TRUNCUS ARTERIOSUS COMMUNIS (TAC) ASSOCIATED WITH MALFORMATIONS OF OTHER ORGAN SYSTEMS

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Objective: To identify the rate and incidence of congenital defect Truncus arteriosus communis or TAC and its associations with malformations of other organ systems.

Background

Truncus arteriosus (TA) is an uncommon congenital cardiovascular anomaly that is characterized by a single arterial trunk arising from the normally formed ventricles by means of a single semilunar valve (ie, truncal valve). In addition, the pulmonary arteries originate from the common arterial trunk distal to the coronary arteries and proximal to the first brachiocephalic branch of the aortic arch. The common trunk typically straddles a defect in the outlet portion of the interventricular septum (ie, conal septum); however, in rare cases, it may originate almost completely from the right or left ventricle. In patients with a patent and normal caliber aortic arch, the ductus arteriosus is either absent or diminutive.

Embryology

The anomaly is thought to result from incomplete or failed septation of the embryonic truncus arteriosus, hence the persistence of the Latin term *truncus arteriosus* and its variants. Aortopulmonary and interventricular defects are believed to represent an abnormality of conotruncal septation. Because the common trunk originates from both the left and right ventricles, and pulmonary arteries arise directly from the common trunk, a ductus arteriosus is not required to support the fetal circulation.

Accordingly, an inverse relationship between the caliber of the ductus arteriosus (derived from the sixth branchial arch) and that of the distal portion of the aortic arch (derived from the fourth branchial arch) is typically present. Although the hemodynamic consequences of a common arterial outflow may predispose to the development of the fourth or the sixth arch (but not both), anomalous development of the arch system is likely a fundamental aspect of the morphogenetic anomalies that produce truncus arteriosus.

Anatomy

Pulmonary arteries may arise from the common trunk in one of several patterns, which are often used to classify subtypes of truncus arteriosus. Several classification schemes have been proposed, none of which is ideal.

The earliest classification, developed by Collett and Edwards in 1949, includes truncus arteriosus types I-IV, as follows

- **Truncus arteriosus type I** is characterized by origin of a single pulmonary trunk from the left lateral aspect of the common trunk, with branching of the left and right pulmonary arteries from the pulmonary trunk.
- **Truncus arteriosus type II** is characterized by separate but proximate origins of the left and right pulmonary arterial branches from the posterolateral aspect of the common arterial trunk.
- **In truncus arteriosus type III**, the branch pulmonary arteries originate independently from the common arterial trunk or aortic arch, most often from the left and right lateral aspects of the trunk. This occasionally occurs with origin of one pulmonary artery from the underside of the aortic arch, usually from a ductus arteriosus.
- **Type IV truncus arteriosus**, originally proposed by Collett and Edwards as a form of the lesion with neither pulmonary arterial branch arising from the common trunk, is now recognized to be a form of pulmonary atresia with ventricular septal defect rather than truncus arteriosus.

Collett and Edwards describe variations of each of these types.

In 1965, Van Praaghs proposed the other commonly cited classification scheme that also includes 4 primary types, as follows

- Type A1 is identical to the type I of Collett and Edwards.
- Type A2 includes Collett and Edwards type II and most cases of type III, namely those with separate origin of the branch pulmonary arteries from the left and right lateral aspects of the common trunk.
- Type A3 includes cases with origin of one branch pulmonary artery (usually the right) from the common trunk, with pulmonary blood supply to the other lung provided either by a pulmonary artery arising from the aortic arch (a subtype of Collett and Edwards type III) or by systemic to pulmonary arterial collaterals.

- Type A4 is defined not by the pattern of origin of branch pulmonary arteries, but rather by the coexistence of an interrupted aortic arch. In the vast majority of cases of type A4, which fall into the type I of Collett and Edwards, the pulmonary arteries arise as a single pulmonary trunk that then branches. In any of these patterns, intrinsic stenosis, hypoplasia, or both may be present in one or both branch pulmonary arteries, which may have an effect on management and outcome.

Mortality/Morbidity

The natural history of truncus arteriosus without surgical intervention is not well characterized. In numerous earlier series, the median age at death without surgery ranged from 2 weeks to 3 months, with almost 100% mortality by age 1 year. Cases of patients surviving into adulthood with unrepaired truncus arteriosus are reported, but they are extremely uncommon. Cause of death in unrepaired patients is usually cardiac arrest or multiple organ failure in the face of systemic perfusion that is inadequate to meet the body's metabolic demands; progressive metabolic acidosis and myocardial dysfunction results.

Currently, for patients undergoing complete repair in the neonatal or early infant periods, early postoperative mortality is generally less than 10%. This represents a substantial improvement from earlier eras; as recently as 20 years ago, the early mortality rate after complete repair was higher than 25% in most series. Among patients surviving the initial postoperative period, the survival rate at a 10- to 20-year follow-up is higher than 80%, with most deaths resulting from sequelae of late repair (pulmonary vascular obstructive disease), reinterventions, or residual/recurrent physiologic abnormalities.

Although rarely used today, surgical palliation by banding of the pulmonary artery to protect the pulmonary vascular bed was a frequently used strategy until the 1970s and early 1980s. This practice resulted in only minor improvement in the natural history of the disease, with substantial early and intermediate mortality rates.

MATERIALS AND METHODS: At the Institute of Pathological Anatomy at the Medical Faculty, which is a part of UKIM, in the period from 2007 to 2010, 1739 autopsies were made and in 70 cases complex congenital cardiopathies were diagnosed. Using a profound morphological analysis, five cases with TAC were separate, three of which were fetuses with induced abortion, because the clinical diagnosis Cardiopathia congenital was set, and two lived do 2,5 months. Two of them were female, and three male. RESULTS In the analysis of TAC a Collett & Edwards schematic representation and classification is used with the following variations of TAC type II (5), in one case there are 4 valves located at the ostium, and in another case the

right pulmonary artery comes out while the left is missing. The deeper morphological analysis showed accompanying heart defects and VSD in all five cases of TAC. By A-V channel they are found at (1), HLHS (2), EFE (1) and LHSVP (1).

The pathoanatomic analysis of the other organic systems of the TAC cases is combined with severe anomalies of other systems in 4 cases:

1. TAC + hydrocephalus internus+ omphalocoelae + hypoplasio extremitas superioris + syndactilia + agenesio pulmonum sin (1),

2. TAC + pulmo unilobatum bill. + hypoplasio thimy + ren polycysticus (1),

3. TAC + thymomegalia + agenesio renum + cheilognathopalathoshisis + hexodactilia mani bill. + arrhinia (1)

4. TAC + pulmo dexter billobatum + lien accesorius + pes equinovarus bill (1).

CONCLUSION: The deepened morphological analysis showed that the most common type of TAC is a type II according to Collett & Edwards classification, with mild male predominance (3:2) and a high percentage (80%) of association with malformations of other organ systems. The knowledge of the morphology TAC and its connection to other anomalies is very important for understanding and early detection of this congenital cardiopathy which enables well-timed and successful further surgical treatment.

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