

## **Dextrocardia with cardiac anomalies: a rare Case experience: chief surgeon on assistant side.**

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### **I. Introduction**

Every person when talked about in this world to locate his heart points towards the left side of his chest but what if the heart is on the right side? And that too with a complex heart disease. The situation would be tricky and difficult to manage for each and every one including the operating surgeon. Here in this study we present to you a series of 6 different cardiac lesions with Dextrocardia and our experience. The intra thoracic location of the heart with base to apex axis directed towards the right not because of any disease causing pulling and pushing but because of congenital malposition is known as Dextrocardia. Mostly Dextrocardia is a common incidental finding but its association with complex congenital cardiac defects make it a challenging task to reach a definitive diagnosis and then to surgically correct the defect. Because of this it has become the subject of interest for pathologist and embryologist also recently including surgeons.

### **II. Materials and methods**

All patients were admitted in Swai Man Singh hospital, Jaipur, Rajasthan, India and were thoroughly screened with all blood investigations, 2d echocardiography with colour flow imaging, X ray chest, ecg and even CT angiography in required cases. Informed consent was taken as per protocol of our hospital from each and every patient's parent in case of minor and from the patients itself if the patient was an adult. Patients were taken in between the period of 2016 to till date.

Presentation of cases

We had 7 different cases which we operated and would like to share our experience.

#### **Case no 1**

We had a case of 8 year old male with symptoms of dyspnoea on exertion, cyanosis, on and off fever and palpitation was diagnosed with Dextrocardia, Situs Ambiguous with right isomerism, and Double Outlet Right Ventricle with complete A-V canal defect and polydactyly in both hands and feet. Diagnosis was proven by x ray chest, ecg, 2 d echocardiography and CT pulmonary angiography which stated Right sided aortic arch, inlet VSD of 24mm, common A-V canal, both the great vessels arising from right ventricle, Aorta on the left to pulmonary artery and slightly anterior with severe valvular pulmonary stenosis (70mmhg), CT pulmonary angiography stated main pulmonary artery is small in calibre in sub pulmonary region (diameter 6.5mm), right pulmonary artery (7.8mm) and left pulmonary artery (10mm). Patient was taken up for surgery and bidirectional Glenn shunting (superior vane cava to left pulmonary artery was anastomosed) was done.

#### **Case no 2**

One case we had of 7 year male came to us with complaints of dyspnoea on exertion and cyanosis and was diagnosed to be a case of Dextrocardia, situs solitus, CCTGA, L-loop ventricles, subaortic VSD, severe pulmonary stenosis, mapcas. Diagnosis was confirmed with 2 d echo and CT pulmonary angiography which stated it to be a case of Dextrocardia, Situs Solitus, L-loop ventricles, CCTGA, right side aortic arch, AV-VA discordance, subaortic VSD, severe pulmonary stenosis. Patient was taken up for surgery and procedure involved pulmonary valvotomy, VSD closure with DVD patch. Patient came difficult off bypass.

#### **Case no 3**

One case we had of 12 year female girl who came to us with complaints of bluish discoloration and breathlessness since early childhood, occasional complaints of cold and cough, she was vitally stable and had cyanosis on examination. Patient was diagnosed on 2d echo to be a case of Dextrocardia, Situs Solitus, CCTGA, VSD, severe pulmonary stenosis, AV-VA discordance. Initially patient was planned for Sennings procedure and

a conduit from right ventricle to pulmonary artery but on table we found opening of pulmonary veins in right atrium (papvc) so Fontans procedure was planned and intra cardiac PTFE conduit baffle inside right atrium connecting IVC to SVC to RPA was created. The other SVC end also anastomosed to RPA. Patient was smooth off bypass.

#### **Case no 4**

Another case we had of 22 year female who came to us with dyspnoea on exertion, cyanosis and weakness and was diagnosed to be a case of Dextrocardia with tricuspid atresia. The diagnosis was confirmed with 2d echocardiograph ,x ray chest, ecg and CT pulmonary angiography which stated Dextrocardia ,SitusSolitus, Tricuspid atresia ,Atrial septal defect, Ventricle septal defect ,severe pulmonary stenosis(80mmhg),right sided aortic arch. Patient was taken up for surgery and Fontan procedure with intracardiac PTFE conduit baffle inside right atrium connecting from IVC to SVC to right pulmonary artery was created. Other end of SVC was anastomosed to right pulmonary artery .patient was smooth off bypass.

#### **Case no 5,6**

2 cases we had were of mitral stenosis who came to us with complaints of dyspnoea on exertion. Diagnosis was confirmed by x ray chest , ecg and 2d echo which stated SitusSolitus ,Dextrocardia, sinus rhythm , mitral stenosis with no clots, no mitral regurgitation ,no atrial fibrillation with Wilkinsons score to be less than 8 with no calcification .both these 2 patients were taken up for closed mitral valvotomy through right thoracotomy and mitral valve was opened to area around 3.5 cm. Patients recovered well in post-operative period and are in follow up .

### **III. Discussion**

Dextrocardia was first recognised in 1643 by Marco severino<sup>1</sup>.3 different cardiac malposition with Dextrocardia are SitusSolitus with Dextrocardia, SitusInversus with Dextrocardia , Situs Ambiguous with dextrocardia .Situs means position/site. Situs refers to Atrias and other body viscera such as Lung , Liver, Spleen, Gastrointestinal tract. Situs can be Solitus ,Inversus , Ambiguous. Solitus means usual/normal, inversus means opposite/reverse, ambiguous means uncertain/indeterminant<sup>2</sup>.situs solitus means morphologic right atrium is on the right of the morphologic left atrium,liver is on right stomach and spleen are on the left side,right lung is trilobed and an eparterial bronchus whereas left lung is double lobe with hyperarterial bronchus<sup>3</sup>. Situsinversus means the morphologic right atrium is to the left of the morphologic left atrium, liver is on the left side of the body, stomach and spleen on the right side of the body, the left lung is trilobed with an eparterial bronchus and right lung is bilobed with an hyperarterial bronchus<sup>3</sup>.situs ambiguous which is also known as heterotaxy means unorganised or uncertain arrangements that means the relationship between atria and viscera is inconsistent. Situs ambiguous can be of 2 types that is asplenia syndrome (right isomerism )orpolysplenia syndrome( left isomerism)<sup>3</sup> .

Incidence of dextrocardia is believed to be around 1 in 12000 people<sup>4</sup>. In our study we used x ray chest , 2d echocardiography ,colour flow imaging and in some cases ct angiogram also to study the cardiac anatomy and haemodynamic using segmental analysis of the patients with dextrocardia and to share our experience of different cardiac lesions with dextrocardia.as per my knowledge and research this is the second series on dextrocardia with different cardiac lesion in India and this part of the continent till date using 2d echo and colour flow imaging , also may be the largest series involving CT angiogram also till date . In our study situsolitus with dextrocardia was the commonest (83.33%),situsambiguous with dextrocardia(16.66%).

(A) Dextrocardia with Situsolitus: this condition is having incidence close to 100% to be associated with CHD<sup>5</sup> but in our study the incidence was of 60% of CHD. In our study 40% under this category didn't have CHD which is contrast to the study done in 203 on 125 patient having dextrocardia which had 7% of incidence of not having CHD<sup>5</sup>.we had 5 cases under this category out of which 2 cases were of rheumatic heart disease with mitral stenosis in which we underwent closed mitral valvotomy as we already discussed in cases study above.1 case we had of dextrocardia with situsolitus with tricuspid atresia,pulmonary stenosis with DOLV with large OS ASD( R-L SHUNT) WITH AV-VA CONCORDANT, another 2 cases we have of dextrocardia with situsolitus with cctga ,vsd,severeeps with av-vadisconcordant , so discordant avconnecons were present in 66.66% and all cases had cctga . av-va concordant was present in 33.33% which is in contrast to the study done in 2003 of 125 patients which had av discordant in 41.9% and av connection concordant in 51.2%<sup>5</sup>. Out of the above 3 CHD cases 1 patient had d-loop and 2 patients have l-loop. All the above cases are rare in their entity.

(B) Dextrocardia with Situs ambiguous: we had a case of 8 year old male diagnosed to be dextrocardia, situs ambiguous with right isomerism,commona-vcannal defect with DORV ,polydactyly in both hands and feet . Patient had severe pulmonary stenosis and inlet VSD ,ostiumprimumASD,main pulmonary artery and

bronchus were hypoplastic. We planned and did glenn shunting. Finally we lost the patient post operatively. Case was a rare entity with polydactyly in both hands and feet.

Out of 6 cases we lost 2 patients in post operative period and a mortality rate of 33.33%. Still we were able to save 4 patients with a successful rate of 66.66%. In a developing centre for complex congenital cardiac surgery cases that too with Dextrocardia which itself is a challenge to operate as chief surgeon is on assistant side and due unusual cardiac anatomy we feel we had good successful rate and hope to improve it further. All above cases were rare in their entity and also the series itself is rare in this part of world till date. Study was just to present our experience at our centre with Dextrocardia in different cardiac lesions.

#### **IV. Conclusion**

dextrocardia is a rare entity and a challenge to operate with different cardiac lesions including complex congenital cases and it needs to be thoroughly investigated and needs expert hands and experience to deal with .

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