Early re-operations in a 5-year national cohort of congenital heart disease patients

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Objective: 1) To determine the quantitative burden of early congenital reoperations; 2) To evaluate if reoperation within the first 30 days is a suitable metric of quality of care.

Methods: Anonymised data on early reoperations were extracted from the UK National Institute for Cardiovascular Outcomes Research (NICOR) for 2005-2010.

Results: 19239 procedures were identified in 15552 patients. During data cleaning 723 re-operations were excluded (3.8%) and the remainder were adjudicated to predefined categories. 676 early reoperations (3.5%) were recorded in 593 patients, ranging from 1-7/patient (median 1/patient). The cases excluded a priori are likely under-represented as practices vary between centres (e.g. for reopening) and for submitting minor procedures data to NICOR. There are many complex scenarios where surgical teams choose operative adjustment (e.g. in palliative procedures) or planned/complex operative sequences to mitigate survival. For retained patients the median age and weight were 4.0 kg and 0.19 years and 18.2% of them were readmitted for reoperation. Independent risk factors were sought by multivariate analysis. The most common reoperations were in patients palliated by shunting.

Conclusions: Reoperations within 30-days are infrequent. Those that can be accurately included in a retrospective analysis are no more prevalent than death (3.2%). ‘Unplanned’ reoperation is a misnomer as only a minority can be classified as such. Prospective studies are under way to establish the definitions and true prevalence of reoperations. Until these studies are completed, inclusion of unplanned surgery on quality of care dashboards is debatable.

Cardiovascular compromise of Fabry disease

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Fabry disease is a progressive X-linked disorder of glycosphingolipid metabolism caused by a deficiency of the alfa-galactosidase lysosomal enzyme. The partial or complete deficiency of the lysosomal enzyme leads to accumulation of neutral glycosphingolipids in the vascular endothelium and visceral tissues throughout the body. In the heart, glycosphingolipids deposition causes progressive left ventricular hypertrophy (LVH). We can confirm the disease by demonstration of a low plasma alfa-galactosidase A (alfa-Gal A) activity. Also, diagnose and make follow-up of the patients with electrocardiogram, chest radiograph, echocardiogram, coronary arteriography or cardiac magnetic resonance imaging. On electrocardiogram we can find: prolongation of the QTc interval (>440 ms), widening of corrected QRS, left ventricular hypertrophy, bundle branch block, ativoventricular block, premature atrial contraction, premature ventricular contraction and Wolff-Parkinson-White syndrome. We can also make analysis of genomic DNA and observe alfa-Gal A gene mutation. The availability of enzyme replacement therapy (ERT) for this debilitating condition has led to the need for a deep knowledge from the pediatric and cardiology pediatric groups in order to diagnose, treat efficiently and rapidly, to improve the quality of life in pediatric patients with Fabry Disease.