

# Multimodality Imaging and 3-D Model of Anatomically Corrected Malposition of the Great Arteries (S,L,D)

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## Abstract

Anatomically corrected malposition of the great arteries (ACMGAs) is a rare condition characterized by abnormally related great arteries that do, despite their abnormal position, arise from the anatomically correct ventricles. We describe a neonate with ventricular inversion, an anterior aorta, and a ventricular septal defect (VSD) who underwent multi-modality imaging, including a 3-D printed model, which clarified the complex segmental anatomy as a rare subtype of ACMGA – the S,L,D heart – based on Van Praagh's classification system.

## Introduction

We present multi-modality images of a rare form of anatomically corrected malposition of the great arteries, an S,L,D heart, by Van Praagh's classification system: S = normal abdominal and atrial situs; L = L-looping of the ventricles resulting in ventricular inversion; D = dextro-malposition of the aorta.

Our case demonstrates a very rare situation, in which the L-looped ventricle is NOT associated with L-TGA.

## Clinical Summary

A cyanotic neonate was initially diagnosed with complex TGA, a VSD, and suspected ventricular inversion by echocardiography (Figure 1).

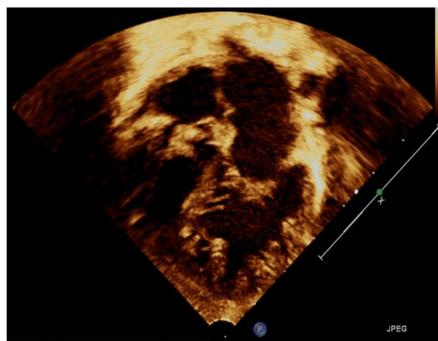


Figure 1. Apical 4-chamber view by 2D echocardiography demonstrates anatomy consistent with L-looped ventricles. There is normal atrial situs with a right-sided, morphologic right atrium, connecting to a smooth-walled ventricle with an AV valve with no septal attachments, consistent with a mitral valve and morphologic left ventricle. The left-sided, morphologic left atrium connects to a trabeculated ventricle with a more apically positioned AV valve with chordal septal attachments, consistent with a tricuspid valve and morphologic right ventricle.

## Clinical Summary

To better define the anatomy and plan surgical intervention, cardiac magnetic resonance imaging (MRI) was obtained.

This confirmed L-looping of the ventricles with a left-sided, morphologic right ventricle giving rise to a pulmonary artery.

The MRI also confirmed the right-sided, morphologic left ventricle, giving rise to the aorta with a hypoplastic arch (Figure 2).

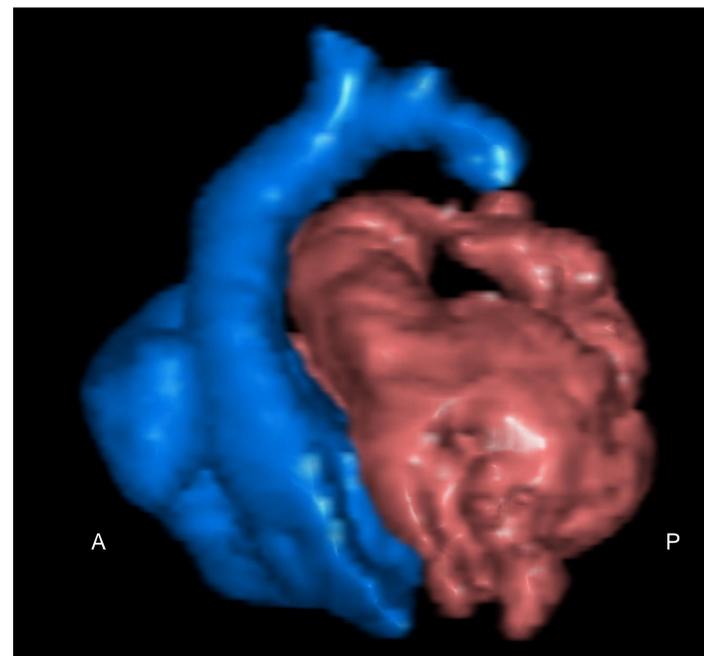


Figure 2. 3D reconstruction from cardiac MR confirms the echocardiographic findings. The right-sided ventricle (blue) is somewhat hypoplastic and smooth walled, consistent with a morphologic left ventricle, and gives rise to an aorta. The left-sided ventricle (red) is trabeculated and gives rise to a pulmonary artery. These findings are consistent with a rare S, L, D segmental relationship.

A = anterior; P = posterior.

A 3-D printed model of the patient's heart was generated from the MRI data and allowed for a more detailed understanding of the intracardiac anatomy (Figure 3).

## Clinical Summary

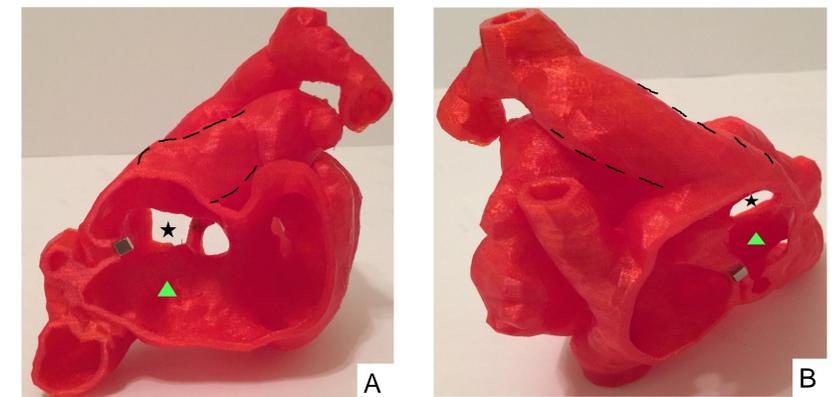


Figure 3. 3D printed model constructed from MR data.

(A) Left-sided morphologic right ventricle free wall is cut away exposing the right ventricular cavity (green triangle) and the large VSD (black star). The right ventricle gives rise to the pulmonary artery (dashed outline).

(B) Right-sided morphologic left ventricle free wall is cut away exposing the hypoplastic left ventricular cavity (green triangle) and the large VSD (black star). The left ventricle gives rise to the aorta (dashed outline).

The patient underwent successful aortic arch reconstruction with resection of abnormal (right-sided) mitral valve tissue and pulmonary artery banding as a palliative step to allow growth and plan for final surgical repair.

## Conclusion

Multimodality imaging allows us to understand this unique and complex congenital anatomy and properly plan surgical repair.

## References

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