A 47-year-old female with pulmonary atresia (Panel E) and ventricular septal defect (VSD) presented to our hospital with severe chest pain and dyspnoea. Her diagnosis had been established in early infancy at which time it was deemed inoperable, and she had since been declined transplantation. She had a single giant major aortopulmonary collateral artery (MAPCA) arising from an aneurysmal brachiocephalic artery and connecting distally to the right pulmonary artery (Panels A, previous CXR and C, three-dimensional reconstruction from a posterior view), which had increased in size on serial imaging and now measured 8.9 × 7.7 cm. On presentation, our patient had circulatory shock and respiratory failure with evidence of a new large right pleural effusion (Panel B). Laboratory data showed severe relative anaemia (haemoglobin 150 g/L compared with baseline of 210 g/L). Urgent chest computed tomography (CT) angiogram revealed focal rupture of the distal anterior MAPCA wall (Panel G) and secondary massive hemothorax with complete right lung compression (Panels D and F, star—hemothorax). An extensive intramural haematoma (high attenuation crescent sign on non-contrast CT) was also present (Panel D). Despite support, the patient arrested and died shortly after.

Pulmonary atresia with VSD represents 1–2% of all congenital heart defects, the natural history of this complex lesion being poor. While progressive MAPCA stenosis is well described, spontaneous rupture is exceedingly rare. Once significant pulmonary arterial hypertension has developed, complete repair is no longer possible and transplantation remains the only definitive treatment option. In select surgically uncorrected patients who are deemed to be at a high risk of rupture, palliative surgical intervention to close the MAPCA and create replacement systemic to pulmonary shunt(s) should be considered to prevent this catastrophic complication.

Panels A–G. AoA, aortic arch; AoR, aortic root; LPA, left pulmonary artery; RPA, right pulmonary artery; MPA, main pulmonary artery; BCA, brachiocephalic artery; RSC, right subclavian artery.

Supplementary material is available at European Heart Journal online.