

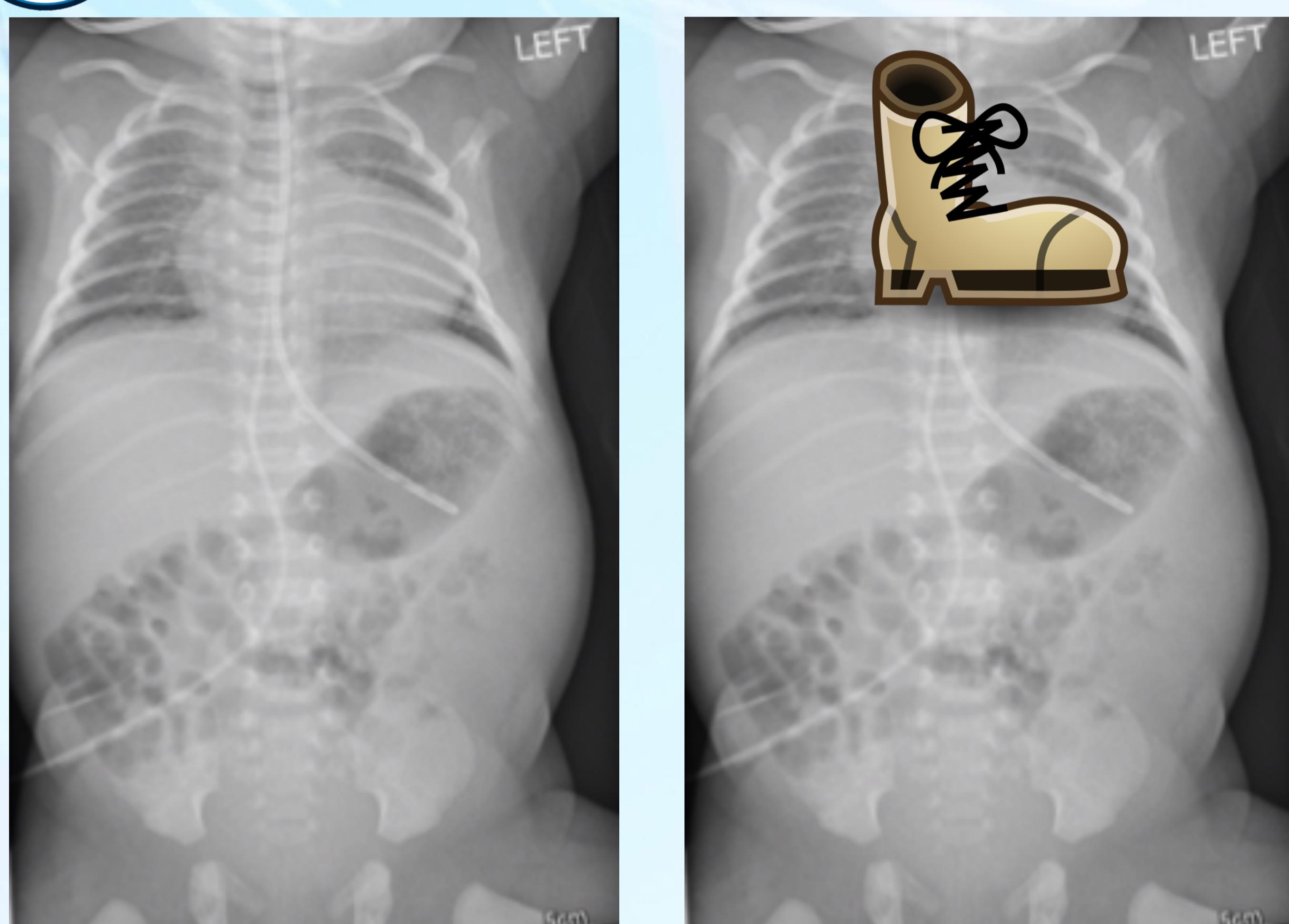
# Coeur-En-Sabot

## A 'Boot'-Camp in Cardiotoracic Surgery

### 1 Case

An antenatal diagnosis of DiGeorge syndrome, presenting with congenital heart disease (CHD: Tetralogy of Fallot). A characteristic boot-shaped (Coeur-en-sabot) appearance was found on chest x-ray (see below). The neonate was referred to the cardiothoracic surgeons for open heart surgery, having a right ventricle to pulmonary artery conduit inserted. Following this, a year later, the patient underwent a successful transcatheter closure of two muscular ventricular septal defects using an Amplatzer Duct Occluder II Device.

### 2 Coeur-En-Sabot



1. Classical 'Boot-Shaped' Heart<sup>1</sup>

2. Upturned Cardiac Apex (right ventricular hypertrophy)

3. Concaves Pulmonary Arterial Segment

4. Pulmonary Oligemia (decreased pulmonary arterial flow)

### 3 DiGeorge

#### 22q11.2 Deletion Syndrome

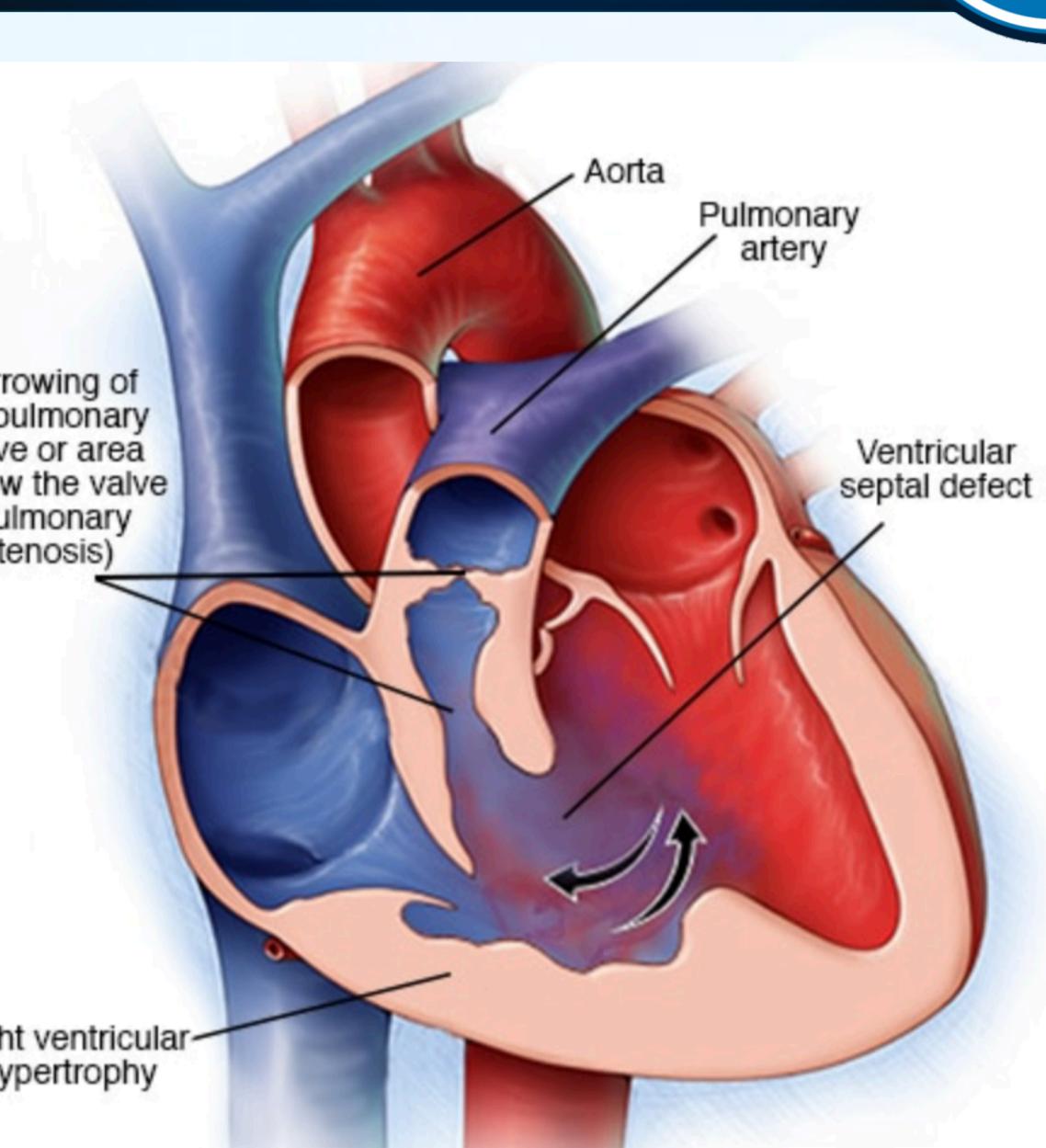
Autosomal dominant inherited condition, resulting from the deletion of a region containing 30 to 40 genes.<sup>4</sup>

Of those with DiGeorge syndrome 80%-99% of people have the symptoms listed below.<sup>8</sup> Our patient presents with those highlighted in blue.

Eye + Ears Defects	Facial Defects	Cardiac Defects	Others Defects
Epicantus	Bulbous Nose	Abnormal Aortic Arch	Abnormality Of The Pharynx
Telecanthus	Cleft Palate	Abnormal Pulmonary Valve	Immunodeficiency
Upplanted Palpebral Fissure	Wide Nasal Bridge	Atrial Septal Defect	Muscular Hypotonia
Conductive Hearing Loss	Dysphasia	Tetralogy Of Fallot	Platybasia
Low-set Ears	Prominent Nasal Bridge	Truncus Arteriosus	Hypoplasia Of The Thymus
	Nasal Speech	Ventricular Septal Defect	

Structural abnormalities:<sup>2</sup>

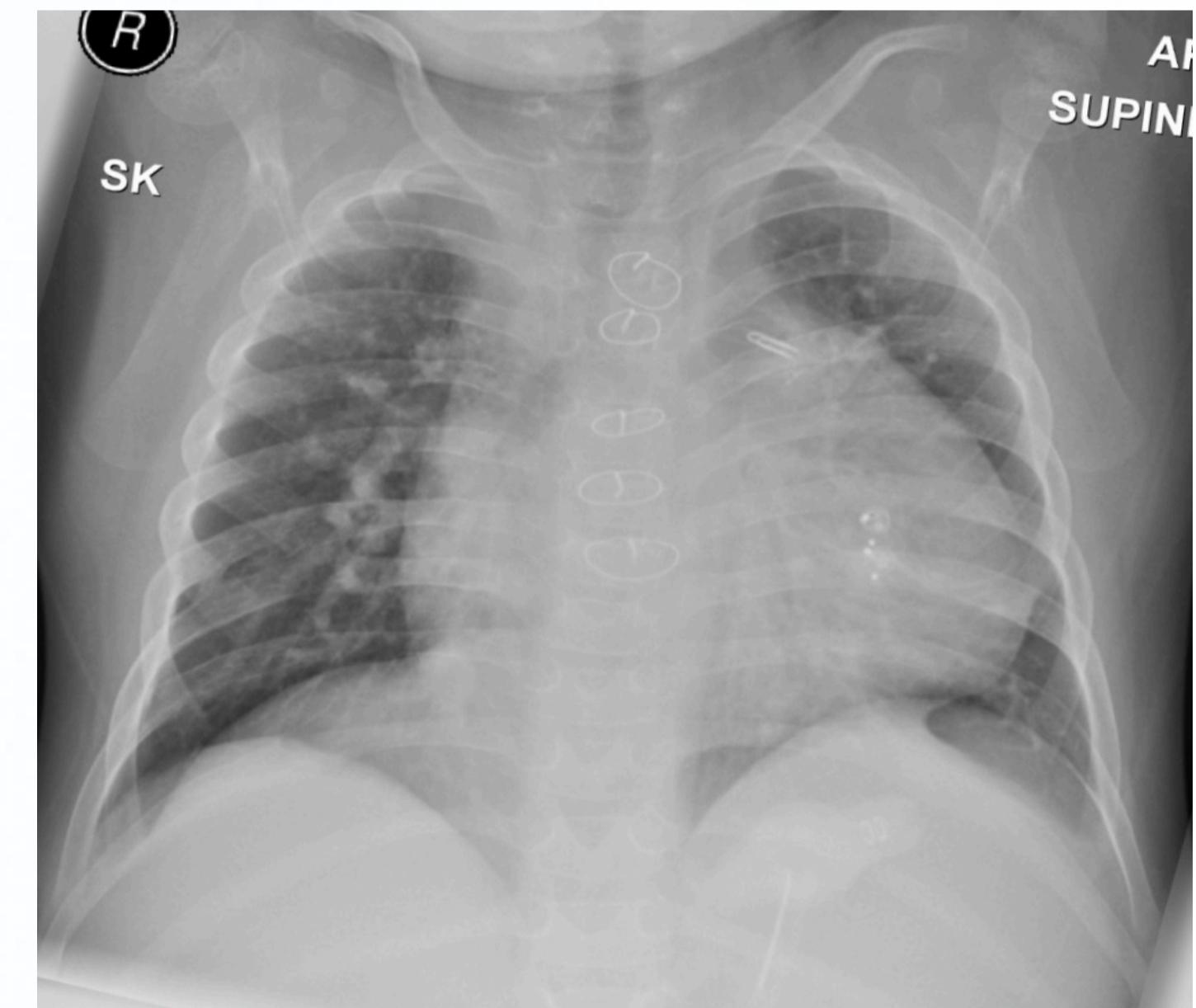
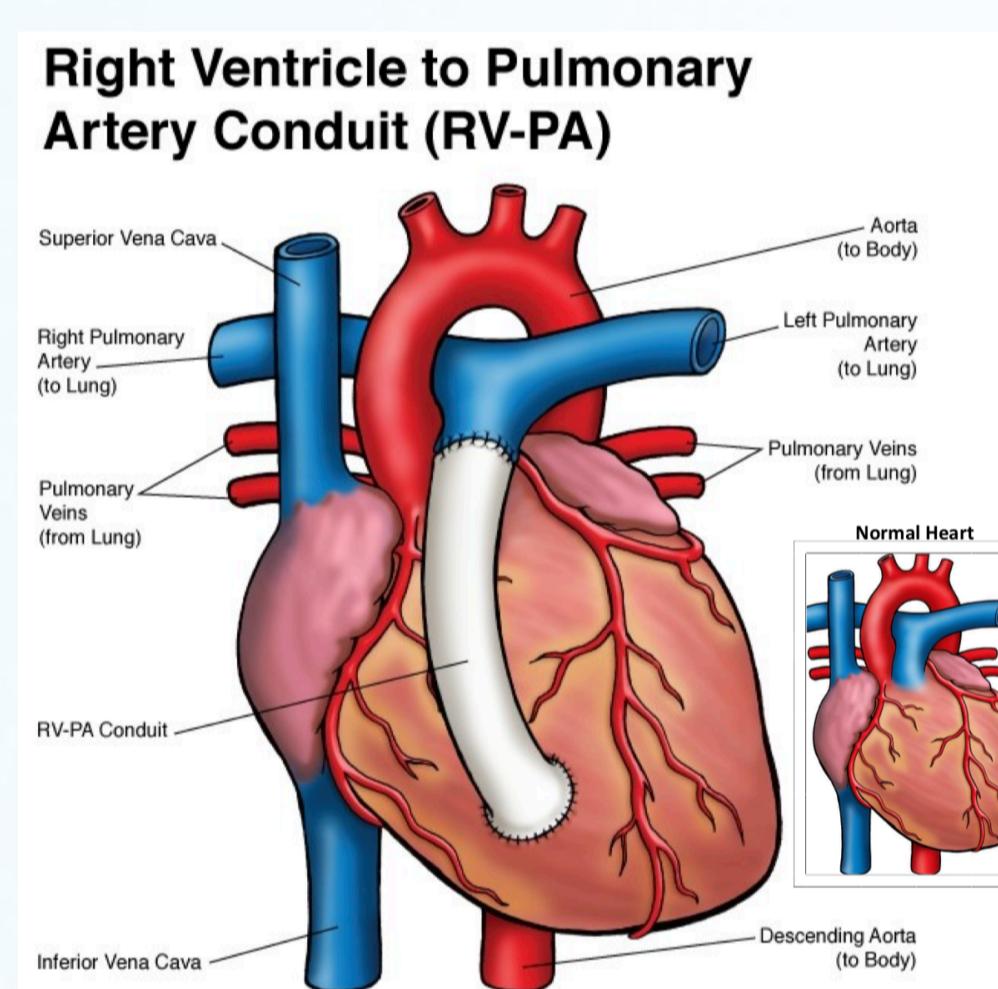
Tetralogy of Fallot is a congenital heart disease that presents with cyanosis, dyspnoea with feeding, irritability and harsh ejection systolic murmur at left mid and upper sternal border.<sup>3</sup>



### 4 RV-PA

#### Right Ventricle to Pulmonary Artery Conduit

A RV-PA was inserted to treat the patient's stenotic pulmonary valve, allowing blood to flow into the lungs. A median sternotomy was preformed and the patient was placed on cardiopulmonary bypass. An incision was then made on the pulmonary artery and right ventricle and the RV-PA conduit was inserted to connect them.<sup>6,7</sup>



X-ray Taken After Surgeries

### 5 VSD Closure

#### Ventricle Septal Defect Closure

An Amplatzer Duct Occluder (ADO) was used to treat the patient's muscular ventricular septal defects. The device is made from 2 disks with an articulated connecting waste.<sup>8</sup> The ADOs as inserted via a minimally invasive transcatheter approach. With a small incision made in the groin and a catheter inserted. The ADO was passed to the site of the VSD and positioned across the defect.



### 7 Patient Now

3.5 years later the patient is currently under investigations, as an inpatient, for suspicion of infective endocarditis of their RV-PA.

### 8 Fun Facts

- TOF was 1st described in 1671 by Niels Stensen.<sup>9</sup>
- Coeur-en-Sabot literally translates to "heart in shoe".
- In 1944, TOF was the 1st surgically managed.<sup>10</sup>

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