First report of successfully palliating a hypoplastic left heart syndrome patient with anomalous left coronary artery from the pulmonary artery beyond Fontan

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ABSTRACT
We report a case of hypoplastic left heart syndrome with an anomalous left coronary artery from the pulmonary artery (ALCAPA) identified intraoperatively during the Stage-II palliation. Due to recurring ventricular fibrillation on sternotomy, a hybrid Stage-I palliation was performed. During comprehensive Stage-II, the ALCAPA was reimplanted in the neoaorta and measures, including a nontraditional Damus connection/arch reconstruction and classic bilateral Glenn procedures, were taken to avoid compression of the coronary artery. After a successful Fontan procedure, he continues to do well at 5 years old, becoming the first patient reported in the literature to survive all the three stages of single-ventricle palliation.

Keywords: Anomalous left coronary artery from the pulmonary artery, bidirectional Glenn, Fontan, hypoplastic left heart syndrome

INTRODUCTION
Hypoplastic left heart syndrome (HLHS) and anomalous left coronary artery from the pulmonary artery (ALCAPA) are each rare cardiovascular anomalies infrequently encountered in the same patient. A recent case series reported six patients with ALCAPA identified among 552 patients with HLHS with high mortality; a few other case reports have also documented this rare combination.[1-4] This patient is the first documented HLHS–ALCAPA to survive all the three stages of single-ventricle palliation.

CASE REPORT
A female infant was born at 39 weeks gestation, prenatally diagnosed with HLHS. Postnatal echocardiogram confirmed HLHS with mitral and aortic atresia and bilateral superior vena cavae (SVC). Initial palliation with a Norwood procedure was aborted due to intraoperative ventricular fibrillation on minimal pericardial manipulation followed by conversion back to sinus rhythm. Thus, a hybrid approach to Stage-I palliation was undertaken with bilateral pulmonary artery (PA) band placement and patent ductus arteriosus stenting, and the patient was discharged on the postoperative day 6.

Pre-Stage II catheterization revealed excellent hemodynamics with the new diagnosis of partial anomalous pulmonary venous return (PAPVR) of the right upper pulmonary vein to the right SVC. During comprehensive Stage II at 5 months old, once on cardiopulmonary bypass and arrested, the stented ductus arteriosus was clamped, and the main PA was incised. A large coronary artery was noticed to be emerging from the left PA ostium and coursing tautly to the myocardium where it immediately bifurcated so ostial cardioplegia...
was given. The coronary’s course prevented significant mobilization, and hence, it was attached to the proximal main PA using a coronary button. After the review of prior angiography which confirmed the diagnosis, it was felt that a proximal Damus–Kaye connection, and thus a slight turning in of the PA, may cause torsion of this taut coronary connection [Figures 1 and 2]. Therefore, the PA branches were excised from the aorta individually and closed medially. Under antegrade cerebral perfusion, the remainder of the stented duct was anastomosed to the proximal main PA with patching of the anterior stented duct, and its native connection to the proximal arch and the ascending aorta preventing torsion of the proximal neoaorta. The aortopulmonary window was significantly narrowed by the coronary artery, and hence, bilateral classic Glenn anastomoses were completed. The known PAPVR was addressed by transecting the right vena cava superior to the PAPVR and oversewing the SVC’s cephalad end to maintain continuity of the PAPVR to the common atrium. The postoperative course was uncomplicated, and the patient was discharged on the postoperative day 8 with oxygen saturations in the low 80s. At the age of 4 years, she underwent pre-Fontan evaluation with catheterization and a cardiac magnetic resonance imaging which demonstrated excellent hemodynamics, normal ventricular function without wall motion abnormalities, and symmetrical flow to the bilateral SVCs and PAs. The reimplanted left coronary artery was unobstructed. Uncomplicated fenestrated intracardiac tunnel Fontan procedure with placement of a Gore-Tex PA interposition graft was performed subsequently. Modification with an interposition graft from the left PA to the right PA allowed for a united Fontan circulation while opening the posterior pericardium allowed more space to prevented vascular compression and venous obstruction [Figure 3].

**DISCUSSION**

A case series of six infants from Boston Children’s Hospital with a combination of ALCAPA and HLHS reported high mortality with only one patient surviving to discharge following Stage-I palliation.\(^1\) Four of these patients’ ALCAPA was not identified until late in the Stage I operation, and they died postoperatively. In the surviving patient, cardioplegia was administered through both the ascending aorta and main PA after its discovery and it was reimplanted to the neoaorta. This same approach was performed for the sixth patient, who was diagnosed preoperatively; however, the patient died on the postoperative day 43 due to heart failure and fungal sepsis.\(^2\) Other case reports document the rare finding with similarly poor surgical outcomes.\(^1,3\)

The decision to perform a hybrid Stage-I palliation because of the patient’s suspicious clinical course led to addressing the ALCAPA at the second-stage palliation, which may have provided a valuable alternative to the Norwood operation in a neonate who unknowingly had a coronary anomaly. However, this technique should still
be considered if ALCAPA is diagnosed preoperatively. A similar approach was also successful in getting a patient through the second-stage palliation after PA banding at 3 days of life.\textsuperscript{[4]} Even avoidance of the Norwood procedure with the use of hybrid Stage-I palliation is not a guarantee of safety. We know of one case where the patient suffered cardiac arrest and mortality immediately following PA band placement in the setting of undiagnosed ALCAPA with unfortunate placement of the left PA band immediately proximal to the origin of the anomalous coronary (personal communication with another hospital). Due to the difficulty of diagnosing ALCAPA in the setting of HLHS, one should always remain cognizant of the remote possibility of ALCAPA intraoperatively. Similarly, angiographic interrogation at the time of hybrid Stage-I palliation ought to be carefully reviewed for the presence of anomalous vascular connections, including coronary origins, before PA band placement. Furthermore, the course of the ALCAPA, in this case, made the aortopulmonary window space limited and thus did not allow for the PAs to pass posterior to the aorta but bilateral SVC made PA continuity unnecessary. Perhaps, the poor outcomes in other reports have been secondary to PA continuity being maintained leading to compression of the coronary or because traditional Norwood reconstruction has caused limited space posterior to the aorta. In the current case, it was felt that creating PA continuity at Stage II would have caused coronary compression despite trying a nontraditional arch reconstruction to increase the aortopulmonary window space and importantly, maintaining proximal neoaortic position to minimize the movement of the reimplanted coronary. However, it is hard to make any recommendations with our limited experience. In summary, the combination of ALCAPA and HLHS is extremely rare and is associated with a high mortality. Outcomes may, in part, be driven by the presence or absence of preinterventional diagnoses. When the reimplanted left coronary button forms the inferior border of the aortopulmonary window, the authors feel nontraditional surgical techniques that maximize the space posterior to the aorta to avoid crowding of this space and maintain the proximal neoaortic position are important.

\textbf{Declaration of patient consent}

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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\textbf{Conflicts of interest}

There are no conflicts of interest.

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