Medical Management of Hypertension in Coarctation of Aorta

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Coarctation of aorta

- The most common causes for referral were hypertension or a heart murmur.
- The correct diagnosis of coarctation was made by the referring physician in only 4% of cases.
Neonates with critical coarctation

- Continuous IV infusion of PGE1
- IV inotropic support (eg, milrinone, dopamine, or dobutamine)
- Supportive care to correct metabolic acidosis, hypoglycemia, respiratory failure, and anemia
Intervention for CoA should be performed in infancy or early childhood to prevent the development of chronic systemic hypertension.
Causes of hypertension

- Abnormalities in peripheral vascular physiology also occur in patients with coarctation.
- Systolic arterial hypertension is a manifestation of the coarctation stenosis, but it also reflects changes in vascular reactivity, arterial wall compliance, and baroreceptor function.
Studies of patients after coarctation repair have demonstrated abnormal arterial vascular function, as well as resetting of the baroreceptor reflex in some patients with persistent hypertension.

Such abnormalities in arterial physiology, which may be present after successful anatomic relief of coarctation, help to explain the occurrence of systolic hypertension in some patients many years after coarctation repair.
The etiology of late postoperative hypertension in patients without a residual/recurrent coarctation may relate to anatomic and functional changes in the arterial vasculature.

Animal studies document abnormal intimal thickening and medial hypertrophy in the proximal aortic arch late after successful relief of experimental coarctation.
Such morphologic changes would be expected to decrease arterial compliance and provide an anatomic basis for the functional abnormalities in vascular reactivity and baroreceptor function that have been reported following coarctation repair.
Even in the absence of a residual coarctation gradient, patients may exhibit late systolic and diastolic hypertension; this is most common in patients whose coarctation repair is delayed beyond late childhood.

The risk for late hypertension may be as high as 10–20% however, even if a coarctation is repaired in infancy.
Hypertension with exercise

- Systolic hypertension after coarctation repair also may occur during dynamic exercise, even in patients without resting hypertension or a resting coarctation gradient.
- Although alterations of vascular physiology may play a role, exercise–induced upper–extremity hypertension often is associated with an increase in the coarctation pressure gradient during exercise.
Hypertension with exercise

- The increase in blood flow across a relatively non-distensible aortic repair site that occurs with dynamic leg exercise may be primarily responsible for exercise-induced elevations in coarctation gradient and upper-extremity systolic pressure following coarctation repair.

- Patients with exercise hypertension, but without a significant residual coarctation gradient at rest, may benefit from beta-blocker therapy.
Mild Coarctation of Aorta is an Independent Risk Factor for Exercise-Induced Hypertension

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Abstract—Exercise-induced hypertension is a predictor of cardiovascular events in patients with coarctation of aorta (COA). However, it is unclear whether mild COA diagnosis is an independent risk factor of exercise-induced hypertension. We hypothesized that for every unit increase in exercise, patients with COA (without hemodynamically significant coarctation) will have a higher rise in systolic blood pressure (SBP) compared with matched controls. One hundred forty-nine patients with COA (aortic coarctation peak velocity <2 m/s) who underwent exercise testing were matched 1:1 to controls using propensity score method based on age, sex, body mass index, hypertension diagnosis, and SBP at rest. We compared exercise-induced change in SBP between patients with COA and controls and also assessed the correlation between Doppler-derived aortic vascular function indices (effective arterial elastance index and total arterial compliance index) and exercise-induced changes in SBP. Compared with controls, patients with COA had a greater change in SBP per unit metabolic equivalent ($\beta$=2.86; 95% CI, 1.96–4.77 versus 1.07, 95% CI, –0.15 to 1.75; $P=0.018$) and per unit oxygen pulse ($\beta$=4.57; 95% CI, 2.97–7.12 versus 1.45, 95% CI, –0.79 to 2.09, $P<0.001$). There was a correlation between $SBP_{\text{peak}}-SBP_{\text{rest}}$ and elastance index ($r=0.38$, $P=0.032$) and between $SBP_{\text{peak}}-SBP_{\text{rest}}$ and total arterial compliance index ($r=-0.51$, $P=0.001$), suggesting an association between vascular dysfunction and exercise-induced BP changes. Patients with COA, without significant obstruction, had higher exercise-induced changes in SBP after adjustment for other risk factors for hypertension. Considering the already known prognostic importance of exercise-induced hypertension, the current study highlights the potential role of exercise testing for risk stratification of patients with mild COA. (Hypertension. 2019;74:1484-1489. DOI: 10.1161/HYPERTENSIONAHA.119.13726.)
Treatment (general advices)

- Standard lifestyle modification for hypertension in children and adults with both unrepaired and repaired disease is recommended including weight control, regular aerobic exercise, low-fat and low-sodium diet, smoking cessation, and avoidance of alcohol.

- High-intensity static (power lifting), heavy weight lifting, sudden stop-start, or isometric exercises and sports should be avoided to reduce the risk of strain on the aorta that can lead to aneurysm formation or dissection.
Competitive sports

- Careful, case–by–case evaluation involving ETT and echocardiogram is required for appropriate risk stratification.
- Participation in competitive sports without limitations may be considered for patients with mild CoA, in the absence of significant pressure gradient and significant collateral vessels, normal ETT without hypertension, and small pressure gradient at rest (≤20-mmHg differential between upper and lower limbs).
Hypertension is controlled using standard therapies, and first-line treatments include beta blockers and the vasodilators, ACE inhibitors, and ARB. The specific choice of agent is patient-specific and must consider the patient’s ascending aortic size (beta blocker preferable) and presence of aortic insufficiency (beta blockers not recommended).
Follow up

- Monitoring via 24–h ambulatory blood pressure measurement is helpful for patients who have borderline measurements.
- ETT to follow the rise in blood pressure with physical activity, are also used when considering initiating or uptitrating medical therapy.
Chronic hypertension remains present in 35–68% of the patients with CoA, even in the presence of an anatomically satisfactory repair.

Furthermore, exercise–induced hypertension occurs in over one-third of the normotensive patients.

Prevalence of systemic hypertension is significantly lower in patients treated in the neonatal period or infancy and in patients who underwent CoA resection with end-to-end anastomosis.
Conclusions: The majority of patients were hypertensive at long-term follow-up after coarctation repair. This is caused by restenosis, defined by a gradient of greater than 20 mm Hg, in only a few patients. Even in those without prosthetic material or minimal-grade restenosis, there is a substantial incidence of arterial hypertension.
In one retrospective study of 126 pregnancies in 54 women with repaired CoA, there were 98 successful pregnancies, 22 miscarriages, and 6 abortions. There were no serious cardiovascular complications during pregnancy and delivery. Hypertension alone was reported in 17% of pregnancies and preeclampsia in 4%. Five of eight patients who had serial echocardiographic assessments had a $\geq 15$ mmHg increase in the CoA gradient. Though serious pregnancy–related complications are rare in women with repaired CoA, aortic dissection has been described in case reports.
Women who present with an unrepaired CoA during pregnancy should receive careful surveillance of the adequacy of blood pressure control with regular follow-up.

A reasonable blood pressure goal during pregnancy is 120 to 140/80 to 90 mmHg, although a lower target may be warranted if there is ascending or other aortic aneurysmal disease.

Care should be taken to avoid placental hypoperfusion in those with residual coarctation.
In the absence of an obstetrical indication for early delivery, women who are normotensive can carry the pregnancy to term and pursue CoA intervention after the physiologic changes of pregnancy have resolved (typically approximately three months after delivery).

For women with poorly controlled hypertension during pregnancy, the decision to perform surgical or transcatheter intervention during pregnancy is made on a case–by–case basis by a multispecialty team.

Every effort should be made to manage the patient medically and then intervene after the pregnancy is complete.

Percutaneous intervention for recoarctation (using a covered stent) is possible during pregnancy but should only be performed for severe refractory hypertension or maternal/fetal compromise.
Pregnancy

- For patients with CoA (unrepaired or repaired), particularly those with hypertension, spontaneous vaginal delivery with use of epidural anesthesia is generally preferred.
- Cesarean delivery is generally reserved for obstetric indications.
- However, it is suggested for gravidas with advanced heart failure and hemodynamic instability despite medical management.
Calcium channel blockers are generally avoided in the setting of aortic dilation, based on animal and limited human data suggesting that they may increase the risk of aortic complications.

If there is severe aortic regurgitation, beta blockers are typically avoided since they prolong diastole and theoretically may increase aortic regurgitation.

Patients with refractory hypertension may require a combination of drugs to effectively control blood pressure.
Hypertension is more common in patients whose repair was performed after 20 years of age compared with those who were repaired in early childhood; however, the risk of hypertension increases over time in all patients.
Paradoxical hypertension

- The postcoarctectomy syndrome may occur during the first 2–5 days following coarctation repair, with systolic and diastolic pressures rising above pretreatment levels.
- In severe cases, mesenteric arteritis and bowel ischemia may develop.
- The mechanism is related to rebound activation of the sympathetic nervous system and the renin–angiotensin system which leads to mesenteric arterial vasoconstriction.
Paradoxical hypertension

- Postoperative it can be prevented with beta-blocker therapy and by aggressive antihypertensive therapy during the immediate postoperative period.
- It is uncommon following balloon angioplasty of coarctation.
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