Uncontrolled hypertension in a child with Hurler syndrome

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A 15-year-old girl with Hurler syndrome was evaluated for recurrent hypertensive episodes with peak systolic pressures in excess of 175 mmHg. CT angiography demonstrates post-ductal long-segment aortic stenosis and beading of the descending aorta extending to the iliac bifurcation (Figs. 1 and 2).

Hurler syndrome is a mucopolysaccharidosis secondary to a deficit of lysosomal enzyme, alpha-L-iduronidase sulfate with characteristic skeletal, visceral, neurological and cardiovascular anomalies [1]. Hypertension has been described in Hurler syndrome in up to 32% of patients [2]. Mechanisms suggested are congenital or acquired architectural anomalies, atherosclerotic aortoiliac occlusive disease and

Fig. 1 Sagittal CTA

Fig. 2 Volume-rendered 3-D reformatted CTA

chronic upper airway obstruction [2]. Coarctation is a rare but described phenomenon in the presentation of Hurler syndrome [2]. Glycosaminoglycan accumulation in intimal cell and macrophage lysosomes leads to myointimal proliferation and aortic narrowing [1]. Although uncommon, aortic stenosis should be included in the differential diagnoses in children with Hurler syndrome and poorly controlled hypertension [2].

References

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