Compensation for external iliac vein hypoplasia via an inherent suprapubic shunt

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A 13-year-old girl was referred to our vein center by pediatricians owing to hypertrophic superficial venous circulation in her right groin, associated with local heaviness and the presence of two enlarged superficial venous branches emerging from her right medial thigh. The patient had previously undergone numerous examinations to exclude gynecological and gastrointestinal causes.

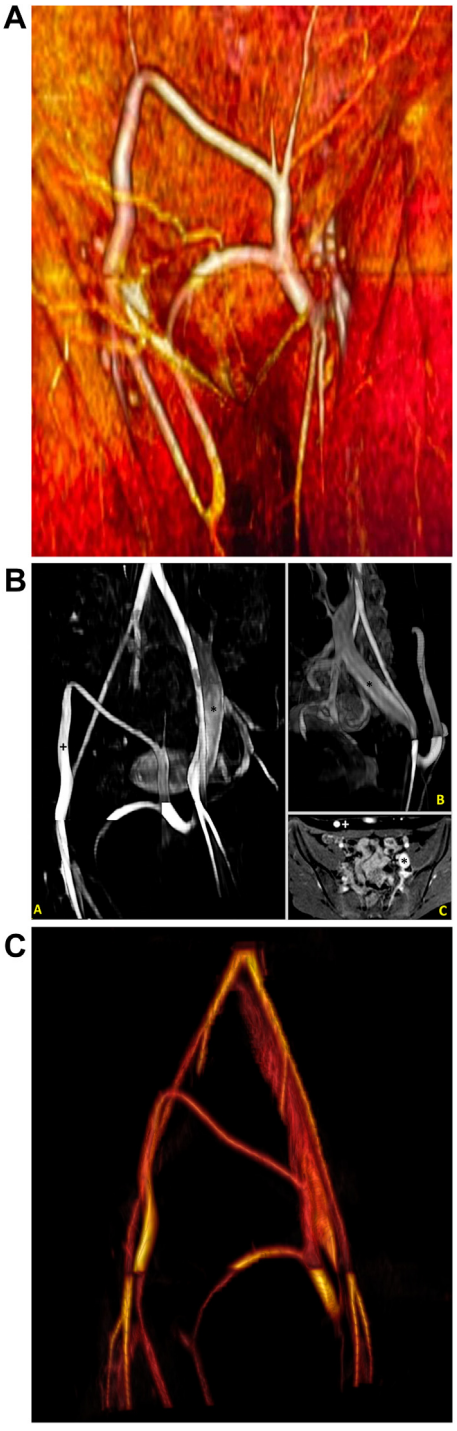
A duplex ultrasound scan revealed reflux in the right common femoral vein with competent femoral valves. Notably, the right great saphenous vein (GSV) did not show significant reflux in the calf, but a severe reflux was detected in the proximal thigh with an enlarged ascending collateral branch directed towards the suprapubic area. In the left limb, duplex ultrasound examination revealed common femoral vein competent valves and modulated flow.

Further exploration of the abdomen led to the diagnosis of external iliac vein agenesia. To better define the anatomy, she underwent contrast-enhanced magnetic resonance venography, which revealed incomplete agenesis or chronic occlusion of the left external iliac vein with aberrant venous drainage (*A*/Cover and *B*). In *B*, the asterisk (*) represents the left common iliac vein (CIV), and the plus sign (+) represents the right GSV merging into the left CIV through a suprapubic collateral, owing to complete right CIV agenesis.

Two main branches were identified, sprouting from the left common femoral vein, and connecting respectively to the right external iliac vein and right GSV through a suprapubic collateral (*C*).

Venous malformations can manifest as hypoplastic or hyperplastic vessels, leading to obstruction or dilation, depending on the case.[1] Embryologically, iliac veins develop from the posterior cardinal veins, which progressively regress and leave remnants like the renal segment of the inferior vena cava and the iliac veins.[2]

External iliac vein agenesia is typically associated with Klippel-Trenaunay syndrome, which shows an incidence of 8%.[3,4] However, this young lady did not present with the typical associated triad of varicose veins, asymmetric limb growth, and arteriovenous malformation, increasing the likelihood of isolated left external iliac vein agenesia, presenting with an incidence of less than 0.09%.[5] Remarkably, the patient did not show signs of deep vein thrombosis; therefore, she was recommended a conservative treatment using compressive stockings, and



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normal lifestyle owing to her young age. She signed the institutional informed consent for the publication of his clinical information and images.

DISCLOSURES

None.

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