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Case Report Open Access

Left Subclavian Artery Stenosis in Neonatal Coarctation: Diagnosis and Treatment

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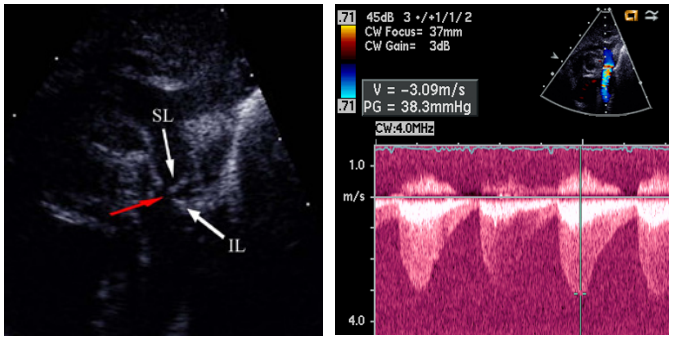
Coarctation of the aorta is the fourth most common congenital cardiovascular anomaly requiring intervention in the first year of life [1]. Although coarctation of the aorta may be a very complex lesion, it usually occurs as a discreet posterior shelf created by an infolding of the aortic media in the upper thoracic aorta at the insertion of the ductus arteriosus with most being located distal to the origin of the left subclavian artery [1]. We report two cases of neonatal coarctation of the aorta in which the left subclavian artery (LSA) was incorporated in the posterior coarctation shelf. To our knowledge this finding has not been previously reported in neonates.

Case Reports

Patient 1

A 6 day old male infant was noted to have discrepant pulses between the upper and lower extremities with four extremity blood pressures as follows: right arm 84/50, left arm 64/48, right leg 61/42, and left leg 64/41. An echocardiogram demonstrated a discreet juxtaductal coarctation with the coarctation ledge narrowing not only the descending aorta but also the origin of the somewhat inferiorly displaced LSA (Figure 1) as well as a very small patent ductus arteriosus (PDA) with bidirectional shunt. Additional findings noted on the echocardiogram included two small left to right atrial shunts and a hypoplastic (z score -3.19) bicuspid aortic valve without stenosis or insufficiency. He was placed on a prostaglandin infusion and subsequently taken to the operating room for coarctation repair.

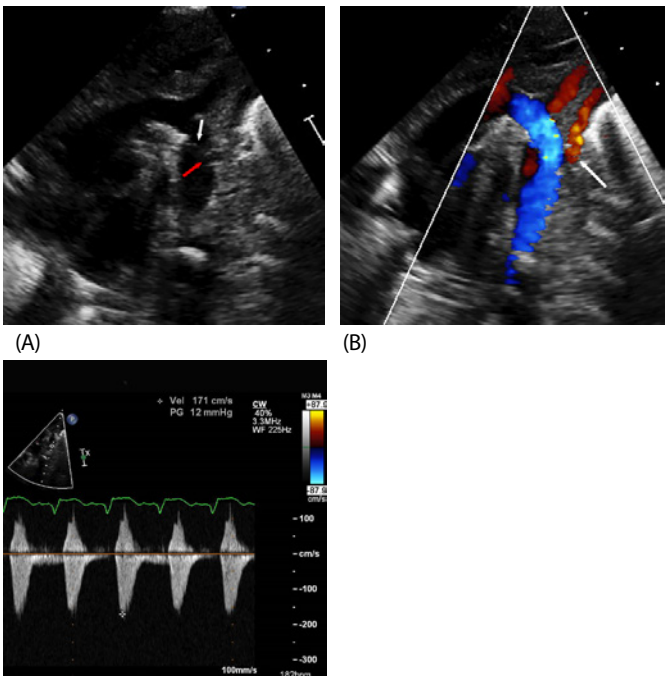
Surgery was performed through a left lateral thoracotomy and inspection of the aortic arch demonstrated narrowing of the transverse arch from the innominate artery to the origin of the PDA with minimal inferior displacement of the LSA. The LSA and vertebral arteries originated separately from the transverse arch and the PDA was located directly opposite the LSA such that there was no true aortic isthmus.



(A) (B)

Figure 1: A. 2D TTE demonstrating the coarctation ledge (white arrows) encompassing and narrowing the origin of the LSA (red arrow). B. Spectral Doppler interrogation demonstrating the double velocity envelope with antegrade flow in diastole consistent with coarctation of the aorta. The peak gradient across the coarctation is 38 mmHg. Key: 2D TTE – two dimensional transthoracic echocardiogram, IL – inferior aspect of the coarctation ledge, LSA – left subclavian artery, SL – superior aspect of the coarctation ledge.

Surgery consisted of left lateral thoracotomy with coarctectomy and extended end-to-end anastomosis. The coarctation ledge on the posterior descending thoracic aorta just distal to the LSA was excised. However, the arterial wall adjacent to the prominent ledge between the origins of the LSA and the left vertebral artery was so thin that the ledge could not be incised or excised. A long incision was made in the descending thoracic aorta to allow for these to be re-implanted



(A) (B)

(C)

Figure 2: A. 2D TTE performed after coarctation repair. The origin of the LSA is widely patent (red arrow). The arch repair is widely patent and the suture line is clearly visible (white arrow). B. 2D TTE with color Doppler interrogation demonstrating laminar flow in the left subclavian artery (white arrow). B, C. Color Doppler snows laminar flow in the descending aorta and spectral Doppler velocities are normal indicating no residual coarctation. Key: 2D TTE – two dimensional transthoracic echocardiogram, LSA – left subclavian artery.

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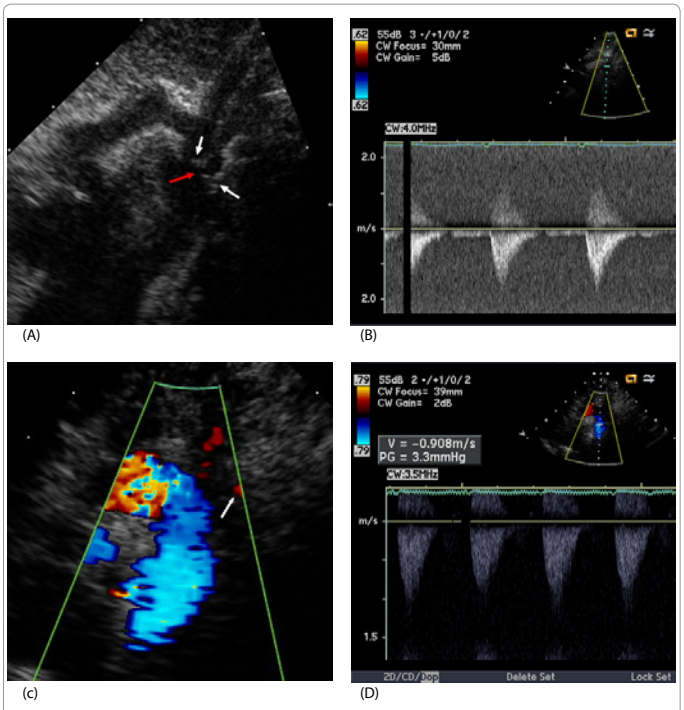

Figure 3: A. 2 D TTE demonstrating the coarctation with involvement of the left subclavian artery. Note that as the posterior coarctation ledge (white arrows) extends into the aortic lumen that it includes and narrows the origin of the LSA (red arrow). B. Spectral Doppler interrogation demonstrates antegrade flow in diastole but no gradient with the PDA open. C. Post operative 2D TTE with color Doppler interrogation shows flow in the region of the LSA (white arrow). C, D. Color Doppler snows laminar flow in the descending aorta and spectral Doppler velocities are normal indicating no residual coarctation. Key: 2D TTE – two dimensional transthoracic echocardiogram, LSA – left subclavian artery. PDA – patent ductus arteriosus.

almost to the side of the aorta in such a manner that would not cause any obstruction. The postoperative echocardiogram demonstrated no residual coarctation with a widely patent LSA with laminar flow (Figure 2 A, B, C). Post operative four extremity blood pressures were as follows: right arm 87/42, left arm 72/35, right leg 71/30, left leg 75/29. On follow-up physical examination brachial and femoral pulses were 2+ without significant delay.

Patient 2

A newborn female had an echocardiogram performed shortly after birth due to a family history of maternal coarctation and a fetal echocardiogram suggesting coarctation of the aorta. Physical examination demonstrated a I/VI systolic ejection murmur at the lower left sternal border, 2+ brachial pulses, 2 + femoral pulses, and four extremity blood pressures as follows: right arm 55/30, left arm 54/37, right leg 56/24, left leg 45/26. A transthoracic echocardiogram confirmed a normal ascending aorta, mildly hypoplastic transverse arch, and a discreet juxtaductal coarctation that also narrowed the origin of the inferiorly displaced LSA (Figure 3). Additional findings noted on the echocardiogram included a large PDA with right to left shunt, a hypoplastic (z score -3.24) bicuspid aortic valve with no stenosis or insufficiency, and mild hypoplasia (z score – 2.10) of the aortic sinotubular junction.

The patient was started on a prostaglandin infusion and surgery was performed at 6 days of age through a median sternotomy due to the hypoplastic transverse arch. Cardiopulmonary bypass was instituted with arterial cannula in the base of the innominate artery and the venous cannula in the right atrium, this cannulation strategy allows regional cerebral perfusion during the arch reconstruction. During the repair a significant amount of thickened tissue (coarctation ledge) was noted around the origin of the LSA. Since this could not be resected without detaching the LSA, it was left intact. The repair was completed by anastamosing the distal transverse arch to the descending thoracic aorta and augmenting the underside of the transverse arch with a pulmonary allograft patch.

Flow to the left arm was preserved even though the coarctation tissue around the origin of the LSA could not be completely resected. Post operative four extremity blood pressures were as follows: right arm 62/29, left arm 57/36, right leg 75/20, left leg 60/19. Flow was noted in the region of the LSA on the post operative echocardiogram and there was a palpable, but decreased, left brachial pulse. At hospital discharge the patient was noted to have 2+ pulses in all four extremities.

Discussion

The embryology of coarctation of the aorta is not completely understood but is associated with abnormal development of the embryonic fourth and sixth aortic arches as well as the dorsal aortae. During normal development the ascending aorta, innominate artery, and proximal aortic arch arise from the aortic sac while the distal transverse arch is formed from the fourth aortic arch. As arch development proceeds, the left seventh segmental artery will migrate cephalad and become the left subclavian artery with the aortic isthmus and descending thoracic aorta arising from the left dorsal aortae. The proximal portion of the left sixth arch forms the proximal portion of the left pulmonary artery and the distal portion forms the ductus arteriosus which passes from the left pulmonary artery to the dorsal aorta. The portion of the arch from the left subclavian artery to the insertion of the ductus arteriosus is the aortic isthmus and the aorta distal to the ductal insertion becomes the descending thoracic aorta.

The development of coarctation of the aorta is proposed to develop via one of the following mechanisms: 1. Extension of ductal tissue into the wall of the aorta; as the ductus closes the ductal tissue in the aorta contracts pulling the posterior wall of the aorta into the aortic lumen resulting in coarctation. 2. Abnormal involution of a small segment of the dorsal aortae distal to the left seventh segmental artery that moves cephalad with the left subclavian artery to the region of the ductus arteriosus. 3. During fetal life the aortic isthmus (the segment of the aortic arch between the left subclavian artery and the ductus arteriosus) is normally narrow because it carries little blood. Following closure of the ductus arteriosus this region (isthmus) normally enlarges until it is the same diameter as the aorta. If the narrowing persists, a coarctation forms [2].

Inclusion of the region of the dorsal aortae adjacent to the origin of the left seventh segmental artery (the embryologic precursor of the left subclavian artery) in whichever pathoembryologic process causes coarctation would account for the anomaly noted in these two patients.

Although coarctation of the aorta is a fairly common congenital cardiovascular malformation (1) occurring in 6-8% of patients with congenital heart disease, coarctation with the posterior shelf encompassing the origin of the left subclavian artery is not. Only sporadic reports appear in the literature all of which are in adult patients. Nishizawa [3] reported a case of atypical coarctation in an 18 year old female with aplasia of the left subclavian artery which may represent the extreme form of this anomaly. Sousa Uya [4] and Vuong

[5] each report a case of atypical coarctation with stenosis of the LSA in two adults with fibromuscular dysplasia, Bastarrika [6] describes a 46 year old with juxtaductal coarctation and stenosis of the LSA, and in a review of 13 adult patients with coarctation and coexisting arterial anomalies, Rokkas [7] describes only one patient with this anomaly. In a review of 96 pediatric patients ranging in age from 3.7 months to 12 years of age, Liu [8] describes one patient with the coarctation occurring at the opening of the LSA but does not report that the origin of the LSA was encompassed by the posterior coarctation shelf.

Diagnosis and proper management of this anomaly are important to prevent compromising flow to the left arm that may result in the sequelae noted in patients who have undergone coarctation repair utilizing the subclavian flap technique: decreased left systolic brachial pressure, decreased core temperature of the left hand, and significant differences in the length and circumference of the left upper arm compared to the right arm [9].

Although diagnosis of this rare anomaly can be readily made by two dimensional (2D) transthoracic echocardiography, imaging of the aortic arch in a sick neonate can be challenging and care must be taken to insure that both the posterior coarctation ledge and the origin of the LSA are clearly seen on 2D imaging. The finding of a posterior coarctation ledge extending into the aortic lumen that narrows the descending thoracic aorta as well as the origin of the LSA is diagnostic, Figure 1.

Surgical repair in adult patients has consisted of repair of the coarctation and reimplantation of the left subclavian artery [7] but this approach is very challenging in the neonate. We feel that it is important to preserve circulation to the left arm and, unless the arch repair will be compromised, advocate not sacrificing the LSA. We were able to achieve this in one patient by resecting the portion of the coarctation ledge that narrowed the origin of the LSA and reconstructing the arch in such a manner that the portion of the ledge that could not be resected did not cause any narrowing of the aortic lumen. In the other patient, the portion of the coarctation ledge at the origin of the LSA could not be resected and was left intact thus preserving flow to the left arm, although the pulse in the left arm was decreased. Clinically, pulses and blood pressures in both upper extremities, as well as a lower extremity, need to be followed to insure that the origin of the left subclavian artery remains patent. Post operative echocardiographic imaging, in addition to the standard evaluation of the coarctation repair, should include two dimensional, color, and spectral Doppler interrogation of the origin and flow into the left subclavian artery.

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