribute output to both the systemic and pulmonary beds. As a result of this physiology the:

- Ventricular output is the sum of pulmonary blood flow (Qp) and systemic blood flow (Qs)
- Distribution of systemic and pulmonary blood flow is dependent on the relative resistances to flow (both intra and extra-cardiac) into the two parallel circuits
- Oxygen saturations are the same in the aorta and the pulmonary artery

This physiology can exist in patients with one well-developed ventricle and one hypoplastic ventricle as well as in patients with two well-formed ventricles.

In the case of a single anatomic ventricle there is always obstruction to either pulmonary or systemic blood flow as the result of complete or near complete obstruction to inflow and/or outflow from the hypoplastic ventricle. In this circumstance there must be a source of both systemic and pulmonary blood flow to assure post-natal survival. In some instances of a single anatomic ventricle a direct connection between the aorta and the pulmonary artery via a patent ductus arteriosus (PDA) is the sole source of systemic blood flow (hypoplastic left heart syndrome or HLHS) or of pulmonary blood flow (pulmonary atresia with intact ventricular septum or PA/IVS). This is known as ductal dependent circulation. In other instances of a single anatomic ventricle intra-cardiac pathways provide both systemic and pulmonary blood flow without the necessity of a PDA. This is the case in tricuspid atresia with normally related great vessels, a nonrestrictive VSD and minimal or absent pulmonary stenosis.

In certain circumstances single ventricle physiology can exist in the presence of two well-formed anatomic ventricles:

- Tetralogy of Fallot (TOF) with pulmonary atresia (PA) where pulmonary blood flow is supplied via a PDA or multiple aortopulmonary collateral arteries (MAPCAs)
- Truncus arteriosus
- Severe neonatal aortic stenosis and interrupted aortic arch; in both lesions a substantial portion of systemic blood flow is supplied via a PDA
- Heterotaxy syndrome

Despite the fact that patients with totally anomalous pulmonary venous return (TAPVR) have complete mixing of pulmonary and systemic venous blood at the atrial level they do not manifest the other features necessary to create single ventricle physiology. This holds true as well for lesions in which a common atrial or ventricular chamber exists due to bidirectional (both L-R and R-L) anatomic shunting across a large defect (atrial septal or ventricular septal) and where there is no obstruction to ventricular outflow.

All patients with single ventricle physiology who have severe hypoplasia of one ventricle will ultimately be staged down the **single ventricle pathway to Fontan physiology**. Patients with single ventricle physiology and two well-formed ventricles will be able to undergo a two-ventricle repair. In some cases (truncus arteriosus, Type 1 and 2 tetralogy of Fallot with pulmonary atresia, severe aortic stenosis) the two-ventricle repair will be complete. In others (type 3 and 4 tetralogy of Fallot with pulmonary atresia) significant residual lesions (VSD, aortopulmonary collaterals) may remain.

With single ventricle physiology the arterial saturation (SaO_2) will be determined by the relative volumes and saturations of pulmonary venous and systemic venous blood

flows that have mixed and reach the aorta. This is summarized in the following equation: Aortic saturation = [(systemic venous saturation) (total systemic venous blood flow) + (pulmonary venous saturation) (total pulmonary venous blood flow)]/[total systemic venous blood flow + total pulmonary venous blood flow].

From this equation, it is apparent that with single ventricle physiology, three variables will determine arterial saturation:

1. The ratio of total pulmonary to total systemic blood flow (Qp:Qs). A greater proportion of the mixed blood will consist of saturated blood (pulmonary venous blood) than of desaturated blood (systemic venous blood) when Qp:Qs is high. An increase in arterial saturation that occurs in complete mixing lesions with increases in pulmonary blood flow relative to systemic blood flow. An arterial saturation approaching 100% is possible only with an extremely large Qp:Qs.
2. Systemic venous saturation. For a given QP:QS and pulmonary venous saturation, a decrease in systemic venous saturation will result in a decreased arterial saturation. Decreases in systemic venous saturation occur as the result of decreases in systemic oxygen delivery or increases in systemic oxygen consumption. Systemic oxygen delivery is the product of systemic blood flow and arterial oxygen content. Arterial oxygen content, in turn, is dependent on the hemoglobin concentration and the arterial saturation.
3. Pulmonary venous saturation. In the absence of large intrapulmonary shunts and/or V/Q mismatch pulmonary venous saturation should be close to 100% breathing room air. In the presence of pulmonary parenchymal disease, pulmonary venous saturation may be reduced. The V/Q mismatch component of pulmonary venous desaturation will be largely eliminated with a FiO_2 of 1.0 while the intrapulmonary shunt contribution will not be eliminated. For any given systemic venous saturation and QP:QS a reduction in pulmonary venous saturation will result in a decreased arterial saturation.

SUPERIOR CAVOPULMONARY SHUNT OR BIDIRECTIONAL GLENN (BDG) PHYSIOLOGY

The BDG directs systemic venous blood from the SVC directly to the pulmonary circulation. The BDG is normally undertaken at 3-6 months of age at which point the PVR has decreased to point where pulmonary blood flow can be provided with systemic venous pressure as the driving pressure. Patients who have outgrown their PA band, RV to PA conduit, or MBTS and have a low SaO_2 and patients who are not tolerating the additional volume on their ventricle with a loose PA band or large MBTS will be staged to a BDG earlier in this interval.

The original Glenn shunt involved an end-to-side anastomosis of the cranial end of the transected superior vena cava (SVC) to the distal end of the transected right pulmonary artery (RPA). Both the proximal SVC and RPA were over sewn. This procedure was performed through a right thoracotomy without CPB. Currently the bidirectional Glenn is used in which the cranial end of the transected SVC is anatomosed end-to-side to the RPA (which is in continuity with the main and left PAs) and the cardiac end of the SVC is over sewn. This creates SVC continuity with both the left and right pulmonary arteries and bidirectional pulmonary blood flow. The main PA is over sewn if it is not atretic. Unless the IVC is interrupted with azygous continuation the azygous vein is ligated so that the SVC does not decompress retrograde to the IVC thereby reducing the quantity of blood delivered to the PAs. This procedure is performed on CPB through a median sternotomy. The previous aortopulmonary shunt is ligated or the PA band taken down. Normally all upper extremity and cerebral venous drainage reaches the SVC. In some patients with congenital heart disease (particularly those with heterotaxy syndrome) there may be bilateral SVCs that are not in continuity via a connecting vein. In this case a **bilateral BDG** must be done with one SVC anastomosed end-to-side to the RPA and the other SVC anastomosed end-to-side to the LPA. LPA-RPA continuity is maintained.

The differences between an aortopulmonary shunt and a BDG are summarized. The most obvious difference between the two is that for a given cardiac output, SaO_2, $SpvO_2$, and SvO_2 the volume load on the systemic ventricle will be twice as large with an aortopulmonary shunt as it is with a BDG. Under these conditions effective pulmonary blood flow will be equal in the aortopulmonary shunt and BDG circulations but in the aortopulmonary shunt circulation there will be recirculated pulmonary blood while there is no recirculated pulmonary blood in the BDG circulation. A child with a BDG will have the same SaO_2 as a child with an aortopulmonary shunt with a significant reduction in the volume load on the systemic ventricle.

Modifications of the BDG have been devised to potentially simplify the ultimate conversion to a Fontan. The **hemi-Fontan** refers specifically to a procedure in which atriopulmonary anastomosis is constructed between the dome of the right atrium at the RA/SVC junction and the inferior surface of the right pulmonary artery. A Gortex baffle or «dam» is used to supplement the central pulmonary artery area and to isolate the cavopulmonary connection from the RA. Another modification incorrectly called a hemi-Fontan involves creation of a double cavopulmonary anastomosis. The cranial end of the divided SVC is anastomosed to the superior surface of the right pulmonary artery. The cardiac end of the

divided SVC is anastomosed to the inferior surface of the right pulmonary artery. The internal orifice of the superior vena cava is closed with a Gortex patch.

FONTAN PHYSIOLOGY

The Fontan procedure is generally performed in staged patients at 1-2 years of age. Fontan physiology is a series («normal») circulation that can be described as follows:

- There is one ventricle with sufficient diastolic, systolic, and atrioventricular valve function to support systemic circulation. This ventricle must in turn:
  Be in unobstructed continuity with the aorta
  Be in unobstructed continuity with pulmonary venous blood
- There is unobstructed delivery of systemic venous blood to the pulmonary circulation (total cavopulmonary continuity).

The essential function of the RV is not only to provide pulsatile flow through the pulmonary arterial system but to maintain a low pressure in the highly compliant systemic venous system, particularly the splanchnic bed[1]. A single ventricle is capable of pumping through both the systemic and pulmonary circulations arranged in series. However, the lack of an RV reservoir requires that systemic venous pressure be elevated, as there is, in essence, a continuous column of blood from the aorta to the systemic capillaries, systemic veins, pulmonary capillaries, and finally the pulmonary veins. Normally 70% of the total blood volume is contained on the venous side of the circulation with the venous circulation having a capacitance 19 times that of the arterial circulation. Fontan patients adapt to reduce venous capacitance (reduce unstressed volume) so that elevated systemic venous pressure can be maintained with a normal systemic venous volume (increased stressed volume). This makes them particularly vulnerable to stimuli that reduce stressed volume such as increased venous capacitance (loss of muscle tone, venodilation from any source) and reductions in vascular volume (blood loss or dehydration).

Pulmonary blood flow in the Fontan circulation is NOT «passive». This common misconception inhibits the ability to fully understand Fontan physiology. Pulmonary blood flow in Fontan circulation is non-pulsatile, continuous flow; the systemic ventricle provides the driving energy for this flow. The Fontan circulation places the systemic and pulmonary vascular resistances in series with the systemic ventricle.

Unfortunately, non-pulsatile pulmonary blood flow increases PVR by approximately 100% over than seen with pulsatile flow. Approximately 1/3 of the pulsatile energy generated by the RV is absorbed by the proximal pulmonary arterial system and redistributed in diastole to maintain recruitment of distal pulmonary vasculature. Loss of recruitment of distal pulmonary vasculature effectively reduces the area of the pulmonary vascular bed. This PVR increase is further exacerbated by the reduction in endothelial release of NO which accompanies long-term loss of pulsatile pulmonary blood flow[2]. In addition, in the absence of pulsatility the total hydraulic power (mean + pulsatile flow) is converted into a pure pressure gradient, increasing the energy necessary for transmission of blood through the pulmonary circulation[3]. The end result is an increase in the afterload on the single ventricle and a reduction in ventricular efficiency[4-6]. This makes Fontan patients vulnerable to further increases in afterload (PVR or SVR) and to reductions in contractility.

Positive pressure ventilation in Fontan patients is generally considered to be detrimental. The presumptive mechanism is the mechanical impediment of pulmonary blood flow with reduced delivery of blood to the systemic ventricle (reduced preload). Mechanical ventilation with reduced tidal volumes and low mean airway pressure may not be as detrimental to cardiac output in these patients as the factors generally associated with intubation and ventilation, specifically anesthesia/sedation (reduction of sympathetic output) and muscle relaxation (loss of muscular tone contribution to venous tone). Management of Fontan patients is further complicated by global impairment of cardiac autonomic nervous activity with reduced heart rate variability and baroreceptor sensitivity[7].

REFERENCES

1. Furey SA, 3rd, Zieske HA, Levy MN. The essential function of the right ventricle. Am Heart J 1984;107:404-10.
2. Khambadkone S, Li J, de Leval MR, Cullen S, Deanfield JE, Redington AN. Basal pulmonary vascular resistance and nitric oxide responsiveness late after Fontan-type operation. Circulation 2003;107:3204-8.
3. Mace L, Dervanian P, Bourriez A, Mazmanian GM, Lambert V, Losay J, Neveux JY. Changes in venous return parameters associated with univentricular Fontan circulations. Am J Physiol Heart Circ Physiol 2000;279:H2335-43.
4. Senzaki H, Masutani S, Kobayashi J, Kobayashi T, Sasaki N, Asano H, Kyo S, Yokote Y, Ishizawa A. Ventricular afterload and ventricular work in fontan circulation: comparison with normal two-ventricle circulation and single-ventricle circulation with blalock-taussig shunts. Circulation 2002;105:2885-92.
5. Tanoue Y, Sese A, Imoto Y, Joh K. Ventricular mechanics in the bidirectional glenn procedure and total cavopulmonary connection. Ann Thorac Surg 2003;76:562-6.
6. Senzaki H, Masutani S, Ishido H, Taketazu M, Kobayashi T, Sasaki N, Asano H, Katogi T, Kyo S, Yokote Y. Cardiac rest and reserve function in patients with Fontan circulation. J Am Coll Cardiol 2006;47:2528-35.
7. Davos CH, Francis DP, Leenarts MF, Yap SC, Li W, Davlouros PA, Wensel R, Coats AJ, Piepoli M, Sreeram N, Gatzoulis MA. Global impairment of cardiac autonomic nervous activity late after the Fontan operation. Circulation 2003;108:II180-5.