

# Coeur-En-Sabot

## A 'Boot'-Camp in Cardiothoracic Surgery

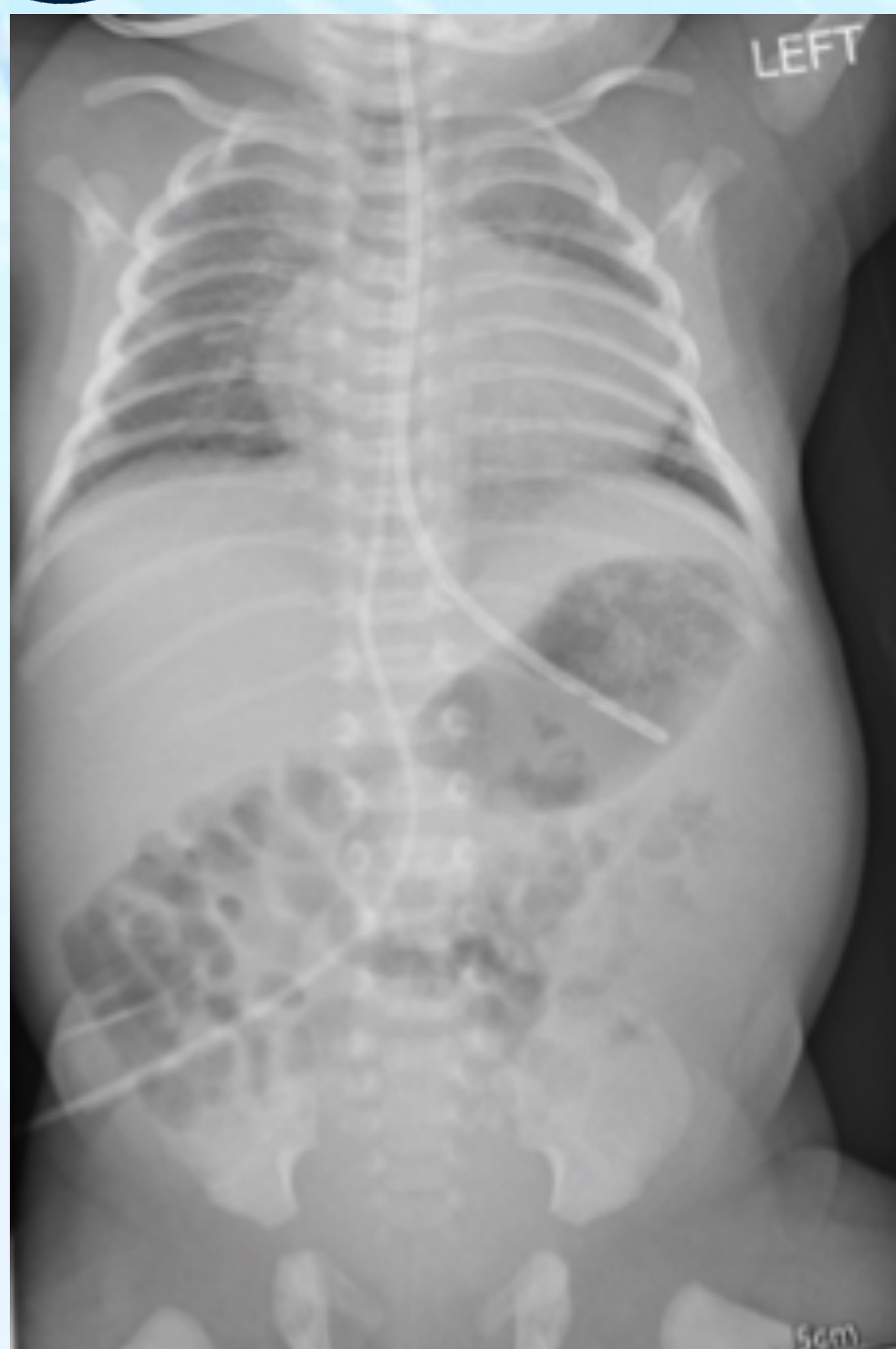
1

### 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. Concave 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.

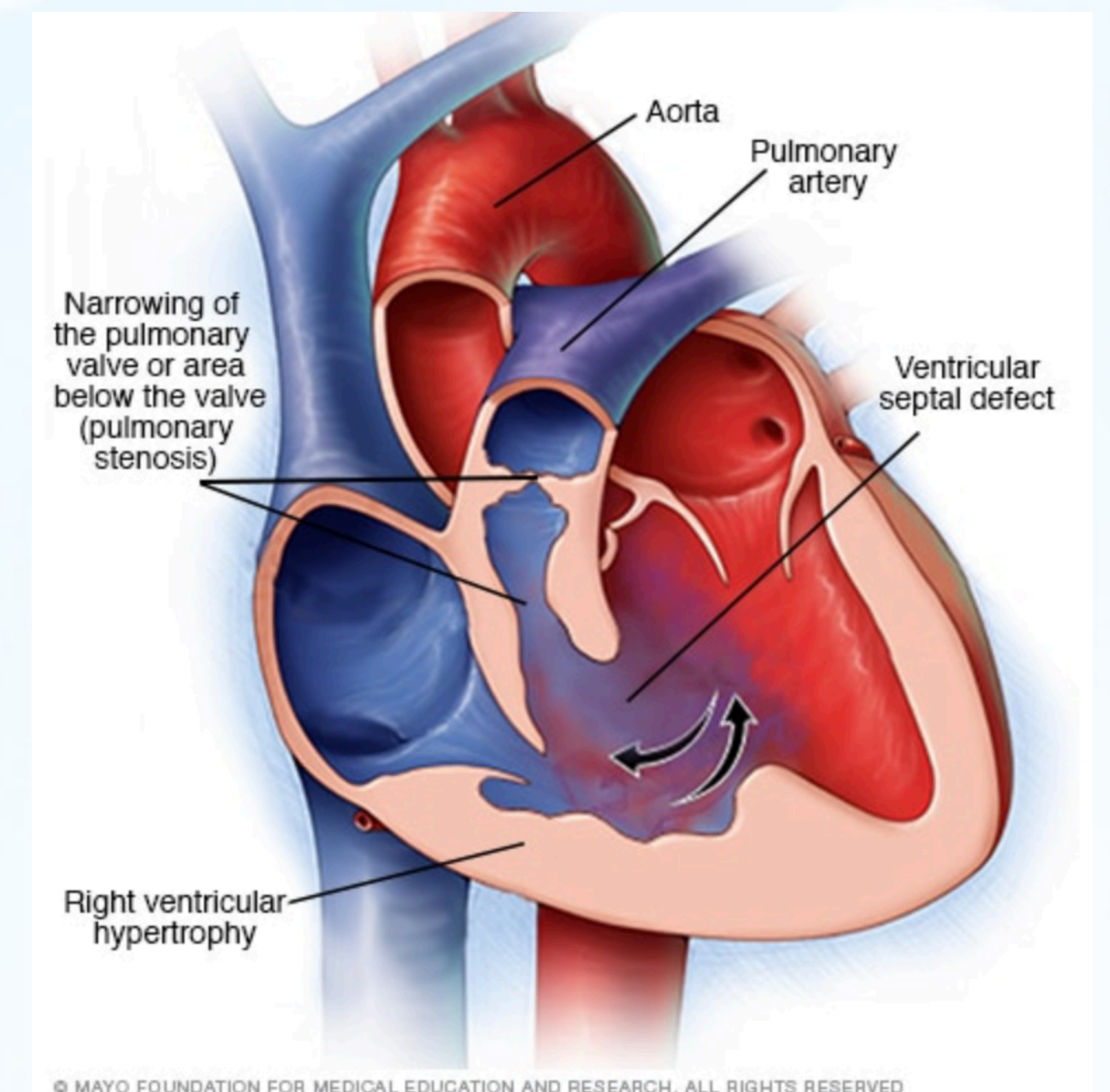
Eye + Ears Defects	Facial Defects	Cardiac Defects	Others Defects
Epicanthus	Bulbous Nose	Abnormal Aortic Arch	Abnormality Of The Pharynx
Telecanthus	Cleft Palate	Abnormal Pulmonary Valve	Immunodeficiency
Upslanted Palpebral Fissure	Wide Nasal Bridge	Atrial Septal Defect	Muscular Hypotonia
Conductive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

### Tetralogy of Fallot

4

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>

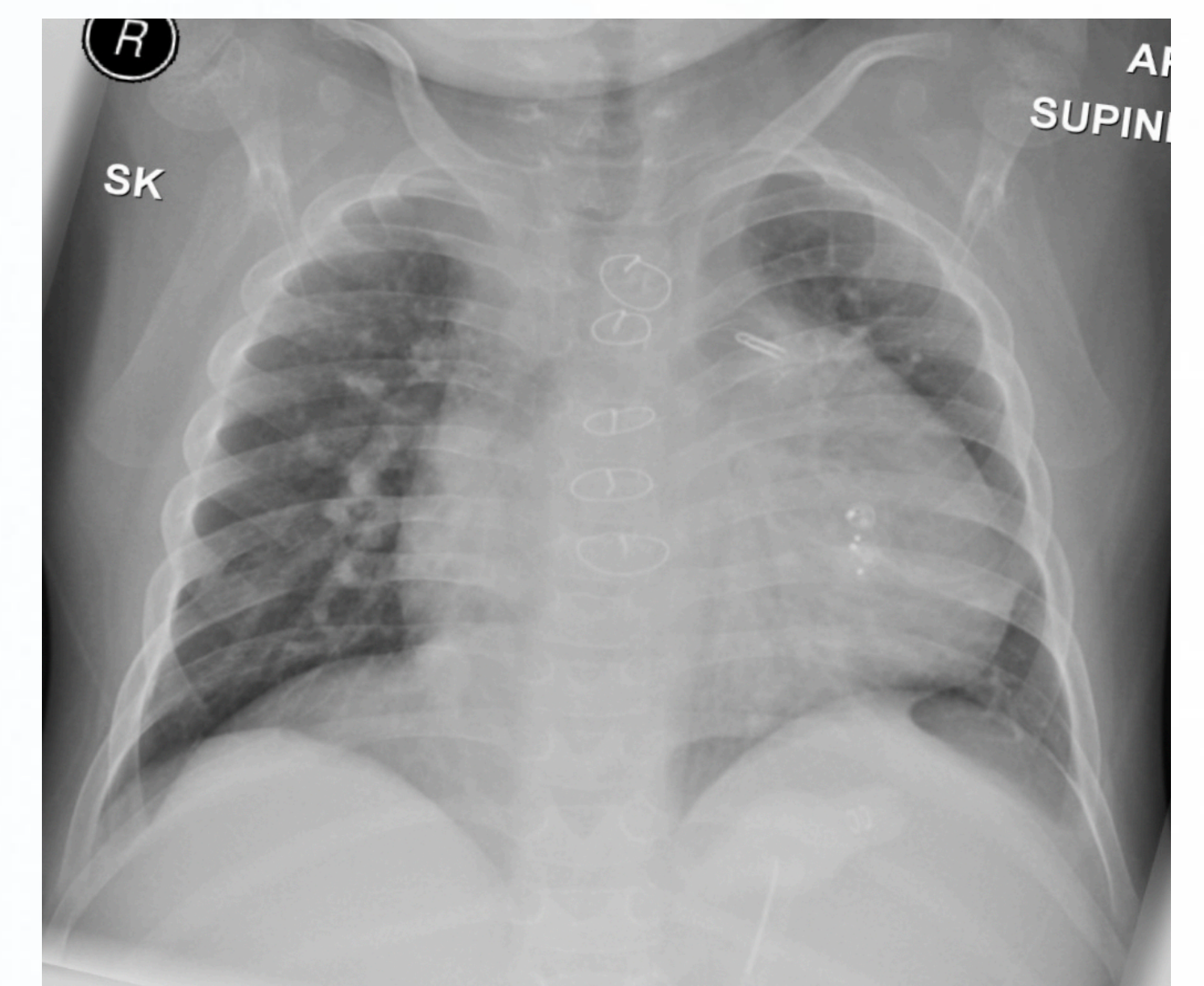
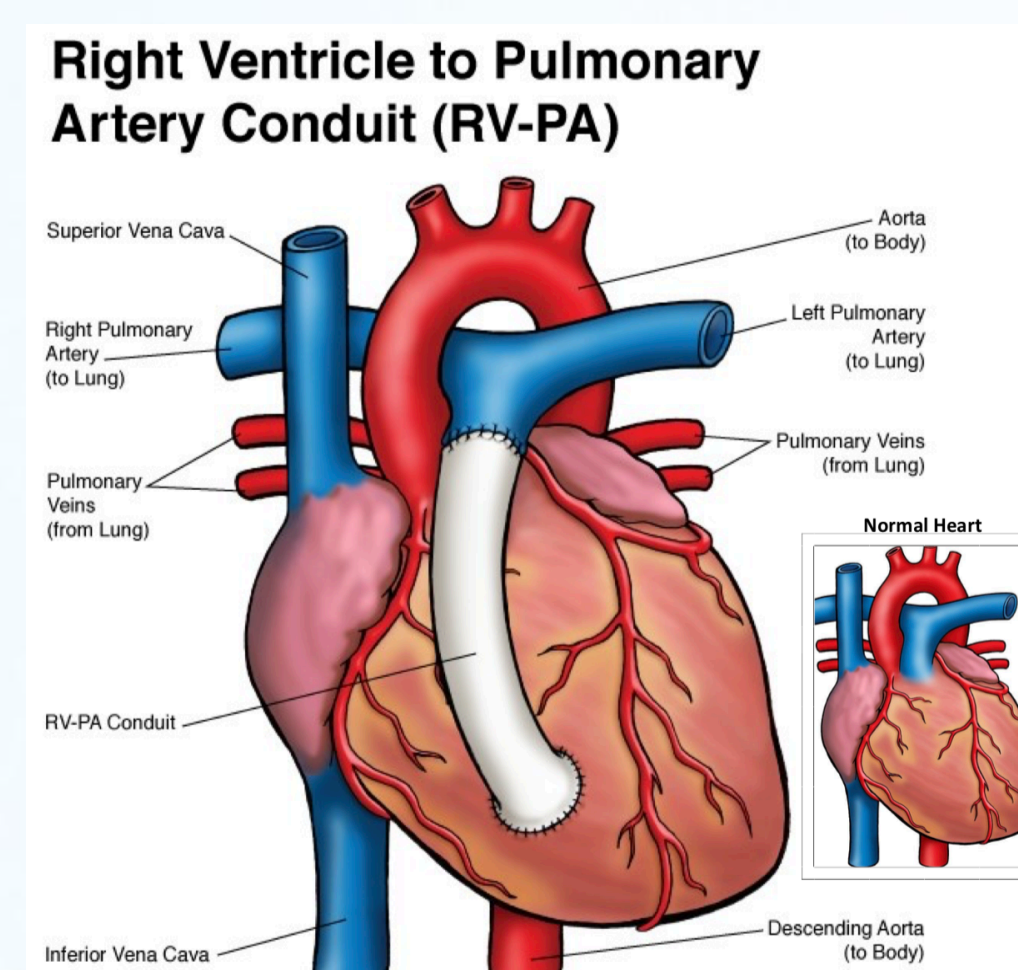


### RV-PA

5

#### 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

### VSD Closure

6

#### 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.



### Patient Now

7

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

### Fun Facts

8

- 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 manged.<sup>10</sup>

Christina Apthorp

1. Weerakkody Y. Tetralogy of Fallot | Radiology Reference Article | Radiopaedia.org [Internet]. Radiopaedia.org. 2019 [cited 20 June 2019]. Available from: [https://radiopaedia.org/articles/tetralogy-of-fallot?lang=gb#nav\\_radiographic-features](https://radiopaedia.org/articles/tetralogy-of-fallot?lang=gb#nav_radiographic-features)

2. tetralogy of Fallot - Symptoms and causes [Internet]. Mayo Clinic. 2019 [cited 24 June 2019]. Available from: <https://www.mayoclinic.org/diseases-conditions/tetralogy-of-fallot/symptoms-causes/syc-20353477>

3. Tetralogy of Fallot - Pediatrics - MSD Manual Professional Edition [Internet]. MSD Manual Professional Edition. 2012 [cited 20 June 2019]. Available from: <https://www.msdmanuals.com/en-gb/professional/pediatrics/congenital-cardiovascular-anomalies/tetralogy-of-fallot>

4. DiGeorge syndrome | Children's Hospital of Wisconsin [Internet]. Chw.org. 2019 [cited 20 June 2019]. Available from: <https://www.chw.org/medical-care/endocrine/endocrine-conditions/disorders-affecting-calcium-metabolism/digeorge-syndrome>

5. Childrensheartclinic.org. 2019 [cited 20 June 2019]. Available from: <http://www.childrensheartclinic.org/downloads/8/2018/10/right-ventricle-to-pulmonary-artery-conduit.pdf>

6. Melbourne T. Cardiology - Pulmonary Atresia with VSD [Internet]. Rch.org.au. 2019 [cited 20 June 2019]. Available from: [https://www.rch.org.au/cardiology/heart\\_defects/pulmonary\\_atresia\\_with\\_vsd/](https://www.rch.org.au/cardiology/heart_defects/pulmonary_atresia_with_vsd/)

7. Childrensheartclinic.org. 2019 [cited 24 June 2019]. Available from: <http://www.childrensheartclinic.org/downloads/8/2018/10/right-ventricle-to-pulmonary-artery-conduit.pdf>

8. SALIBA Z, EL-RASSI I, ABI-WARDE M, CHEHAB G, DAOU L, KHATER D et al. The Amplatzer Duct Occluder II: A New Device for Percutaneous Ductus Arteriosus Closure. Journal of Interventional Cardiology. 2009;22(6):496-502.

9. "Fallot's tetralogy". Whonamedit?. Archived from the original on 3 October 2016. Retrieved 2 October 2016.

10. Lillehei CW, et al. Direct vision intracardial surgical correction of the tetralogy of Fallot, pentalogy of Fallot, and pulmonary atresia defects: report of the first ten cases. Ann Surg 1955;142:418-45.