Anomalous left coronary artery arising from the pulmonary artery in hypoplastic left hearts: Case series and review of literature

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Anomalous left coronary artery arising from the pulmonary artery (ALCAPA) occurring in the setting of single ventricle disease is rare. Preoperative recognition of the lesion is critical for intraoperative cardiopulmonary bypass and surgical management. We present a 6-case series from Children's Hospital Boston with a review of literature.

CLINICAL SUMMARY

A retrospective review of 552 patients undergoing stage I palliation between 1995 and 2010 was performed. ALCAPA was identified in 6 patients.

Six cases of ALCAPA in hypoplastic left heart syndrome (HLHS) were found in this retrospective review (Table 1). In 5 patients, the diagnosis was made intraoperatively. In the first 4 patients, ALCAPA was identified late in the procedure (ie, after ductal ligation), and all 4 patients died of myocardial failure in the postoperative period. In the fifth patient, ALCAPA was identified during dissection of the right pulmonary artery (RPA). Care was taken to preserve coronary perfusion by snaring the branch pulmonary arteries and delaying ductal ligation. Cardioplegia was delivered to both coronary arteries via the ascending aorta and the controlled main pulmonary artery (MPA). This patient survived. In the sixth patient, diagnosis was made preoperatively by echocardiography and catheterization (Figures 1 and 2). Perfusion and cardioplegia were handled as described above, and this patient died of myocardial failure.

DISCUSSION

ALCAPA in the absence of significant congenital heart disease occurs in 0.25% to 0.5% of live births.¹ The incidence when associated with complex congenital heart disease is unknown, especially as it pertains to HLHS. In our series, a preoperative diagnosis was made in only 1 patient. With the recent findings of this anomaly in HLHS, our center has become vigilant in defining the coronary arteries

before surgery. Obviously, this can be difficult to define with certainty by standard imaging modalities, such as echocardiography, catheterization, magnetic resonance imaging, or computed tomography scan. Thus, the true prevalence could be higher even as indicated in this retrospective review. Nonetheless, the presence of ALCAPA in patients with HLHS undergoing staged repairs will certainly affect medical and surgical management and ultimate prognosis.

Operative stage I palliation is the treatment of choice, and in the majority of cases ductal ligation is performed as the initial maneuver after initiation of cardiopulmonary bypass before dissection of posterior aspect of the MPA and branch pulmonary arteries (PAs). This can result in 1) coronary ischemia from a vascular steal from the anomalous coronary after ductal ligation and 2) accidental injury to anomalous coronary during dissection.

There have been several small case series documenting outcomes of this abnormality.

Saroli et al² reported 4 patients with HLHS or Shone complex and coronary anomalies. Diagnosis was made intraoperatively in 2 of these patients, resulting in coronary injury, and they ultimately died. Diagnosis was made preoperatively in 2 of these patients (ALCAPA in 1, high origin of left coronary artery in 1), and they were successfully managed.

Nosal and colleagues³ describe a patient with HLHS in whom ALCAPA from the RPA was diagnosed intraoperatively because of left ventricular ischemia on initiation of cardiopulmonary bypass. Additional coronary perfusion was initiated with a second cannula in the MPA and snaring of distal branch PAs. Stage I palliation and reimplantation of ALCAPA were performed, and the patient was discharged on day 14 and subsequently underwent a successful bidirectional Glenn.

Bartram and colleagues,⁴ in their review of causes of death after the modified Norwood procedure, which involved 122 cases, reported only 1 case of accidental ligation of the left circumflex off RPA.

Daebritz and colleagues⁵ reviewed the outcomes of the stage I procedure at a single institution in the early phase of the procedure over a 9-year period. There were 131 cases of HLHS and 63 cases of NHLHS, with 26 deaths in the first 24 hours, of whom 15 had autopsy. Of interest, 10 were noted to have coronary ischemia. However, none of the 9 patients with HLHS had ALCAPA, whereas the single patient with NHLHS had an anomalous left circumflex from the PA.

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TABLE 1. Diagnosis, procedure, and outcomes

Patient	Diagnosis	Procedure	ALCAPA diagnosis	ALCAPA management	Outcomes
1	HLHS, AA/MA, PAPVR	Stage I, BTS ligation of anomalous coronary	LCx from RPA on transection of MPA	Ligation 1-mm diameter vessel with length insufficient for reimplantation	Mortality 1.5 h postoperatively
2	DORV, MA, sub-AS, CoA, preoperative NEC	Stage I, BTS ALCAPA reimplantation	LCx from RPA on transection of MPA	Reimplantation into neo-aortic root	Mortality 9 h postoperatively
3	HLHC, AS, MS, multiple VSD	Stage I, Sano, ALCAPA reimplantation, transitioned to ECMO	LCx from LPA LV ischemia on ductal ligation	Reimplantation into neo-aortic root	Mortality POD 13 despite ECMO support
4	DISRV, DORV, common atrium, CoA, arch hypoplasia, TAPVR	Stage I, BTS, TAPVR repair, ALCAPA reimplantation transitioned to ECMO	LCA off RPA LV ischemia on ductal ligation	Reimplantation into neo-aortic root	Mortality POD 21 despite ECMO support
5	HLHC, MA, AS, VSD	Stage I, BTS, ALCAPA reimplantation	LCA off RPA noted during RPA dissection before CPB	Reimplantation into neo-aortic root	Survival to discharge
6	HLHC, dysplastic MV with MS, AS, hypoplastic LV, VSD, ALCAPA	Stage I, BTS, MV repair, ALCAPA reimplantation	LCA off distal RPA close to RUL branch take off, diagnosed on preoperative echocardiogram and confirmed by catheterization	Reimplantation into native aortic root	Postoperative ECMO for myocardial failure LCA stenting, died of fungal sepsis on POD 43

AA, Aortic atresia; ALCAPA, anomalous left coronary arising from pulmonary artery; AS, aortic stenosis; BTS, Blalock–Taussig shunt; CPB, cardiopulmonary bypass; CoA, coarctation; DISRV, double-inlet single right ventricle; DORV, double-outlet right ventricle; ECMO, extracorporeal membrane oxygenation; HLHC, hypoplastic left heart complex; HLHS, hypoplastic left heart syndrome; LCA, left coronary artery; LCx, left circumflex; LPA, left pulmonary artery; LV, left ventricle; LVDCC, left ventricle-dependent coronary circulation; MA, mitral atresia; MPA, main pulmonary artery; MS, mitral stenosis; NEC, necrotizing enterocolitis; PAPVR, partial anomalous pulmonary venous return; POD, postoperative day; RPA, right pulmonary artery; RUL, right upper lobe; RVDCC, right ventricle-dependent coronary circulation; TAPVR, total anomalous pulmonary venous return; VSD, ventricular septal defect.

On the basis of our clinical experience, preoperative identification of the coronary artery anatomy is essential. This might necessitate a combination of imaging strategies, including echocardiography, catheterization, magnetic resonance imaging, and computed tomography

angiography. Careful surgical examination of coronary anatomy before initiation of cardiopulmonary bypass remains key.

With the diagnosis established, management of cardiopulmonary bypass can be modified to include the following:

- maintenance of ductal patency with snaring of distal branch PAs to avoid coronary steal and myocardial ischemia;

FIGURE 1. Anteroposterior view of ALCAPA arising from the RPA. RPA, Right pulmonary artery; ALCAPA, anomalous left coronary arising from pulmonary artery.

FIGURE 2. Echocardiographic images of left coronary artery arising from the RPA, 6mm distal to the pulmonary arterial bifurcation. RPA, Right pulmonary artery; ALCAPA, anomalous left coronary arising from pulmonary artery.

- cardioplegia administered simultaneously into the ascending aorta and isolated MPA/proximal branch PA; and
- reimplantation of ALCAPA with generous buttons to avoid distortion.

Alternative options would be a hybrid approach with PA banding distal to ALCAPA and reimplantation at time of comprehensive stage II, or neonatal transplantation.

CONCLUSIONS

The preoperative diagnosis of this rare entity is key because it allows for adequate surgical planning.

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Percutaneous mitral valve repair as a bail-out strategy for patients with severe mitral regurgitation after cardiac surgery

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With excellent long-term results, surgical valve repair is the standard treatment in patients with mitral valve (MV) disease, replacement being an option if reconstruction is not feasible.^{1,2} In patients with prohibitive operative risk, the MitraClip system (Abbott Laboratories, Abbott Park, Ill) has accomplished promising preliminary results reducing mitral regurgitation (MR).^{3,4} We performed this procedure as a bail-out approach in high-risk patients with severe MR not responding to intense medical therapy in the early postoperative period after cardiac surgery.

CLINICAL SUMMARY

From March to October 2009, 3 patients underwent coronary artery bypass grafting, tricuspid valve replacement, or

aortic valve replacement combined with tricuspid valve repair at the University Heart Center Hamburg (Table 1). Preoperatively, mild to moderate MR was noted. Postoperatively, severe functional MR impeded hemodynamic stabilization necessitating prolonged inotropic support. To avoid early redo surgery associated with disproportional operative risk, an interventional approach was chosen after interdisciplinary discussion.

Percutaneous MV repair was performed with the MitraClip device, a polyester-covered metal clip, as previously described.⁵ Briefly, after introduction through the femoral vein and transseptal puncture, the steerable device was advanced across the MV. The clip was then positioned, and the leaflets were captured at the origin of the regurgitant jet creating a double-orifice simulating Alfieri's technique. Clip implantation was performed under fluoroscopic and transesophageal echocardiographic guidance (Figure 1). Follow-up echocardiographic evaluation was performed at discharge and 3 months after the procedure.

RESULTS

All 3 female patients (age, 71–78 years) were considered high-risk surgical candidates as predicted by standard risk-stratification tools (Table 1). Percutaneous MV repair was performed 5 to 7 days after primary surgery. Two clips were used in 2 of the patients, creating a triple-orifice area. MR was reduced from grade III–IV to I–II allowing for hemodynamic recompensation. There were no signs of mitral stenosis after clip implantation.

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