

Updates from FMD group including UK first FMD patient information day

Patient information day

The auspicious date of 19.01.19 saw the UK's first patient information day on Fibromuscular Dysplasia (FMD) held at Salford Royal Hospitals NHS Foundation Trust.

The day was aimed at providing an overview on FMD, an update on international registry data and the most cutting edge clinically relevant research. We were privileged and honoured to have guest speaker, Sally Bee (<https://www.sally-bee.com/>) share her journey, first with spontaneous coronary artery dissection (SCAD) and then FMD ; and Prof Alexandre Persu, an international FMD expert and head of the European FMD Registry join us from Belgium. Dr Tina Chrysochou provided an overview of FMD and the UK collaborative effort of the FMD specialist interest group under the Renal Rare Disease Registry (RaDaR).

Following patient request ahead of the meeting, the day also benefitted from local neurology and neuro-radiology expertise by SRFT colleagues, Dr Martin Punter and Dr Amit Herwadkar. Prof David Adlam, lead for the UK SCAD research group, discussed the latest developments in SCAD. This included his latest collaboration showing genetic susceptibility to FMD and SCAD : PHACTR1 is associated with an increased risk of FMD, cervical artery dissection, SCAD and migraine. Dr 'Bean' Dhaun, consultant nephrologist from Edinburgh shared a case study on an unusual presentation of FMD to highlight how this condition can present in multiple ways, and how clinicians should have a high index of suspicion when cardiovascular events occur outwith the traditional risk factors or atherosclerosis.



16:17 · 19/01/2019 from Salford, England · Twitter

The day finished off with a workshop during which patients and carers shared their experiences and devised a wish list for management within clinical services and research ideas for FMD in the UK.

Have a look at **#FMD2019** for tweets from day. Feedback from the meeting was excellent, with all individuals finding it extremely or very worthwhile. We are very grateful to Kidney Research UK (KRUK) and Kidney Care UK (KCUK) who kindly sponsored the event. Their sponsorship also went towards filming the event which will be made publically available.

Update from the literature

FMD is a non-atherosclerotic, non-inflammatory disease of the blood vessels that causes abnormal growth within the wall of an artery.

In the UK, FMD was historically thought to carry a benign prognosis. As such, research in this area has lagged behind. Yet, FMD can be associated with severe hypertension, ischaemic or haemorrhagic stroke, myocardial infarction and end stage renal disease. It can lead to invasive procedures such as percutaneous angioplasty, reconstructive surgery, or intracranial aneurysm clipping. Thus, both the disease and its treatment can lead to significant morbidity and mortality.

Through information gleaned from the American and European registries, we are now challenging previously held information on FMD e.g:

- 1.** Common clinical manifestations of FMD are actually more diverse. In the US FMD registry, hypertension was the most common presenting manifestation, affecting 63.8% of patients; however, other common signs and symptoms, included headache (52.4%), pulsatile tinnitus or “swooshing” noise in the ears (27.5%), dizziness (26%), cervical bruit (22.2%), and neck pain (22.2%).
- 2.** Two thirds of patients have additional arterial bed involvement.
- 3.** Men and women present differently. While women outnumber men 9:1 in registries, renal and visceral involvement with dissection and aneurysm is more common in men.
- 4.** FMD remains under-diagnosed in European and American registries and diagnoses often delayed
- 5.** The etiology remains unclear
- 6.** Research is underway, but more is needed to glean how genetics, environmental stressors and hormones may play a role

The recent publication of the international consensus guidelines could not have been timed better for the patient information day to help health care professionals navigate clinical management of FMD:

https://journals.lww.com/jhypertension/Fulltext/2019/02000/First_international_consensus_on_the_diagnosis_and.2.aspx?fbclid=IwAR1P09reGBuTl1rXPxUQWtu3PMJsNwKmjYI_Bv75BP-FHwAserFlmkMT9OY

The UK initiative

In the UK, the FMD RaDaR Specialist Interest Group is aiming to raise awareness and provide advice about the best strategy for the diagnosis and management of this still underdiagnosed disease. Patients with FMD in any arterial bed can be uploaded to FMD RaDaR - rarerenal.org/rare-disease-groups/fibromuscular-dysplasia-rdg/ (**@RenalRadar**). We have linked with a patient group who formed the FMD Society of UK and Ireland (**@FmdsUK**) in 2018 and are a valuable resource to signpost patients to.

Next steps

The RaDaR FMD group, led by Dr Tina Chrysochou are in the process of securing funds to enable the start of a national epidemiological study. Patients recruited to RaDaR will be approached for inclusion into this registry amongst other registration means. This database will align with the well-established European registry to ensure seamless collaboration and meaningful comparison of data.

Dr Tina Chrysochou (on behalf of the FMD RaDAR specialist interest group)