# Complete isolation of left innominate artery in a patient with CHARGE syndrome: case presentation and review of reported cases

Rossana Zanoni, MD[(a)]; Emanuela Concetta D'Angelo, MD[(a)]; Gabriele Egidy Assenza, MD[(a)]

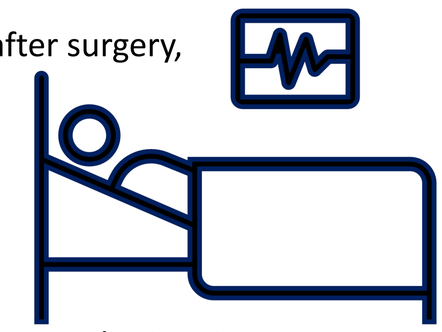
Francesco Petridis, MD[(b)]; Luca Ragni, MD[(a)]; Daniela Palleri, MD[(a)]; Elisabetta Mariucci, MD[(a)]; Emanuela Angeli, MD[(b)]; Gaetano Domenico Gargiulo, MD[(b)#]; Andrea Donti, MD[(a)#];

a) Pediatric Cardiology and Adult Congenital Heart Disease Program, Department of Cardio-Thoracic and Vascular Medicine IRCCS Azienda Ospedaliero-Universitaria di Bologna, Italia (Italy)

b) Pediatric Cardiac Surgery and Adult Congenital Heart Disease Program, Department of Cardio-Thoracic and Vascular Medicine IRCCS Azienda Ospedaliero-Universitaria di Bologna, Italia (Italy)

7-year-old male child affected by CHARGE syndrome referred for evaluation of persistent patent ductus arteriosus.

## Clinical history:

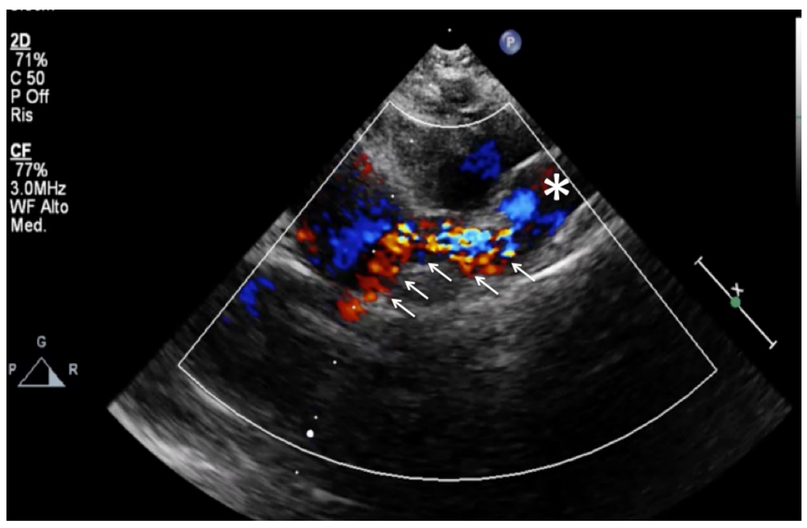


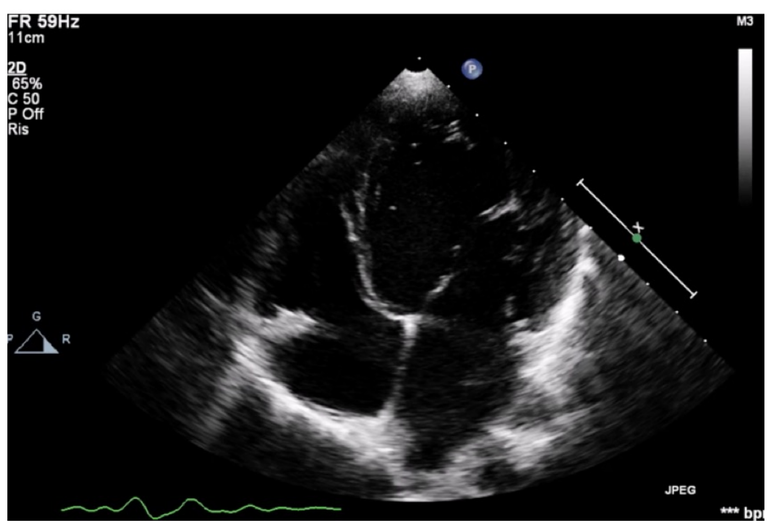
- corrective surgery of esophageal-tracheal fistula at neonatal age, tracheostomy due to laryngomalacia and bilateral vocal cord paralysis after surgery,
- paralysis of the VII cranial nerve,
- gastro-esophageal reflux,
- flat hemangioma of the right hand fingers.
- dyspnea and cyanosis during activity. No history of diminished left arm strength or any cerebrovascular insufficiency.

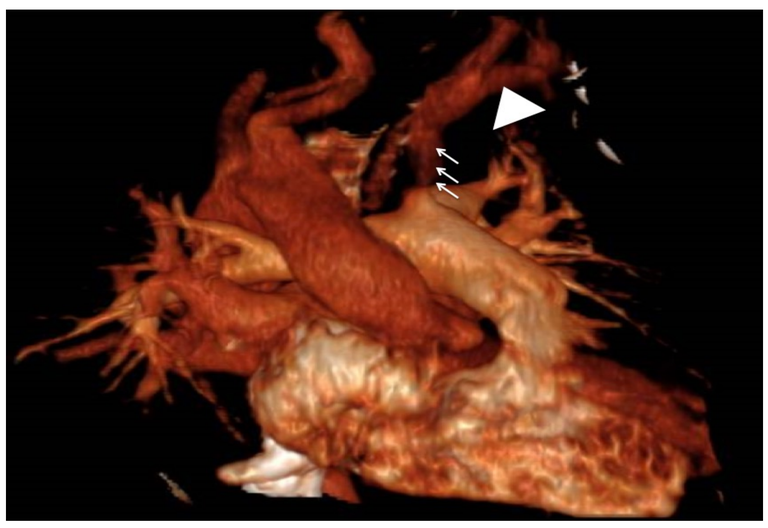
**Physical examination:** continuous murmur at the mesocardium, with normal peripheral arterial pulses, weak pulse in the left arm and no cyanosis at rest.

## Diagnostic work-up:

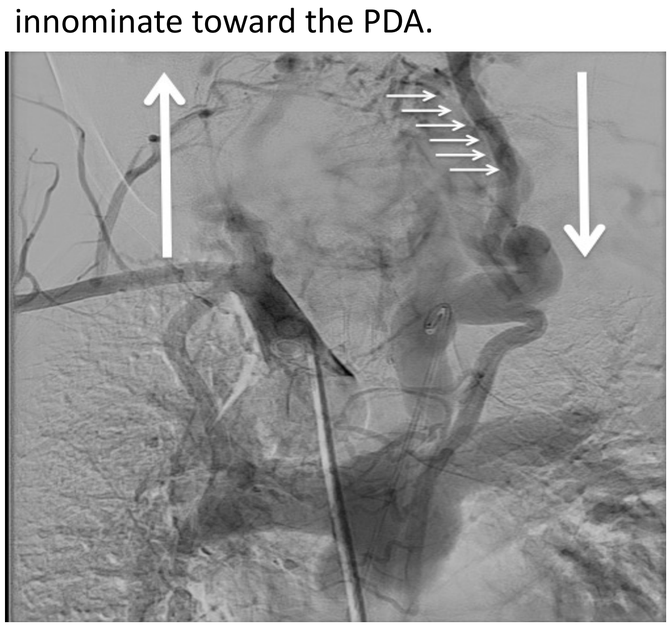
- **Chest radiograph:** prominent first right arch of the cardiac silhouette with depressed first left arch, mild cardiomegaly, slightly prominent pulmonary vascular markings.
- **Echocardiogray:** large-sized PDA, suspected origin from left innominate artery, mild enlargement of left cardiac chambers. No evidence of elevated right ventricular systolic pressure.
- **Contrast enhanced computed tomography:** right aortic arch with complete isolation of left innominate artery. Left innominate artery originating from proximal portion of left pulmonary artery by the wide left PDA .

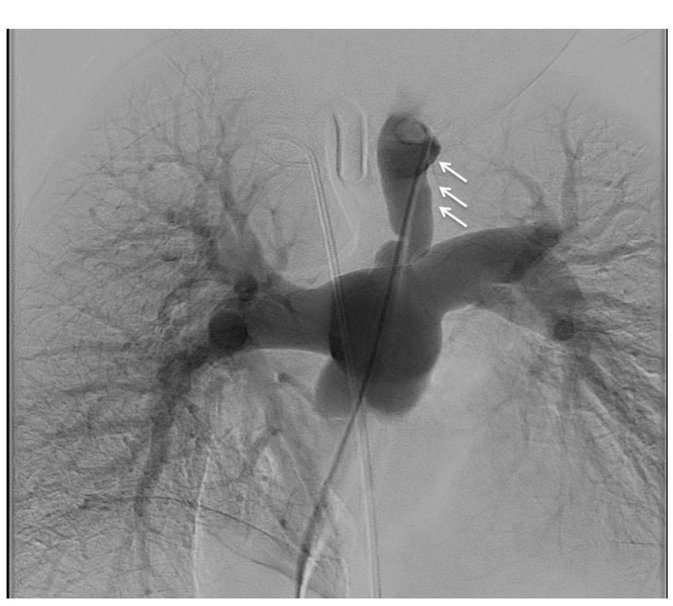


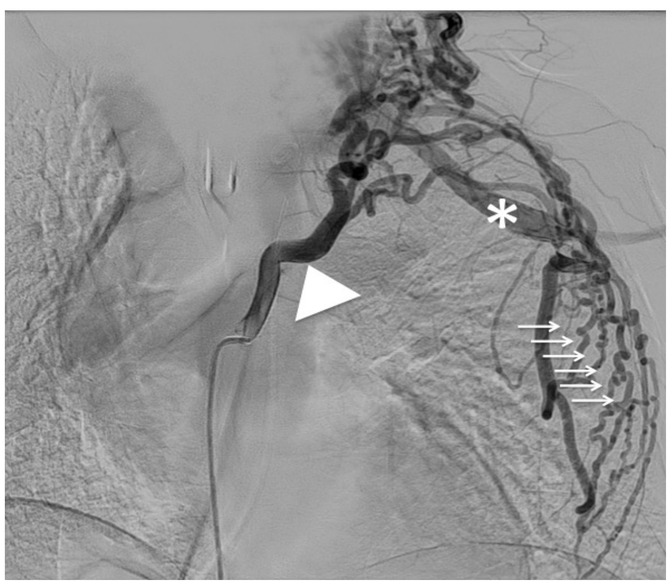



- **Cardiac catheterization and selective angiography :** mean pulmonary pressure 21 mmHg, wedge pressure 14 mmHg. Large competitive flow. Good retrograde opacification of the left common carotid through intracranial circulation. Two collateral arterial vessels feeding the left innominate artery with retrograde filling of left innominate toward the PDA.





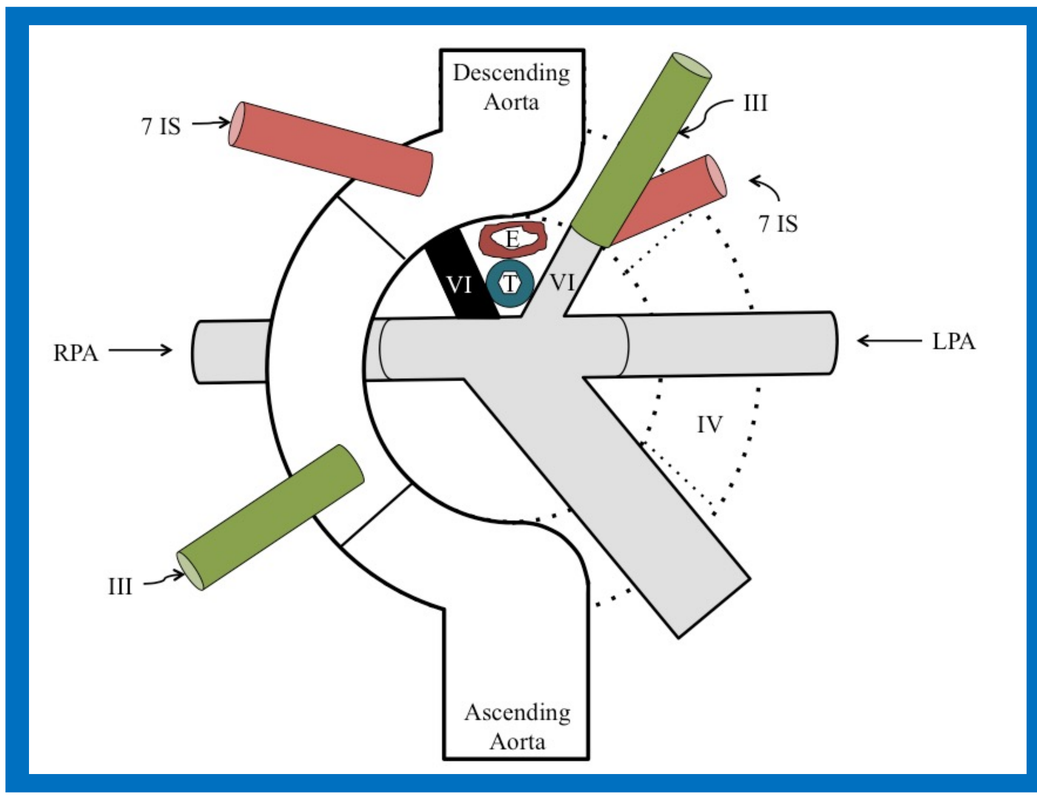


## Heart-team discussion :

Left ventricular volume overload + pulmonary overcirculation + left innominate arterial stealing

→ **transcatheter closure of the PDA:** Release of an Abbott Vascular Plug II 12mm. No residual shunt, no neurological or muscular.

→ **Our patient appears to be the first child with CHARGE syndrome and isolated left anonymous artery successfully treated by percutaneous closure of the left PDA.**




**Embryological explanation :** Rathke's diagram + Edwards's concept of double aortic.

**Six similar cases of patients with CHARGE syndrome are reported:**

- 3 complex cardiac diseases + isolated left carotid artery
- 3 anomalous origin of the brachiocephalic trunk from the left pulmonary artery

→ All surgical correction.