Hybrid Strategy for High-Risk Neonates with Interrupted Aortic Arch: A Can Well Worth Kicking?

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Abstract

Keywords

- interrupted aortic arch
- hybrid procedure
- low birth weight

The use of hybrid techniques to avoid neonatal cardiopulmonary bypass in high-risk individuals is well reported in the setting of hypoplastic left heart syndrome. We describe the use of that technique as a bridging procedure in high-risk neonates with an interrupted aortic arch. We report three cases where hybrid branch pulmonary artery banding and ductal stent implantation has been successfully used to defer complete repair, allowing recovery, maturity, and weight gain. This strategy may be considered for patients deemed at high risk for primary neonatal repair.

Interrupted aortic arch (IAA) is a rare congenital heart defect that continues to be a surgical challenge, especially when associated with complex cardiac abnormalities or noncardiac comorbidities including prematurity and low birth weight.¹ Large retrospective analyses suggest that in-hospital mortality and early postoperative mortality is in significant excess to that expected with most complex surgical pathology in the modern era.^{2,3} With advances in fetal echocardiography, most infants are diagnosed antenatally, allowing planning and discussion of different treatment options.⁴ Preoperative, operative, and postoperative care advances have significantly improved the outcome of patients undergoing neonatal repair.¹ While neonatal direct arch anastomosis is the preferred surgical approach, in many cases associated comorbidities confer very high surgical risk causing clinicians to seek alternative strategies. Neurodevelopmental follow-up studies have suggested that hospital survivors from neonatal repair have a significantly higher incidence of recordable psychomotor and neurodevelopmental deficit.⁵ Should a temporizing hybrid strategy akin to that performed in patients with hypoplastic left heart syndrome be considered in such high risk interrupted arch patients?

Case Reports

Between July 2013 and March 2016, a diagnosis of the interrupted aortic arch was confirmed in 16 infants admitted

to the Evelina London Children's Hospital. Out of the 16 infants 13(81%) were felt to be suitable for a primary surgical repair, but 3 patients were felt to be very high risk due to associated comorbidities, leading us to perform hybrid bilateral pulmonary artery (PA) banding and ductal stent implantation as a temporizing measure (►Fig. 1).

Our technique of hybrid palliation for left heart hypoplasia is well described elsewhere. Briefly, in all three cases, the procedure was performed through a midline sternotomy. After placing bilateral PA bands, a 6F short sheath was placed in the proximal main pulmonary artery via a purse-string suture. Following the angiographic assessment, the arterial duct was stented with self-expanding sinus-SuperFlex stents (OptiMed, Ettlingen, Germany). No additional procedures were performed.

All three procedures were uncomplicated, and each patient had an uneventful recovery and discharge. They all grew well at home, with each baby more than double their weight before full repair (►Table 1).

They were readmitted electively for full repair at time intervals based on a combination of clinical progress and echocardiographic follow-up. Patients 2 and 3 had standard repair of IAA with a combination of reapproximation of the aortic tissue augmented with a pulmonary homograft. Subjectively although the stents were easily removed from the ductus, reconstruction of the arch was felt to be more challenging than if performed as a primary procedure.

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Fig. 1 (A and B) Ductal angiography in a patient with IAA during stent implantation. Hybrid angiography via a sheath placed directly in the main pulmonary artery shows a long uniform ductus arteriosus supplying the descending aorta and the left subclavian artery. The orientation of the duct is quite different to that seen in standard hypoplastic left heart syndrome with no flow seen into the aortic arch, which is disconnected. IAA, interrupted aortic arch.

Patient one required the mobilization of the left common carotid and the left subclavian artery to restore continuity of the aortic arch.⁶ All three patients also had patch closure of their VSD. Patient one required surgical relief of left ventricular outflow tract obstruction 2 years following the complete repair followed by left pulmonary artery stent implantation. Neither of the other patients required any other surgical intervention since the complete repair. Follow-up posthybrid has been 39, 24, and 15 months, respectively, with no evidence of neurological or extracardiac concerns to this point.

Discussion

Hybrid palliation of hypoplastic left heart syndrome approach was developed as an alternative to the neonatal Norwood surgery, providing a less invasive, lower morbidity first stage.^{7–9} This approach has also been used as a bridge to allow decision making and potential growth toward biventricular repair in patients with borderline left structures.⁷ Outcomes up till now have demonstrated comparable results with primary Norwood procedures despite having sicker, smaller patients in the hybrid groups.^{10,11} Recently, some small studies have reported an extended application to other cardiac lesions, especially left heart obstruction lesions.¹² Karimi et al described a similarly high-risk IAA case in 2010 using an unusual strategy for hybrid sheath placement.¹³ Since then hybrid techniques have become better defined; as well as consensus regarding sheath placement, the availability of softer self-expanding stents for the ductus arteriosus minimize the intimal ingrowth and maximize the ability of the surgical team to mobilize and reconstruct the aortic arch.¹⁴

Our center has a 10-year experience with the hybrid procedure in the management of high-risk patients with hypoplastic left heart syndrome.¹¹ In this cohort study, we adopted the hybrid procedure in selected, high-risk patients with interrupted aortic arch. As well as the well-documented advantage of avoiding neonatal cardiopulmonary bypass surgery, with associated deep hypothermic circulatory arrest, delaying the repair to achieve a weight of approximately 4 kg, improves the operating field and the maturity of the tissues. One commonly discussed disadvantage of hybrid palliation in HLHS is the tenuous retrograde dependence of coronary blood supply in the setting of aortic atresia. This concern becomes irrelevant in the setting of IAA given the obligatory antegrade flow to the ascending aorta. Although repair at a later date and increased weight sounds inherently attractive; this has to be balanced with some potential negatives.^{15,16} There is undoubtedly a place for cross-sectional imaging in the early procedural planning, interim assessment, and postoperative assessment to inform the best timing and method for each stage. This application has been extensively described for delineation of complex arch anatomy in infants.¹⁷ Aortic tissue becomes less flexible over time; hence the potential to pull the descending aorta cranially and achieve a direct anastomosis may be limited. This is further hampered by the presence of the stent in the ductus, which may have generated a significant local tissue reaction, further limiting the mobility of the tissues in this region. Interestingly the softer self-expanding stents used by our group generate very little radial force, minimizing tissue trauma and reaction, hence providing only a minimal challenge to removal for our surgical team.¹⁴ Another consideration is the presence of adhesions given the redo nature of the repair, but in our experience, this is minimized by minimal

Table 1 Patient characteristics and follow-up data

No.	Gestation (wk)	Age at presentation (d)	Mode of presentation	Weight at hybrid (kg)	Age at hybrid (d)	Post-hybrid intubation days (hospital days)	Primary echo diagnosis	Extracardiac comorbidities	Weight at complete repair (kg)	Time between hybrid and complete repair (mo)	Post-Sx repair PICU days	Key echo findings 6–8 wk postoperatively	Long-term problems
1	33	7	Murmur	1.7	22	1 (34)	Normal situs and cardiac connections. Muscular VSD with a posterior deviation of the outlet septum. Type B IAA. Aortic valve Z score = -2.1 Mitral valve Z score = -0.4	IUGR	3.6	2	5	LVOTO Vmax = 2.8 AAo, ascending aorta Vmax = 2.5	LVOTO resection 18 mo postoperatively
2	40	3	Collapse	2.7	10	3 (10)	Normal situs and cardiac connections. Doubly committed VSD. Type B IAA. Aortic valve Z score = -2.8 Mitral valve Z score = -2.4	IUGR	6	8	3	LVOTO Vmax = 2.2 m/s AoArch Vmax = 2 m/s	None
3	35	0	Antenatal	2.1	7	1 (30)	Normal cardiac connections. Muscular outlet VSD. Type B IAA. Aortic valve Z score = -1 Mitral valve Z score = +0.1	22q11	4.9	4	5	LVOTO Vmax = 1.8 m/s AoArch Vmax = 1.8 m/s	None

Abbreviations: AoArch, aortic arch velocity measured in the distal arch directed toward the descending aorta; AAo, ascending aorta; IAA, interrupted aortic arch; IUGR, intrauterine growth retardation; LVOT, left ventricular outflow tract; MV, mitral valve; PICU, pediatric intensive care unit; VSD, ventricular septal defect.

Note: Velocities quoted are from continuous wave Doppler interrogation.

dissection at the time of the PA band placement. An objective discussion on the surgical behavior of tissues in the neonatal heart compared with those in older infants is not feasible. Homogeneity of the anatomy and individual tissue characteristics added to the confounding presence of the ductal stent; PA bands some degree of adhesions introduces too many variables to come to a meaningful conclusion.

All our patients had a successful secondary full repair during infancy with short, uncomplicated postoperative intensive care unit stays, and short overall hospital stays. Interval CT imaging was used in each case to help with surgical planning. In one patient the left common carotid artery was sacrificed along with the left subclavian artery to achieve continuity of the aortic arch (a technique occasionally required in a primary repair); after 3 years' postoperative follow-up this has not caused any neurological sequelae. It is difficult to know if this maneuver would have been required if a primary repair had been undertaken, but it should serve as a caution about the ability to easily achieve arch continuity later in infancy.

Conclusions

A staged approach to repair of complex patients with interruption of the aortic arch is a reasonable strategy when faced with the challenges of a primary repair in some instances. Modern protocols and approaches to hybrid palliation of critical left heart lesions provide a predetermined robust pathway for managing these patients. The trade-off between a neonatal primary repair and a temporizing procedure followed by secondary repair is difficult and gathering adequate patient numbers and reliable evidence to justify a change in approach is fanciful. Although our standard approach will continue to be a single stage definitive surgical repair, significant comorbidities are taken into consideration for each patient's individualized plan. In this small, selected high-risk series, a two-stage hybrid strategy is associated with good medium-term outcome. As such we would recommend considering this strategy when faced with such challenging circumstances.

Competing Interests and Funding
None.

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