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CLINICAL AND EPIDEMIOLOGICAL ASPECTS IN COARCTATION OF THE AORTA IN CHILDREN

Review Article

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Abstract

Coarctation of the aorta is the fifth as frequency among congenital heart defects, representing 6-8% of all of them. The incidence of coarctation of the aorta is 1: 2,500 births. It is a more commonly encountered pathology in males with a ratio between male and female sex of 2: 1 Patients have different clinical aspects depending on the age at which they are diagnosed. Newborns are most commonly found with heart failure and cardiogenic shock, and large children and adolescents address the pediatrician due to symptoms of headache, epistaxis, lower limb pain, fatigue. To diagnose coarctation of the aorta it is important to measure blood pressure in the upper and lower limbs, and to find a difference of 20 mmHg between them. Pulse is weaker or even absent in the femoral artery whereas in the brachial artery and radial pulse is strong.

Aortic coarctation is a congenital heart malformation that occurs in 98% of cases in the arterial duct, under the origin of the left subclavian artery, but it can still be found in any part of the aorta.
Often it can accompany a Turner syndrome; it is associated in over 70% of cases with bicuspidia aortica. (Kliegman, 2016). Diagnosis could be made using MRI, CT and echocardiography, which is more useful in younger patients as they have acoustic transthoracic images that are easier to assess. (Hoffman, 2014).
Correction treatment may be surgical or endovascular. Surgical methods are: resection and terminal anastomosis, aorticoplasty through the flap of the subclavicular artery or with a synthetic graft. Endovascular methods consist of balloon angioplasty and stenosis. A study by Xianglin Yeaw compared the method of surgical repair and endovascular stenosis in children over 15 years with the following result: both types of interventions had a similar effect in lowering blood pressure and relieving symptoms, but stenosis required a lower number of days of hospitalization with fewer complications. (Yeaw, 2016)
The natural history of patients with this malformation shows an average survival age of 31 years (Campbell, 1956), but death may occur during the first year of life in 90% of complicated patients and in 60% of symptomatic patients. (Gatzoulis et al, 2017)

EPIDEMIOLOGY

Aortic coarctation accounts for 6-8% of all cardiac malformations and is the fifth most frequent condition out of all congenital heart diseases with an incidence of 1 to 2500 births. It is believed that this pathology is even more common in newborn babies who died shortly after birth (Kenny, 2011) it was found in 1: 1550 of necropsies (Jenkins, 1999).

MORTALITY AND MORBIDITY

A study based on autopsy reports that in the absence of surgical correction of coarctation until the age of 14, the survival rate after 20 years is 79%, and in case of interventions performed until the age of 14, the chance of survival after 20 years increases to 90%. Mortality in patients whose aortic coarctation is not surgically treated until the age of 50 is 90% (Campbell, 1970).

GENDER

Aortic coarctation affects more males compared to females, with a reported ratio of 1.74: 1 (Kenny, 2011), and in a study conducted by Campbel, it was even four times more common in boys. (Campbell, 1956). However, predominance differs in case of abdominal aortic coarctation, where the lesion is in equal proportion in both genders (Cohen, 1988).

RACE

Studies report that the incidence of aortic coarctation does not differ by patient's race, however some authors suggest that this is less common in Asians and Africans compared to the white race. It is believed that the apparent lower incidence in Asian and African patients is due to lower level of living conditions and neglect of symptoms or sometimes the frequent presence of asymptomatic patients leading to late diagnosis. (Van der Host, 1972)
Also, some studies suggest that the incidence of both aortic coarctation and other congenital heart diseases differs also by geographical area, which may indicate the lower risk of malformation in specific environments, different genetic susceptibility, and cultural and socio-economic variations. (Botto, 2001)

AGE

The average age at which the patient presents to the pediatric cardiologist is 5 years or according to other authors between 5.8 and 8 years.(Ing, 1996). Although studies show that the optimal age for surgical repair is between 1 and 5 years, the most common age intervals at which the patient is diagnosed are under one year and between the age of 11 and 19.
Symptomatic patients aged under one year usually have congestive heart failure (approximately 67%), and adolescents develop hypertension (Liberthson, 1979). The new studies recommend more frequent prenatal diagnosis as it is associated with longer survival and a better preoperative clinical balance. (Franklin, 2002)

OBJECTIVE CLINICAL EXAMINATION

Clinical examination of newborn babies and infants

Most commonly newborns are presented for examination during the first few weeks of life with dyspnea, eating difficulties, palpitations, and in severe cases with congestive heart failure, cardiogenic shock and its specific signs - tachypnoea, acidosis, oliguria, cyanosis, hepatomegaly. (Dumitriu, 2007) They may rarely be asymptomatic.

In a classic case, aortic coarctation occurs with the disappearance or reduction of the pulse in the lower limbs, but if cardiogenic shock is present, low-pulse hypotension occurs in all arteries, with cardiomegaly, and the presence of a left parasternal systolic murmur, and accentuated noise two. (Garfunkel, 2007), (Kliegman, 2011). There may also be detected pulmonary static rallies.
Newborns may also have reversed differential cyanosis if it is detected prior to closureof the aetial duct, being represented by superior cyanotic limbs and lower limbs normally stained with normal oxygen saturation.
Reversed differential cyanosis is rarely visible at inspection and may be demonstrated by pulsoximetry. (Kliegman, 2016).
If aortic coarctation is associated with large left-right shunt lesions (*e.g.* DSV), the saturation of the pulmonary artery may get closer to the saturation of the aortic artery, with lower pulse oximetry differences. Reversed differential cyanosis may also occur in right-left ductal shunt malformations such as arterial vessel transposition, patent ductus arteriosus, and pulmonary hypertension.

Clinical examination in child and adolescent
Clinically, children and adolescents develop epistaxis, headache, lower limb pain especially when doing physical exercises, fatigue and angina pectoris. (Garfunkel, 2009)
In the objective examination, upper limb hypertension is detected, reduced pulse in the lower limbs, and if the origin of left subclavicular artery is involved, the pulse is diminished also in the left upper limb (Garfunkel, 2009). Concomitant palpation of the brachial or radial pulsewith thefemoral pulse should routinely be performed. A delay in the femoral pulse in a hypertensive patient may be a clue for aortic coarctation (Rao, 1992). Sometimes the child shows a normal somatic development in the upper half of the body and is more fragile in the lower half. (Dumitriu, 2007)
From the auscultation perspective, we may ofteb detect an ejectional murmur, 2-4/6 grade on the right upper side right of the sternum or in the middle and inferior part of the sternum and at the left interscapulovertebralae (Dumitriu, 2007) a higher upper limb pressure of 20 mmHg than in the lower limbs. A galloping pace occurs in the presence of the hypertrophied and noncompliant left ventricle. Usually, there may be heard a murmur in the suprasternal notch. (Rao, 1992). Cardiac noises are normal if the aortic coarctation is not accompanied by other heart diseases. Aortic bicuspidia, as we have already noted often accompanies aortic coarctation, so an ejection click will be heard at ascultation, which does not change with breath in the apex and left parasternally in the upper half or in a diastolic breathing due to aortic regurgitation (Dumitriu, 2007). We may sometimes hear a continuous interscapular left breathing due to the passage of blood through the collapsed area (Rao, 2005).
When symptoms occur in the adolescent, it is usually due to complications such as headache hypertension, dyspnea due to left ventricular dysfunction, exercise intolerance. Due to the hypoperfusion of the inferior limbs in some patients, claudication may occur. (Nance, 2016)
Aortic coarctation can be rarely detected also due to severe intracranial haemorrhage (Curtis, 2012) as a result of a rupture of aneurysmal brain arteries, aortic dissections, spinal and intercostal artery aneurysms. Premature atherosclerotic disease leads to myocardial infections and cerebrovascular accidents (Nance, 2016). It is also important to perform an ophthalmologic examination, which can detect anomalies of the retinal vessels. (Rao, 2005)
A study conducted by Ing Frank concluded that the most common clinical signs in patients with aortic coarctation were systolic blood pressure and a pressure of more than 10 mmHg in upper limbs compared to lower limbs and a higher limb hypertension greater than 95 percentile for the age, and 50% of the patient lot had a blood pressure higher than 140 mmHg. Also, 12% had no pulse at the femoral artery. (Ing, 1996).
Recognition of clinical signs in aortic coarctation is of great importance as it is a congenital malformation commonly found often in one out of 2500 newborns. Early diagnosis enables better record of patients and the initiation of a surgical repair of coarctation prior to the occurrence of various complications including decreased incidence of hypertension.

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