

## Sudden Death Due to Aortic Coarctation

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Aortic coarctation (CoA) is one of the most frequent and severe congenital heart diseases. If it is diagnosed and treated in time, which rarely leads to sudden cardiac death. In recent years a few new cases were reported by forensic pathologists reviving the interest of this community about this disease. We present in this paper a 25-year-old male of sudden cardiac death caused by coarctation of the aorta and discuss some morphological characteristics useful in diagnosing this condition during the autopsy.

**Key Words:** Aortic coarctation • Congenital heart disease • Sudden cardiac death in young adults

### INTRODUCTION

Aortic coarctation (CoA) is one of the most frequent and well known congenital heart diseases counting for 7% of the cases. Most of them are diagnosed in time and treated by surgery or balloon angioplasty citation before; only rarely (usually the asymptomatic forms) remain undiagnosed, from which only 15% live more than 50 years, the cause of death usually being congestive heart failure or acute complications such as aortic dissection/rupture, bacterial endocarditis/endarteritis, rupture of a cerebral aneurysm, etc. As nowadays aortic coarctation is only exceptionally a cause of sudden death, only a few cases being reported in English literature in the last years, most morphological features and the natural history of this disease are known from articles written before the development of corrective medical interventions in the 1940s.<sup>2</sup> Its presence may also suggest a possible collagen disorder like Marfan or Ehlers Danlos syndromes with medical-legal, ethical and genetic impli-

cation un other family members.<sup>3</sup>

CoA is more frequent in Caucasians than Asians (7:1),<sup>2,4,5</sup> more frequent in men than women (1,4-4:1) and is often associated with other pathologies, such as aortic bicuspid valve disease (BAV) (50-75%), hypoplasia or congenital absence of the aortic arch, ventricular septal defect (VSD), transposition of the great arteries, mitral valve abnormalities, Taussing-Bing abnormality, tricuspid atresia, hypoplastic left heart, atresia of the pulmonary artery, abnormal origin of right (or both) subclavian artery (arteries), aneurysms in the Circle of Willis,<sup>3,6</sup> hemangiomas, etc.

### CASE REPORT

A 25-year-old male, without significant personal history (including cardiac) was found dead in his bath tub. External examination found the following: advanced putrefaction and internal organs in advanced autolysis (mainly due to the warm water in the bath tub). Internal examination revealed, at the opening of the pericardial sac, approximately 500 ml of liquid, black blood and blood clots. Heart volume was normal for his age, weight and sex. Heart examination revealed advanced myocardial fibrosis and advanced autolytic changes; maximum thickness of the LV was 1.5 cm, and of the RV was 0.4 cm.

Examination of the aorta revealed an adventitious

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constriction distally from the origin of the brachycephalic branch (BB), left carotid artery (LCA), and left subclavian artery (LScA) at about 1.2 cm from the insertion of an obliterated ductus arteriosus (DA); at 3 cm from the aortic origin was found a rupture of the aortic wall with infiltration, with a length of 1.1 cm, on the convex part of the aortic arch. (Figure 1). After opening the aorta we found a narrowing stenosis corresponding to the adventitial constriction; starting from here (and ascending) upwards, we found a 4 cm long Stanford A type aortic dissection between media and the adventitia; the ascending aorta diameter distally from the coarctation was normal. The aortic valve was tricuspid; other vascular and congenital abnormalities were not found. Examination of other organs and systems could not find other significant pathologic changes.

Histological examination found the following: lungs – intense fibrosis around pulmonary vessels and bronchi; heart – extensive subendocardial, perivascular and interstitial myocardial fibrosis; in the coarctated area – marked medial and adventitial fibrosis and disorganization of the medial elastic lamina. Cause of death was Acute cardiac failure triggered by cardiac tamponade, caused by a non-traumatic rupture and dissection of coarcted aorta. IHC for elastic fibers and apoptotic factors was tempted but proved unsuccessful due to increased tissue autolysis.<sup>7,8</sup>

## DISCUSSION

Aortic coarctation is characterized by a focal narrowing (before, after or around the insertion of the ductus arteriosus - DA), mainly caused by abnormalities of the medial aortic layer; the decrease of aortic diameter is variable from mild to complete obstruction; when the lesion is preductal and DA is patent the heart does not present significant changes but atherosclerotic areas or unspecific aortic calcifications may be found.

There are several classifications for CoA, the most used being: (1) adult and infant aortic coarctation and (2) preductal, ductal and postductal. Nowadays, all these types are considered as juxtaductal CoA. Two other types, less encountered in clinical practice are: descending aortic coarctation, with four subtypes (type I – supra-renal coarctation with renal gland stenosis, type II –

infra-renal coarctation with adrenal gland stenosis, type III – supra-renal coarctation with normal renal arteries, and type IV – infra-renal coarctation with normal renal arteries) and aortic pseudocoarctation (lengthening of the distal aortic arch due to an abnormal growth of the preductal aorta).

Main complications occurring at this level are: aortic dissection/rupture of aorta and bacterial endarteritis.<sup>9</sup> Campbell for example<sup>5</sup> from 34 cases of CoA found dissection/rupture of the aorta to be the cause of death in 21% cases, the average age of death being 25 years.

Heart morphology is highly dependent on the morphological characteristics of the coarctation but also on the presence of associated congenital diseases, with the left ventricle being either hypoplastic (in preductal CoA) or hypertrophied (caused by increase afterload). Left atrium is usually dilated in AoC, secondary to high atrial telesystolic pressure and increased backflow from LV, whilst right ventricle (RV) becomes hypertrophied due to increased circulating volumes and pressures needed for aortic perfusion throughout DA; when pulmonary hypertension (PHT) develops, it adds a supplemental increase of the RV pressure leading to right ventricle failure (RVF). Valvular complications are often found, especially valvular insufficiency and subaortic stenosis. CoA is associated with aortic bicuspid valve disease in 50-85% cases. Circulation and vascular abnormalities may lead to local infective processes, bacterial endocarditis being more frequent in persons with CoA than



**Figure 1.** Aortic rupture secondary to a dissection originating in the coarcted area.

the general population. Elevated blood flow favors the development of berry aneurysms which are predisposed to rupture – the resulting intracranial hemorrhages also being important causes of death in patients with CoA.

Collateral circulation (Table 1) is a significant adaptive mechanism by which the lower parts of the body are vascularized. Clinically, they can be diagnosed by palpating these dilated arteries at the level of the anterior or posterior thorax or by hearing systolic murmurs at these levels; radiological examination reveals the presence of specific intercostal notches. At necropsy, collateral circulation can be highlighted through several methods: 1) determining the presence of intercostal notches; 2) identifying dilated and tortuous anastomotic vessels; 3) post-mortem angiography, 4) postmortem computed tomography or magnetic resonance imaging. Complications of collateral circulation are rarely the cause of death in patients with CoA – Haberer<sup>10</sup> described a case in which a dilated anterior spinal artery triggered medullar compression at the second thoracic vertebral level with subsequent transverse myelitis which got complicated and caused death.

There are two types of intercostal notches: superficial, with symmetric entrance and exit slopes and deep/profound with asymmetric entrance and exit slopes.

Typically they are bilateral, posterior and lateral on ribs III to IX (the blood flow goes in especially through posterior and lateral branches; anterior branches, resulting from internal mammary are less important in these cases), sometimes double (especially on the III-V ribs). Ribs I and II are not affected as their origin is proximal to the coarctated area (their origin is from the subclavian artery - ScA); they can be affected in cases with an associated abnormal, distal origin of the subclavian artery.

Main pathological findings are enlargement of the subendothelial space, with an increase in the number of collagen fibers at this level and the presence of smooth muscular fibers; internal elastic lamina is characterized by an increased fragmentation, hyperproduction of collagen fibers and a radial orientation of the smooth muscular fiber (SMF) layer; in media and external elastic lamina, SMF are settled in circular packages; in areas of intimal thickening, the structure of the media becomes disorganized, with randomly distributed SMFs, a very well represented collagen matrix and a decreased number of elastic fibers.<sup>3,7</sup> External third of the media has an increased concentration of acid mucopolysaccharides best viewed using the periodic acid-Schiff technique. The aortic arch and isthmus (if hypoplastic) have a reduced number of elastic lamellas; distally the number is similar to

**Table 1.** Collateral circulation in aortic coarctation (modified after [7])

Anastomotic group	Branches involved
Scapular and cervical	Descending branches from suprascapular and transversalis colli (from thyroid axis) with dorsal and lateral branches from aortic intercostals Descending branches from posterior scapular and superficial cervical (from transversalis colli) with dorsal and lateral branches from aortic intercostals Descending branches from long thoracic and subscapular with its dorsalis scapula branch (from axillary artery) with dorsal and lateral branches from aortic intercostals
Internal mammary	Superior epigastric with deep epigastric branch of external iliac Musculo-phrenic with phrenic branches from thoracic and abdominal aorta Mediastinal branches with mediastinal branches from the aorta Anterior intercostals with terminal branches from aortic intercostals
Intercostal	Terminal branches with intercostal branches from internal mammary Lateral branches with the subscapular and long thoracic arteries Dorsal branches with posterior scapular First and second intercostal (arising from the subclavian artery with superior intercostal arteries Each intercostal with those above and below
Spinal	Spinal arteries from vertebral artery with spinal branches of aortic intercostals Inferior thyroid branches passing through intervertebral foramina with spinal arteries; from there blood can flow through branches of aortic intercostals

the one found in normal subjects.<sup>8</sup> Histological changes determined by CoA are often intricately associated with those determined by BAV. The latter is frequently associated with a decrease of the SMF in the ascending aorta and with elastic fiber degeneration in the media (cystic necrosis of the media). The most probable cause is apoptosis caused by enhanced parietal stress, which is also demonstrated by the variation of Bcl-2 expression on the convexity versus concavity of the aortic arch;<sup>7,8</sup> the presence of BAV augments up to nine times the risk for acute dissection of aorta and decreases the average age when it occurs, in comparison to individuals suffering from tricuspid aortic valve.<sup>2,10</sup>

Sudden death caused by aortic rupture as a consequence of CoA is rare nowadays, as CoA is relatively easy to diagnose and treat. From an etiological point of view, our case belongs to the typology described by Campbell (male, 25 years old).<sup>4-6</sup> From an etiopathogenic point of view, this case presents certain particularities, such as: 1) absence of associated cardiovascular congenital abnormalities, including the absence of BAV, which favors dissection of the aorta in patients with CoA,<sup>2,10</sup> 2) absence of proximal aortic dilatation/aneurysm, which could also favor aortic dissection/rupture; 3) absence of cardiomegaly/ventricular hypertrophy, also often associated with CoA (50-85%). However, LV had significant fibrotic changes suggestive for a chronic cardiovascular pathology (in this case, a possible explanation could be the initial presence of a left ventricular hypoplasia, with subsequent hypertrophy).

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