Two cases of Noonan syndrome: aortic coarctation causing a giant aneurysm of the descending aorta

Noonan sendromlu iki olgu: İnen aortta büyük anevrizmaya neden olan aort daralması

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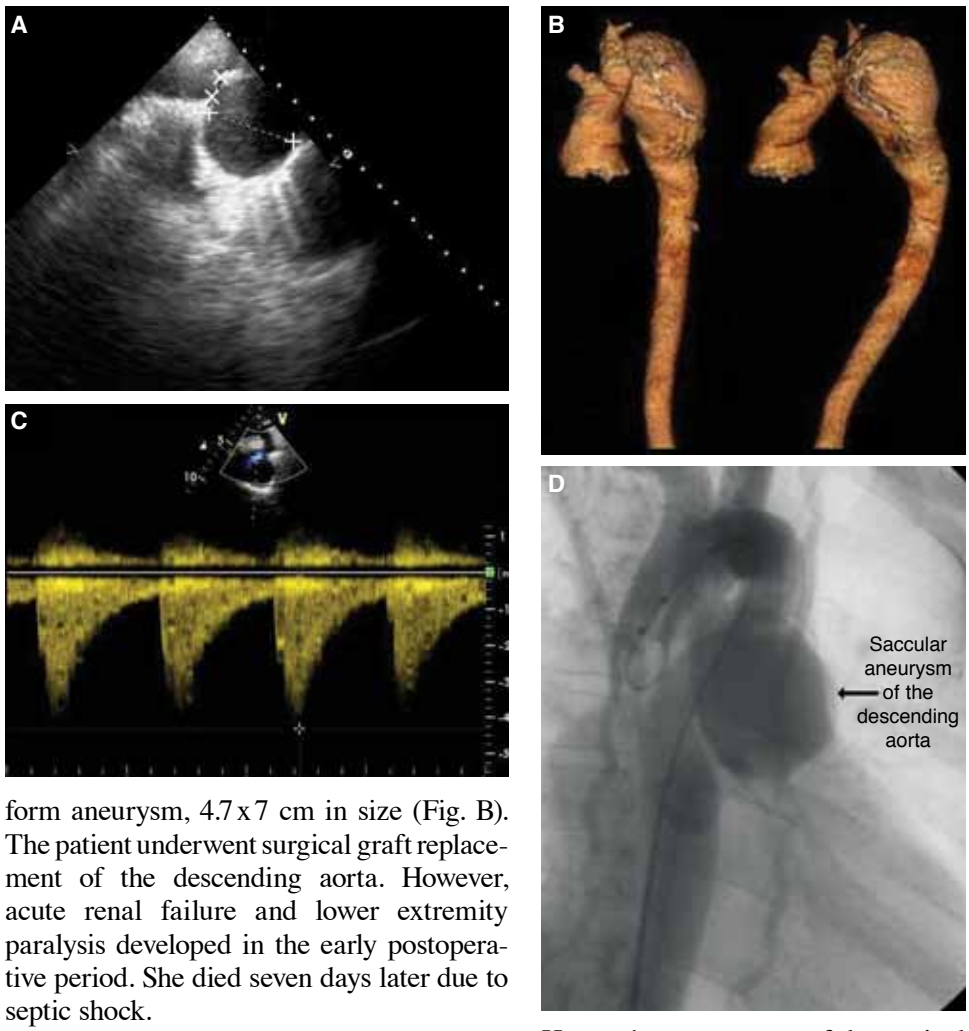
Aortic aneurysm of the descending aorta is a very rare condition in childhood and is one of the most serious complications of aortic coarctation. Aortic dilatation is not a typical condition in Noonan syndrome, it may develop in later stages of the postoperative period.

◆ A 16-year-old girl was referred with the diagnosis of Noonan syndrome. She underwent surgical closure of a patent ductus arteriosus when she was 10 months old. On physical examination, there was a grade 2/6 systolic murmur on the lower left sternal border. Echocardiographic examination revealed aneurysmal dilatation of the descending aorta (Fig. A). Computed tomography demonstrated a large fusiform aneurysm, 4.7 x 7 cm in size (Fig. B). The patient underwent surgical graft replacement of the descending aorta. However, acute renal failure and lower extremity paralysis developed in the early postoperative period. She died seven days later due to septic shock.

◆ A 15-year-old boy was admitted with chest pain and systemic arterial hypertension. Physical examination revealed high blood pressure (140/90 mmHg), a grade 2/6 systolic murmur, and weak femoral pulses. Two-dimensional and Doppler echocardiography demonstrated a saccular aneurysm of the descending aorta, 5.6 x 5 cm in size, associated with aortic coarctation, with typical continuous wave Doppler display across the coarctation. The patient underwent left heart catheterization, during which a pressure gradient of 75 mmHg was measured across the coarctated segment (Fig. C, D). He underwent successful surgical replacement of a Dacron tube graft interposed to the involved aortic segment. Postoperative period was uneventful and echocardiography showed a normal flow pattern.



Figures. (A) Echocardiography and **(B)** computed tomography showing fusiform aneurysmal dilatation of the descending aorta. **(C)** Typical continuous wave Doppler display across the coarctation obtained from the suprasternal view. **(D)** Catheter angiography showing a large saccular aneurysm after the narrowed segment in the descending aorta.