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The chest x-ray in congenital heart disease 2

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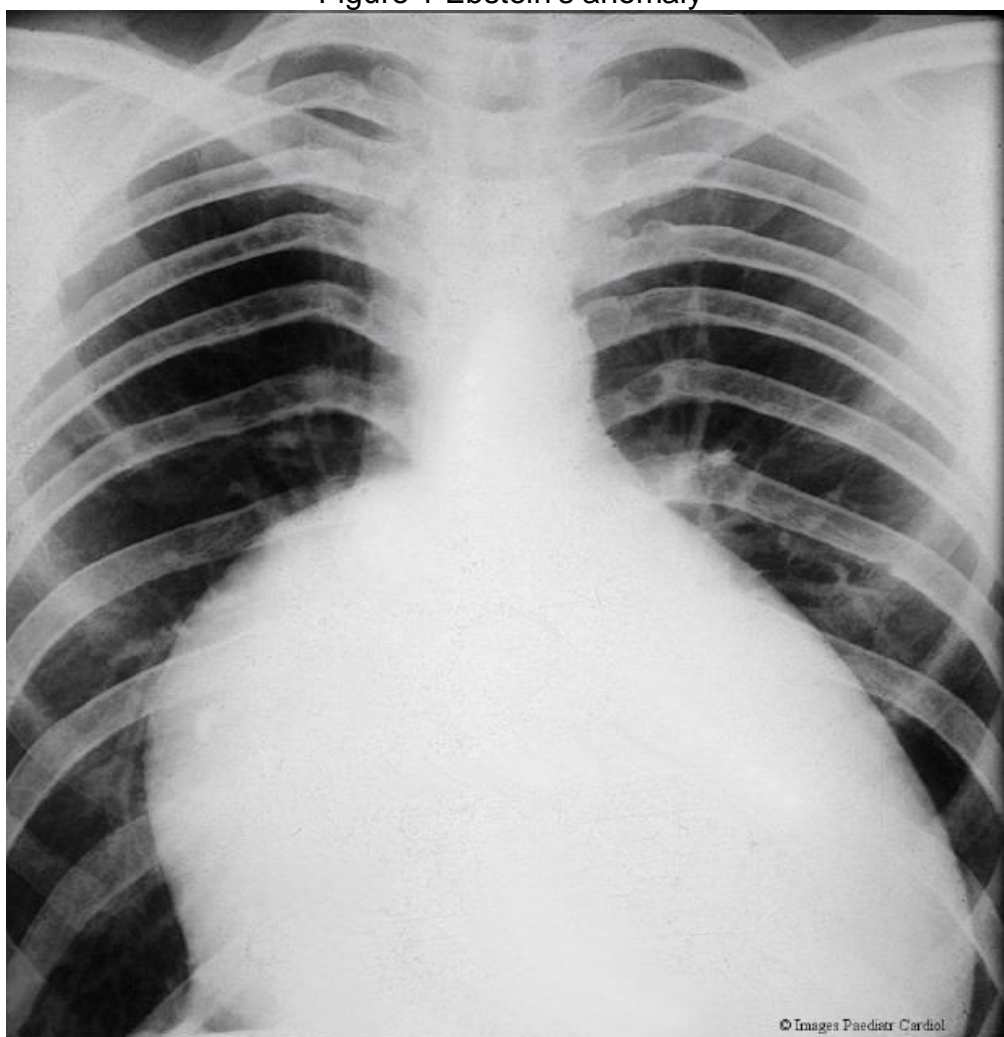
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This paper is the second of our series focusing on the chest x-ray (CXR) as a useful investigation in congenital heart disease.¹ We remind the readers that the CXR is a simple, quick and cheap test that yields useful diagnostic information, heart size, lung pathology as well as providing a readily available documentation of these facts for serial comparison.

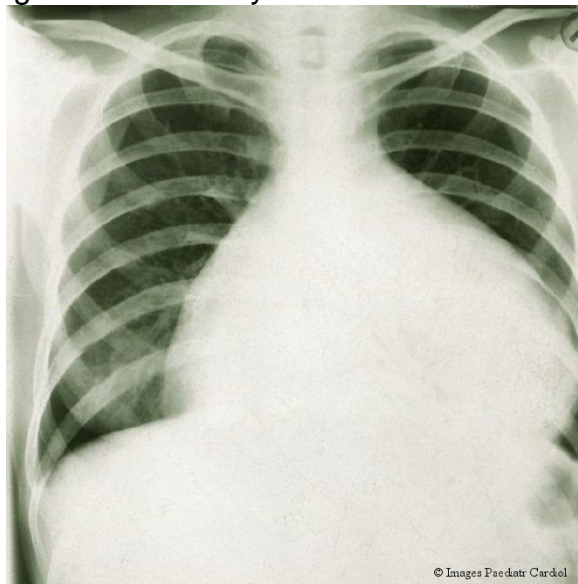
Ebstein's anomaly showing the classic appearance of cardiomegaly, small aortic knuckle, small pulmonary arteries giving the appearance of underfilled lungs, dilated inferior vena cava and huge right atrium. The right ventricular outflow is prominent and pushed to the left by the dilated right atrium, which is dilated due to tricuspid regurgitation.

Figure 1 Ebstein's anomaly



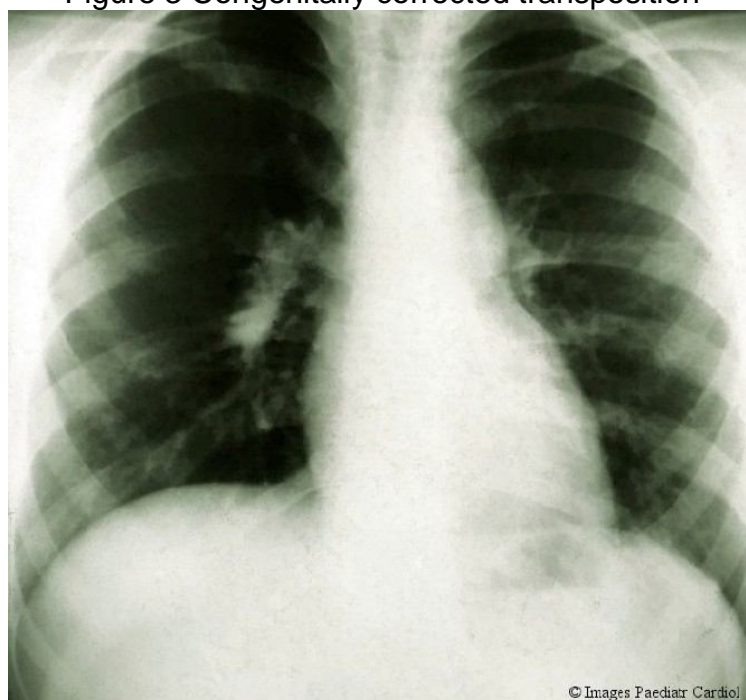
Huge cardiomegaly, oligoemic lungs, dilated right ventricular outflow and right ventricle: this is the classic appearance of pulmonary stenosis in heart failure and it is difficult to distinguish from chronic pericardial effusion but the absence of left sided congestion or dilated pulmonary veins suggests pulmonary stenosis.

Figure 2 Pulmonary stenosis in heart failure



Congenitally corrected transposition with the aorta in the left position where the pulmonary artery should be. The identification of the ascending aorta is made by the bulge in the pulmonary artery smoothly running into the aortic knuckle at the left upper edge of the mediastinum. The right pulmonary artery is slightly dilated and this suggests that pulmonary stenosis could be present. Note that post stenotic dilatation involves the right and not the left pulmonary artery when the great arteries are malposed or transposed.

Figure 3 Congenitally corrected transposition



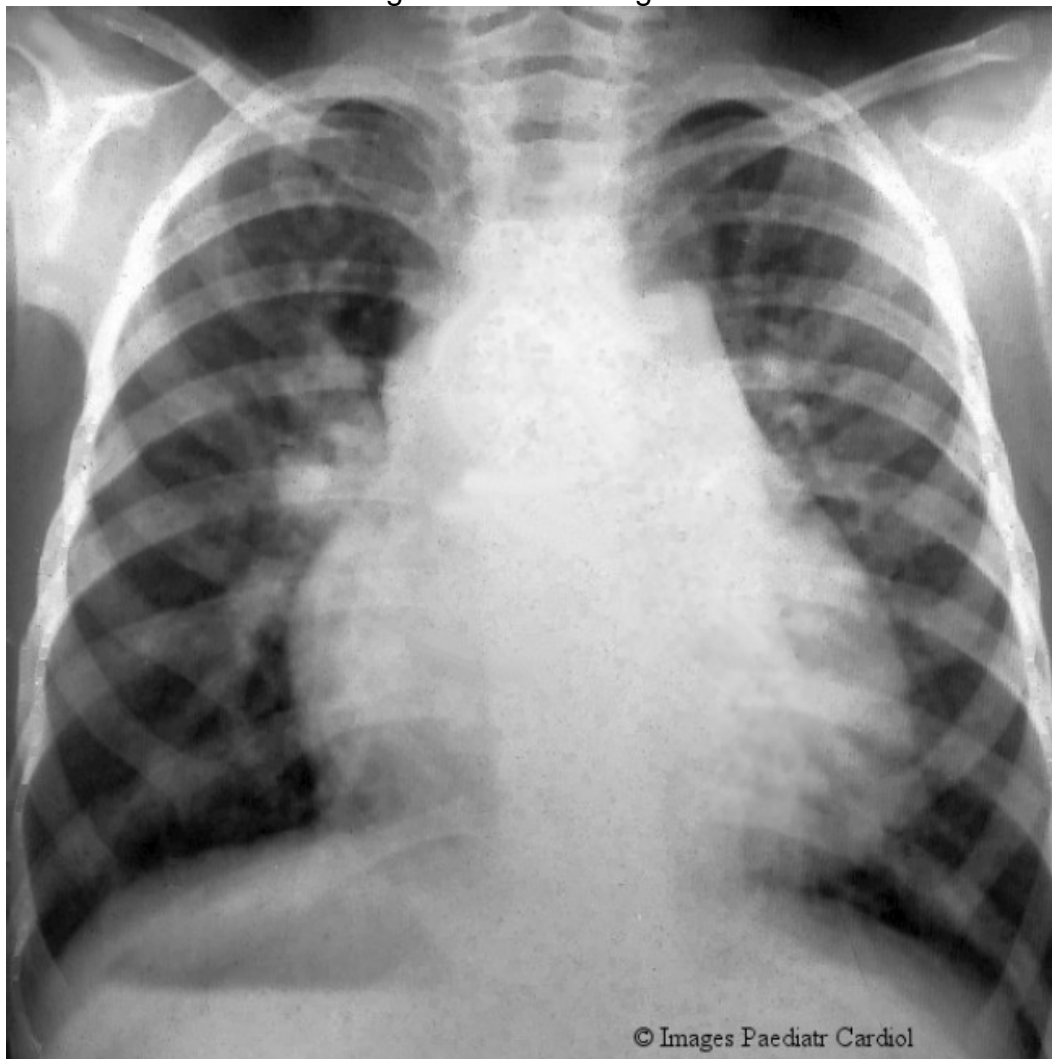
Angiogram showing the position of the aorta and subaortic right ventricle in corrected transposition as seen in figure 3.

Figure 4 Congenitally corrected transposition - angiogram



Cardiomegaly and pulmonary plethora implies a large left to right shunt from a congenital defect. Note that the stomach bubble is under the right diaphragm which signifies liver on left as is the apex of the heart. This has several names: isolated levocardia or situs ambiguus. Its importance is that it is associated with certain intracardiac lesions such as atrioventricular defects and single atrium. In addition there is evidence of venacaval abnormalities shown by the bulge on the upper right mediastinal border in the region of superior vena cava/right atrial junction which is the dilated inferior vena cava end on joining at that point instead of lower right atrium. There is another bulge on the left upper border of the mediastinum where the pulmonary artery is expected and in the region of straight shadow of probable left superior vena cava, which is a left hemiazygos (like a left inferior vena cava joining from the abdomen) and this forms the left bulge. Anomalies of systemic venous drainage are common with single atrium. The diagnosis of left isomerism would be made were the bronchi visible as these vena caval anomalies are associated with left isomerism.

Figure 5 Situs ambiguus



A patient with tricuspid atresia, transposition and pulmonary stenosis who has had a Glenn shunt 20 years previously, and this has formed fistulous communications in the right lower lobe. The left pulmonary artery is dilated as the result of a Blalock shunt.

Figure 6 Tricuspid atresia, transposition and pulmonary stenosis post Glenn



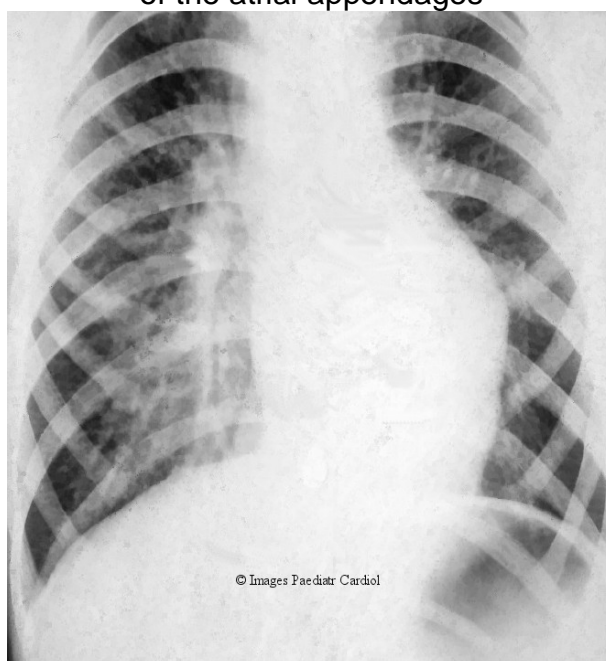
Classic appearance called coeur en sabot once said to suggest Fallot's tetralogy. In fact it is often seen with pulmonary atresia with ventricular septal defect. In this picture there is a right aortic knuckle from the arch on that side and deflecting the trachea to the left. The upturned apex making the point of the sabot is the right ventricle. There is a bay where the pulmonary artery should be seen but is absent. The pulmonary artery distribution is abnormal with small vessels.

Figure 7 Pulmonary atresia with ventricular septal defect



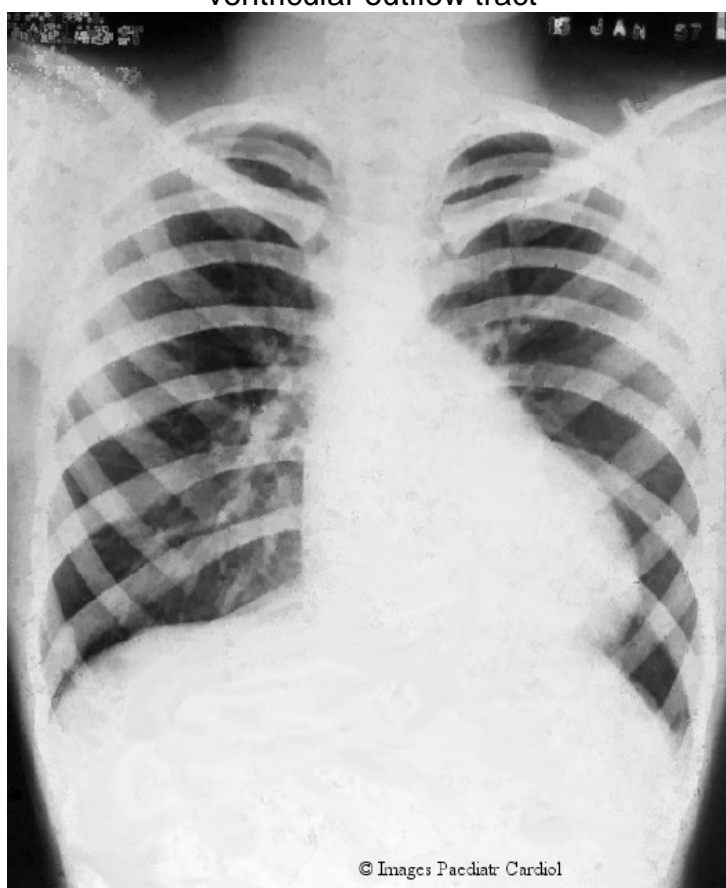
Classic appearance of tricuspid atresia and transposition of great arteries with juxtaposition of the atrial appendages, which give rise to pointed bulge on the left border of the mediastinal shadow below the region where the pulmonary artery should be seen. The right border of the right atrium is straight because the absence of the right ventricle pulls the border toward the right atrium. The mediastinal pedicle is narrow because there is transposition of the great arteries. The vascularity of the lungs is normal because the pulmonary stenosis is not severe.

Figure 8 Tricuspid atresia and transposition of great arteries with juxtaposition of the atrial appendages



This patient has a sternal depression which pushes the heart to the left, displacing the apex leftwards. There is an accompanying straight right border of heart shadow in the region of the pulmonary artery. On left border, there is unusually prominent left bulge and this is due to a pericardial defect which permits prolapse of the right ventricular outflow and main pulmonary artery through it.

Figure 9 Sternal depression and pericardial defect with prolapse of the right ventricular outflow tract



References

1. Somerville J, Grech V. The chest x-ray in congenital heart disease 1. Total anomalous pulmonary venous drainage and coarctation of the aorta. Images Paediatr Cardiol. 2009;38:7–9.

