

Rebecca Green BA, Xiaokui Gu MA, Eva Kline-Rogers MS, RN, James Froehlich MD, Pamela Mace RN, Jeffrey W Olin DO, Heather Gornik MD, MHS, Kevin E Meyers MD

**Title:** The US Registry for Fibromuscular Dysplasia (USFMD): Results in the First 24 Patients < 18 Years

**Background:** FMD has been described in virtually every arterial bed. The prevalence and natural history of FMD in children is unknown. This is the first multicenter description of 24 children with FMD.

**Objective:** Describe the demographics, presenting signs and symptoms (s&s) of FMD in children.

**Methods:** We reviewed the clinical features, presenting s&s and vascular beds involved in the first 24 patients <18 years in the USFMD registry from 3 sites from Jan 2009 until Oct 2014. We compared them with 999 adults.

**Results:** Mean age at diagnosis was  $6.8 \pm 4.3$  yrs (16 d to 16 yrs). FMD is more common in males <18 yrs 37.5% vs. 6% ( $p < 0.0001$ ). Fewer children with FMD are Caucasian 63.6% vs. 91.5% adults ( $p = 0.00039$ ). Family history is > in children (19%) vs. adults (4.7%) ( $p = 0.019$ ). Family history of stroke is < in children (19.0%) vs. adults (48.2%) ( $p = 0.008$ ). Children reported prior history of HTN 91.7% vs. 69.9% ( $p = 0.022$ ). Pediatric & adult FMD patients report a similar history of headache 61.9% vs. 65.6% ( $p = 0.82$ ). Presenting s&s of FMD in children are shown in Table 1, with 4.7% asymptomatic at presentation. Children rarely present with carotid bruits 5% vs. 34.6% ( $p = 0.0036$ ). Renal diagnostics include US 77.8% vs. 81.4% ( $p = 0.76$ ), CTA 38.1% vs. 37.4% ( $p = 1$ ), MRA 10% vs. 13% ( $p = 1$ ) and angiography 75% vs. 41.7% ( $p = 0.0014$ ) in children vs. adults. FMD affects the renal vasculature in children 95.8% vs. adults 69.7% ( $p = 0.0029$ ), extra-cranial carotid vessels less in children 22.2% vs. 73.3% ( $p = 0.0022$ ). The aorta is involved in 28.6% of children vs. 2.4% of adults ( $p = 0.00056$ ) and mesenteric arteries 46.2% vs. 16.2% ( $p = 0.012$ ).

**Conclusion:** FMD affects more boys than adult males; is seen in more minorities in children in this sample; and a family history is more common. Most children present with HTN; many present with headache and abdominal bruits. More children undergo angiography to diagnose FMD. The renal vasculature including the abdominal aorta is involved in more children than adults. That there are few children with carotid involvement may be ascertainment bias.

Table 1 Presenting Symptoms and Signs

Symptoms	Age < 18	Age $\geq$ 18	p-value
Headache	11/22 (50.0)	551/918 (60.0)	0.38
Dizziness	2/19 (10.5)	283/837 (33.8)	0.046
Flank/Abdominal Pain	2/20 (10.0)	126/848 (14.9)	0.75
<b>Signs</b>			
Hypertension	23/24 (95.8)	601/942 (63.8)	0.00072
Abdominal Bruit	3/20 (15.0)	74/832 (8.9)	0.41
Carotid Bruit	1/19 (5.3)	218/850 (25.6)	0.057