Successful Repair of Complete Atrio-ventricular Septal Defect at the Beginning of the Second Decade

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Abstract
Complete atrio-ventricular septal defects (CAVSD) are present in about 3% of children born with congenital heart pathologies. They usually require early surgical correction, mostly in infancy, and surgery is considered to be the gold standard. It is unlikely that anyone would survive beyond the first years without severe morbidity. However, we report a case of a Pakistani girl who underwent successful surgical repair of CAVSD at the age of 11.

Introduction
An estimated 2–3.5% of children born each year with congenital heart defects (CHD) have a complete atrio-ventricular septal defect (CAVSD), a third of whom have associated Down’s syndrome.\textsuperscript{1}

CAVSD constitutes an anomaly of endocardial cushion development that results in the absence of atrial and ventricular septa with a common annulus for the atrio-ventricular (AV) valves.\textsuperscript{2,3}

Surgical correction of CAVSD is the gold standard. With improved surgical and cardiopulmonary bypass technique, and advanced peri-operative care in the current era, infancy is recommended as the optimum time for the repair. However, we report a case of an adolescent girl with CAVSD who underwent successful surgical repair at the age of 11.

Case summary
An 11 year old girl with a height of 118 cm, weight of 18.1 kg, and a known case of Down’s syndrome, presented with recurrent respiratory tract infections, and bouts of dyspnea and cyanosis since birth. On physical examination, her pulse was regular at 81 bpm, blood pressure was 93/59 mmHg and peripheral cyanosis was evident. Pulse oximetry was done which revealed an $O_2$ saturation of 93%. $S_1$ and $S_2$ were audible with a pan-systolic murmur heard throughout the cardiac auscultatory areas. The patient was categorized as New York Heart Association (NYHA) Class I, and a chest X-ray was advised that showed cardiomegaly with increased lung markings due to congestion in the pulmonary vasculature.

Further workup involving transthoracic echocardiography (TTE) revealed a moderate-sized primum atrial septal defect (left to right shunt) with a large inlet ventricular septal defect (bi-directional flow, favouring a right to left shunt). A diagnosis of Rastelli type ACAVSD, along with severe pulmonary artery hypertension, due to the left to right shunt, was made. There was severe AV valvular regurgitation, and moderate pulmonary regurgitation. The diagnosis was confirmed upon right heart catheterization (RHC) that was performed for a hyperoxia study and to ascertain the reversibility of pulmonary arterial hypertension which was found to be reversible. RHC revealed $Qp/Qs$ ratio of 2.4:1 on room air and $Qp/Qs$ ratio of 4.25:1 on 100% $O_2$.

The patient was advised to undertake elective repair of the CAVSD. A median sternotomy was performed and bi-caval cardiopulmonary bypass was established. A right atriotomy allowed a visual confirmation of Rastelli type A CAVSD and a patent foramen ovale (PFO). Double patch closure of the septal defect was performed using the pericardium. The PFO was left open. Post-bypass aortic and pulmonary artery pressures were 86/40 mmHg and 36/12mmHg respectively. On TTE no residual ASD, VSD, or AV valve regurgitation was appreciated and the PFO was evident.

A TTE performed on the 4\textsuperscript{th} post-operative day showed a residual VSD with a bi-directional shunt mostly left to right. The flap-like PFO had bi-directional flow, mostly from left to right. There was mild mitral, tricuspid, and pulmonary regurgitation. Right and left ventricular systolic functions were 50% and 45–50% respectively.

Discussion
Complete atrio-ventricular septal defects arise as a result of abnormal development of the endocardial cushion, resulting in a large ostium primum atrial septal defect, a large...
ventricular septal defect in the upper muscular septum, and a common AV valve straddling the ventricular septum. This results in intercommunication between the chambers leading to significant left to right shunting which manifests as cardiomegaly. Pulmonary hypertension develops as a consequence leading to pulmonary vascular disease in the first year of life. Without surgical intervention, considered as the gold standard, most patients die within two to three years, usually from congestive heart failure or respiratory tract infections. The argument in favor of early repair is that it ensures that the correction is performed before pulmonary vascular changes become irreversible. Pulmonary vascular disease sets in within the first year of life in CAVSD patients. However, with natural CAVSD survivors into the second decade of life, such as in our case, pulmonary vascular disease, as a significant threat to life, is already ruled out because of possible pulmonary stenosis. Those survivors might also possibly have a relatively competent left ventricular valve. Late repair of CAVSD, well beyond infancy, would then not compromise patient survival as it might in patients who develop pulmonary vascular disease. By avoiding surgical CAVSD repair in such patients, the inherent difficulties and complications of cardiopulmonary bypass in a complex procedure in infants are avoided.

**Conclusion**

In conclusion, surgical repair of CAVSD beyond infancy can be considered as safe and effective as in infants. The strong advocacy for surgical repair in infancy should only be limited to patients who do not have a relatively competent left ventricular valve or natural pulmonary stenosis. We insist that other institutions should review the procedure in adolescents and young adults and carefully follow the results. Further studies could establish the definition of a ‘competent’ mitral valve to preclude surgery in infants.

**Ethical approval**

Consent was obtained.

**Conflict of interest**

No conflicts of interest have been declared by the author.

**Author contribution**

AA – Conception and design of study, acquisition and analysis of data, initial draft of the article.

NS – Draft and revising article for critical intellectual content.

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