Unusual Spectrums Related to Coronary Artery in Surgery for Congenital Heart Diseases

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\section*{Abstract}

Background: Uncommon events in cardiac surgery may increase morbidity and mortality. We present an institutional experience of management of unusual events related to coronary artery in pediatric cardiac surgery to expand knowledge about them aiming to avoid related unexpected complications.

Patients and methods: Among 837 pediatric cardiac surgery cases, we recorded six cases who presented with unusual anomalies and complications related to coronary arteries between May 2017 and June 2019. Pre-operative echocardiography was the standard investigation for all cases and other specific work-ups were ordered according to each case. Management by surgical or non-surgical intervention was planned according to each unusual anomaly or complication. Post-operative hemodynamics and mortality rate were the main determinants of the outcome.

Results: Unusual scenarios related to coronary artery recorded 0.71\% among all cardiac operations in children. Early management of complicated coronary artery after arterial switch operation was the common procedure. Post-operative extracorporeal membrane oxygenator was indicated for 3 cases. One death was recorded.

Conclusion: Early management of unusual series related to coronary artery during cardiac surgery could lower risk and achieve favorable outcome.

Key words: Unusual; Coronary artery; Pediatric Cardiac surgery

\section*{Introduction}

Unexpected anomalies or complications related to coronary artery increase risk of cardiac surgery. Shetty [1] performed successful surgery for rare anomalous single coronary artery from pulmonary artery. He concluded that late incidental diagnosis leads to worse prognosis [1]. Our experience presents unusual anomalies or complications related to coronary artery in pediatric cardiac surgery to expand knowledge and set out clear safeguards about these rare scenarios.

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Patients and Methods

Among 837 cases of cardiac operations in children, we retrospectively analyzed six cases who were managed for unusual events related to coronary artery between May 2017 and June 2019. Pre-operative diagnosis, management, and post-operative assessment were recorded according to each scenario.

Unusual Coronary Artery Anomaly

1. Aberrant single coronary artery from pulmonary artery (ASCAPA): A full-term 1-month old male baby presented severe biventricular dysfunction with suspicion of aberrant left coronary artery from pulmonary artery (ALCAPA). Multidetector computerized tomography (MDCT) angiography and aortic angiography revealed ASCAPA (Figures 1 A, B). Cardiac surgery was done but unfortunately the location of the single coronary ostium was posterior so it was difficult for translocation. The surgeon tried to extend the coronary artery but the difficult position made the anastomosis stretched. Beside the extensive ischemic myocardium, the patient was unable to be weaned from CPB and was connected to an extracorporeal membrane oxygenator (ECMO). The patient died 5 days later due to an unrecovered severely depressed myocardium.

2. Coronary arteriovenous fistula (CAVF) from single coronary artery to pulmonary artery (PA) (Figure 2A): A full-term 1-month old female baby presented with pulmonary atresia, large sub-aortic ventricular septal defect (VSD) and overriding aorta. A striking large vessel was seen originating from the right coronary sinus of the aorta and gave rise to the right coronary artery (RCA), then finally inserted into the main pulmonary artery (MPA). Aortic angiography revealed a large fistula originating from the right coronary sinus and supplying both coronary arteries, and continuing to the MPA. Intra-operative assessment revealed no coronary vessel arising from the left coronary sinus; instead a large vessel originating from the right coronary sinus (single coronary artery) giving rise to the RCA, then curved to supply the MPA and ended by giving rise to the left coronary artery (LCA). VSD was closed, proximal MPA was separated from the coronary fistula, and a 12-mm right ventricle to pulmonary artery (RV-PA) conduit (Contegra) was performed. The post-operative period was uneventful and the baby was discharged home in good condition.

Figure 1. A: Aortic angiography Anomalous origin of a single coronary artery from the pulmonary artery (ASCAPA). B: Multidetector computerized tomography (MDCT) of the same patient (yellow arrow). C: Selective angiography for a fistula between the neo-aorta after arterial switch operation (ASO) (yellow arrow). D: Neo-aortic angiography revealed device occlusion management for the same patient (yellow arrow) with good coronary flow (white arrow).
Unusual Complication Related to Coronary Artery Surgery

1. Ostial RCA stenosis post aortic root replacement: A 12-month old female was born with severe aortic valve stenosis, coarctation of aorta and mitral stenosis (MS). The patient underwent balloon dilatation for both the coarctation and the stenotic aortic valve early in life. She then underwent open valvotomy for the aortic and mitral valves at the age of 8 months. After that, the patient had severe MS and severe aortic insufficiency. She was accepted for a high-risk operation of aortic root replacement by aortic homograft after Konno-type enlargement plus mechanical mitral valve replacement. After 20 minutes of weaning off CPB, the heart developed ischemic changes, so the patient was placed on ECMO and transferred directly to the catheterization lab. Angiography showed ostial stenosis of RCA that was stented successfully (Figures 2 D, E). After stenting, cardiac function dramatically improved and the patient was weaned from ECMO support and discharged home.

2. Complicated intramural LCA during arterial switch (ASO): A one-year old boy presented with double outlet right ventricle (DORV), subpulmonic VSD, transposition of great arteries (TGA) and hypoplastic aortic arch. He was accepted for high-risk total correction. The VSD was closed, and arterial switch and arch repair were performed. The left coronary artery has an intramural course, so it was anastomosed using aortic wall pedicle patch. Post-operatively, the patient was unable to be weaned from bypass and required ECMO support. He was sent back urgently to the OR to check for the coronary anastomosis, which was problematic due to the intramural nature of the left coronary. The left coronary anastomosis was revised, moving the site more lateral and using a piece of autologous pericardium to patch the proximal half of the left main coronary artery. However, we failed to come off CPB, so the patient was switched back to ECMO and transferred urgently to the catheterization lab to check for coronary patency and flow.
Surprisingly, cardiac catheterization showed no compromise to coronary patency but large neo-aorta to RV fistula (iatrogenic) was seen producing coronary steal. The fistula was closed using 4 vascular plugs after which cardiac function improved over time and the patient was weaned from ECMO a few days later (Figures 1 C, D).

3. **Missed RCA transfer after ASO**: A one-year old baby had DORV, TGA, and coarctation (COA). The patient underwent pulmonary artery banding and coarctation repair in the neonatal period. Redo surgery was done for VSD closure, debanding, and ASO. Due to extensive adhesions, only a single coronary artery was identified intra-operatively. Hence, single coronary button reimplantation was performed. Post-operatively, the patient developed severe RV dysfunction. Aortic angiography and MDCT angiography revealed a missed non-transferred RCA (Figures 2 B, C). Redo surgery was performed to transfer the RCA from the neo-pulmonary to the neo-aorta. After surgery, echocardiography revealed progressive improvement of RV function and the patient survived.

4. **Difficult coronary artery transfer during ASO**: A 20-day old female baby weighing 2.3 kg was referred to our hospital as a case of DORV, TGA, subpulmonary VSD and interrupted aortic arch type B. The decision was to do total repair. After aortic arch reconstruction using equine pericardium and Le Compet technique to accomplish ASO, we identified a very small ascending aorta 3 mm in diameter, which made the regular ASO impossible due to difficult coronary artery transfer, especially the coronary button technique. It was controversial to do single versus biventricular repair. Finally, the surgical team decided to do biventricular repair in the form of VSD closure without baffling, Damus Key Stansel (DKS)-Rastelli repair and direct stitch closure of the sub-aortic area (Figures 3-5). This unusual idea achieved the arterial level repair for TGA without coronary artery transfer plus the following benefits: Closure of VSD without baffling kept the RV cavity within optimum size, avoiding bad post-operative RV function; Closing the subaortic area prevented possible steal of blood to the low-pressure RV. The patient did well post-operatively after one week of open sternum. The new surgical technique described herein that can be called Ellassal-Radi technique

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Figure 3. Preoperative TTE showing DORV, large sub pulmonic VSD, TGA, severe subaortic stenosis, hypoplastic ascending aorta, interrupted aortic arch. A: Parasternal long axis view. B: Apical 4 chamber view for the same patient showing the left sided pulmonary artery and the right sided hypoplastic aorta.
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Figure 4. Post-operative TTE for the same patient in Figure 3.A, B: 2D and color mode for apical 4 chamber view showing DKS-Rastelli repair: The previous hypoplastic ascending aorta connected to the right side of the neo aorta as a single coronary artery with good flow from the neo aorta to the single coronary (yellow arrow).

Figure 5. 1: preoperative diagnosis. 2: postoperative repair of the same case: A: Small ascending aorta component of DKS. B: Main Pulmonary artery. C: RV-PA conduit (Contegra). D: VSD patch. E: Stitch closure of subaortic area. F: Aortic arch reconstruction by equine pericardial patch.

**Table 1. Demographic, diagnostic, and management data**

<table>
<thead>
<tr>
<th>No.</th>
<th>Age</th>
<th>Sex</th>
<th>Unusual diagnosis</th>
<th>Specific work up</th>
<th>Management</th>
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<tbody>
<tr>
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<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>1</td>
<td>30 d</td>
<td>F</td>
<td>CAVF from single coronary to PA</td>
<td>Aortic angiography</td>
<td>Ligation &amp; Division</td>
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<tr>
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<td>30 d</td>
<td>M</td>
<td>ASCAPA</td>
<td>MDCT</td>
<td>Translocation</td>
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<td>B- Unusual complication related to coronary artery surgery:</td>
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<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>1 y</td>
<td>F</td>
<td>Missed RCA transfer post ASO</td>
<td>MDCT</td>
<td>Reoperation</td>
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<tr>
<td>4</td>
<td>1 y</td>
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<td>Complicated intramural LCA post ASO</td>
<td>Aortic + PA angiography</td>
<td>Device occlusion</td>
</tr>
<tr>
<td>5</td>
<td>1 y</td>
<td>F</td>
<td>RCA stenosis post aortic root surgery</td>
<td>Aortic angiography</td>
<td>RCA stent</td>
</tr>
<tr>
<td>6</td>
<td>20 d</td>
<td>F</td>
<td>Difficult coronary artery transfer during ASO. Intraoperative assessment</td>
<td>DKS + Rastelli repair.</td>
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</table>

Results

Unusual series related to coronary artery recorded 0.71% among all patients indicated for pediatric cardiac surgery operations. Both the aortic angiography and MDCT were needed to confirm coronary artery anomaly or hidden coronary artery complication. ECMO was conducted in 3 cases. Delayed sternal closure was indicated for all cases. One case diagnosed ASCAPA died from cardiogenic shock due to pre-operative myocardial ischemia and intra-operative difficult coronary translocation. Other data are shown in Tables 1 and 2.

Table 2. Outcome

<table>
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<td>Mean duration of ICU stay (days):</td>
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<tr>
<td>Mean duration of hospital stay (days):</td>
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<tr>
<td>Number of patients required mechanical circulatory support (ECMO):</td>
<td>3</td>
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<tr>
<td>Number of patients required delayed sternal closure:</td>
<td>5</td>
</tr>
<tr>
<td>Mortality n:</td>
<td>1</td>
</tr>
</tbody>
</table>


Discussion

Unusual Coronary Artery Anomaly

- CAVFs: Only 0.1%–0.2% of all patients who undergo selective coronary angiography are diagnosed with a CAVF [2]. It accounts for 0.2–0.4% of all congenital cardiac defects. It is rarer in adults (incidence only 0.11%) than in children [3]. The association of coronary artery pulmonary fistula (CAPF) in children with pulmonary atresia plus VSD is variable. Collison et al. [4] reported 8% and Sathanandam et al. [5] reported 1.3%. We recorded one case of single coronary artery with CAPF that was managed by surgical ligation and division.
- ASCAPA: Anomalous origin of a single coronary artery from the pulmonary artery (ASCAPA) is rare and a potentially fatal malformation. It is rarer than anomalous left coronary artery from pulmonary artery. Echocardiography is not as helpful as expected in some patients and more tools like CT angiography or even cardiac catheterization are essential to diagnose such cases [1, 6]. In our patient with ASCAPA, it was difficult to be diagnosed by echocardiography, and MDCT was needed to confirm the diagnosis.

Unusual Complication Related to Coronary Artery Surgery

- Ostial RCA stenosis post aortic root surgery: Although coronary ostial stenosis after aortic root replacement has been reported for over 40 years, it remains a challenge and requires emergent management to save patients. PCI as a treatment for iatrogenic coronary stenosis after aortic root surgery has acute and long-term effectiveness [7]. We followed the same management for the 12-month-old female with ostial RCA stenosis after aortic root surgery.
- Missed RCA transfer after ASO: A single common coronary ostium was present in 16 patients (2.2%) in one cohort study [8], none of whom experienced early or late death. In our patient with redo surgery for arterial switch after PA band and arch repair, visual identification of the RCA was missed and the patient was considered as having a single coronary ostium. The stormy sequela raised the importance of identifying the coronary anatomy before arterial switch particularly in redo surgery. Such redo surgery resulted in adhesions and anatomical distortion, which made visual identification of coronary arteries intra-operatively a difficult mission. Cardiac cath or preferably CT angiography will help to avoid catastrophic post-operative complications.
- Complicated intramural LCA during ASO: Intramural coronary artery course is associated with a higher incidence of arterial injury or stenosis during ASO [9]. Our patient with intramural coronary artery went on post-
operative ischemia related to coronary stenosis. Surgical trials to relieve any compression on the coronary artery and to ensure its patency failed to improve myocardial ischemia. In the cath lab, it was surprising that ischemia was a result of fistula from aorta to RV (coronary steal phenomenon). Occlusion by vascular plugs avoided further surgical manipulations and rescued the baby.

- **Difficult coronary artery transfer during ASO:** DORV with a subpulmonic VSD is a very rare (<1%) form of congenital heart disease. It represents about 30% of all cases of DORV. It is not uncommonly associated with aortic arch anomalies, aorticstenosis, subaorticstenosis and coronary artery anomalies [10]. Kumar et al. [11] did not consider that an arterial switch was possible due to a remote non-facing coronary sinus. Instead, a DKS-Rastelli repair was performed to achieve anatomic correction without manipulation of the coronary arteries [12]. We did the same DKS-Rastelli repair for the same complex anomaly but we did not consider an arterial switch due to the hypoplastic ascending aorta (3 mm) that prohibited coronary button technique. Additionally, we closed subaortic RVOT and the VSD was closed without baffling across the RV that avoided reduction in cavity size. We consider that this later new modification makes our repair better than the Kumar technique, so we called this novel modification as Elassal-Radi technique [13].

### Conclusion

While early management of common anomalies and complications related to coronary artery may decrease risk of cardiac surgery, unexpected coronary artery events during cardiac surgery need immediate capture of diagnosis and rapid management to avoid life-threatening complications. Surgery that needs coronary artery manipulations in children is still at risk of terrible outcomes related to coronary artery. Aortic angiography confirmed the diagnosis. Percutaneous intervention plays a major and effective role to treat them. Success to address these unexpected scenarios is a real gauge to assess the efficacy of multidisciplinary coordination between pediatric cardiology and cardiac surgery services.

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### Ethical Compliance

The authors have stated all possible conflicts of interest within this work. The authors have stated all sources of funding for this work. If this work involved human participants, informed consent was received from each individual. If this work involved human participants, it was conducted in accordance with the 1964 Declaration of Helsinki. If this work involved experiments with humans or animals, it was conducted in accordance with the related institutions’ research ethics guidelines.

### References


