

ATTACHMENT TO LAËNNEC IN IDENTIFICATION OF THE REASON FOR ARTERIAL HYPERTENSION

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The subject of this paper "Coarctation of the aorta" was chosen with the purpose of attempting to suggest the importance of early detection of coarctation of the aorta in the medical care of infants, children and adolescents by pediatricians and physicians and, at the same time, to point out the clinical importance of skilled physical examination in the care of patients. A diagnosis of the coarctation of the aorta can be made by using only the auscultation method. Findings indicating elevated blood pressure and minutuous auscultation of the heart can be absolutely sufficient to confirm diagnosis of coarctation of the aorta. The importance of early diagnosis of the disease lies in the aim to prevent all the severe consequences that coarctation of the aorta can involve. Coarctation of the aorta is associated with premature cardiovascular disease in adult life. Early detection and treatment is important to prevent shortening of the life-span of these individuals.

Descriptors: COARCTATION OF THE AORTA, ARTERIAL HYPERTENSION, EARLY DIAGNOSIS, PHYSICAL EXAMINATION

Introduction

Coarctation of the aorta (CoA), a juxtaductal type, is not a rare reason for arterial hypertension in pediatric clinical practice. Diagnosed and then repaired early in life, aortic coarctation is a potentially curable cause of hypertension (1). Diagnosis of CoA can be readily made by using the elementary skills of physical examination that include inspection, auscultation, and palpation (2-4). With their knowledge, talents, experience and primordial use of their senses - of sight, hearing and touch - doctors cannot fail in diagnosis of CoA, and cannot endanger their patients. Coarctation of the aorta is a "simple" heart disease and, at the same time, a very "complex" one, "simple" due to the simple anatomic change, the most frequent being a circular narrowing of the aorta, and very "complex" due to all the physiopathology that occurs

with the narrowing of a segment of the descending aorta (5). In the first place, that includes arterial hypertension in the proximal part of the body, above the obstruction.

The diagnosis of CoA can be made using only one instrument, a stethoscope, by linking the data obtained through measurement of blood pressure, and by the identification of systolic murmur in the left interscapular region. Advanced technology such as ultrasound and magnetic resonance will confirm diagnosis of coarctation established by using elementary medical skills.

Reminiscence

René Théophile Hyacinthe Laënnec, a French physician, invented the stethoscope in 1816, while working at the Hôpital Necker-Enfants Malades in Paris. Paris was the capital of the medical world in that period, from 1800 to 1850. The Parisian medical community was the bright light that attracted physicians from all over the world and medicine was influenced profoundly by what has come to be known as the French School (4, 6). French medicine still attracts phy-

sicians from all over the world. Laënnec presented his invention in a talk at the Academie de Médecin in the February of 1818, later publishing the classic treatise "De l'Auscultation Médiante". René Laënnec named the instrument for mediate auscultation the *stethoscope*, which is derived from the Greek "*stethos*"-chest, and "*skopéō*"-to examine, inspect. (6, 7). We are now in the 21st century, while the name of Laënnec's invention has remained unchanged, that is, we still use the same term - *stethoscope* - even though a period of almost 200 years has elapsed since its first presentation to colleagues in Paris.

Coarctation of the aorta

Lesions of the aortic arch or proximal portion of the descending aorta are common and account for about 5-9% of congenital disorders of the heart and great vessels (1, 8, 9). Etymologically, coarctation means narrowed, constricted, or compressed and is of Latin derivation. Latin *coarctatus* "co" + "artare" to compress (from *artus*-tight) (7). CoA can be defined as a discrete area of narrowing of the aorta at the point of insertion of the ligamentum arteriosum (1).

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In some earlier descriptions, the lesions were classified as the "infantile" type of coarctation, which, it was thought, usually produced severe symptoms in infancy and the "adult" type, which usually produced symptoms in older children or adults. The infantile type of coarctation, also known as tubular hypoplasia of the aorta, or preductal coarctation, was characterized by diffuse narrowing of the aorta proximal to the ductus arteriosus. Because the ductus arteriosus is patent during infancy, and blood may flow from the right ventricle and pulmonary artery to the descending aorta, the term *coarctation of the aorta with systemic right ventricle* was used to describe this anomaly. The adult type of coarctation was defined as a localized narrowing of the aorta in the region at the junction of the ductus arteriosus or ligamentum arteriosum with the aorta, and the term *juxtaductal coarctation* described this localized narrowing of the aorta. There is usually a circumferential narrowing that often produces a greater degree of obstruction of the lumen than suggested by the external appearance (8).

The classification of coarctation into infantile and adult types is no longer acceptable, because infants can have also a localized aortic narrowing in the region of the ductus arteriosus and may develop severe cardiac failure in the newborn period (8). *Morphogenesis and the etiology* of the CoA are not clearly known. Existing diverse hypotheses about the disturbance in the aortic arch are as follows (9):

- The hemodynamic theory proposed by Rudolph includes the alteration in fetal hemodynamics related with tubular hypoplasia of the aorta.
- The mechanical theory proposed by Škoda of abnormal extensions of the ductus arteriosus into the aortic wall as the factors responsible for closure of the ductus arteriosus after birth, with constriction of the ductus tissue in the aorta resulting in coarctation.
- Other theories:
 - genesis of coarctation could be due to abnormal cell migration of the neural crest;
- CoA as a disease inherited in an autosomally dominant way pattern.

Pathological anatomy

- *Narrowing*: The mere term "coarctation of the aorta" indicates that the aortic lumen is narrowed. Found more often as localized and circular, just distal to the left subclavian artery. Histological examination evidence is hyperplasia of the media cells, and the sclerosis of the intima.
- *Dilatation* of the descending aorta below the lesion.
- *Consequence of the coarctation*. CoA should be regarded as a diffuse arteriopathy (8):
 - Collateral circulation such as between the intercostal and epigastric vessels, scapular and mammary vessels, vertebral and spinal vessels, for revascularisation of the descending aorta.
 - Aneurysmal formations such as aneurysms of the circle of Willis, so-called "berry aneurysms", are thought to occur in about 10% of individuals with CoA, but their true incidence is not reliably known. They appear to occur in adult life and to be associated with longstanding hypertension. It is proposed that there is an underlying degenerative change in the media.
 - A modification of the coronary vessels and the histologically proliferative and degenerative changes in the vessel's intima-media hypertension, and the hypertrophic left ventricle due to the arterial.
- *Associate cardiovascular lesion*:
 - Tubular hypoplasia and coarctation may both occur in the same individual.
 - Aortic atresia has associate juxtaductal coarctation in about 75-80% of infants.

Physiopathology

The fundamental problem lies in the mechanism of blood pressure elevation, arterial hypertension. The cause of hypertension above the coarctation and hypotension below the coarctation is, in the first place, a *mechanical* issue. The other factors that interfere to raise the blood pressure are:

- activation of the *renin-angiotensin-aldosterone* (RAS) system provoked by renal hypoperfusion;
- dysregulation of the aortic arch *baroreceptors*;

- elevation of *systemic peripheral vascular resistance* (1, 8, 10).

Clinical semiology

The first detection of the disease is from the newborn period till late childhood, often before the age of 15 years (8). *Circumstance of discovery of CoA:*

- Most often by auscultation of systolic murmur.
- Or, by finding arterial hypertension; the condition of coarctation of the aorta may first be recognized when it is appreciated that a child or adolescent has hypertension.
- The eye being caught by a particular morphology of the body, unusual habitus, athletic thorax on thin legs.
- By presented symptom links to cephalic hypertension - epistaxis, vertigo, headache.
- By presented symptom links to hypotension of an inferior limb - a cold foot.

Physical examination

The contrast between signs of hypertension above the obstruction and signs of hypotension below the obstruction of the aorta is clearly evident.

- *Inspection:*
 - The upper part of the trunk and arms are frequently well developed, whereas the pelvis and the lower extremities are slender.
- *Auscultation:*
 - Elevation of the systolic and diastolic blood pressure in the arms. Usually, the arm blood pressures are moderately increased from between 130 to 160/90 to 110 mmHg and the leg pressures range from 75 to 100/65 to 85 mmHg, but higher arm pressures may occur, particularly in adults (8). If the left subclavian artery is involved in the coarctation, the left arm pulse may be weak and the pressure low.

- The heart is not usually enlarged clinically but there may be a prominent apical impulse.
- The first sound is normal; the second sound may be increased at the upper right sternal border when hypertension is marked.
- A systolic ejection click is frequently heard along the left margin of the sternum, and it usually indicates the presence of a bicuspid aortic valve.
- A soft continuous murmur present over the left side of the spine along the scapular margin, which could be due to flow through the coarctation, but could be related to flow through large collateral arteries such as the intercostal or scapular vessels.
- A harsh systolic ejection murmur of grade 2 to 3/6 intensity may be heard at the upper right sternal border if aortic stenosis is present.

- *Palpation:*

- The pulses in the arms are strong but the femoral and dorsalis pulses are decreased.
- The quality of the pulses in the lower limbs depends on the severity of the lesion and the collateral circulation.
- In severe obstruction, femoral pulses may not be palpable.
- If obstruction is less severe, or with good collateral circulation, the pulses may be weak but delayed as compared with the arm pulses.
- The large collateral arteries may be palpated over the lateral and inferior scapular margins and above the clavicle.

Complementary examinations

- *Electrocardiography:* in children is variable, usually it shows a normal pattern or an increase in left ventricular forces.

- *Chest Radiography:* the heart size is normal or moderately increased and there is prominence of the left ventricle. An indentation may be seen along the left margin of the aortic shadow just beyond the arch; this has been termed as 3 sign.
- *Ultrasound Examination:* in the hands of skilled echocardiographers, the diagnosis can be made with assurance, so that is usually possible to refer the patient for surgery without additional studies (8).

The important information that should be obtained by ultrasound includes:

- definition of the presence of and site of obstruction;
- site of origin and morphology of the large arteries arising from the aorta;
- estimation of the severity of obstruction;
- patency of the ductus arteriosus;
- definition of associated intracardiac or great vessel malformation;
- Doppler flow studies are often helpful in defining the site of coarctation;
- continuous wave Doppler flow may demonstrate two flow velocities within a single envelope.
- *Magnetic resonance imaging (MRI)* is a very elegant technique for exposing the CoA. By providing the morphologic image, this technique can describe perfectly the aortic arch (9).

Principles of Management

The conventional treatment of coarctation in children and adults is surgical. Surgical repair of CoA has a very low risk of mortality. Operative complications include hemorrhage from injury, chylothorax from injury to the thoracic duct, and spinal cord ischemia resulting in paraparesis or paraplegia. Of these, the most feared is paraplegia (1, 8, 9). The use of balloon angioplasty is not recom-

mended as a primary procedure for relief of coarctation because of the high incidence of complications, it is now considered to be appropriate treatment for recoarctation (1, 8-10).

If the cardiac failure can be controlled by medical therapy, it has been recommended that surgery be delayed to the age of 3 to 5 years. The aortic diameter reaches about 50-60% of adult diameter by 3 to 4 years of age. Based on studies, a significant gradient did not develop until the aortic diameter was reduced by 50% and it was hypothesized that, even if the aorta did not grow after surgery, its diameter would be adequate even in adult life (8). So, the timing of the repair is most important. When aortic repair is performed in patients younger than 5 years, blood pressure normalizes in up to 80-90% of cases. When CoA is repaired later in life, in teenagers or adults, two-thirds of patients may have persistent chronic hypertension despite successful surgical repair with no residual pressure gradient. Hypertension tends to be "fixed" on account of structural vessel abnormalities (1).

Surgery should not be delayed if there is marked hypertension or if there is evidence of marked left ventricular hypertrophy on the electrocardiogram, particularly if associated with ST and T wave changes (8). Repair of coarctation does, however, reduce the risk of cerebral hemorrhage or cerebral vascular complications, as well as the risk of aortic rupture. Although the effects of relief of coarctation on the risk for infective endocarditis are not known, it is recommended that antibiotic prophylaxis for infective endocarditis be continued after surgery. This is particularly important because many of these individuals have bicuspid aortic valves (8).

Postoperative evolution in the distance of time

Despite operative repair, subjects who have had coarctation of the aorta still remain at high risk of cardiovascular morbidity and mortality. Systemic hypertension at rest and during effort is a well-known complication of patients after successful coarctation of the aorta

repair. The pathophysiological course of hypertension in this condition is not fully understood, and several functional and anatomic factors have been proposed as possible explanations (5, 11-13). Anatomic factors, such as narrowing at the site of intervention, aortic arch hypoplasia, and type of surgery, as well as functional abnormalities, such as baroreceptor dysfunction, altered responsiveness of the renin-angiotensin system, abnormal response to sympathomimetic agents, and increased arterial resistance of the upper limb associated with structural alterations of the arterial wall, have been proposed as the causes of this finding.

Early vascular remodeling in upper limb conduit arteries has been detected in children and young adults, even after successful and early surgery. The determinants of vascular remodeling in post-coarctectomy subjects are unclear. Three parameters for functional arterial wall alterations were calculated: distensibility, stiffness and elasticity (12). There is now a large body of literature that describes a consistent vascular phenotype of the conduit arteries proximal to the CoA repair site. This phenotype includes increased intima-media thickness, increased stiffness, and reduced vasoreactivity of the pre-coarctation arteries (5, 11-13). Recently, aortic arch geometry was reported in association with hypertension at rest in patients with successful CoA repair.

A study by French authors from 2004 is the first to report the role of postoperative aortic arch geometry in vascular remodeling in subjects having undergone successful CoA repair (11, 12). These novel findings have clinical relevance, as aortic arch geometry appears to identify subjects at high risk of abnormal blood pressure response and adverse vascular remodeling. Aortic arch morphology by the shape on magnetic resonance images is qualitatively classified as "Gothic", "Crenel", or "Romanesque". Gothic arch had a triangular form, Crenel arch had a rectangular form, and Romanesque arch had a semicircular rounded form. Gothic arch geometry observed after successful CoA repair is a strong predictor of large-artery abnormality in the pre-CoA arterial bed, including vascular thickening, stiffening, and dysfunction (11).

Assessment of aortic arch geometry using MRI and studying vascular function in these subjects may be important in predicting the risk of late hypertension, a major cause of cardiovascular morbidity and mortality late after CoA repair. Aortic imaging might consider arch geometry, as well as being a screening tool for detection of discrete re-CoA or late aneurysm formation (11, 12). Isolated systolic hypertension, influenced by the stiffness of large arteries, is the most common subtype of hypertension after CoA repair and is a major risk factor for stroke, coronary artery disease, and total cardiovascular mortality (5, 11, 12).

Conclusion

Coarctation of the aorta is a lesion of the aortic arch or proximal portion of the descending aorta and accounts for about 5-9% of congenital disorders of the heart and great vessels. Symptoms are attributed to upper body arterial hypertension. A systolic vascular murmur can be heard over the region of stenosis. The importance of early diagnosis of the disease lies in the aim to prevent all the severe consequences that coarctation of the aorta can involve. The diagnosis of the aortic coarctation can be made using elementary medical skills. In present-day times we are witnesses to the fact that the time pressures of modern practice, combined with advances in diagnostic technology, seem to undermine the value of these skills.

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Sažetak

PRIVRŽENOST LAENNEC-u PRI OTKRIVANJU UZROKA ARTERIJSKE HIPERTENZIJE

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Tema članka "Koarktacija aorte" izabrana je sa svrhom ukazivanja na važnost rane dijagnoze koarktacije aorte u dojenčadi, djece i adolescenata koje medicinski nadziru pedijatri, liječnici školske i obiteljske medicine te istovremeno odabranom temom isticanje kliničke važnosti primjene uobičajenih medicinskih vještina fizikalnog pregleda. Sama metoda auskultacije može biti dovoljna za postavljanje dijagnoze. Nalaz povišenog krvnog tlaka na gornjim ekstremitetima i minuciozna auskultacija srca će biti sasvim dovoljne u potvrđivanju dijagnoze. Značaj rane dijagnoze bolesti leži u potrebi sprječavanja svih teških posljedica koje koarktacija sa sobom nosi. Rano otkrivanje i rano liječenje koarktacije spriječiti će skraćivanje životnog vijeka osoba koje pate od ovog oblika bolesti srca, jer je koarktacija aorte vezana uz prerano nastupajuću kardiovaskularnu bolest odraslih.

Deskriptori: KOARKTACIJA AORTE, ARTERIJSKA HIPERTENZIJA, RANA DIJAGNOZA, FIZIKALNI PREGLED