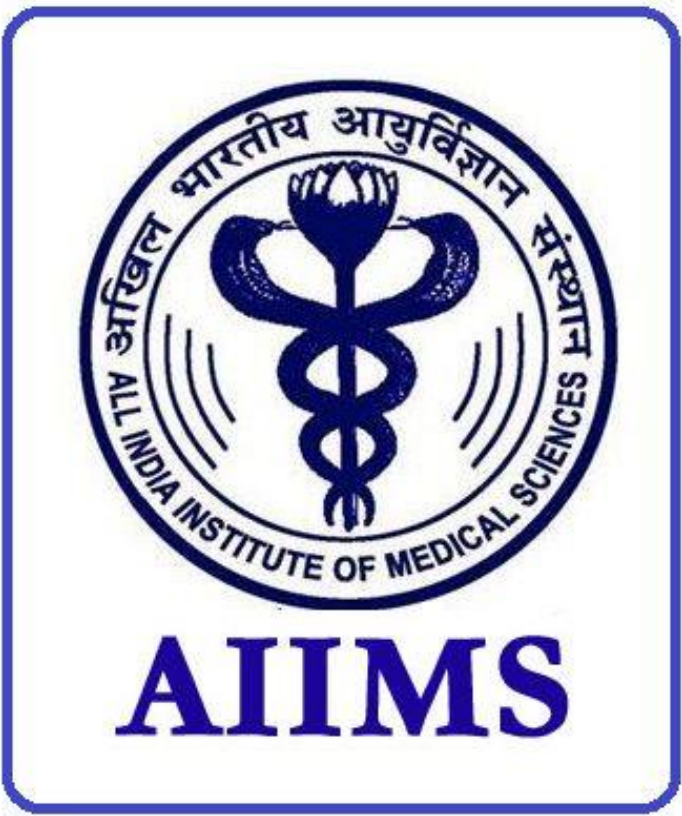




Cranio-Cervical Fibromuscular Dysplasia In Pregnancy Presenting With Hypertensive Emergency Crisis : Case Report And Review Of Literature

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BACKGROUND

- The exact prevalence of Fibromuscular Dysplasia (FMD) is unknown; however the prevalence of renal FMD is 0.4 %, whereas the estimated prevalence of cranio-cervical FMD is much lower at 0.1%.
- There is a paucity in the management protocol of FMD in pregnancy.

CLINICAL PRESENTATION

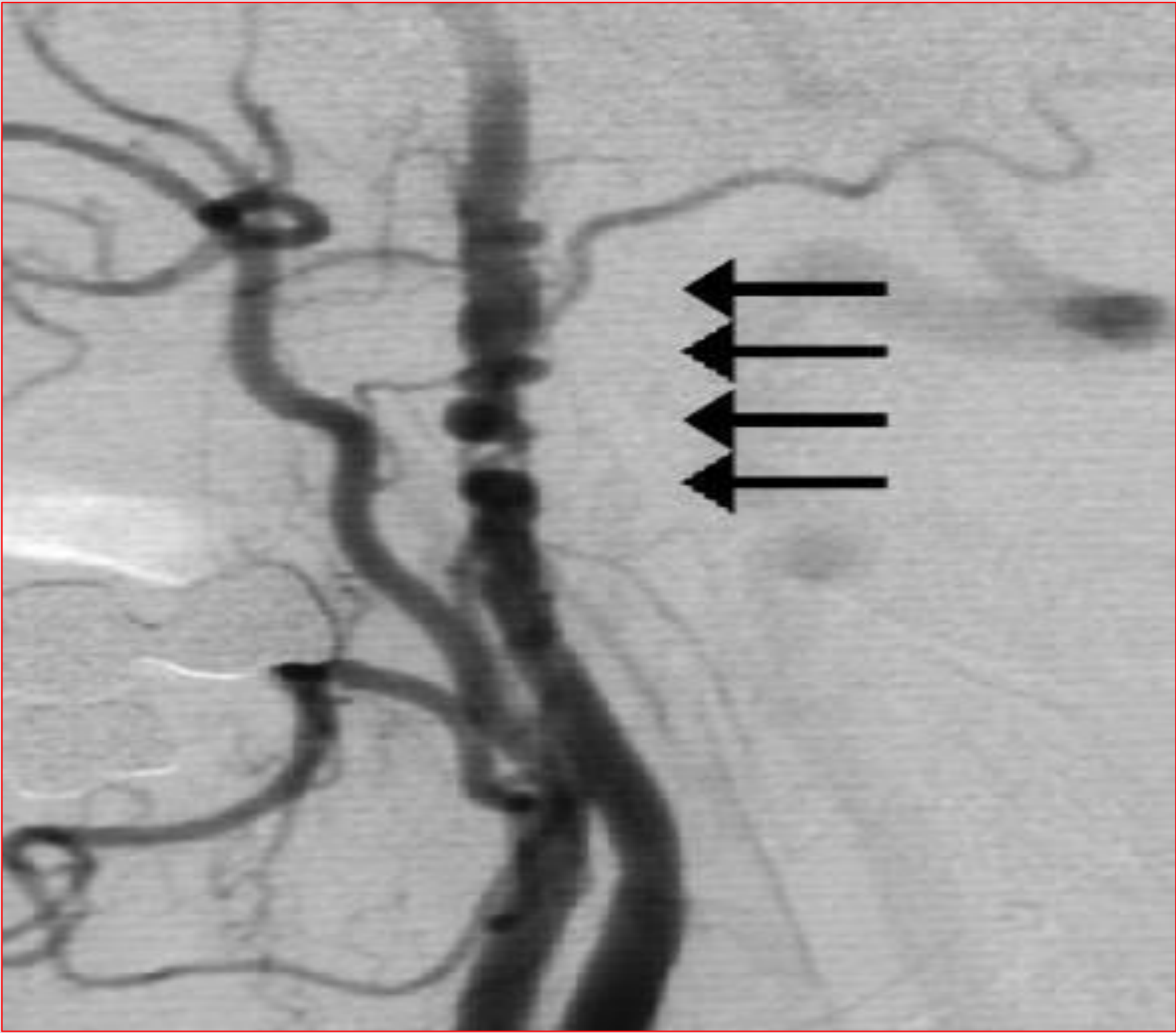
- 38 year old primigravida woman, a diagnosed case of cranio-cervical FMD was started on 100mg Labetalol tablet thrice a day in 1st trimester which was later increased to 200mg thrice a day in 2nd trimester.
- She had a history of sub-arachnoid hemorrhage with right posterior inferior cerebellar artery and right superior cerebellar artery aneurysm, bilateral vertebral artery stenosis, and residual right oculomotor nerve paresis.
- She presented with hypertensive emergency crisis at 32 weeks and was managed by a team of obstetricians, neurologists, and cardiologists.
- She then underwent an emergency caesarean section with copper T 380A insertion for contraception.
- A premature baby which was small for gestation age was delivered.
- In post-partum period she had uncontrolled hypertension and was started on 10mg Amlodipine tablet twice a day and Metoprolol 50mg once a day.

EXAMINATION

- 3rd cranial nerve paresis was noted, however there was no carotid bruit.
- Cardiovascular and respiratory examinations were normal.
- Per abdominal examination revealed a uterus which was at 28 weeks gestation with a relaxed fetus in breech presentation; and fetal heart rate of 135 beats per minute.

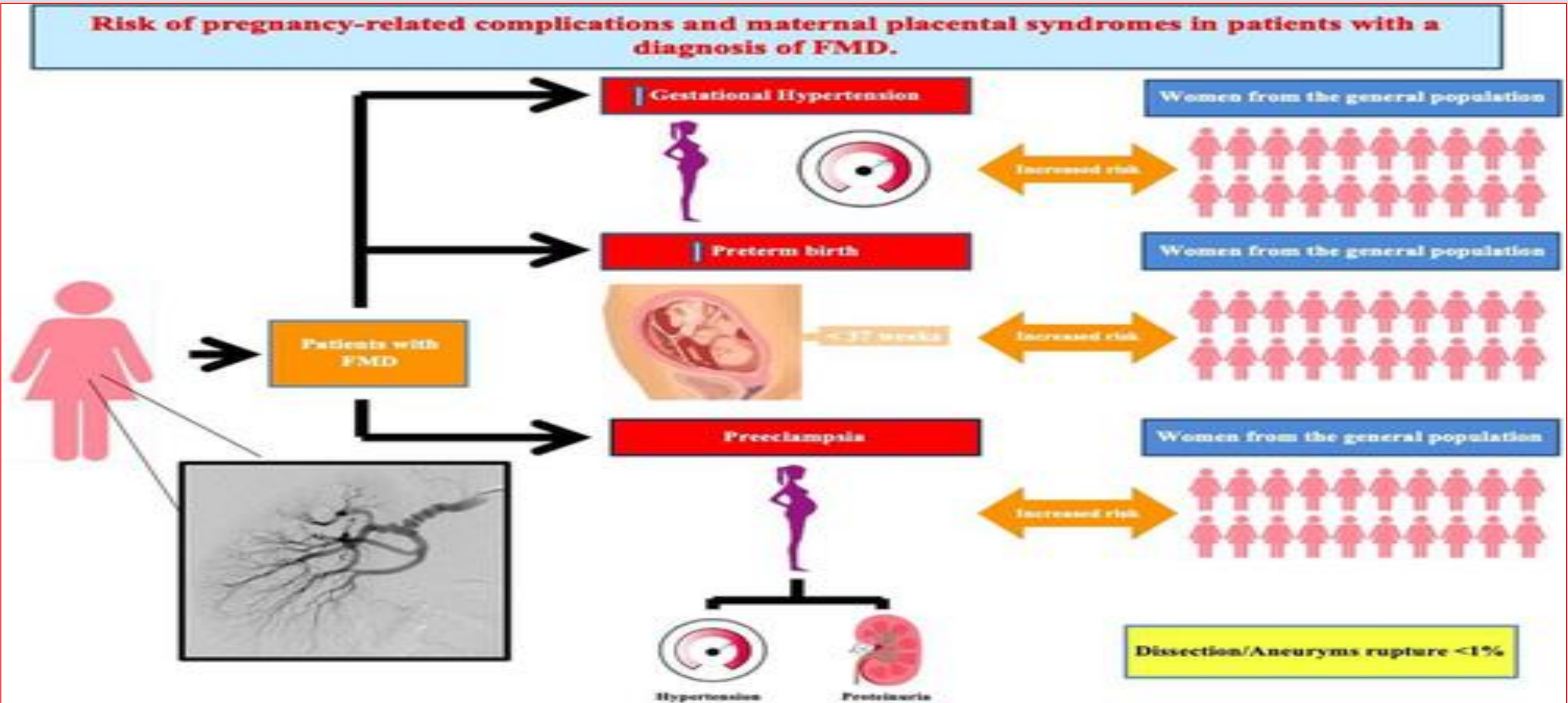
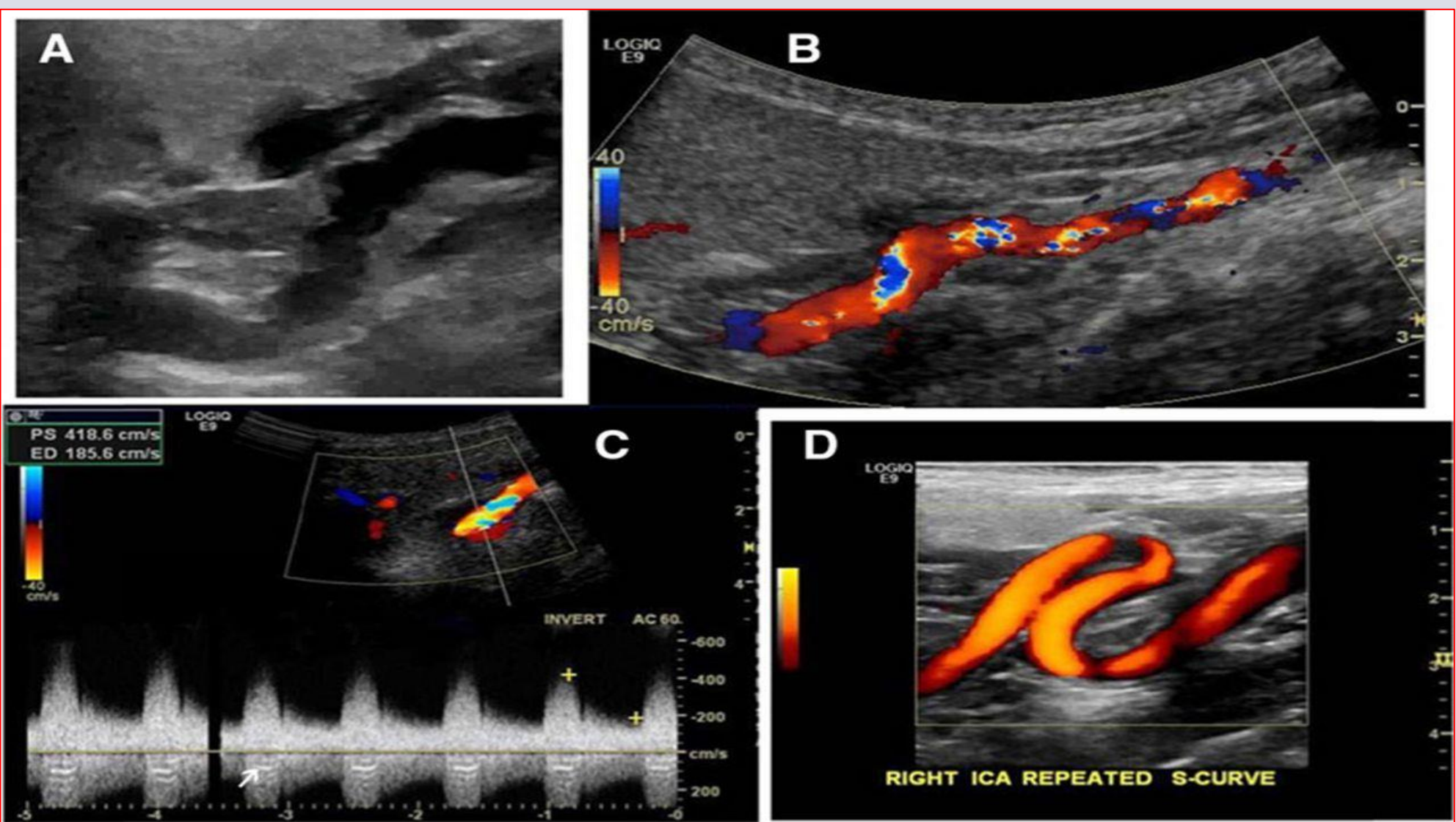
Table 3. Presenting Signs and Symptoms Among Patients in the United States Registry for Fibromuscular Dysplasia⁵

Symptoms/Signs	n (%) Divided by 447
Hypertension	285 (63.8)
Headache	234 (52.4)
Current headache	135 (30.2)
History of headache	173 (38.7)
Pulsatile tinnitus	123 (27.5)
Dizziness	116 (26)



The string-of-beads sign with alternating regions of lumen narrowing and vessel dilatation on angiogram of the ICA

IMAGES



Typical duplex ultrasound findings of carotid fibromuscular dysplasia. **A**, B-mode imaging showing the beading and tortuosity of the mid and distal internal carotid artery. **B**, Color Doppler of the distal internal carotid artery exhibiting the typical pattern of tortuosity and marked turbulence. **C**, Color Doppler showing turbulence and spectral analysis demonstrating high peak systolic (419 cm/s) and end-diastolic velocities (186 cm/s). The “seagull” sign (**arrow**) indicates that the stenosis is quite severe. **D**, Color power angiography demonstrating severe tortuosity and redundancy (S curve) of the internal carotid artery

MANAGEMENT

- On admission the blood pressure (BP) was 180/100 mm of Hg.
- Injection Labetalol 20 mg was given stat intravenously followed by 40mg, then 80mg. BP remained uncontrolled at 170/100 mm of Hg.
- Injection Labetalol infusion was started at 5mg /hour.
- A loading dose of injection Magnesium sulfate 4 grams was given, followed by 1mg/kg/hour infusion.
- She was planned for emergency caesarean section in view of severe pre-eclampsia crisis uncontrolled on medication.

CONCLUSION

- Patients with cranio-cervical fibromuscular dysplasia need a close BP monitoring in the antenatal and also in the post-partum period to avoid catastrophic complications like stroke.
- They also warrant a strict foetal surveillance and a multi-disciplinary team has to manage FMD patients in pregnancy to avoid adverse perinatal outcome.

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