

**A rare cause of central cyanosis and nose bleeding: hereditary haemorrhagic telangiectasia with large pulmonary arteriovenous fistula**

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**Introduction:** Pulmonary arteriovenous fistula (PAVF) is a rare cause of right-to-left shunting. The most of them are congenital, 50% of these cases are associated with hereditary haemorrhagic telangiectasia (HHT). We reported a 10-year-old boy who was presented with cyanosis and clubbing and diagnosed PAVF associated with HHT.

**Case:** A 10-year-old boy patient admitted to our clinic with cyanosis and clubbing for 6 years. There was frequent nose bleeding in family history (in his father, sister, grandmother, uncle, aunt and cousin) (Figure A). Physical examination revealed telangiectasia in the lips, central cyanosis, and clubbing. Oxygen saturation was 78 %. The telecardiogram revealed irregular opacity in the middle and basal zone of the left lung. Echocardiographic examination was normal; contrast echocardiography showed bubble contrast in the left chambers, raising suspicion of PAVF. Computed tomographic arteriography confirmed multiple PAVF originating from the left inferior segmental pulmonary artery and draining into the left lower pulmonary vein. (Figure B). The PAVF was closed successfully with multiple coils by transvenous embolization. Oxygen saturation was 95% after embolization.

**Conclusion:** In this case, the patients who are presented with cyanosis and frequent nose bleedings in the family history should be considered as HHT. PAVF should be suspected during a contrast echocardiography study. Confirmation of micro bubbles entering the left atrium directly from a pulmonary vein suggests that a PAVF is present. Contrast computed tomographic arteriography is the modality of choice for defining the precise anatomy of PAVF.

