

## **Fibromuscular Dysplasia (FMD) and children**

The most prevalent form of FMD identified in children and young adults is intimal fibroplasia, which is described by long, irregular or smooth, focal stenosis. Persons with FMD may be asymptomatic and only diagnosed at routine medical visits. There seems to be no pre-pubertal sex predilection.

### **Clinical presentations**

Children often report various non-specific symptoms including headache, insomnia, fatigue and chest or abdominal pain, which may be indicative of hypertension, depending on which artery(s) is affected. A minority of children with hypertension present with neurological symptoms including seizures, transient ischemic attacks, cerebral infarctions (stroke), subarachnoid hemorrhages, and cranial nerve palsies. FMD of the head and neck arteries is increasingly recognized as a cause of stroke in children.

### **Classifications of FMD**

FMD lesions are classified according to the arterial layer they affect: intima, media, or adventitia. As stated previously, the most common form of FMD in children is intimal fibroplasia, which can occur in any arterial bed. Intimal fibroplasia may present as either a focal band-like narrowing, or a long, tubular narrowing.

### **Diagnosis of FMD**

FMD can be diagnosed by invasive means and strongly suspected by non-invasive means. Non-invasive testing includes duplex ultrasonography, magnetic resonance angiography (MRA), and computed tomographic angiography (CTA). The accepted gold standard remains conventional angiography, however, by virtue of its invasive nature and risk involved its place in initial diagnosis continues to be challenged.

## **Treatment of FMD in children**

Both medical and surgical treatment options exist. Treatment involves controlling high blood pressure, re-establishing vascular flow, preventing clotting of the affected vessel(s), and eliminating factors that contribute to further vessel damage (e.g. smoking in teenagers). Use of a low estrogen dose birth-control pill in teenagers with controlled blood pressure should be weighed against a possible increased risk of vascular clotting.

Medical therapy in children includes anti-hypertensive medications to control blood pressure. Aspirin therapy should be considered in children with FMD as anti-platelet therapy may reduce the risk of thrombosis in affected vessels.

Percutaneous transluminal renal angioplasty (PTRA) remains the treatment choice for renal-artery FMD. Indications for this intervention include recent or rapid onset of hypertension and difficulty in controlling high blood pressure with anti-hypertensive medications. Associated adverse events may include recurrent stenosis, arterial occlusion with renal loss, and arterial rupture and pseudo aneurysm formation. Due to the various medical therapies and percutaneous balloon angioplasty surgical reconstruction is reserved for patients with complex FMD.

## **Prognosis**

There are presently no *specific* studies or reports on the long-term prognosis and outcome of FMD in children. The causes, natural history, management and long-term outcomes of FMD in children require further research and evaluation.