CASE REPORT

Staged interventional and surgical management for complex low birthweight tetralogy of Fallot: another imperfect but legitimate strategy

Georgia May Connolly,¹ Catherine Armstrong,² Massimo Caputo,³ Gareth J Morgan⁴

¹Bristol Royal Infirmary, Bristol, UK

²Department of Paediatric Cardiology, Bristol Royal Hospital For Children, Bristol, UK

³Department of Congenital Heart Disease, Bristol Heart Institute, Bristol, Alabama, UK

⁴Department of Paediatric Cardiology, Guy's Hospital, London, UK

Correspondence to

Dr Georgia May Connolly,
gc7837@my.bristol.ac.uk

SUMMARY

Management of spelling low weight infants with complex tetralogy of Fallot is often fraught. We present a 2.8-kg infant with DiGeorge syndrome and spelling tetralogy with non-confluent pulmonary arteries, the left pulmonary artery being supplied by an aberrant ductus arteriosus from the left common carotid artery. We performed stenting of the outflow tract and the ductus to allow this patient to achieve a successful elective surgical correction.

BACKGROUND

Management of complex neonatal tetralogy of Fallot (TOF) has multiple approaches and there is no overall consensus as to the best strategy.^{1–3} The use of neonatal outflow tract stenting has added a potential temporising procedure to our armamentarium.^{4–8} This case illustrates the extremes of complex tetralogy with the left pulmonary artery (LPA) supplied by an aberrantly arising ductus arteriosus; providing a management dilemma in an unstable low weight infant.

CASE PRESENTATION

An ex-31 week gestation, 8-week-old infant (2.8 kg) with DiGeorge syndrome presented with tetralogy spells after a course of ibuprofen had failed to close what was thought to be a simple neonatal patent ductus arteriosus (PDA). Echocardiography revealed normal situs, levocardia, atrioventricular concordance, ventriculoarterial concordance, TOF with tight infundibular stenosis and valve stenosis (valve annulus=3 mm; z-score of -8), hypoplastic main pulmonary artery (MPA) and right pulmonary artery (RPA). The pulmonary arteries were non-confluent with the LPA supplied by an aberrant PDA which arose high on the left common carotid artery. The aortic arch was right sided. The LPA measured 2.8 mm (z-score -3.9) and the RPA 2.9 mm (z-score -3.9). The patient was started on prostaglandin and esmolol infusions. Saturations were in the low 70s and after repeated cyanotic spells we elected to undertake stenting of the right ventricular outflow tract (RVOT) and PDA.

TREATMENT

Catheterisation: All catheter procedures were carried out according to local protocols with systemic heparinisation.

Procedure 1

At 8 weeks and 2.8 kg, catheterisation was performed under general anaesthesia (GA) through the right femoral vein (RFV). Following an angiography in the RVOT, a balanced, middleweight coronary guide wire was positioned in the distal RPA and a 15×4 mm Multi-link Vision coronary stent (Guidant Corporation; Santa Clara, California, USA) was deployed, with satisfactory resolution of the infundibular and valvar stenosis (figure 1A,B). The catheter was advanced through the ventricular septal defect and using a 035" Terumo J-Tip Glidewire (Terumo medical; Somerset, New Jersey, USA) the origin of the PDA was intubated with a 5F right coronary guide catheter (figure 2A,B). After angiographic interrogation, the balanced middleweight coronary guide wire was passed into the distal LPA and three stents were sequentially placed along the length of the PDA starting from the origin of the LPA (Multilink 4×15 mm, 4×18 mm and 4×23 mm). Following deployment of each stent angiography demonstrated narrowing in the vessel immediately proximal suggesting that the ductal tissue ran at least as far back as the last stent deployed (figure 2C,D). The stents were left as far distal to the carotid artery as possible to avoid damage to the carotid and decrease the risk of embolic stroke. Angiography showed satisfactory patency of the ductal stents with filling of all LPA segments (figure 2C,D). Saturations improved into the low 90s and the baby was haemodynamically stable. The prostaglandin infusion and esmolol infusions were then weaned successfully.

Procedure 2

Three weeks later the saturations again decreased into the 60 s therefore a further catheterisation was performed. Again under GA the RFV was accessed with a 5F sheath and a 5F right coronary guide catheter again positioned in the PDA origin. The proximal vessel arising from the carotid had narrowed in keeping with ductal constriction; therefore after positioning a balanced middle weight coronary guide wire through the existing stents and into the distal LPA, an additional 22×4 mm Multi-link stent was positioned just distal to the carotid origin of the PDA. Saturations improved to the mid 80s and the patient was discharged home from the neonatal unit 2 weeks later on 5 mg/kg of aspirin.

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Figure 1 (A) The coronary stent in position across the right ventricular outflow tract (RVOT) with the 5F guide catheter and the balanced mid weight (BMW) wire distally positioned in the right pulmonary artery. (B) The critically narrowed RVOT (delineated with arrows); the BMW wire is across the RVOT and the guide catheter is in situ to deliver the stent.

Procedure 3

The third procedure was performed electively 3 months later after a general decrease in saturations to 70% in order to assess and redilate the RVOT and ductal stents. The infant's weight at this stage was 6.1 kg. Access was again through the RFV (5F sheath). Right ventricular angiography revealed significant RVOT stent in-growth with narrowing of the stent lumen. Over a 018" SV-5 wire (Boston Scientific Corporation, Natick, Massachusetts, USA) a Maverick XL monorail (5.5×20 mm) balloon was used to dilate this restenosis to 14 atmospheres multiple times. Repeat angiography showed a satisfactory result with evidence of a stable fracture at the distal aspect of the stent. The stented PDA was then angiographically assessed, identifying significant instent restenoses which were serially dilated along the length of the PDA length using the same Maverick XL balloon (5.5×20 mm). This gave a satisfactory angiographic result with saturations again rising into the mid 80s.

Surgical repair

At 13 months 8.7 kg a complete repair was performed. The saturations at this point had remained at 78–84% on room air however we were conscious of the potential detriment of further delaying complete repair of TOF. The patient's weight gain had also tailed off and it was difficult to determine how much this was due to his heart condition and how much was due to his genetic syndrome.

The operative repair was uncomplicated. The majority of the RVOT stent was removed through a transannular patch and the double outlet ventricular septal defect (VSD) was closed. The duct was divided and the LPA opened vertically. After removal of most or the ductal stent, the LPA was attached to the back of the MPA directly; a porcine patch was used to augment the LPA. The

patient recovered without complication and was discharged home 6 days later.

Post surgical catheterisation

Approximately 9 months following surgical repair (22 months, 14.5 kg) the patient underwent an elective exploratory cardiac catheterisation during which we electively dilated the LPA (figure 3). The distal arborisation of left and right pulmonary arteries was encouraging. The relative LPA size has increased from an initial z-score of −3.9 to a z-score of +1.8, with the Nakata index increasing from 66 to 260 (figure 3A,B).

Complications

The second stent to be deployed in the PDA migrated distally acutely, resulting in the distal margin of the complex of stents protruding into the main LPA without obstructing flow into any of the LPA branches.

Two days following the second procedure, the baby was noted to be having brief localised, left-sided seizures and cross-sectional cranial imaging; an MRI revealed two small areas of infarction in the right temporal and frontal lobes, in keeping with an embolic source.

DISCUSSION

This case demonstrates one of the current major dilemmas in management of TOF. Current management strategies in symptomatic neonates are:

1. Primary neonatal repair^{1–3};
2. Surgical palliation with a modified Blalock-Taussig shunt^{1 9};
3. Interventional catheterisation palliation followed by complete repair.^{4–8}

Figure 2 The patent ductus arteriosus (PDA; posteroanterior cranial projection) arising from the left common carotid (A). The catheter is positioned through the VSD in the proximal PDA. The tortuosity and length of the PDA is delineated with this view and the left anterior oblique (LAO) view (B). The initial discrete ductal narrowing can also be seen proximal to the true LPA. In both views the previously positioned right ventricular outflow tract stent can be seen. LAO projections of the stent complex in the PDA; the length of which was determined by the extent of ductal narrowing seen during the procedure (C and D).

Figure 3 Following complete surgical repair the left pulmonary artery (LPA; which was detached from the stented patent ductus arteriosus and reimplanted into the reconstructed main pulmonary artery) was angiographically examined and noted to have a significant stenosis close to its origin. The arborisation of the LPA can be well seen as can some stent remnants in and around the vessel. The final result following balloon dilation is seen in (B) and appears satisfactory (LPA z-score=+1.8). The projection is left anterior oblique.

The morbidity and mortality associated with either surgical shunts or neonatal repair are well documented in patients with uncomplicated tetralogy, not withstanding the additional complexity posed in this case.^{10 11} The UK's National Institute for Cardiovascular Outcomes Research website quotes the 30-day survival for all patients having a modified Blalock-Taussig shunts at only 90%; before loading for disease complexity and low weight or considering that this patient would have needed bilateral shunts.¹²

We carefully considered performing a complete repair prior to the third cardiac catheterisation at 5 months of age and 6.1 kg; however due to the perceived complexity of incorporating the disconnected LPA into a reconstructed RVOT and MPA, it was felt that a low-risk stent dilation procedure to stabilise saturations and allow further weight gain would be beneficial. At 8.7 kg and 12 months of age the full repair was challenging but achievable for the surgical team with only a 2-day stay in intensive care and four further days inpatient care necessary.

The potential complication of a cerebrovascular accident (CVA) in this case is concerning; however, the location of the MRI abnormalities are on the right side whereas the stent was implanted from the left carotid artery. We also intentionally left the origin of the aberrant PDA from the carotid unstented in order to decrease any thromboembolic risk. Nonetheless, we must consider that the cause of the CVA was thromboembolic and related to cardiac catheterisation. Thankfully, the patient's neurological symptoms abated and he is developing appropriately. Repeated cardiac catheterisations at an early age have their own intrinsic problems; the unquantifiable individual risk of radiation exposure, the possibility of loss of venous access vessels which will be essential in this child's future management and the risk of specific complications related to stent placement and dilation. Successful navigation through this gauntlet of problems may still however be more palatable than the mortality and morbidity associated with either neonatal complete repair or bilateral modified Blalock-Taussig shunt insertion which would have been necessary in this case.

An echocardiography during presurgical follow-up continued to show good flow to both pulmonary arteries with an RVOT continuous wave velocity of 3.2 m/s and good flow in the long PDA stent complex. Pulmonary venous return could be easily seen from two right-sided and two left-sided pulmonary veins.

The success of the eventual surgical repair with encouraging pulmonary artery development in this case may be seen as the end justifying the means. Predictably the patient returned at the age of 22 months for dilation of the LPA, however the arborisation and quality of the distal PA's was very satisfactory.

We would recommend consideration of an aggressive interventional strategy in such patients where neonatal surgery bears a high risk. In our case the patient has now reached 3 years of age and has had elective surgical correction followed by one catheterisation to dilate the LPA. We do not suggest that this approach offers an answer to the difficult conundrum of complex neonatal TOF however in this difficult situation we suggest that this model of care should be considered. The potential benefits are to promote growth and development of the pulmonary arteries, avoiding the need for neonatal cardiopulmonary bypass or the high mortality and morbidity associated with a modified Blalock-Taussig shunt.

Learning points

- ▶ An aggressive interventional strategy in patients where neonatal surgery bears a high risk should be considered.
- ▶ The benefits of stenting as a temporary management strategy include allowing time for the growth and development of the pulmonary arteries, which avoids the need for neonatal cardiopulmonary bypass.
- ▶ When undertaken at 13 months of age, the surgical outcome was successful in this case.

Contributors GMC cowrote the first draft and gathered the clinical details. CA cowrote the first draft. MC and GJM conceived the plan for the patient management; edited and structured the paper; reviewed the literature and approved the final draft.

Competing interests None.

Patient consent Obtained.

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