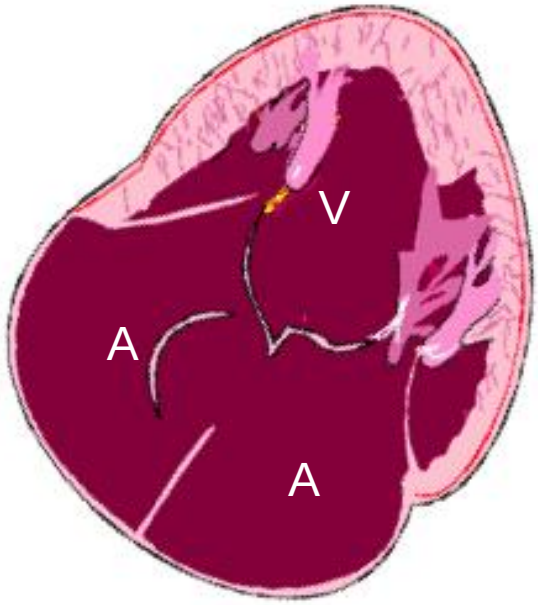
Single ventricle

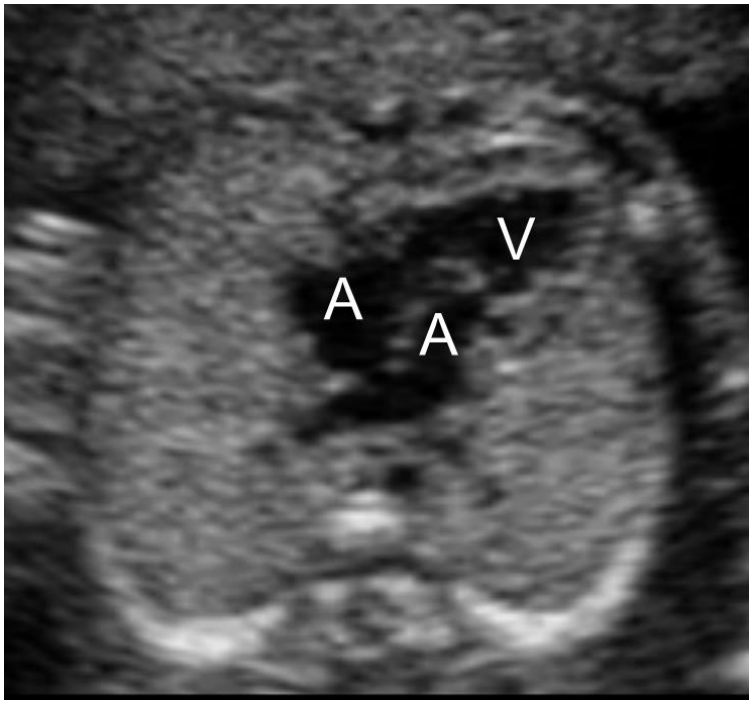
Double inlet single ventricle

Single inlet single ventricle

This is a group of anomalies in which the entire atrioventricular junction is connected to only one chamber in the ventricular mass; anatomy is variable but there are mainly two types: double outlet single ventricle and atresia of one atrioventricular valve (or single inlet single ventricle)

Double inlet single ventricle





In *double inlet* single ventricle each atrium has a separate atrioventricular valve and both empty into a single ventricle;

Images by Gianluigi Pilu and Philippe Jeanty

Double inlet single ventricle: the apical view demonstrates two atrioventricular valves open into one single ventricular cavity

Clip SV_01

Images by Gianluigi Pilu

Double inlet single ventricle: color Doppler demonstrates blood flow from the right and left atrium into a single ventricle

Clip SV_09

Images by Paolo Volpe

Double inlet single ventricle

Clip SV_15

Images by Paolo Volpe

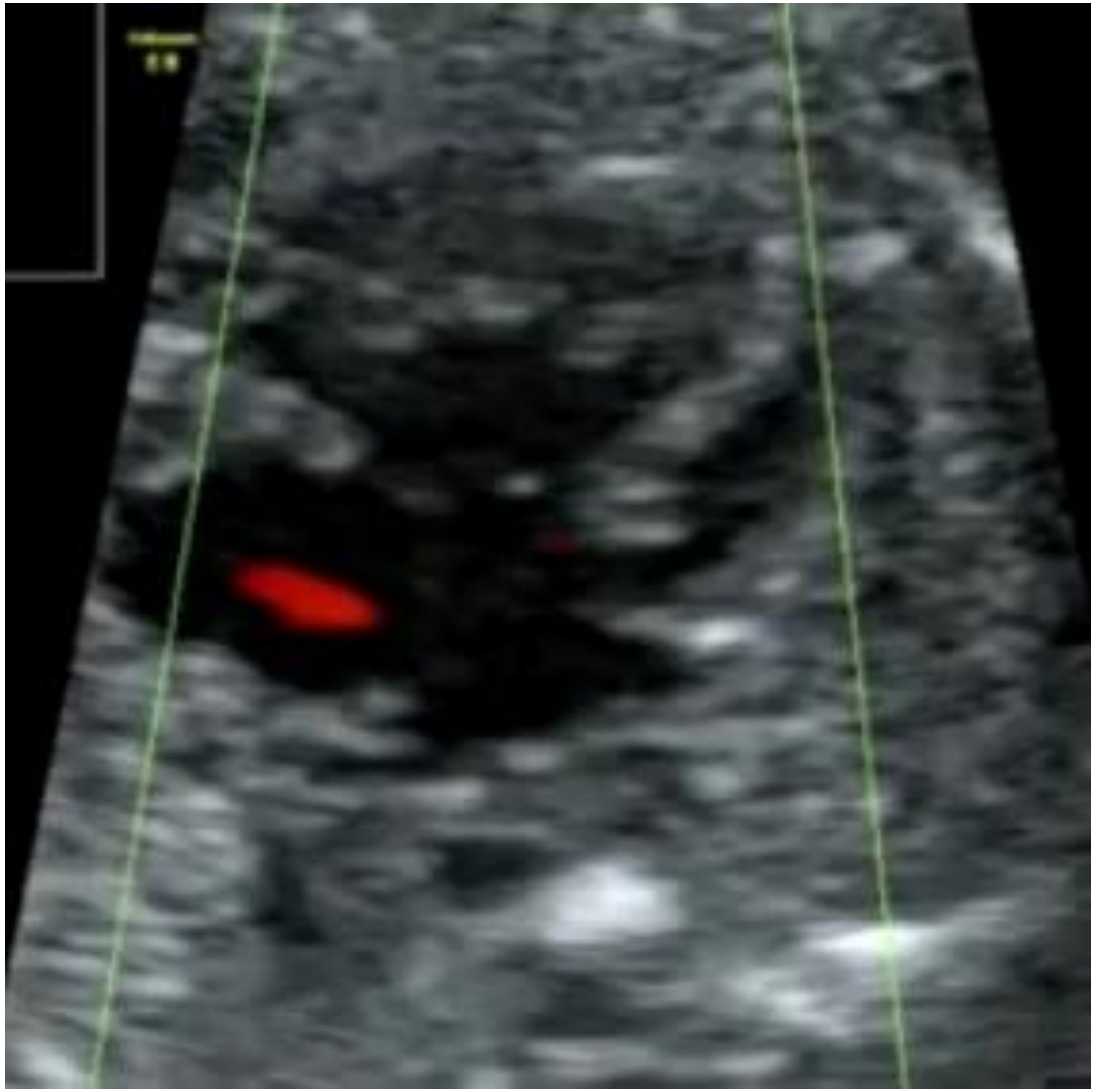
A single inlet with a common atrioventricular valve is also possible. In this case of unbalanced atrioventricular septal defect note the small size of the left ventricle

Clip SV_11

Images by Paolo Volpe

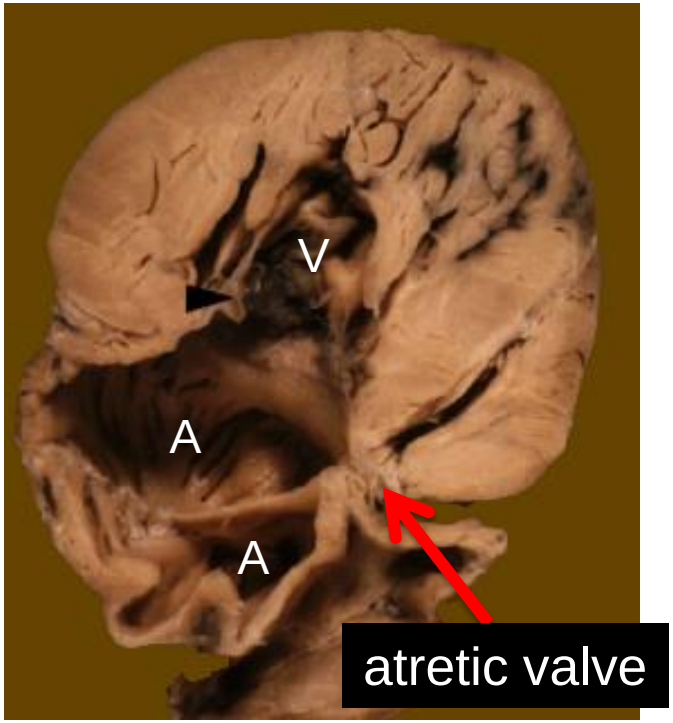
Same case of the previous image. Color Doppler highlights the small size of LV

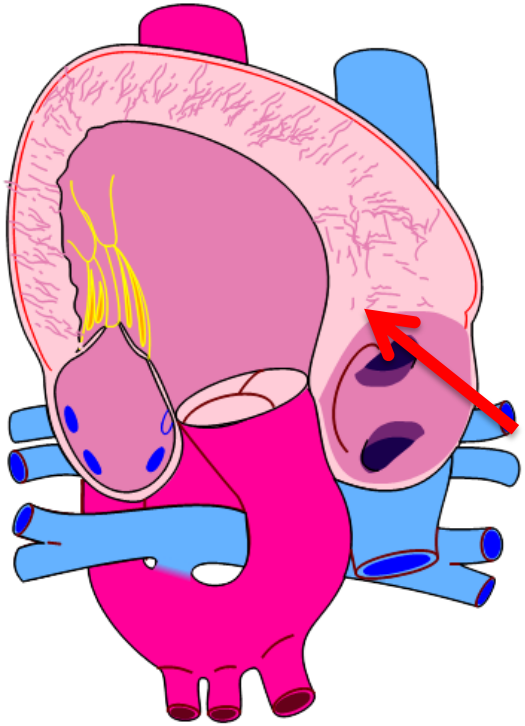
Clip SV_12

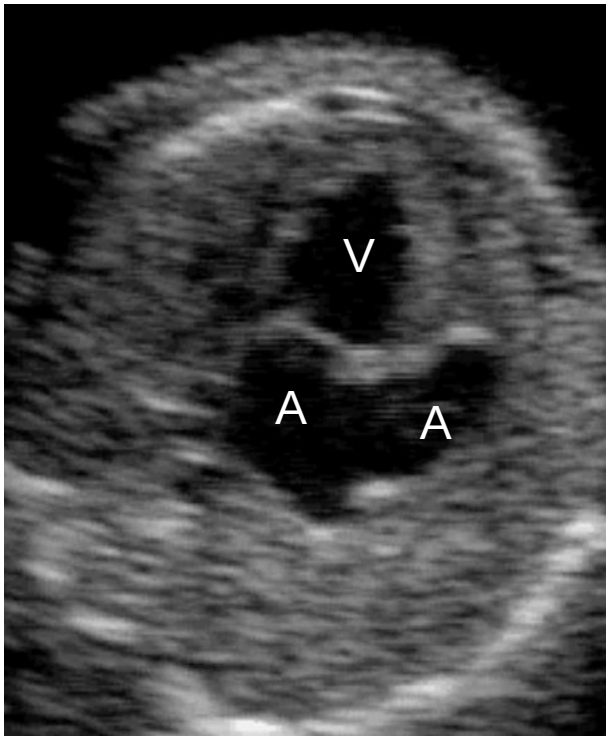


Images by Paolo Volpe

Single inlet single ventricle



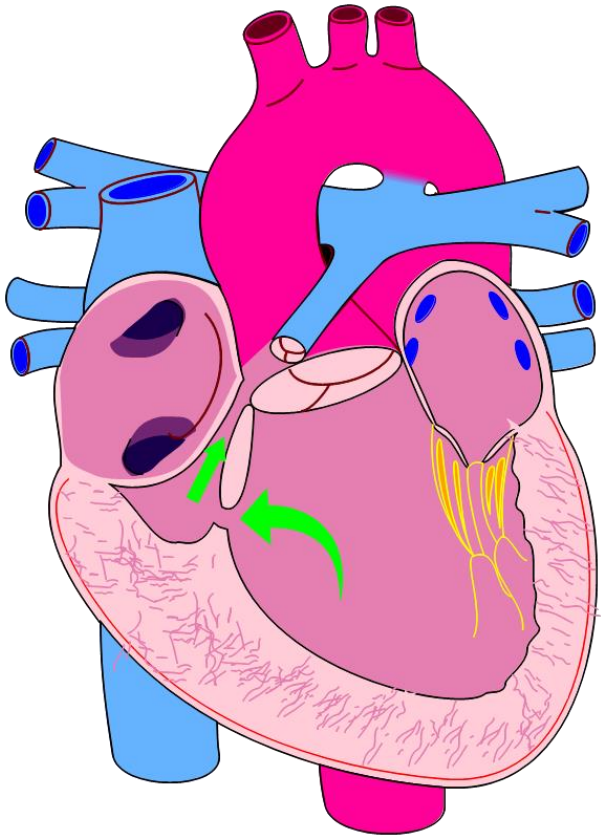


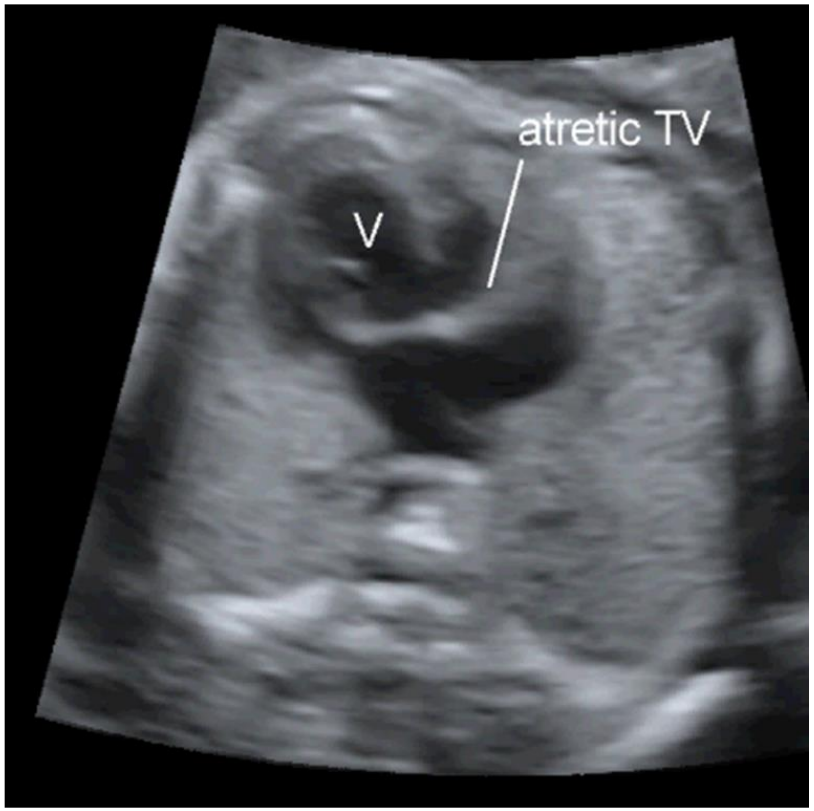


In *single inlet* single ventricle one atrioventricular connection has not developed, is atretric and the relative ventricle is absent or severely underdeveloped; there is only one patent atrioventricular valve and one functional ventricle and the two atria communicate via the foramen ovale

Images by Gianluigi Pilu and Philippe Jeanty

Tricuspid atresia

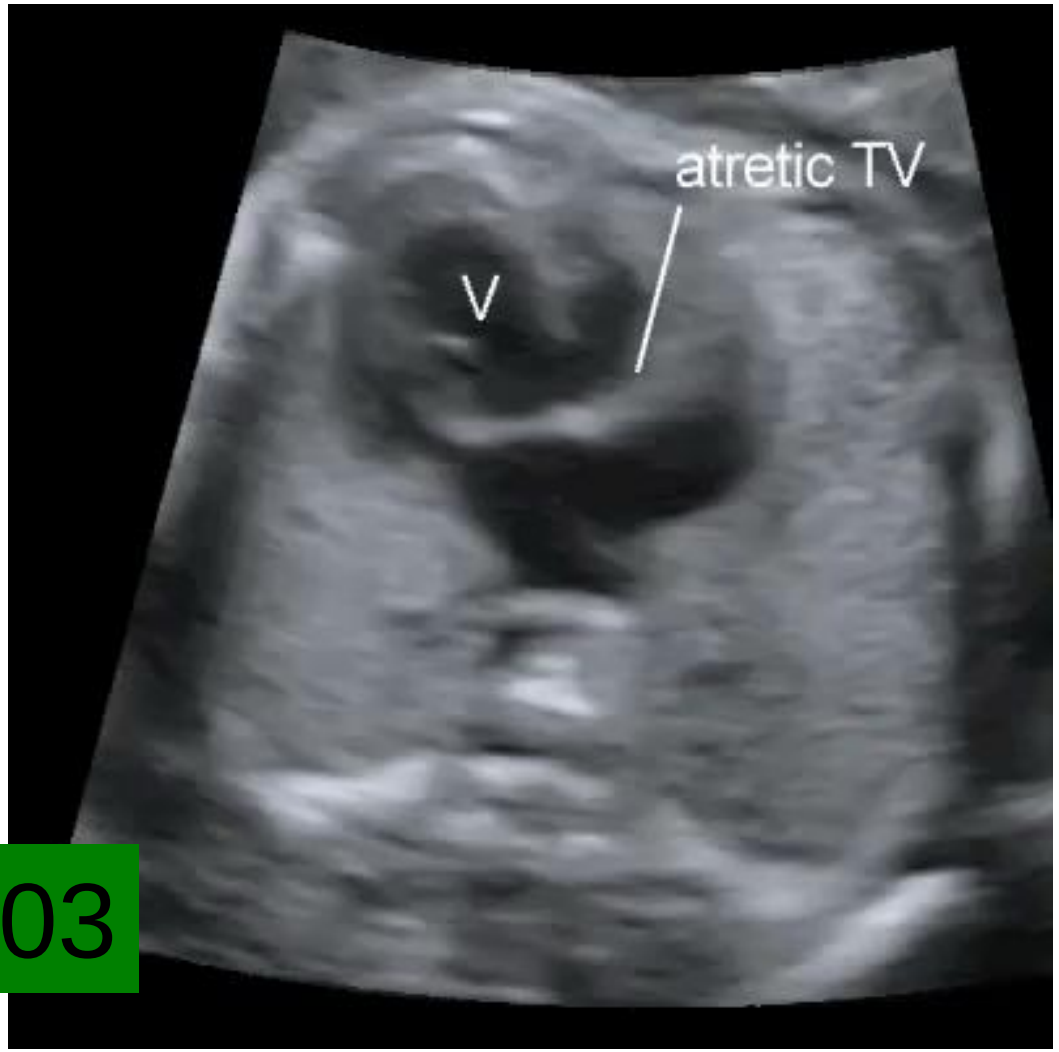




Many different variations of single ventricle anatomy are encountered, but the most common is *tricuspid atresia*; the functional ventricle is of left type, with a rudimentary right ventricular cavity that lacks connection with the atria but usually communicates with the main ventricular chamber through a ventricular septal defect.

Images by Gianluigi Pilu and Philippe Jeanty

Tricuspid atresia: the tricuspid valve is typically fixed, thick and echogenic; a small ventricular chamber is seen



Clip SV_03

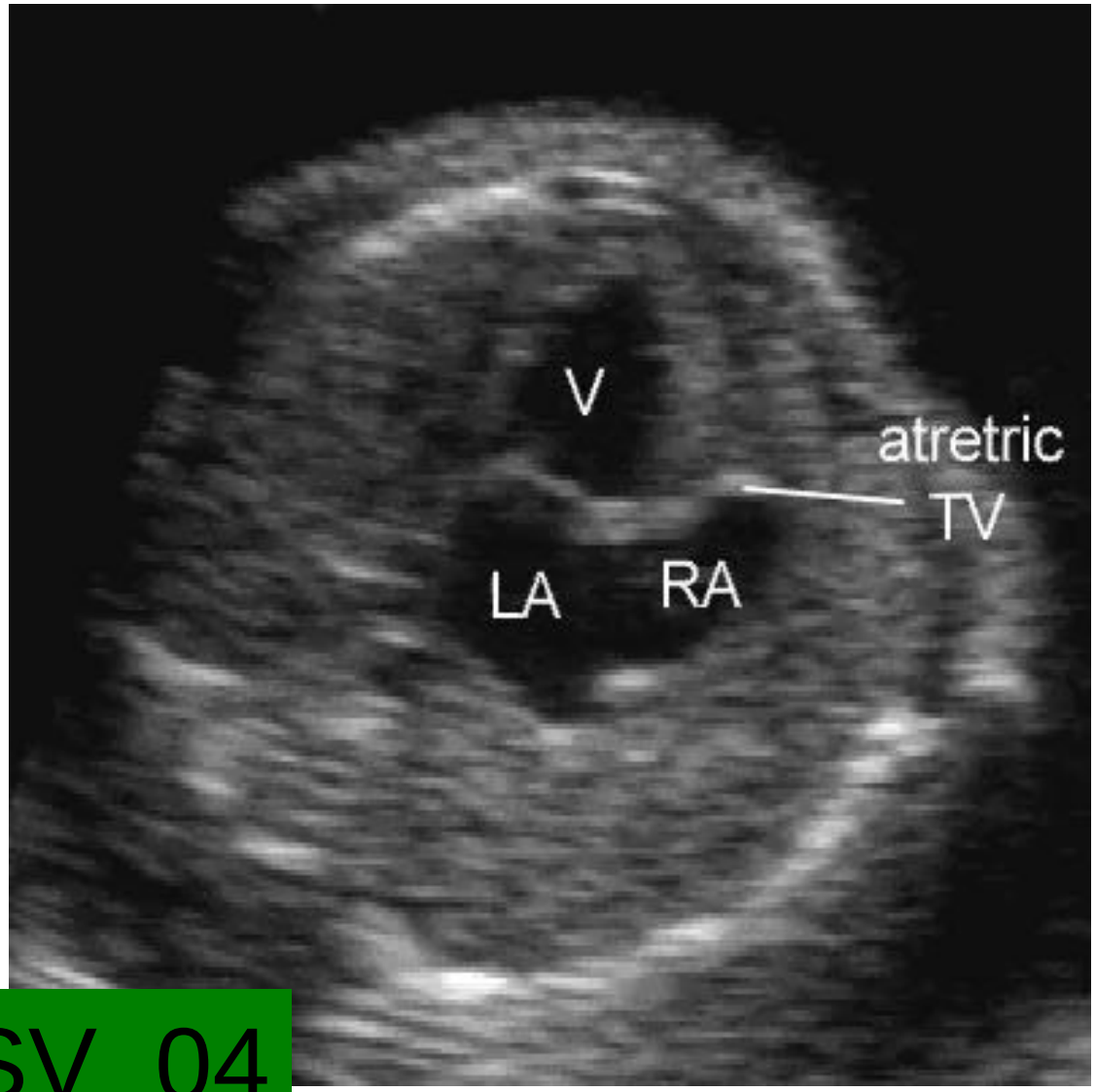
Images by Gianluigi Pilu

Tricuspid atresia: color Doppler shows blood flow passing across the mitral valve and demonstrates absence of flow across the tricuspid valve

Clip SV_10

Images by Paolo Volpe

Tricuspid atresia: in a minority of cases the right ventricle is a virtual cavity and can not be seen; this is usually seen in association with pulmonary atresia and/or in case of restrictive VSD



Clip SV_04

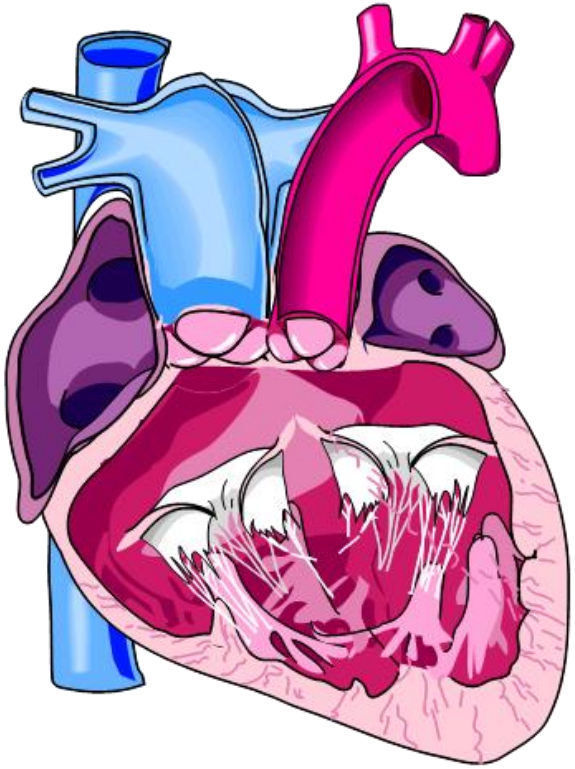
Images by Gianluigi Pilu

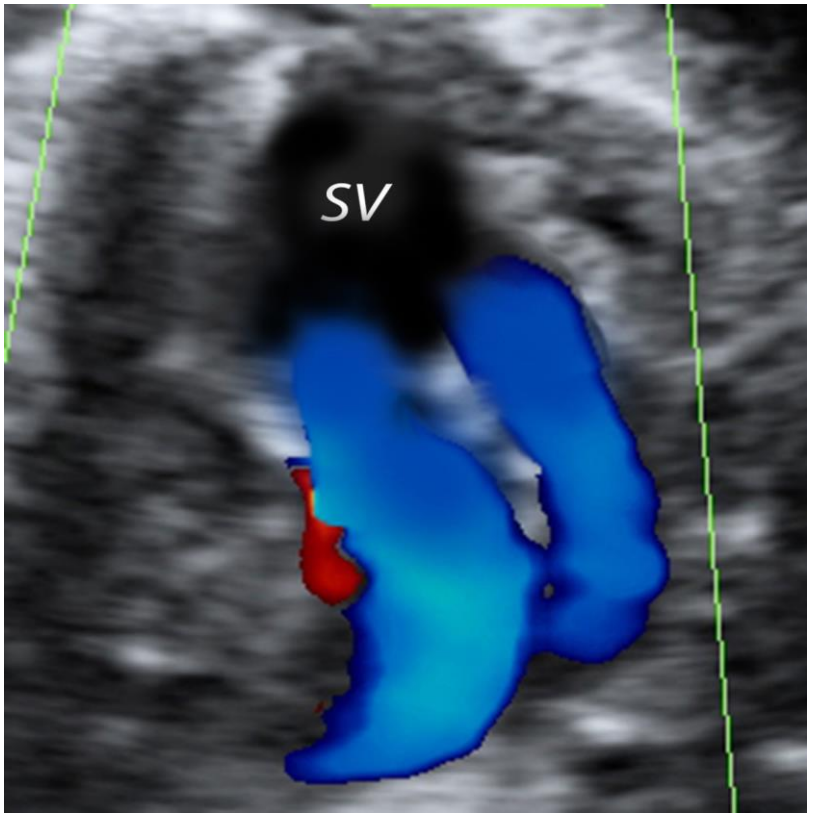
A rare variety of single ventricle: mitral atresia with a small left ventricle and a dominant right ventricle

Clip SV_05

Images by Gianluigi Pilu

Great vessels with single ventricle





With single ventricle the great vessels are frequently transposed. Outflow obstruction, either pulmonary or aortic, is frequently seen and has a major influence on the final outcome.

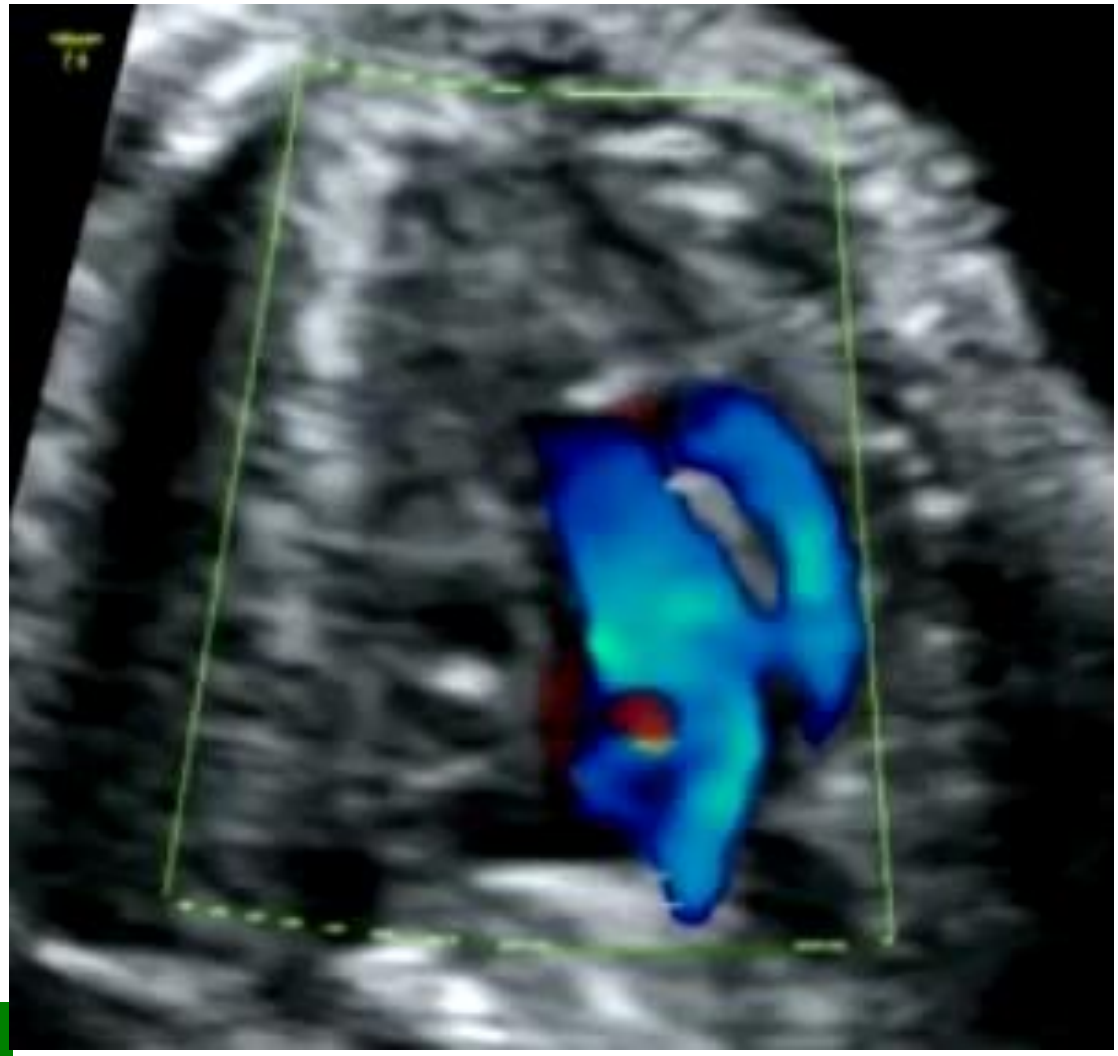
Images by Paolo Volpe and Philippe Jeanty

Double inlet single ventricle with two vessels of normal size arising in a parallel fashion

Clip SV_09

Images by Gianluigi Pilu

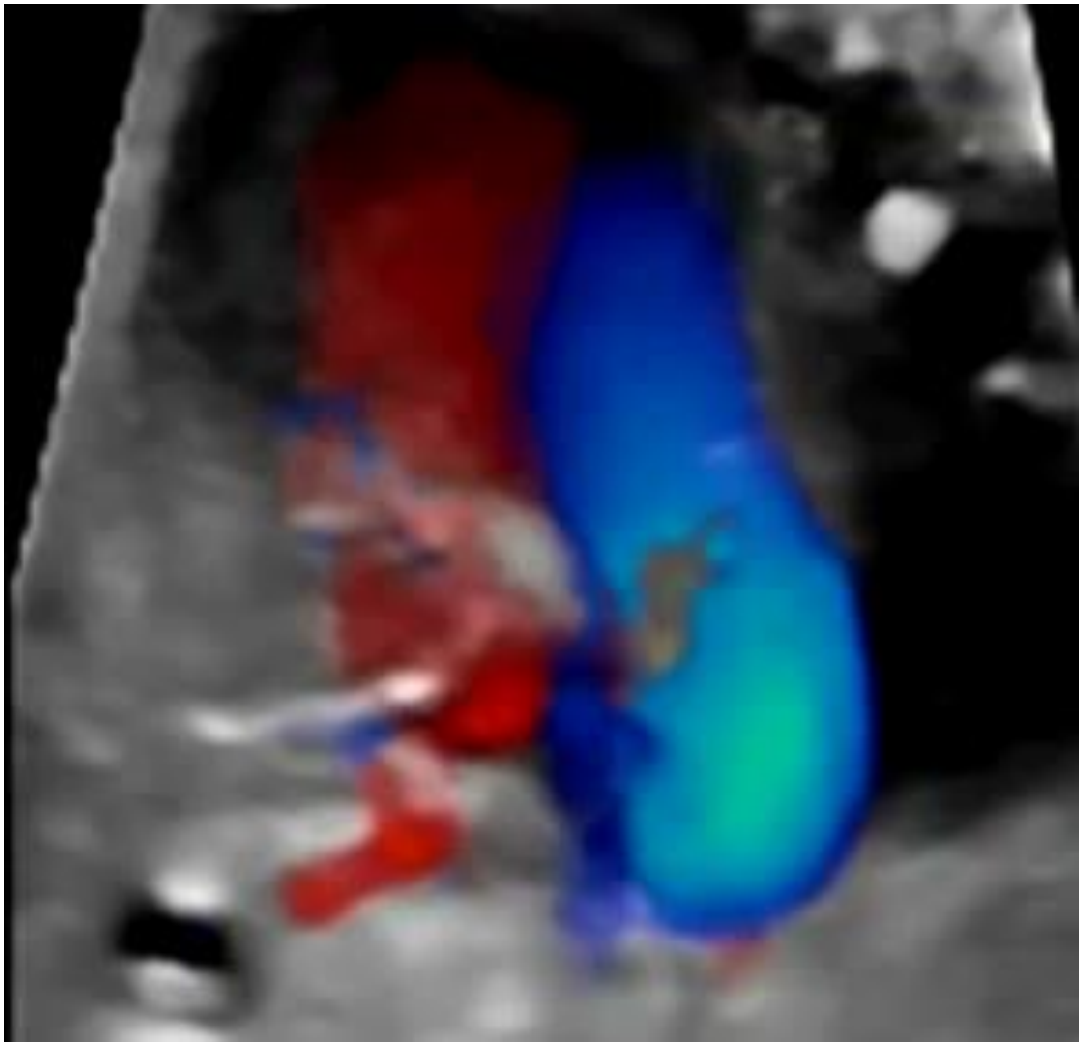
Color Doppler demonstrating the aorta and pulmonary artery arising in a parallel course



Clip SV_13

Images by Paolo Volpe

Glass-body rendering demonstrates ventricular filling of the single ventricle and blood flow in aorta. Note the small size of the obstructed pulmonary artery.



Clip SV_14

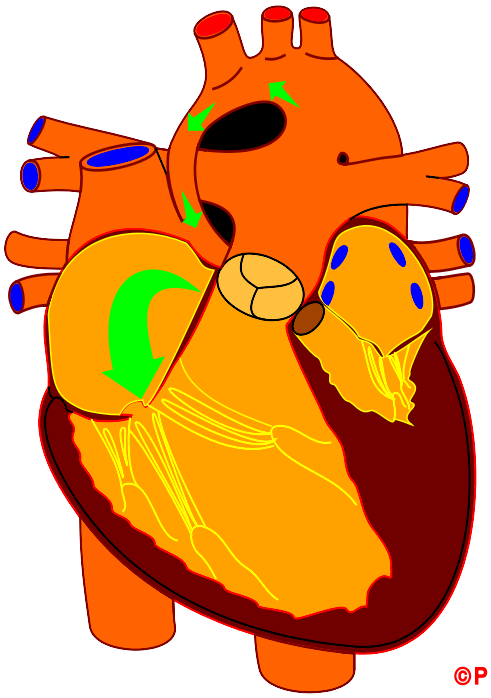
Images by Paolo Volpe

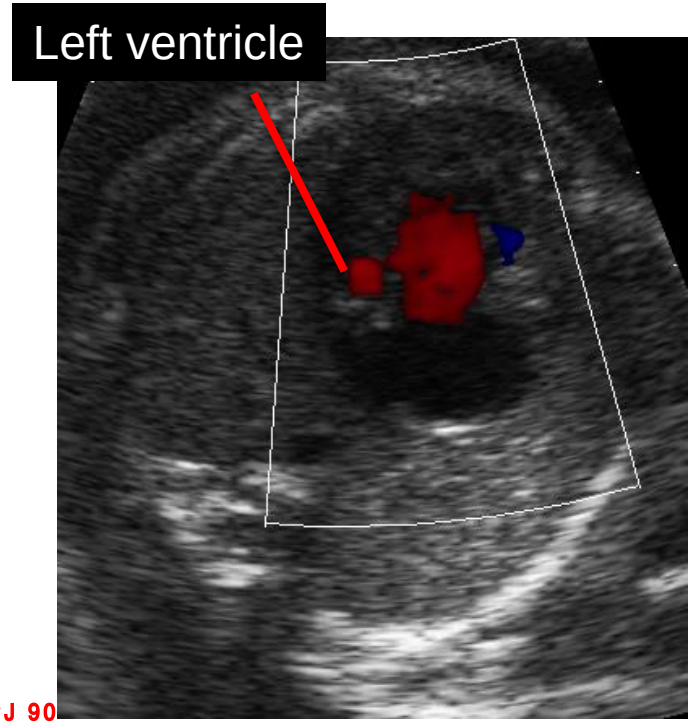
A rare presentation of single ventricle: the great vessels are normally related; pulmonary stenosis is present

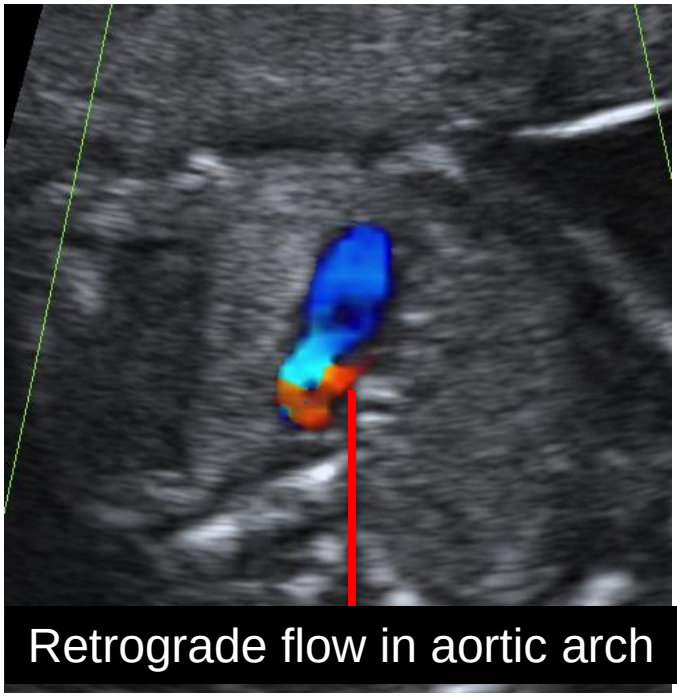
Clip SV_16

Images by Paolo Volpe

Different presentations of single ventricle:
hypoplastic left heart syndrome



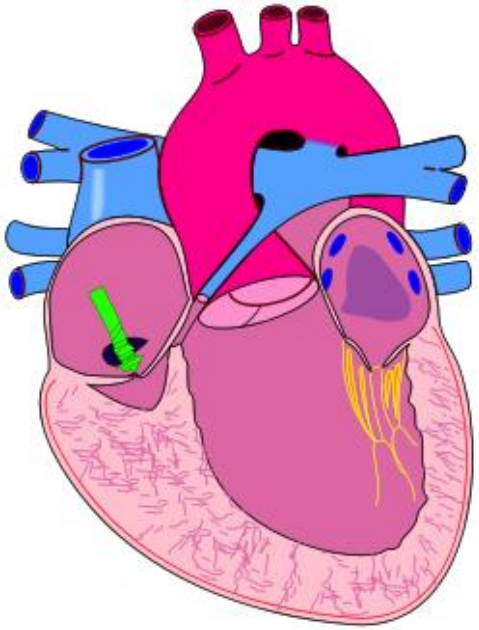


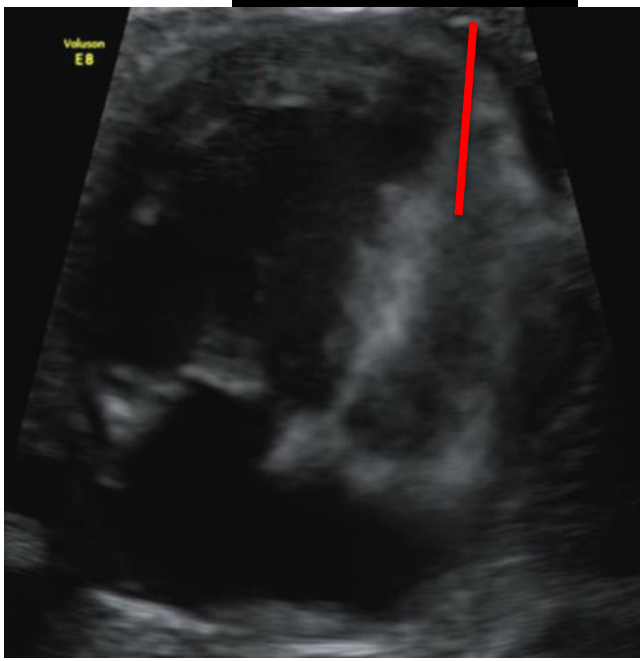


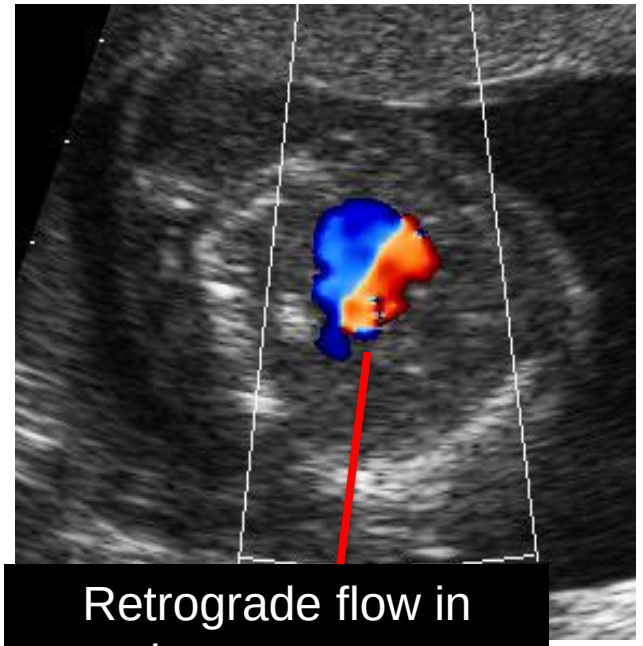
Hypoplastic left heart syndrome is usually associated with a rudimentary left ventricle and a dominant right ventricle; however the presence of severe aortic obstruction or atresia identifies a specific entity with a particularly severe outcome

Images by Gianluigi Pilu and Philippe Jeanty

Different presentations of single ventricle:
pulmonary atresia with intact ventricular septum



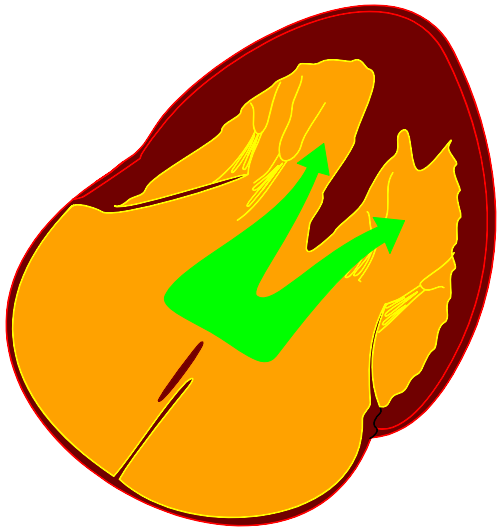


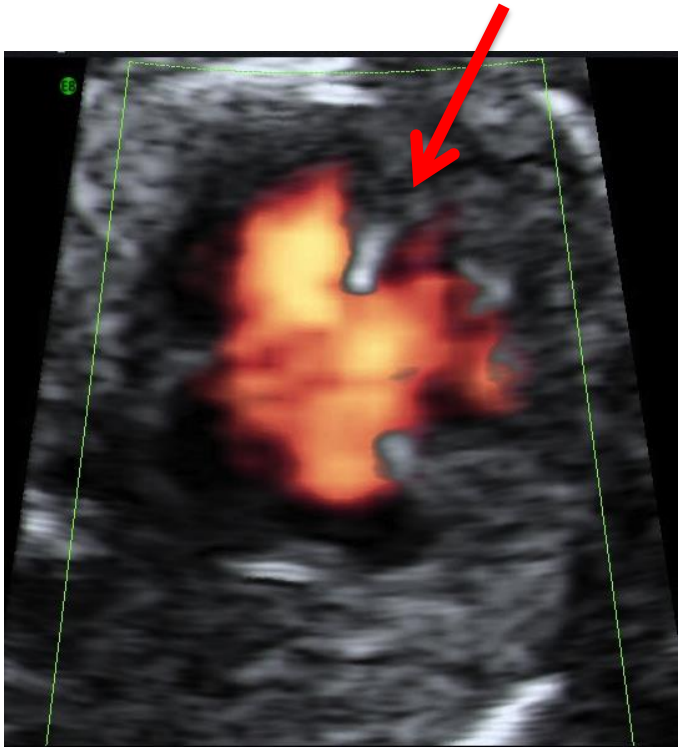


Pulmonary atresia with intact ventricular septum may be associated with an underdeveloped right ventricle, but osbtruction to pulmonary blood flow identifies an anomaly with a more specific prognosis

Images by Gianluigi Pilu, Philippe Jeanty, and Paolo Volpe

Differential diagnosis of single ventricle:
complete atrioventricular septal defect





In atrioventricular septal defects the ventricular septal defect may be large but remnants of the septum are usually well visualized

Images by Gianluigi Pilu and Philippe Jeanty

Transplant free survival of infants diagnosed in utero with different types of single ventricle

From Beroukhim et al: UOG 2014 (DOI: 10.1002/uog.14634)