# Perspectives in Paediatric Cardiology: Perspektive u pedjatrijskoj kardiologiji

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# COARCTATION OF THE AORTA

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## Abstract

This "simple" condition still raises many questions. Johan Meckel published 1768 "narrowing of the aortic conduit". 1944 was the first successful surgical repair performed by Clarence Crafoord and Robert Gross independently. In the 70-ties balloon angioplasty started and the development of transcatheter treatment is still in progress. The transcatheter treatments have become the treatment of choice for "simple" coarctation but surgical repair still has an important role.

Congenital coarctation (CoA) involves with very rare exceptions thickening of the media forming a ridge protruding from the posterior and lateral wall into the lumen. Further narrowing occurs with additional thickening of the intima. This narrowing occurs usually juxtaductal, opposite the position of ductus arteriosus. The constriction is almost always distal to the left subclavian artery but the origin of this vessel could be involved. Left ventricular afterload is increased by the mechanically induced resistance and the resistance associated with high blood pressure proximal to the constriction. The higher wall tension causes left ventricular hypertrophy. Proximal systolic hypertension and distal hypertension in varying degree is always present as well as change in the arterial waveforms above and below. The arterial pressures are not affected until reduction of lumen by 45–55%. Distally the systolic and diastolic blood pressure decreases gradually with less change in diastolic pressure producing narrow pulse pressure. Development of collateral vessels makes the alterations in blood pressures less pronounced. Primarily they arise from branches of the subclavian vessels, internal mammary arteries, intercostal and spinal arteries. Besides the mechanical obstruction with reduced aortic distensibility not fully compensated by proximal vessels, also resetting of baroreceptors and renal factors most likely contributes. The clinical picture is different in newborns and infants compared to older individuals.

Treatment: In the neonate prostaglandin dilates the arterial duct permitting flow via the ductus beyond the obstruction. Congestive heart failure is treated with furosemid, digoxin, vasoactive amines, phosphodiesterase inhibitors and calcium sensitizers. If blood pressure elevation is so severe that it requires treatment by pharmacological means, surgery is preferable.

Three methods are preferred 1) Resection and end-to-end anastomosis (standard method) 2) subclavian flap aortoplasty 3) synthetic patch aortoplasty. The surgical approach is usually from left posterolateral thoracotomy through the fourth left intercostal space. If additional

intracardiac repair is necessary the approach is through sternotomy. Balloon-angioplasty has been performed since approximately 1982. Percutaneous transluminal angioplasty was first demonstrated to be effective in restenosis after surgery. There may be protection against rupture of the aorta by post-operative scarring and in the aortic wall. A tear in the intima and partially in the media is required for a good result. The inflation diameter should be 2.5 times the narrowest diameter or 1–2 mm greater than the aorta diameter at the base of the subclavian artery but should not exceed the aortic diameter of the descending aorta at the level of the diaphragm. The dilation of the aorta is painful. Two balloons could be used to keep the femoral vascular trauma low.

Stenting of recoarctation as well as primary coarctation has become more and more popular. With the introduction of covered stents the risk of bleeding from wall rupture has been reduced and the stents also permit exclusions of aneurysms. Essentially the risks are stent-migration, damage to the femoral arteries and the same as with surgery. The stents need to be expandable to the normal size of the vessel in adulthood and the size of the equipment does not allow stenting of coarctation in children younger than approximately 10 years. Bare metal stents have also been used proximal to the isthmus across the subclavian and carotid artery. In rare cases, covered stents have been used also across the offspring of the subclavian artery when there has been proof of capacity of the circle of Willisi to provide collateral flow to the arm.

This "simple" condition still raises many questions. This spans from the aetiology and pathophysiology to the treatment.

## HISTORY

Johan Meckel published 1768 "narrowing of the aortic conduit". 1944 was the first successful surgical repair performed by Clarence Crafoord and Robert Gross independently. In the 70-ties balloon angioplasty started and the development of transcatheter treatment is still in progress. The transcatheter treatments have become the treatment of choice for "simple" coarctation but surgical repair still has an important role.

## Etiology and Anatomy

**Congenital coarctation** (CoA) involves with very rare exceptions thickening of the media forming a **ridge** protruding from the posterior and lateral wall into the lumen. Further narrowing occurs with additional thickening of the intima. This narrowing occurs usually juxtaductal, opposite the position of ductus arteriosus. The constriction is almost always distal to the left subclavian artery but the origin of this vessel could be involved.

**Tubular hypoplasia** is a combination of small diameter and abnormal length involving the transverse arch and/or the isthmus. Isolated tubular hypoplasia does not include the ridge in isthmus and the media in this region is normal. In infants The

length of the distal arch-segment (between left subclavian and left carotid artery) the proximal arch segment (between the left carotid and innominate artery) and isthmus should each not exceed 5 mm. Compared to the ascending aorta, the relative diameter of the segments should be: proximal arch 60%, distal arch 50% and isthmus 40%.

## Associated Malformations

Bicuspid aortic valve: The most frequent lesion (13–85% in lit.) Mitral valve malformation of a wide spectrum to Shones syndrome with left heart obstruction in multiple sites.

Ventricular septal defect(s): The clinical manifestations when the VSD is moderate or large shift from high left ventricular afterload to more of left to right shunting and failure. This should be considered when examining newborns. The incidence of CoA is also increased with AVseptal defects and cyanotic VOC as Truncus arteriosus, TGA and DORV of Taussig-Bing type

## Epidemiology and Genetics

CoA is present in 5–8% of VOC patients. Males dominate (2/3), less so in more complicated Coarctation syndrome, and abdominal CoA is more frequent in females. Seasonal peaks indicate some possible environmental influence. In full XO Turners syndrome an incidence of 15–20% is reported but mosaic forms have less incidence. Rare familial occurrence has been reported.

## Embryology and Pathogenesis

Smooth muscle from the ductus extends into the aorta and ductus closure can unmask a CoA but is unlikely the sole cause of aortic constriction. The shelf in the aorta is a constant finding and is proposed to develop from pathological intrauterine conditions. Theoretically intrauterine shift in flow from the left to the right ventricle decreases the flow in isthmus which may become underdeveloped.

## Physiology

Left ventricular afterload is increased by the mechanically induced resistance and the resistance associated with high blood pressure proximal to the constriction. The higher wall tension causes left ventricular hypertrophy. Proximal systolic hypertension and distal hypertension in varying degree is always present as well as change in the arterial waveforms above and below. The arterial pressures are not affected until reduction of lumen by 45–55%. Distally the systolic and diastolic blood pressure decreases gradually with less change in diastolic pressure producing narrow pulse pressure. Development of collateral vessels makes the alterations in blood pressures less pronounced. Primarily they arise from branches of the subclavian vessels, internal mammary arteries, intercostal and spinal arteries. Besides the mechanical obstruction

with reduced aortic distensibility not fully compensated by proximal vessels, also resetting of baroreceptors and renal factors most likely contributes. Experimental work in rats showed an increased blood pressure response to angiotensin II bolus, increase in renal vascular resistance and change in regulation of renal angiotensin II receptors. Constriction of the aorta distal to the takeoff of the renal arteries resulted only in decrease in the blood pressure in the femoral artery but the proximal pressure was normal. In dogs the blood pressure was normalized when the kidneys were transplanted to vessels proximal to the induced constriction of the aorta. Inconsistent data regarding the renin activity and actual angiotensin II levels in man indicate a more complex role of the Renin-Angiotensin system.

## Clinical features

The clinical picture is different in newborns and infants compared to older individuals.

### *Coarctation past infancy*

Symptoms: Most patients with isolated CoA have no cardiac symptoms. Minor complaints as cold feet and leg cramps seem more frequent. Par aesthetic pain and muscle weakness may occur in the lower extremities reflecting more likely compression of the spinal cord by a dilated anterior spinal artery or one of its branches impinging upon a nerve root exiting the vertebral canal rather than poor blood flow. Nose bleeds and headaches are seen. Claudicatio with sharp pain in the lower extremities limiting exercise makes abdominal coarctation more likely. Collaterals are more difficult to develop in the abdomen.

### *Physical examination*

Reduced femoral pulses and hypertension are the most common findings that lead to further examination. In the lower extremities the blood pressure is low or hard to measure. Hypertension is almost always present. Turners' syndrome is recognized by short stature, webbed neck and shield chest and is associated with CoA. Difference in blood pressure between arms occur and depends on the relation of origin of the vessels in relation to the CoA and possible involvement of the left subclavian artery in the constriction – both left and right arm pressure as well as the pressure in the leg can in rare occasions be low and the pulses should then be compared with the carotid pulses. Murmur are best heard posterior on the left chest medial to the scapula, left infraclavicular area and axilla. Continuous murmurs due to collaterals can also be heard both anteriorly and posteriorly. Murmurs could be absent of atresia or low cardiac output and severe obstruction. Pulsating interscapular arteries with palpable or visible collaterals in the interscapular region are rare in childhood. Simultaneous palpation of brachial and femoral vessels may reveal delay of femoral pulses even if well developed collaterals or mild obstruction may permit palpable femoral pulses.

### *ECG*

ECG is often normal in children even with significant CoA. Signs of left ventricular hypertrophy with increased voltage in left precordial leads may develop. Right bundle branch block is seen in approximately 50% of adults and older children and possibly reflects progression of neonatal ventricular hypertrophy.

### *Chest Roentgenography*

The size of the heart is normal or mildly increased with left ventricular contour. In some patients the *3-sign* or double aortic curve as a result of pre- and poststenotic dilatation can occur. Rib notching is practically pathognomonic of thoracic CoA and its incidence increases with age.

### *Echocardiogram*

Left ventricular wall thickness is increased reflecting the increased afterload. Suprasternal and high left views may show the CoA. It is important to examine the mitral valve, left ventricular thickness and shortening fraction, left ventricular outflow tract and the abdominal aorta. Magnetic Resonance Imaging and CT-scan:

This gives information of the degree of the coarctation, the localization and relation to the subclavian and carotid arteries and any collaterals. This is used to plan any catheter intervention or surgery. Any concomitant aneurysms, heart malformation and possible left ventricular hypertrophy could be detected. When considering more complex interventions, the course of vertebral arteries and the patency of circulus willisii are valuable information. The spinal blood supply is usually not seen but can hopefully be elucidated in a simple way in the future.

### *Cardiac catheterization and Angiography*

Usually the indication for any treatment can be established by non-invasive studies and catheterization and angiography (in more modern labs - rotational angiography) is used for guiding and ascertaining the indication for intervention by estimation of the gradient. All factors with reduced cardiac output such as severe aortic valve obstruction, myocardial disease or anaesthesia per se, will reduce the gradient even if the obstruction is severe.

### *Coarctation in infancy*

Often occurs in combination with other malformations e.g. VSD. In the majority of symptomatic cases there is varying degrees of pulmonary hypertension though seldom to super systemic levels. Different criteria are used to differentiate normal tapering of the aorta and coarctatio. Taussig suggest that the isthmus is not abnormal until it is narrowed to the width of one carotid artery. Moulaert concluded that the diameter of the isthmus should be less than 40% of the ascending aorta. The haemodynamics vary depending on site and severity of aortic obstruction, associated defects and any contribution of flow in the duct. In isolated CoA with a ledge

proximal to the duct a critical resistance to left ventricular emptying occurs only after birth when the ductus closes and runoff to the lower resistance pulmonary bed is abolished, resulting in severe left-sided congestive failure. This results in a clinical presentation of an infant in respiratory distress with severe cardiac failure. If the cardiac output is seriously impaired, a murmur may not be present until improvement of the cardiac output. With right to left shunt over the duct differential cyanosis occur with cyanosis more pronounced in the lower part of the body. With a coexisting VSD the symptoms are mainly those of large left to right shunting. The combination in an infant with CoA and heart failure imposes a high suspicion of associated defects. The anatomical and physiologic complexity of CoA is high in infancy. ECG: In isolated CoA there are signs of right ventricular hypertrophy or right bundle branch block. Left ventricular ECG-changes are uncommon in the neonate but left ventricular strain may reflect the increase in left ventricular afterload occurring with closure of the duct.

Echocardiogram: Increase in LV wall thickness may be seen. In failing children a dilated poorly contracting left ventricle is seen which also could be the case in cardiomyopathy or endocardial fibroelastosis. The changes are reversed after relief of the obstruction. Chest X-ray: The changes vary with associated defects from increased vascularisation as a sign of left to right shunting to venous congestion seen in severe coarctation or lesions with left heart obstruction. Rib notching is extremely rare.

Cardiac catheterizations are risky and should be avoided unless non-invasive examinations are not conclusive.

### *Unusual Coarctation*

Atypical locations are ascending aorta, transverse arch and abdominal aorta. Multiple sites of obstruction are extremely rare. Abdominal CoA is more common in females. There are sometimes murmurs over the abdomen. Leg cramp and Claudicatio occurs. The obstruction is usually longer and is usually associated with *Takayasus syndrome* with marked intimal hypertrophy alternating with areas of aneurysm. When involving the upper extremity pulses and renal arteries, but sparing the femoral arteries, it can give reduced upper pulses but palpable femoral pulses and lower extremity hypertension – reversed coarctation. In *Pseudocoarctation or "kinked aorta"* the aorta is elongated and tortuous producing a kinking in the vessel. There is a medial ridge and probably a mild form of coarctation. Multiple sites of obstruction are rare as well as functional coarctatio produced by extrinsic compression.

## Natural history

The lifespan is reduced with reported 90% mortality by age of 50 and the mean age of death being 35 years. Causes of death were intracranial haemorrhage (11%), aortic rupture or dissection (23%), endocarditic (22%) and congestive heart failure (18%). Berry aneurysms in the Circle of Willis are thought to occur in 10% of

patients with CoA. Cerebral haemorrhage in neonates with CoA is extremely rare. Dissection and rupture during pregnancy in patients with CoA is rare but is the main hazard in this situation.

## Treatment

### *Medical*

In the neonate, prostaglandin dilates the arterial duct permitting flow via the ductus beyond the obstruction. Congestive heart failure is treated with furosemid, digoxin, vasoactive amines, phosphodiesterase inhibitors and calcium sensitizers. In children, medication may be discontinued as collaterals develop. If blood pressure elevation is so severe that it requires treatment by pharmacological means, surgery is preferable.

### *Surgical Therapy*

Earliest method of repair in an experimental animal was reported by Blalock and Park 1944. Surgical correction in humans was first performed by Crafoord and Nylin in1944 in Sweden and by Gross and Huffnagel in 1945. Surgical treatment of coarctatio in an infant was first reported by Kirklin et al. 1952. Surgery could be performed when collaterals were well developed. Three methods are preferred 1) resection and end-to-end anastomosis (standard method) 2) subclavian flap aortoplasty 3) synthetic patch aortoplasty. The surgical approach is usually from left posterolateral thoracotomy through the fourth left intercostal space. If additional intracardiac repair is necessary the approach is through sternotomy. When a Dacron patch was used, the late aneurysm was a late and serious complication.

### *Interventional catheterization*

Balloon-angioplasty has been performed since approximately 1982. Percutaneously transluminal angioplasty was first demonstrated to be effective in restenosis after surgery. There may be protection against rupture of the aorta by post-operative scarring and in the aortic wall. A tear in the intima and partially in the media is required for a good result. The inflation diameter should be 2.5 times the narrowest diameter or 1–2 mm greater than the aorta diameter at the base of the subclavian artery, but should not exceed the aortic diameter of the descending aorta at the level of the diaphragm. The dilation of the aorta is painful. Two balloons could be used to keep the femoral vascular trauma low.

Stenting of recoarctation as well as primary coarctation has become more and more popular. With the introduction of covered stents the risk of bleeding from wall rupture has been reduced and the stents also permit exclusions of aneurysms. Essentially the risks are stent-migration, damage to the femoral arteries and the same as with surgery. The stents need to be expandable to the normal size of the vessel in adulthood and the size of the equipment does not allow stenting of coarctation in children younger than approximately 10 years. Bare metal stents have also been used

proximal to the isthmus across the subclavian and carotid artery. In rare cases covered stents have also been used across the offspring of the subclavian artery when there has been proof of capacity of the circle of Willisii to provide collateral flow to the arm.

### *Complications*

Surgery: Recurrent laryngeal nerve injury, phrenic nerve injury, bleeding from collateral vessels and from the high pressure aortic suturline, chylothorax, postcoarctectomi syndrome (mesenteric arthritis), paradoxical post-operative hypertension and spinal cord ischemia with transient or permanent paralysis.

Interventions: Balloon-angioplasty is associated with immediate and late aneurysm formation. When late repair of this is done, the collaterals may have regressed and perfusion to the lower body may be needed. Paradoxical hypertension is rare.