CONGENITAL HEART DISEASE IN FETAL AND PEDIATRIC AUTOPSIES: A 15-YEAR RETROSPECTIVE REVIEW AT A SINGLE TEACHING HOSPITAL IN BANGKOK, THAILAND

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Background and aims: Congenital heart defects carry significant morbidity and mortality in the pediatric age group. This study determined the spectrum of congenital heart defects in our fetal and pediatric autopsy population, and the association with extra-cardiac malformations and chromosome abnormalities.

Methods: Autopsy reports over a 15-year period were reviewed for cases with congenital heart defects. Post-mortem findings were correlated with echocardiography records when available.

Results: Of 608 autopsies, 119 (19%) had congenital heart defects, with the greatest proportion in the neonatal group (53%). Persistent left superior vena cava and hypoplastic left heart syndrome were the most common defects. Extra-cardiac malformations were present in 41% of cases, the most common of which were dysmorphic features. One case each of CHARGE association and Meckel-Gruber syndrome were identified. Chromosome abnormalities were present in 18.5% of cases, the most common of which was trisomy 18. All cases had been identified pre-mortem. Prenatal echocardiography was available in 52 cases and there was 85% correlation between echocardiogram and cardiac autopsy findings. Defects missed by echocardiography were generally of mild severity. One case interpreted as hypoplastic left heart syndrome turned out to be double outlet right ventricle. Discordances between echocardiogram and autopsy cardiac findings did not impact patient management.

Conclusions: Autopsy examination is important to delineate the anatomy of cardiac defects as well as recognize extra-cardiac malformations for identification of possible genetic syndromes. This information can then be used for parental counselling as well as for internal assessment of the accuracy of pre-mortem imaging studies

References:

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2. Gilbert-Barness E, Debich-Spicer D. Congenital malformations of the heart. In: Gilbert-Barness E (ed.) *Potter's Pathology of the Fetus, Infant and Child.* Philadelphia PA: Mosby Elsevier; 2007: pp. 987-1024.

Category	Total	Found in isolated	As a part of complex	p
Septal defects	38	18	20	0.74
Atrial septal defect	7	6	1	
Ventricular septal defect	13	6	9	
Atrioventricular septal defect	8	4	6	
Patent ductus arteriosus	6	2	4	
Left ventricular outflow tract obstruction	36	14	22	0.23
Hypoplastic left heart syndrome	27	8	19	
Interrupted aortic arch	1	1	-	
Coarctation of aorta	5	4	1	
Bicuspid aortic valve	3	1	2	
Right ventricular outflow tract obstruction	24	14	10	0.41
Pulmonary stenosis	3	1	2	
Bicuspid pulmonic valve	4	2	2	
Ebstein anomaly	11	10	1	
Tricuspid stenosis	1	1	-	
Pulmonic atresia	4	-	4	
Tricuspid atresia	1	-	1	
Anomalous great vessels	34	8	26	0.003
Persistent left superior vena cava	26	4	22	
Vascular ring	2	2	-	
Major aortopulmonary collateral arteries	4	1	3	
Varices	1	1	-	
Right-sided aortic arch	1	-	1	
Conotruncal anomaly	20	10	10	1.00
Truncus arteriosus	11	6	5	
Tetralogy of Fallot	2	1	1	
Double outlet of right ventricle	7	3	4	
Connection anomaly	13	3	10	0.51
Transposition of great arteries	7	3	4	
Anomalous pulmonary venous return	6	-	6	
Other anomalies	11	5	6	
Endocardial fibroelastosis	7	3	4	
Absence of ductus arteriosus	1	1	-	
Ectopia cordis	1	1	-	
Cor triatriatum	1	-	1	
Dextrocardia	1	-	1	
Total	176	72	104	0.03

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