Aortenisthmustenose

Title Coarctation of the aorta
In infancy and childhood

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2. Definition

- narrowing at the distal arch to descending aorta
- 3-5% of congenital heart defects
- boys = 2 : 1 = girls

- sometimes combined with arch hypoplasia
- other locations – abdominal coarctation
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Various forms of arch hypoplasia
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abdominal coarctation
Common associated defects:

- bicuspid aortic valve (45-85%)
- valvular AS
- PDA
- VSD
- Shone-Komplex (supravalv. MS, parachute valve subvalvular AS)

Rare associated defects:

- HLHS
- AVSD
- D-TGA
- Taussig-Bing-heart

Vascular anomalies:

- lusoric origin of right subclavian artery
- stenosis of left subclavian artery
- aneurysms of the aorta
- cerebrovascular aneurysms (10%)
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Associates syndroms:
- Turner
- Williams-Beuren
- CHARGE
- VACTERL
- Kabuki-Syndrom
- PHACE-Syndrom
3. Clinic and presentation

- critical Coa of the newborn
- non critical coa

- critical Coa of the newborn:
  - PDA-dependent systemic perfusion
  - diminished femoral pulses
  - differential cyanosis
  - heart failure
  - shock - MOF

- Non critical coa:
  - prestenotic hypertension
    - pressure load of LV
    - arteriosclerosis
    - cerebral aneurysms
  - poststenotic malperfusion
3. Auscultation and presentation:

- critical Coa of the newborn
  - murmur may be absent
  - weak pulses
  - shock

- Non critical coa:
  - weak pulses
  - systolic murmur (back, abdomen)
  - continuous flow murmur (collaterals)
  - head aches, calf pain, nose bleed,
  - cerebral insult
4. Diagnostics

- aim:
  - anatomy
  - severity of stenosis
  - severity of hypertension, collateral flow,
  - additional cardiac defects
  - secondary effects
  - planning of therapy
  - assessment of prognosis

- Clinical examination
  - blood pressure measurement
  - differential cyanosis
Diagnostics:

- blood pressure measurement (arms/legs, 24 hrs, exercise)
- pulse oxymetry
- spiroergometric assessment
- ECG
- CXR
- ECHO
- rarely catheter investigation
- MRI
- CT-scan
- ophthalmologist
Relevant diagnostics:

- Coa newborn
- ECHO
  - anatomy
  - severity of stenosis
  - gradient

- Otherwise
- ECHO
  - rarely MRI (CT)

Note: if diagnosis is clear by noninvasive measures, no additional invasive measures are recommended.
Echocardiography:

- Most important tool
  - LV anatomy
  - LV function (FS, wall thickness)
  - additional defects (left sided obstructions)
  - aortic valve (bicuspid, stenosis)
  - aortic arch anatomy (diameters)
  - anatomy of coarctation (diameter, location)
  - doppler flow (gradient, diastolic run-off, flow in abdominal aorta)
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Other diagnostics:

- 24-hr RR measurement - long term effect/hypertension
- ECG - not important
- CXR - not important for primary diagnosis
- catheter - not important for primary diagnosis only for therapy

- MRI - if ECHO is not diagnostic good anatomy, 3-D

CT - if contraindications for MRI radiation
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5. Treatment:

- aim: establishment of a normal size aorta without gradient

- measures not possible by medical therapy
  - prostaglandin, shock, severe hypertension

- only possible by - surgery
  - intervention
Indications:

- critical coa: treatment always necessary

- non critical coa:
  - if systolic gradient is $\geq 20$ mmHg (I C)
  - if systolic gradient is $< 20$ mmHg (IIa)

  plus:

  - hypertension
  - significant stenosis
    (diameter Coa/desc. Ao $< 0.8$)
Surgery (1):

- simple coa:
  - lateral thoracotomy
  - extended end-to-end resection
  - if necessary arch augmentation

- coa with arch hypoplasia:
  - median sternotomy
  - with bypass, hypothermia, arrest, selective head perfusion
  - patch augmentation
  - simultaneous correction of other defects
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Surgery (2):

Older patients
- difficult mobilisation
- sometimes bypass surgery, prosthesis
- redo surgery
- same techniques

- risks
- simple coa mortality < 3%
- complex cases up to 10%
- nerve injury
- chylothorax
- paraplegia
- postcoarctectomy syndrome
- re-stenosis
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Surgery (2):
- Older patients - difficult mobilisation
- Bypass surgery, prosthesis
- Redo surgery - same techniques
- Risks - simple coarctation mortality < 3%
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Interventions (1):

- newborns – 6 months:
  - no standardised treatment
  - high rate of restenosis
  - in selected cases as palliative measure (NEC, sepsis, shock, MOF, etc.)

- > 6 months:
  - alternative to surgery
  - Balloon angioplasty
    - good for membranous coa
    - less ideal for hypoplasia
    - sequential dilatation
    - 2-5-fold balloon size
    - balloon not bigger than distal arch
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Interventions (2):

- children:
  - additionally stent implantation
  - covered stents
  - good for membraneous coa
  - good for arch hypoplasia
  - redilatation ppossible

- re-coarctation
  - interventional treatment of choice
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Interventions (3):

- complications:
  - arterial access
    - thrombosis, bleeding
    - stenosis, malperfusion
  - coarctation site
    - rupture, aneurysm, dissection
  - balloon
    - rupture
  - stent
    - dislocation, fracture,
    - neointima
    - absolute size
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- without treatment: - reduced life expectancy

- follow-up: - life-long

- annually: - clinical examination, ECHO, pressures,

- exercise testing: - every 3-4 years (if feasible)

- MRI/CT scan: - if ECHO is unclear, every 5 years
Follow-up (2):

- long term risks:
  - aortic aneurysms, rupture, dissection
  - SBE, coronary heart disease, stroke
  - retinopathy
  - restenosis
  - hypertension – medical treatment

- If bicuspid AV:
  - stenosis, regurgitation
  - dilatation of ascending aorta

- sport
  - regular exercise recommended
  - individual assessment with ergometry
  - contraindications with aneurysms
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Prevention

-none