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INTERNAL THORACIC ARTERY RUPTURE IN A CHILD WITH TYPE IV EHLERS-DANLOS SYNDROME

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Ehlers-Danlos syndrome (EDS) is an inherited connective tissue disorder characterized by articular hypermobility, dermal hyperelasticity, and widespread tissue fragility that results from gene mutations affecting the structure or assembly of collagen. Type IV or arterial-ecchymotic type EDS has been associated with rupture of large arteries.[1] Rupture of the internal thoracic artery (ITA) has been previously reported in an adult in this Journal,[2] but not in childhood. Here we present the case of an 8-year-old boy with type IV EDS and ITA rupture.

Clinical summary. An 8-year-old white boy with EDS type IV had a 2-day history of right back and shoulder pain and circulatory collapse. His medical history was significant for subglottic stenosis and tracheostomy as an infant, and there was no history of recent trauma other than minor contact while playing with another child. He had recently been treated with oral corticosteroids and antibiotics for bronchitis with resolution. Initial evaluation at an outside hospital found complete opacification of the right lung field with associated

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atelectasis and a hemoglobin value of 9 g/dL. On arrival in our hospital, examination revealed a thin, frail child with tachycardia, moderate respiratory distress, decreased breath sounds over the right hemithorax, and a hemoglobin level of 7 g/dL. A No. 24 chest tube was placed in the right side of the chest with an initial output of 250 mL of blood and partial clarification of the chest radiograph. Because of persistent bleeding, an aortic arteriogram was obtained. This disclosed an interruption at the origin of the right ITA with distal reconstitution. The interventional radiologist thought that embolization was not indicated because of the anatomy and fragility of the arteries with this disease.

Because the patient continued to have active bleeding, he was taken to the operating room. A right thoracotomy was performed. A retained hemothorax and a large hematoma at the origin of the right subclavian artery with back bleeding from the distal ITA were found. The distal artery was suture ligated and the origin of the ITA at the subclavian artery was subsequently closed with difficulty because of extreme fragility of the vessel. The patient did well after the operation, with a stable hemoglobin value and a clear chest radiograph. The chest tubes were removed on postoperative day 3 and the patient was discharged to his home on postoperative day 6.

Discussion. At least 10 types of EDS are currently recognized and each result from a genetically transmitted disorder in collagen synthesis. Type IV EDS, or arterial-ecchymotic type, is the result of abnormal collagen III formation, which is inherited autosomally.[1] This syndrome is associated with rupture of large arteries, as well as rupture of abdominal viscera.[3,4] Numerous reports of disruption of aortic branches have been reported and may occur spontaneously, as a result of trauma or of pregnancy-induced stress.[4,5] Vascular rupture as the presenting finding in a patient with previously undiagnosed EDS type IV has been reported.[6] Other inherited disorders, including von Recklinghausen disease, Marfan syndrome, and hereditary hemorrhagic telangiectasia, have also been associated with spontaneous vascular rupture. In our patient, the possibility of minor trauma was reported but not believed to be significant. A search of the literature fails to find a reported case of ITA rupture in a pediatric patient. The role of corticosteroids in this case is unclear. However, a case of spontaneous ITA rupture was reported in a 52-year-old woman with type IV EDS who had recently been treated with a course of oral corticosteroids.[2] Repair or ligation of ruptured vessels may be difficult because of tissue friability and inability of tissue to hold suture. Arteriography in patients with type IV EDS is associated with high mortality and complication rates, 17% and 67%, respectively, and the use of fine catheters is advised if arteriography is unavoidable.[5] Long-term survival is unfortunately limited, with mortality rates exceeding 60% before middle age. Death results mainly from vascular catastrophes.[1,5]

Conclusion. Arterial rupture is frequently associated with type IV EDS. However, ITA disruption in a pediatric patient has not been reported in the literature and is believed to be a rare occurrence. The occurrence of hemothorax in this disease should mandate prompt surgical intervention.

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