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Brief Report

Treatment of haemoptysis in pulmonary atresia with tranexamic acid

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Abstract We report the case of a young woman with continuing haemoptysis, pulmonary atresia, previous shunt surgery, and pulmonary hypertension. She was not suitable for further surgery or for therapeutic embolisation of bronchial vessels. Treatment with tranexamic acid resolved the haemoptysis.

Keywords: Haemoptysis; congenital heart disease; tranexamic acid

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Haemoptysis in young people with congenital heart disease usually results from enlarged bronchial collateral arteries associated with pulmonary atresia, or from pulmonary hypertension and pulmonary vascular obstructive disease.[1] It has been reported that haemoptysis is not predictive of mortality in Eisenmenger syndrome.[2] However, it can cause serious decompensation and is an alarming symptom. Mortality does occur,[3] particularly if the amount of blood loss is large.[4]

Case report

Our patient is a 25-year-old woman with pulmonary atresia, a ventricular septal defect, and multiple aorto-pulmonary collateral arteries. She underwent a modified left Blalock–Taussig shunt operation at the age of 2 months and replacement of this shunt at the age of 5 years. At 6 years of age, three stenosed right-sided collaterals were enlarged and a Gortex shunt was inserted. She was subsequently reassessed for full repair, but deemed unsuitable. Pulmonary hypertension was diagnosed at the age of 11 years – mean right pulmonary artery pressure 53 millimetres of mercury. She remained well during her teenage years and early adult life.

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Following three episodes of sudden-onset haemoptysis on board a long-haul flight, she was hospitalised. There were no chest pains, infective symptoms, or dyspnoea. Approximately 600 millilitres of blood was produced in the first 24 hours. She was cyanosed and had episodes of desaturation down to 60% on oximetry. Her coagulation profile was normal, and she was on no regular medications. Her initial haemoglobin level was 201 grams per litre.

Computerised Tomographic Pulmonary Angiography revealed a large overriding ascending aorta, pulmonary atresia, multiple collateral arteries, no pulmonary embolism, and patchy attenuation in the left lower lobe. Echocardiography showed normal left ventricular size and function and moderate right ventricular dilatation with low-normal systolic function. Magnetic resonance imaging defined the pulmonary vessels and shunts (Fig 1).

Our patient continued to experience episodes of haemoptysis in the hospital, despite therapy with morphine as a cough suppressant and sedative. Her haemoglobin level fell to 159 grams per litre but she remained haemodynamically stable. She was given a blood transfusion. Therapeutic embolisation was considered to be too risky because of her pulmonary supply. Eventually, tranexamic acid was commenced by intravenous infusion of 1 gram over 30 minutes, followed by 1 gram orally three times daily. She experienced no further episodes of haemoptysis after commencement of tranexamic acid,

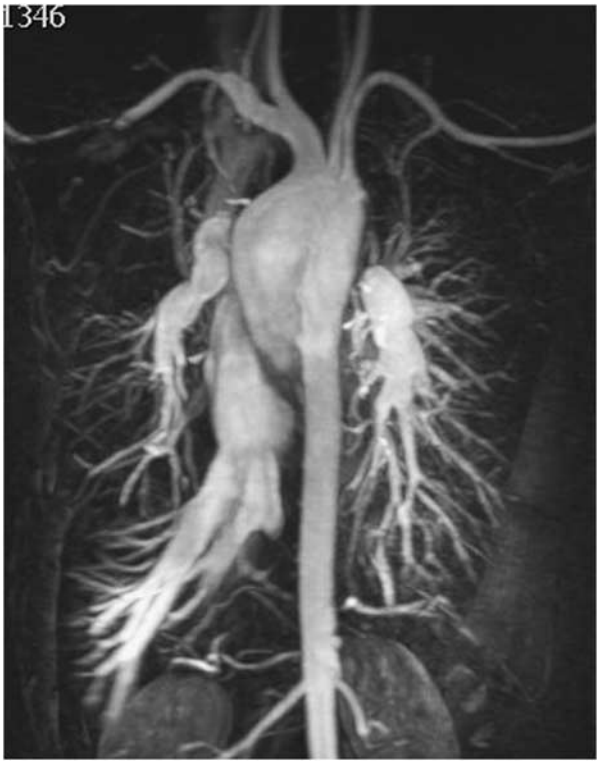

Figure 1.
Magnetic resonance imaging of the aorta and pulmonary vessels. The pulmonary trunk is absent. The left Blalock shunt is patent and is the sole arterial supply to the modified left lung. The right Blalock shunt is occluded and there is focalisation of multiple aorto-pulmonary collateral arteries with a patent connection from the ascending aorta.

which was stopped after 10 days. Bosentan therapy for pulmonary hypertension was commenced before discharge.

Discussion

Tranexamic acid is a lysine derivative that blocks the lysine binding sites on plasminogen, preventing the plasmin–tissue plasminogen activator complex from binding to fibrin, thus inhibiting fibrinolysis. The use of tranexamic acid has been well established in many areas including in the prevention and treatment of postpartum haemorrhage,[5] haemoptysis in cystic fibrosis,[6] and reduction in red blood cell transfusions during surgery.[7] Recent evidence also supports its use in bleeding trauma patients.[8] However, we found no cases reporting tranexamic use in congenital heart disease-related haemoptysis.

For haemoptysis in cystic fibrosis and in congenital heart disease, bronchial artery embolisation has become an accepted and appropriate mode of therapy.[9,10] This was not possible in our patient because her pulmonary supply was dependent on major aorto-pulmonary collateral arteries. Similarly surgical intervention was not an option.

In conclusion, we report a patient with congenital heart disease who had recurrent severe haemoptysis episodes that ceased after commencement of tranexamic acid. There is a good evidence base for the successful use of tranexamic acid in other patient populations. Further studies are needed to determine whether tranexamic acid should be routinely used as therapy in the treatment of haemoptysis in congenital heart disease, particularly when invasive methods are not an option.

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