## Theme: Pediatric Cardiology



## Prevalence of congenital heart disease from Northern India (Ann Pediatr Cardiol. 2016;9:205-9)

This large study (>20,000 newborns) provides important data about birth prevalence of congenital heart disease (CHD) as a whole, and details of individual CHDs. For the purpose of this study, significant CHD was defined as all major CHDs (any CHD that was likely to require intervention within the first year, including newborns with critical CHD that require intervention within the first 4 weeks of life) or specific forms of minor CHD (ASD >5 mm, PDA >2 mm with left ventricle volume overload, restrictive VSD, and valvular aortic/pulmonary stenosis with gradients <25 mmHg).

The birth prevalence of significant CHDs was 8.07 per 1000 live births, and approximately 20% of the defects identified at birth were cyanotic CHDs. Among the acyanotic defects, ventricular septal defect was the commonest; and among the cyanotic defects, transposition of great arteries (TGA) was the commonest. It is important to note that the birth prevalence of TGA was higher that tetralogy of Fallot (TOF). Given the very high mortality of untreated TGA (~90% in the first year), when compared to TOF (25% in the first year), it is not surprising that TOF seems a lot commoner among older patients with CHD.

The study underscores the massive burden of CHD among Indian children. From this prevalence data, it can be estimated that approximately 100,000 babies are born each year with "major" and "critical" CHD in India. The total number of operations in the limited centers in India that perform infant and newborn heart surgery can potentially take care of only about 10% of this disease burden. There is clearly an urgent need to build national capacity to take care of the remaining 90%.



## Late causes of death after pediatric cardiac surgery: A 60-year population-based study (*J Am Coll Cardiol.* 2016;68:487-98)

In the past 4-5 decades, the care of children with congenital heart disease (CHD) has been transformed by advances in pediatric cardiovascular care, especially pediatric cardiac surgery. Corrective surgery offers a prospect of survival with good quality of life for children born with critical life-threatening CHD. The excellent health systems and health records of Scandinavian nations provides a unique opportunity to examine the long-term outcomes after congenital heart surgery.

This study examines the results from a nationwide pediatric cardiac surgery database and Finnish population registry of 10,964 patients undergoing 14,079 operations at <15 years of age in hospitals in Finland from 1953 to 2009. A follow-up rate of 98% was achieved. An early mortality (<30 days) of 5.6% perhaps reflects the fact that the database

includes the earliest era of congenital heart surgery. The late mortality was 10.4%.

As expected, heart failure contributed to most deaths. This decreased significantly after 1990, presumably reflecting major advancements that have occurred in CHD surgery in the past three decades. Sudden death after surgery for atrial septal defect, ventricular septal defect, tetralogy of Fallot, and transposition of the great arteries decreased to zero following operations from 1990 to 2009. Deaths from neoplasms, respiratory, neurological, and infectious disease were significantly more common among study patients than controls. Pneumonia caused the majority of non–CHD-related deaths among the study population.

The results of this study should be viewed as a benchmark for the rest of the world, particularly low- and middle-income countries where comprehensive pediatric cardiac care has only now begun to take shape. Additionally, it is important to recognize that while patients operated for CHD have vastly improved outcomes, they cannot, by any means, considered as "cured".



## Coronary artery complications in Kawasaki disease and the importance of early intervention (JAMA Pediatr. [published online October 17, 2016]. doi:10.1001/jamapediatrics.2016.2055)

This meta-analysis of 16 comparative studies (Intravenous immunoglobulin (IVIG) alone *versus* Steroids plus IVIG) that met predefined criteria involving 2476 patients was carried out with the specific purpose of determining the effect of corticosteroid therapy in preventing coronary artery complications in patients with Kawasaki disease (KD).

This systematic review and meta-analysis reveals that corticosteroid therapy in addition to IVIG was associated with a significantly lower rate of coronary artery complication compared with intravenous immunoglobulin therapy alone, particularly among high-risk patients (OR 0.240; 95% CI 0.123, 0.467). The benefits were maximal for those with a relatively short duration of illness before corticosteroids therapy and progressively diminished as the duration increased. The duration of fever was significantly less in the corticosteroids group. There was no increase in the risk of adverse events as a result of the corticosteroid administration.

The impressive results in reduction of the most serious complication of KD argues strongly in the favor of routine administration of corticosteroids along with IVIG in all high-risk patients with KD.

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