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INVITED EDITORIAL

The bidirectional Glenn shunt for univentricular hearts

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In 1958, working on the principle of partial right heart exclusion elucidated by Carlon et al. [1], Dr. Glenn published the superior vena cava (SVC) to right pulmonary artery (RPA) connection [2]. The initial Glenn shunt was an anastomosis between the side of the ligated SVC and the distal end of the transected RPA, and flowed to the ipsilateral lung alone, the contralateral lung being supplied by antegrade flow from the main pulmonary artery (MPA).

Today, the bidirectional Glenn shunt (BDG) is performed almost exclusively as an (end of) SVC to superior aspect (side) of ipsilateral pulmonary artery (PA) connection, and therefore, the superior caval return has drainage to both the lungs (i.e., it is bidirectional).

The principal indication for a bidirectional superior cavopulmonary shunt today is as staging towards a total cavopulmonary connection after which total exclusion of a deficient/insufficient right ventricle is achieved with complete elimination of mixing between the systemic and pulmonary venous return and normalization of systemic oxygen (O_2) saturation.

The BDG may be performed with or without the use of cardiopulmonary bypass (CPB). Requirement of a concomitant intracardiac procedure is one of the reasons for use of CPB which may also be used electively. However, it is also possible to perform the BDG without CPB; in which case, two angled cannulas connected together are used to drain the venous blood from upstream to the clamp placed on the SVC, to the right atrium with partial heparinization and passive drainage. The BDG can also be performed in a "Clamp and Go" fashion if the anatomy permits and if superiorcaval and ipsilateral PA clamping is tolerated (hemodynamically and with respect to systemic O_2 saturation) for the time it takes, which is usually less than 20 min. When the BDG is performed using decompressing cannulas but without CPB, the SVC pressure is found to be in the range of 20 mmHg, when done with just clamps and no decompression, up to a mean of 40 mmHg. To date, we have not encountered any complication in the BDGs done with just clamping and neither has there been any neurological /radiological neurological sequelae.

A bidirectional Glenn shunt is appropriate first-stage management for a functional or anatomical single ventricle situation (i.e., one not amenable to biventricular repair) and is said to improve candidature for future Fontan conversion.The aim is to achieve superior cavopulmonary diversion along with satisfactory ventricular function and mild or less atrioventricular valve regurgitation (AVVR), without systemic ventricle outlet obstruction, adequate PA size, and no ventricular inflow gradient, so that the requisite pre-Fontan requirements are met to the best extent possible. Pre- Glenn PA pressures are to be considered along with existent systemic arterial saturation to decide the upper acceptable level as they can be expected to come down once antegrade pulmonary blood flow is reduced or eliminated, and are the same as for a Fontan (< 15-mmHg mean).

Suboptimal results may be encountered if the BDG is performed in the setting of elevated PVR. Hence, the lower limit of age for surgery for a BDG has been lowered very cautiously. Today, a BDG is known to have been done even as early as 4–6 weeks after birth [3]. The principal indication for doing a BDG early is as stage II palliation following a Norwood procedure, especially if a systemic artery to pulmonary artery shunt-dependent pulmonary circulation exists. This is because of the known lability of this circulation, combined with the propensity for a small systemic to pulmonary artery shunt to occlude, thereby resulting in interstage attrition. The superior cavopulmonary shunt unloads the single ventricle and, at the same time, provides a more reliable source of pulmonary arterial inflow than the preexistent systemic arterial or systemic ventricle to PA shunt. In the younger ages, routinely doing a BDG below 3 months age is still a matter of debate, but in the absence of the negative prognostic factors for a BDG, the outcomes are, in general, good. Other known incremental risk factors for a BDG are (1) systemic ventricular dysfunction, (2)

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uncorrected AVVR, (3) iatrogenic or congenital pulmonary vein stenosis (PVS), (4) uncorrected systemic ventricular outlet obstruction, and (5) PA arborization abnormalities [4].

Uncorrected or recurrent AVVR and PVS (in the setting of repaired total anomalous pulmonary venous connection—TAPVC) are factors that play a role in the inferior results of BDG done in patients with heterotaxy. Correction of total anomalous pulmonary venous drainage in RA isomerism may result in an anastomotic lumen that is borderline due to the small sizes of the pulmonary veins, a small common chamber, and atypical disposition of the common chamber in the chest that makes appropriate posterior atrial incisions a challenge in a situation where there is tight pulmonary stenosis or pulmonary valvar atresia. Augmentation of pulmonary blood flow by means of a BDG or a complete Fontan will unmask a previously undiagnosed pulmonary vein ostial gradient, thus placing the entire circulation in jeopardy. The above along with the variations in venous and PA anatomy are some of the important reasons for the inferior results of single ventricle repairs in heterotaxy patients compared to non-heterotaxy counterparts.

Bilateral superior cavae are a concern because both are generally smaller in size than a unilateral cava. Hence, there is a concern regarding anastomotic gradients. Our own policy has been to try and delay a bilateral BDG in order to wait for growth of the cava. In our own hands, there have been cases where unilateral gradients have been detected by echo in bilateral BDG patients. The technical advantage in the presence of bilateral cavae is that one can do the anastomosis sequentially with the ipsilateral cava clamped, relying on an unobstructed contralateral cava to provide cerebral drainage. In general, in all superior caval anastomosis, it is our practice to make an extended pulmonary arteriotomy of at least 1.5 times the diameter of the cava to avoid the post anastomotic appearance of "beaking." In addition, we advocate releasing the SVC clamp and occluding a drainage cannula, if any, before tying the continuous suture, with the PA as yet clamped, to distend the anastomosis maximally and avoid purse-stringing of the venous anastomosis.

Antegrade flow is treated depending on post BDG SVC pressure, its pulsatility, and the increment obtained in systemic pressures upon snugging the MPA, which gives an idea regarding the amount of antegrade flow and, of course, the systemic O_2 saturation after the BDG. The advantage of retaining some antegrade flow into the PAs through the native MPA or through a patent ductus arteriosus or a systemic to PA shunt is an augmented pulmonary blood flow and hence better systemic O_2 saturations. However, this advantage comes with a tradeoff of elevation of SVC pressure and hence may need titration by means of a PA band. The other advantage is that this allows the so-called hepatic factor egress to the pulmonary circulation. Absence of exposure to this hepatic factor has been said to set the stage for early development of short-circuiting intrapulmonary vascular channels (pulmonary arteriovenous shunts) that results in systemic desaturation by not permitting pulmonary arterial blood to reach the alveoli [5, 6].

We have noted in our experience (and this is a finding that, to the best of my knowledge, is yet unreported in literature) that patients with unrestricted pulmonary blood flow have smaller diameter SVCs than those with reduced pulmonary blood flow. The possible explanation is unclear but in a patient with previously increased pulmonary blood flow and palliated by means of a PA band, a BDG often requires some degree of antegrade flow to be left open via the band to maintain acceptable postoperative O_2 saturations. In such banded patients where reduction of antegrade flow is also required to keep SVC pressure in limits, we often resort to suturing a Goretex patch to the interior of the MPA with a perforation to allow controlled antegrade flow.

Following a BDG, it is sometimes seen that cyanosis aggravates in the follow-up period disproportionate to that can be explained by the child's growth. Often, venous pop offs that steal the superior caval blood into the inferior caval circulation are the culprit. Rarely, they may communicate with pulmonary veins too. Sometimes an overlooked azygous or contralateral hemiazygous vein or even a vein of Marshall may be the culprit. If the pop offs are not a result of an overlooked preexistent vein, the etiology is usually elevated superior caval pressures arising from either excessive antegrade flow in the MPA, anastomotic narrowing at the Glenn, or elevated PA pressures and resistance. The latter situation is to be guarded against, as a large decompression may give rise to erroneously low-pressure readings and they therefore need to be occluded to give the true PA pressures. Pulmonary arteriovenous malformations as a cause of profound desaturation following Glenn shunt have already been mentioned.

A staging BDG is often avoided in preference to a single-stage Fontan operation in patients who are suitable for a Fontan operation but who present late, as the improvement in O_2 saturation obtained after just a BDG is only miniscule (especially if the main pulmonary artery is interrupted). This is probably because of the different quanta of systemic venous return through the superior and inferior vena cavae (IVC) at different ages. The inflection point is about 4–6 years of age when, due to preferential lower body growth, the inferior caval return outstrips that from the superior cava. In infancy, the metabolic state of the upper body predominates, and hence, the proportion of systemic arterial flow to the upper body is higher, thus enhancing SVC return. Likewise, all factors that increase cerebral vasodilatation (e.g., permissive hypercapnia) also increase SVC return and hence systemic O_2 saturation. As the patient grows, greater lower extremity somatic growth reduces this imbalance and completely inverts it during exertion when the arterial flow to the lower body far exceeds upper body supply. Hence, a BDG performed early

in life provides good saturation for only a limited period in the life of a patient and has to be complemented with IVC diversion to the lungs (i.e., Fontan completion) to maintain good O_2 saturation.

The experience from the All India Institute of Medical Sciences, New Delhi, published in this issue of the Journal [7] is remarkable in many ways. It is a consecutive series of 215 patients with univentricular (anatomically or functionally) hearts who received a superior cavopulmonary shunt between 2003 and 2013. It is one of the larger single-center experiences with this procedure and almost certainly the largest one from the developing world. Age less than 6 months at surgery came out as a risk factor for prolonged intensive care unit stay, while a small left PA and long CPB times also increased morbidity. None of the other known incremental risk factors for poor outcome with a BDG were found to be of any significance. In the follow-up period, 123 (60%) of the operated patients were catheterized and 69 had successful Fontan conversions.

A striking feature is in the demographics. While most published experiences so far are with ages far lower, and usually limited to infancy, the median age in this series is 3 years while the mean is 5 years, the age range being 1 month to 38 years. The other striking feature in contrast to available Western literature is that there is not a single patient of hypoplastic left heart syndrome (HLHS) in the entire series, while in any series from the West, HLHS would be one of the common lesions undergoing this operation. Late presentation probably accounts for both the above peculiarities.

However, there are some points that I would like to raise regarding the above experience of Talwar et al.

1. While the poor natural history of HLHS and lack of timely referral and, finally, reluctance of most families to take the staged approach starting with the Norwood procedure may to a large extent explain the absence of HLHS patients in the experience, single ventricle without protective pulmonary stenosis is also few in number. Only 11 patients had a prior PA band placed. In an experience stretching over a decade, one wonders as to the factors that could lead to such a small number of patients in the subset of single ventricle with (initially) unrestricted pulmonary blood flow being part of this experience.
2. Post Glenn mean O_2 saturation of 92.4% on room air are reported despite the advanced age in their group of patients, and in their paper, Dr. Talwar et al. recommend a bidirectional Glenn for effective palliation "in select patients" even up to adulthood. Now, it is a common knowledge that an aortic O_2 saturation of 80% is expected in a balanced pulmonary to systemic circulation with a Qp:Qs [ratio of pulmonary blood flow (PBF) to systemic blood flow (SBF)] of 1:1, given good mixing. A saturation of > 90% in a single ventricle situation implies a Qp:Qs of > 1.5:1. Given that the superior cava contributes a maximum of 60% of total venous return and assuming the IVC mixed venous O_2 saturation to be 70%, in the presence of free cardiac mixing, the maximum aortic O_2 saturation following a BDG without an accessory source of PBF cannot exceed 85–88% *(Aortic saturation = systemic venous saturation × total systemic venous blood flow + pulmonary venous saturation × total pulmonary venous blood flow / total SBF + total PBF).* An aortic O_2 saturation > 90% would signify an additional source of PBF, other than the SVC. The fact that antegrade flow in the PA was left open in only 16 of the 215 patients, therefore, makes the finding of a *mean* saturation of 92% inexplicably high. The 60% of systemic venous return through the SVC occurs only shortly during infancy and then comes down variably, and finally, down to 30–40% in later childhood and adolescence. The mean age of 5 years makes this high-saturation figure an enigma, indeed.
3. Among the additional procedures required at time of BDG, the commonest associated operation was a pulmonary arterioplasty for discontinuous PAs or origin stenosis of one PA (34/215), and this was presumably a statistically significant factor that prolonged intensive care unit stay (small left PA size is the factor that is mentioned). There is no mention regarding the fate of the pulmonary arterioplasties on follow-up. The authors have used oral anticoagulants in patients who have undergone pulmonary arterioplasty in the follow-up period to keep international normalized ratio (INR) values between 2 and 3, a practice rarely used by other groups. It would have been interesting to know of how the particular techniques of pulmonary arterioplasty used (end to end anastomosis with anterior autologous pericardial patch in the majority, complete tube of pericardium in 6) fared vis-à-vis medium-term patency of the connection and the size of the subserved PA, in the background of the knowledge that the venous pressure head and flow of a unilateral Glenn shunt may not always suffice to maintain good flow into a contralateral repaired/smaller PA [8]. The number of patients with heterotaxy is not mentioned, but going by at least atrioventricular (AV) valve and pulmonary venous drainage problems, right atrial isomerism was a rare diagnosis. By the authors own admission, only patients with low PA pressures were selected to undergo the bidirectional Glenn shunt. Preoperative ventricular function is not mentioned in the demographics and it is assumed that only patients with good ventricular function were selected.

Finally, this is a group of patients from a center that has the reputation of being the premier reference center for the entire country and thus receiving a huge patient load. Significant degrees of AV valve regurgitation were reportedly not

encountered in the single ventricle population that was considered for this intervention, and there were only 12 patients out of a total of 215 who were subjected to AV valve repair. Similarly, only 4 patients required repair of a total anomalous pulmonary venous drainage. It is no surprise, therefore, that none of the above-described incremental risk factors, save small size of left pulmonary artery (probably a surrogate for need for pulmonary arterioplasty), came out with any statistical significance as markers of poor outcome or increased morbidity. One rues the loss of opportunity for a fair investigation of risk factors, and a comparison of different ways of handling them, secondary to use of very strict selection criteria, in such a large series.

References

1. Carlon CA, Mondini PG, Demarchi R. Surgical treatment of some cardiovascular diseases (a new vascular anastomosis). J IntColl Surg. 1951;16:1–11.
2. Glenn WWL. Circulatory bypass of the right side of the heart. IV. Shunt between superior vena cava and distal right pulmonary artery: report of clinical application. N Engl J Med. 1958;259:117–20.
3. Jaquiss RD, Ghanayem NS, Hoffman GM, et al. Early cavopulmonary anastomosis in very young infants after the Norwood procedure: impact on oxygenation, resource utilization, and mortality. J Thorac Cardiovasc Surg. 2004;127:982–9.
4. Scheurer MA, Hill EG, Vasuki N, et al. Survival after bidirectional cavopulmonary anastomosis: analysis of preoperative risk factors. J Thorac Cardiovasc Surg. 2007;134:82–9.
5. Jonas RA. The importance of pulsatile flow when systemic venous return is connected directly to the pulmonary arteries. J ThoracCardiovascSurg. 1993;105:173–6.
6. Srivastava D, Preminger TJ, Spevak PJ, Lock JE. Development of pulmonary arteriovenous malformations following cavopulmonary anastomoses. (Abstract). Circulation. 1993;88:149.
7. Talwar S, Sandup T, Gupta S, et al. Factors determining early outcomes after the bidirectional superior cavopulmonary anastomosis. Indian J Thorac Cardiovasc Surg. https://doi.org/10.1007/s12055-017-0571-5.
8. Bacha EA, Lang P, Mayer JE Jr, McElhinney DB. Connection of discontinuous pulmonary arteries in patients with superior or total cavopulmonary circulation. Ann Thorac Surg. 2008;86:1948–54. https://doi.org/10.1016/j.athoracsur.2008.07.072.