Newborn with tetralogy of Fallot and absent pulmonary valve

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A term newborn girl with prenatal diagnosis of tetralogy of Fallot with absent pulmonary valve syndrome (TOF-APV) and massively dilated main and proximal right pulmonary arteries presented with tachypnea and cyanosis. Chest radiograph demonstrates hyperinflated right lung and a large 2.7-cm dilated right pulmonary artery (Fig. 1, *arrows*). Postnatal echocardiogram also demonstrated discontinuous left pulmonary artery. Contrast-enhanced CT of the chest reveals massively dilated right pulmonary artery (*) with mass effect upon the right main stem bronchus (*short arrow*) (Fig. 2). Right upper lobe atelectasis is also present (Fig. 2, *large arrow*).

TOF-APV is a rare variant of TOF and comprises only 3–6% of all infants with TOF. Patients have massive

Fig. 1 Supine chest radiograph

Fig. 2 Contrast-enhanced CT of the chest

pulmonary insufficiency and aneurysmal dilatation of the pulmonary arteries. Infants might present with mild respiratory symptoms or with significant respiratory compromise requiring early surgical repair [1]. Severe respiratory compromise is caused by tracheobronchomalacia and compression of the large airways by the dilated pulmonary arteries, with resultant air-trapping and further atelectasis. CT is helpful to delineate pulmonary arterial and airway anatomy and is also useful on postsurgical evaluation [2].

References

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