IMAGE IN CARDIOLOGY

Closure of pulmonary arterio-venous malformations in a patient with a novel form of Hereditary Haemorrhagic Telangiectasia

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The patient is a 21-year-old female patient known with Hereditary Haemorrhagic Telangiectasia (HHT) but associated significant hypermobility (to the point where she fulfils the Beighton criteria for Ehlers-Danlos syndrome). This combination has not previously been described and probably represents a new mutation. She has a longstanding history of nosebleeds, gastro-intestinal bleeds and joint dislocations. Her brother died at a young age from pulmonary haemorrhage and this prompted a full body CT-scan to look for arterio-venous malformations (AVM). She had no cerebral AVMs but a number were found in her lungs and gastrointestinal tract. Because of the connective tissue disorder, it was feared that intervention on the AVMs may be hazardous and she was referred to our Unit for evaluation.

A transthoracic echo with agitated saline contrast showed the appearance of a large number of micro-bubbles in the left atrium within three beats of the right atrium, indicating a significant shunt (Figure 1).

A contrasted CT-scan (arterial phase) of the chest and abdomen demonstrated multiple arterio-venous malformations, ranging in size, throughout both lungs. The 2 largest lesions were located in

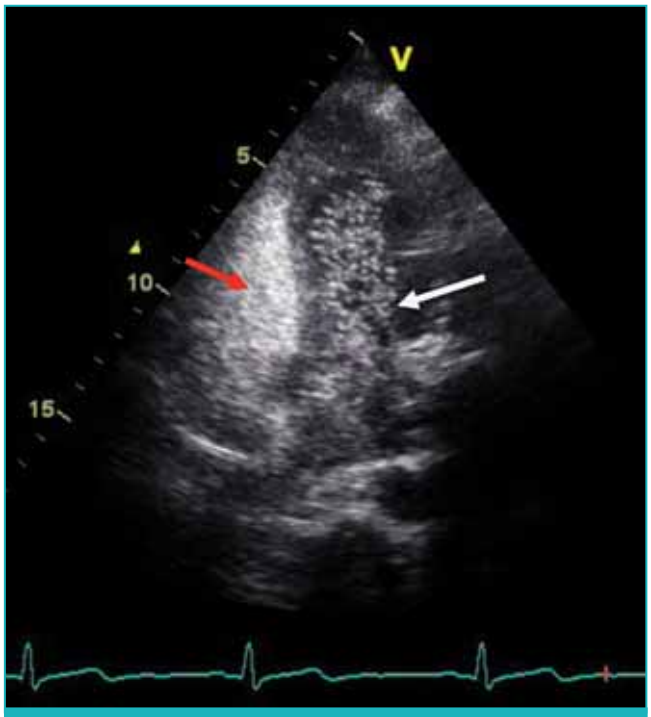

FIGURE 1: Apical 4 chamber view after intra-venous agitated saline injection showing micro-bubbles in the left ventricle (white arrow) and right ventricle (red arrow). Note normal sized cardiac chambers.

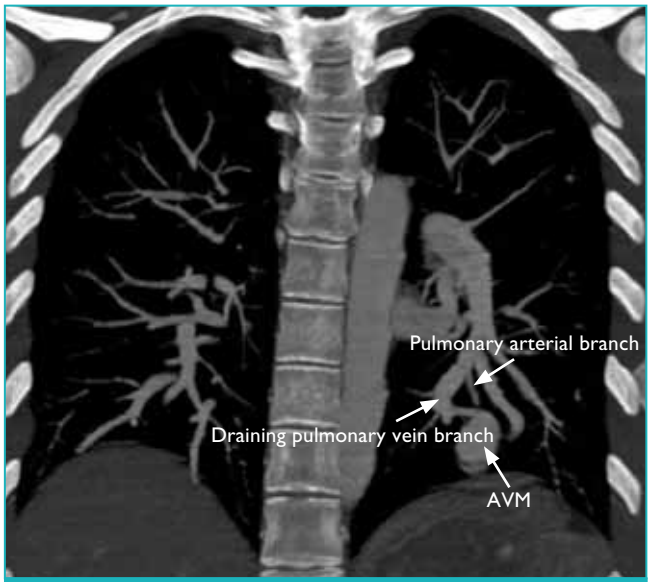

FIGURE 2: MIP (maximum intensity projection) coronal reconstruction of CT chest in arterial phase, demonstrates a large arterio-venous malformation (AVM) in the left lower lobe.

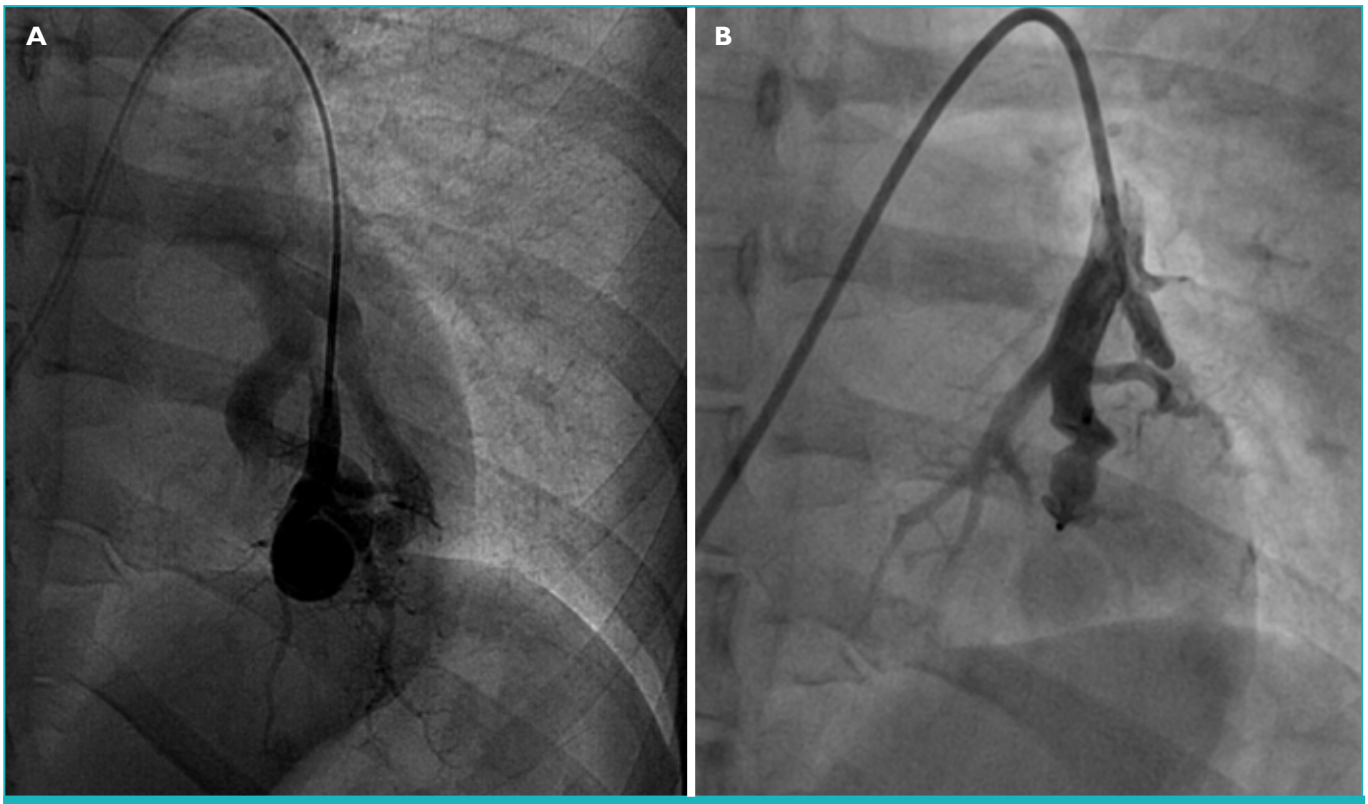

FIGURE 3:
A: Pulmonary angiogram of the left lung showing 1 of the large AVMs prior to closure.
B: Repeat angiogram after closure of the same defect. Image taken 2 months after the implant of an 8mm Amplatzer vascular plug, at the time of closure of 2 AVMs in the contra-lateral lung.

the left lower (Figure 2) and right middle lobes. Early filling of the porto-venous system, due to multiple arterio-venous fistulae in the abdomen, were also identified.

The procedure: because of the associated high risk for cerebral emboli with large pulmonary AVMs, it was decided to attempt closure of the defects. The opinion of a number of international experts was sought and although none had performed the procedure in a patient with abnormal connective tissue, it was felt that the procedure could be performed. Only the 3 large AVMs were considered for closure and we elected to close them in 2 procedures so that if she developed significant haemorrhage, the contralateral lung could be intubated and protected. Pulmonary angiography revealed large feeding arteries and Amplazer vascular plug II's (AVP II) (St.Jude Medical, St. Paul, Minn.) were oversized by 20 - 30% (defects measured 4.6, 6 and 8mm were treated with AVP II sizes 6, 8 and 10mm respectively). We deemed this the ideal compromise between device embolisation if undersized and tearing of vessels if oversized.

Both procedures were successful (Figure 3A and 3B) and uncomplicated apart from mild chest discomfort. A follow up contrast echo showed significantly less bubbles appearing in the left atrium after 5 heartbeats. This simple test can be used to monitor her condition in future and limit exposure to radiation from regular CT-scans.

Conflict of interest: none declared.