Fetal Counseling Provider Information
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Coarctation of the Aorta

Incidence

- 4 per 10,000 live births, 4-6% of all CHD. [1]
- More common in males[2]
- Associated with other cardiac lesions:
    - VSD and ASD
    - Bicuspid aortic valve (in up to 50-66%)[3]
- Occurs in 10-35% of those with Turner's syndrome (XO)[2,4]
- More likely to occur in those with a family history of left-sided obstructive lesions[5]

Fetal Imaging Predictors of Postnatal Interventions/Outcomes

Coarctation is often suspected on screening ultrasounds when there is right ventricle (RV) to left ventricle (LV) size discrepancy with a relatively larger RV. However, there is only moderate sensitivity (62%) and a high rate of false-positives in diagnosing coarctation based solely on the presence of ventricular size discrepancy.[6]

Ideally, multiple measurements of the aortic arch and left-sided structures are made with comparisons to right-sided structures and normative values. Serial follow-up throughout pregnancy can also be helpful to follow changes over time.

Predictors of Coarctation Postnatally:

Study	Predictors
Familiari et al[7] Systematic Review and Meta-analysis 12 studies included (922 fetuses)	• Hypoplastic arch (sens 90%, spec 87%) • Posterior "shelf" (sens 48%, spec 98%) • Multiple factors significant (no cut-off values): Lower MV z-score, higher TV z-score Smaller AoV, smaller isthmus Larger PA diameter, higher PA/Aorta ratio Higher RV/LV ratio Lower isthmus/duct ratio
Beattie et al[8] n=62 (45 w/ CoA)	• Ismthus/duct ratio <0.7 (false + 38%) • Isthmus z-score <-2 (false + 18%)

	• Aorta/PA ratio <0.65 (sens 76%, spec 70%) • Diastolic flow persistence improved sensitivity, but increased false positives
Gomez-Montes[9] n= 115 (52 w/ CoA)	Early dx ≤ 28 wks • Isthmus (3vt) z score ≤ -1.1 (sens 97%, spec 75%) • Isthmus (sag) z score ≤ -1.2 (sens 73%, spec 80%) • AAo z score ≤ -1.1 (sens 86%, spec 77%) Late dx >28 wks • TV/MV ratio ≥ 1.48 (sens 67%, spec 71%) • PA/Ao ratio ≥ 1.85 (sens 88%, spec 76%)
Matsui et al[10] n=44 (20 w/ surgery for CoA)	• Isthmus/duct ratio <0.74 • Isthmus z-score <-2 • Posterior "shelf" (specificity 90%) • Flow disturbance (specificity 94%)
Quartermain et al[11] n=35 (20 w/ CoA)	• Transverse arch ≤ 3 mm at >30 wks (sens 94%, spec 93%) • AoV/PV ratio <0.6 (sens 79%, spec 80%) • MV/TV ratio <0.6 (sens 70%, spec 87%) • LV mid-cavitary width / RV mid-cavitary width <0.6 (sens 70%, spec 67%) • PFO bidirect or L>R (sens 70%, spec 100%)
Toole et al[12] n=62 (27 w/ CoA)	• Isthmus/duct ratio <0.5 (sens 33%, spec 100%) • MV z-score <-1.63 (sens 71%, spec 74%) • MV/TV < 0.75 (sens 56%, spec 77%)

Studies have also looked at other potentially useful measurements:

Left common carotid-to-left subclavian artery distance (LCSA)
LCSA>4.5mm (sens 80%, spec 95%)[13]

Carotid to subclavian index (CSI), ratio of the aortic arch diameter at the left subclavian to the distance between the left carotid and left subclavian
CSI<0.77[14]

The angle between the ascending aorta and descending aorta (AAo-DAo)
AAo-DAo ≤ 20.31° (sens 95%, spec 100%)[13]

The angle between the transverse aorta and the descending aorta (TAo-DAo)
TAo-DAo ≥ 96.15° (sens 90%, spec 100%)[13]

Isthmus to ductal angle (IDA)
IDA <117 (sens 24%, spec 96%)[12]

Therapies

- Critical coarcation: ductal dependent for systemic blood flow will require PGE after birth.
- Many neonates with questionable arch obstruction on fetal echocardiogram will need to be monitored closely while the PDA closes. This monitoring typically includes serial echocardiograms (as available) along with serial 4 extremity BP measurements and pulse examination of the lower extremities in the early transition period.
- Neonatal period: Potential surgical interventions depending on extent of arch narrowing and surgical center
    - Coarctation repar via posterolateral thoracotomy if the obstruction is discrete or involving only the isthmus and distal transverse arch
    - Arch advancement via median stenotomy with cardiopulmonary bypass if there is diffuse tubular hypoplasia of the proximal and distal transverse arch.
- Recurrent obstruction past the neonatal period may benefit from cath based balloon angioplasty and/or stent placement.
- Childhood and adolescence: surgical or catheter intervention

Prognosis

Survival:

- Good long-term survival in the modern era: 98% at 50 yrs and 89% at 60 yrs.[3]

Neurodevelopment:

- Patients requiring congenital surgery in the first 3 months of life are at risk for neurodevelopmental abnormalities, including those with complex coarcation. [15]

QOL:

- Hypertension later in life is common (up to 42%)[3] but incidence may be lower among patients who undergo earlier/neonatal repair.

- Need for re-intervention later in life:[3]
    - Recurrent coarctation ~ 12%
    - Aortic aneurysm ~ 5-10% (aortopathy associated with coarcation as well as in those with bicuspid aortic valve)

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