Clinical Guideline

Coarctation of the Aorta

SETTING	South West England and South Wales
GUIDELINE FOR	Cardiology teams in South West England and South Wales hospitals
PATIENT GROUP	Adult patients with congenital heart disease

GUIDANCE

Follow-up: every two years
annually if pseudo-aneurysm or resistant hypertension

Associated lesions: bicuspid aortic valve (up to 85%), Shone's complex (whole spectrum, including arch hypoplasia), VSD, complex congenital heart defects, berry (intracranial) aneurysms of the circle of Willis (up to 10%), anomalous origin of right subclavian artery (5%). May be seen in up to 35% Turner's Syndrome. Rarely in Williams syndrome.

Inheritance: paternal inheritance risk 3%, maternal inheritance risk of any left sided congenital heart disease up to 10%.

Long-term complications:

1. Hypertension. 50% continue to be hypertensive post coarctation stenting/surgery.
2. Ascending aortopathy especially if BAV, pseudoaneurysms at repair site (especially Dacron patch repairs/balloon angioplasty)
3. Aortic dissection
4. Circle of Willis aneurysm rupture
5. Complications of hypertension
6. Recurrent coarctation

At each visit:

History: usually asymptomatic
if unrepaired may have exertional headaches, leg fatigue, or claudication
haemoptysis may be from a leaking/ruptured aneurysm - life-threatening, requires immediate investigation and treatment.

Exam: blood pressure in both arms or **right** arm, can do leg blood pressures
check leg pulses, assess radio-femoral delay
inter-scapular/left infra-clavicular murmurs due to collaterals
scars (lateral thoracotomy+/- median sternotomy if other lesions)
murmurs secondary to associated lesions

NB. If large collaterals, femoral pulses may be less diminished, and catheter and Doppler gradients may underestimate obstruction.

ECG:	LVH and strain
Echo:	suprasternal: arch dimensions, flow in the proximal descending aorta especially 'diastolic tail' aortic annulus, root and ascending aortic dimensions anatomy of aortic valve left ventricular function, LVH associated lesions

Further investigations:

CXR:	at diagnosis. May see rib notching from collaterals (ribs 3-8),"3 sign" adjacent to area under transverse arch/above MPA silhouette, dilated ascending aorta. Post-repair: evidence of thoracotomy, assess stents (PA and lateral films).
CPET:	not routine. Hypertensive response if SBP >200mmHg, significance unknown.
Holter:	not routine
ABPM:	consider at each visit. Do at least 2 yearly. HT diagnosis and treatment should be guided by this.
TOE:	rarely useful
Catheter:	to assess gradient (resting +/- provocation). >20mmHg peak to peak is significant.
EP study:	not routine
MRI:	5 yearly for re-coarctation, pseudo-aneurysm, collaterals yearly if pseudo-aneurysm 10 yearly brain MRI to look for Circle of Willis anatomy and aneurysms
CT:	to plan complex coarctation procedures 3 months, 1 year and 5 yearly after coarctation stenting as needed for assessment of stent narrowing or fracture (MRI drop-out)
Bloods:	lipid profile
Drugs:	treat hypertension aggressively with usual agents aspirin for 6 months post coarctation stenting
Pregnancy:	if unrepaired, risk of fetal loss, prematurity and IUGR. Avoid if pseudoaneurysm. Risk of pre-eclampsia if pre-existing hypertension.
Contraception:	not for combined pill if hypertensive
Endocarditis:	No antibiotic prophylaxis unless previous endocarditis, for associated high risk lesion or for 6 months post-surgical intervention.

Exercise: Avoid severe isometric exercise. Aerobic exercise recommended for blood pressure control.

Reduction of cardiovascular risk is crucial, even in those without hypertension. Encourage aerobic exercise, avoiding smoking and achieving BMI<25kg/m2.

Discuss if:

- Catheter gradient of ≥ 20mmHg (hypertensive or normotensive)
- Hypertensive and anatomical narrowing of ≥ 50% relative to the aortic diameter at diaphragm, even if catheter gradient < 20 mmHg
- Enlarging pseudo(aneurysm) at previous coarctation repair site
- Intracranial aneurysm on MRI

RELATED DOCUMENTS

Regional Referral Guidance for Adult Patients with Congenital Heart Disease
Regional Referral Pathway for Cardiac Disease in Pregnancy

REFERENCES

- Baumgartner H et al. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2020 00, 1-83.
- Stout et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease. Journal of the American College of Cardiology Aug 2018, 25255; DOI: 10.1016/j.jacc.2018.08.1029
- Canadian Adult Congenital Heart Network (www.cachnet.org)

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SAFETY

None

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