Top Ten Things To Know
Fibromuscular Dysplasia: State of the Science and Critical Unanswered Questions

1. Fibromuscular dysplasia (FMD) is nonatherosclerotic, noninflammatory vascular disease that may result in arterial stenosis, occlusion, aneurysm, or dissection.

2. The cause of FMD and its prevalence in the general population are not known.

3. This statement outlines the historical perspective of FMD as well as the epidemiology and genetic considerations for furthering our understanding of the disease.

4. Histological classification systems of FMD were developed as early as 1971; however, none have been uniformly accepted because of obtuse terminology and uncertainty of the relationship of the histological variants, given that the pathogenesis is fundamentally unknown.

5. Further limiting the utility of all histopathological classifications is the realization that FMD today is a disease almost exclusively diagnosed radiographically.

6. Acknowledging the practicality and appropriateness of an angiographic classification, the American Heart Association proposes a minor modification of the classification recommended by the European Consensus in 2012.

7. Multifocal disease is the classic string of beads on arteriography and is represented by medial fibroplasia in virtually all adults. Focal disease is without regard to lesion length and is usually caused by intimal fibroplasia, but may also be caused by medial hyperplasia or adventitial FMD. Patients may have simultaneous multifocal and focal disease in different vascular territories.

8. The paper discusses clinical manifestation of FMD across various arteries and patient populations as well as differential diagnosis, diagnostic strategies, treatment, and therapy options.

9. There is a great need for additional FMD research in several areas including pathogenesis, diagnostic approach, and natural history and outcomes of FMD.

10. Significant advances in our understanding of FMD will undoubtedly require collaboration across a large network of research and clinical centers in the United States and abroad.