Clinical Features, Management and Outcome of Pre- and Postnatally Diagnosed Common arterial trunk: A Comprehensive Single Institution's Experience Since 1990

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Introduction: Common truncus arteriosus (CTA) is associated with significant morbidity and mortality although improvements in management have been made over time. The purpose of this study was to determine outcome, risks associated with mortality and the impact of prenatal detection on survival.

Methods: We performed a retrospective review of CTA cases diagnosed fetally and postnatally at our institution between 1990 and 2014.

Results: There were 192 CTA cases with 61 fetally diagnosed. Of the fetal cases there were 18 terminations of pregnancies, 2 intrauterine deaths and 4 live-births that were not actively managed. One patient diagnosed postnatally was not actively managed. This left 167 patients with intended postnatal care. Mixed truncal valve disease occurred in 41% of cases with moderate or severe regurgitation or stenosis seen in 23% and 18% of cases respectively. Other associations included aortic arch interruption (16%), right aortic arch (26%), pulmonary arterial interruption (16%) and DiGeorge syndrome (24% of tested cases). Including 17 (9%) infants that died prior to surgery, survival of the 167 actively managed patients was 74%, 62% and 56% at 1 month, 1 year and 10 years respectively. Reoperation rate at last follow up was 37%. Multivariable independent risk factors for mortality included interrupted aortic arch [HR 3.7 (2.1-6.6), p<0.001], moderate or severe truncal valve stenosis [HR 3.3 (1.7-6.4), p<0.001] or regurgitation [HR 2.6 (1.5-4.5), p=0.001], hypoplastic or interrupted pulmonary arteries [HR 2.1 (1.2-3.6), p=0.006], prematurity [HR 1.2 (1.1-1.3), p=0.004], lower preoperative weight [HR 1.6 (1.1-2.4), p=0.02], earlier year of surgery [HR 1.08 (1.03-1.13), p=0.002] and the need for life support preoperatively in the form of inotropes [HR 3.2 (1.6-6.3), p<0.001] or mechanical ventilation [HR 2.1 (1.1-4), p=0.03]. Fetal detection rates improved over time but a fetal diagnosis was not associated with improved survival rates.

Conclusions: The overall outcome of CTA during childhood is poor and comparable to the most severe forms of congenital heart disease. Moderate to severe truncal valve regurgitation or stenosis, interrupted aortic arch and pulmonary artery abnormalities are associated with a worse outcome.