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EDITORIAL

Optimising the surgical management of neonates with hypoplastic left heart syndrome

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Article Mroczek et al., see p. 1697

INTRODUCTION

For several decades, the optimal management of neonates with hypoplastic left heart syndrome (HLHS) and its variants has been the subject of ongoing controversies in congenital heart surgery. The introduction of a staged palliative surgical strategy by Norwood et al. [1] in the 1980s changed the fate of these severely sick patients tremendously. Since then, various modifications of the surgical technique, timing, and perioperative management have been developed, allowing comparable early survival and acceptable morbidity after stage 1. In the majority of cases, a modified Blalock-Taussig shunt (mBTS) was used as the source of pulmonary blood flow. However, significant interstage mortality and morbidity have been recognised, due to volume overload of the systemic right ventricle (RV), diminished coronary reserve, and reduced splanchnic perfusion owing to diastolic run-off to the low-resistance pulmonary vascular bed. The right ventricle to pulmonary artery conduit (RVPAc) as an alternative source of pulmonary flow, as suggested by Sano et al. [2], offered the rationale of limited pulmonary over-circulation due to systolic antegrade flow and absent left-to-right shunting at the arterial level, stable haemodynamic situation, and higher diastolic systemic arterial pressure. In comparison with patients on mBTS, early mortality and morbidity were repeatedly reported as more favourable [3, 4]. The enthusiasm for the use of RVPAc was somehow blurred, however, by the reports of a higher rate of interstage shunt-related re-interventions and questionable decrease in systemic ventricular function and transplant-free survival in the long term [5, 6]. Every effort to optimise the outcome of neonates with HLHS, either by refinements of the surgical technique or by improvements in perioperative care and advanced follow-up protocols, is greatly appreciated.

In the latest issue of *Kardiologia Polska,* Mroczek et al. [7] contribute their extensive experience of the Norwood stage 1 operation using the RVPAc in several modifications, finally proposing a refined surgical technique involving a ring-enforced conduit for “double dunk” implantation. Both the proximal and distal anastomoses of the conduit are fashioned by protruding either a part of the ring-enforced or ring-free margin to the lumen of the RV or pulmonary bifurcation. Although both “dunk” techniques were already described in recent years, the authors have developed their own refinements and demonstrate the direct impact of their strategy on a fair number of patients [8–11]. Furthermore, the authors studied, identified, and described in detail the potential reasons for their own and others’ suboptimal results. The comprehensive approach followed in their current group of patients clearly shows progress in optimising the outcomes of this complex patient cohort.

SURGICAL TECHNIQUE

Ring-enforced conduit

The use of a ring-enforced graft to overcome the potential obstruction of the substernal midportion of the RVPAc was first described by Schreiber et al. in 2009 [8]. In comparison to previous procedures with the use of an expanded polytetrafluorethylene (ePTFE; Gore-Tex) conduit (W. L. Gore & Associates, Inc., Elkton, MD, USA), the rate of substernal compression could be significantly reduced.

The “dunk” technique for RVPAc placement

Proximal dunk anastomosis

Direct anastomosis between the Gore-Tex graft and the epicardium layer of the RV ventriculotomy could lead to an obstruction due to myocardial hypertrophy or ingrowth of “neo-intimal” tissue. Worsening cyanosis was the trigger for unintended re-interventions. An effective strategy to overcome these drawbacks of RVPAc was elaborated and described as the “dunk” technique for proximal anastomosis [9–11].

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The ring-enforced Gore-Tex graft is cut flush with the first external ring at the proximal end of the conduit. A stab incision is made in the RV outflow tract, and the graft is placed into the ventricle to reach the previously marked third external ring. Two 6-0 purse-string sutures, running at the level of the epicardium, are used solely for the fixation of the graft on the RV.

Distal dunk anastomosis

Several measures can be employed to prevent the occurrence of stenosis of the distal anastomosis between the Gore-Tex graft and the pulmonary artery (PA) bifurcation. Extensive resection of the ductal tissue, suturing the graft first to a patch of the pericardium or homograft tissue and subsequently to the PA bifurcation, has been reported to ameliorate the distal stenosis with, however, inconsistent effects. The "distal dunk" technique was recently described by Mascio and Spray [12] as a combination of the previous methods, implementing the ring-enforced graft and "proximal dunk" techniques. The PA bifurcation is closed partially in a vertical fashion, leaving an opening for the distal RVPAc anastomosis. The distal end of the graft is trimmed, leaving 1 mm of PTFE beyond the last ring. The anastomosis is performed between the PA opening and the last ring on the graft using a 7-0 monofilament suture. The rigid ring and the free 1-mm end of the PTFE membrane protruding into the lumen of the PA effectively stent the anastomosis. The authors describe a series of 10 palliative stage 1 procedures using this novel technique, with no catheter-based or surgical intervention needed.

Lessons learned and improvement of outcome

Mroczek et al. [7] started with the use of the non-enforced RVPAc in Group I and II and experienced a higher rate of intervention on the proximal anastomosis, similarly to the evidence in the literature. Regardless of the position of the graft in respect to the neo-aorta the results were similar and logically led to a switch to the use of the re-enforced graft in the current subset of patients (Group III). Further, the authors hypothesised that the more extensive ventriculotomy used to decrease the potential of muscular obstruction or "neo-intimal" ingrowth could be responsible for future ventricular dysfunction and therefore changed again to a less invasive, cylindrical-shaped ventriculotomy using a 4.5-mm aortic punch.

In Group III, Mroczek et al. [7] adopted a similar rationale for the construction of the proximal anastomosis, as described above, and reported simultaneous introduction of a novel technique of fixation of the graft to the RV epi/myocardium. The authors emphasised the delicate positioning of the fixation sutures. Three 5-0 polypropylene sutures are introduced through the full thickness of the RV myocardium (including the endocardium) and fixed only to the third ring of the graft, without perforating the membrane. Using this modification, the "dunk" portion of the graft between the first and the third ring is entered into the RV at a distance equal to the thickness of the RV myocardium. As an additional effect, stitching full-thickness bites should prevent the ingrowth of "neo-intima" due to "everting" of the endocardium under the level of the first ring, i.e. the first ring protrudes freely into the RV cavity.

The construction of the distal "dunk" anastomosis is similar to that in the publication by Mascio and Spray [12]. The impact of the potential bow-tie effect on the geometry of the PA branches by rotating the original PA bifurcation site in a more cephalad direction, supported by computational figures, was briefly described previously by the same group [13]. The authors emphasised the more anterior location of the distal anastomosis by the creation of a new opening on the anterior aspect of the PA. Obviously, depending on the length of the RVPAc (up to 5.5 cm) and the position to the right of the neo-aorta, the appropriate location of the distal anastomosis could play a crucial role in the geometry of the pulmonary arteries.

With all these aspects taken into account, a significant improvement in outcome was achieved in Group III, as documented in the manuscript.

CONCLUSIONS

The current publication by Mroczek et al. [7] presents a comprehensive analysis of their recent process of optimising the surgical management of patients undergoing a stage 1 palliation procedure. Building on their extensive experience in the field, the authors integrated significant innovative approaches into a modern protocol, leading to an obvious improvement in the outcome of a difficult subgroup of patients. The authors are to be congratulated on their outstanding results stemming from their ongoing innovative efforts.

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