Newsletter

HELLO! As the newly elected chairperson of the Marfan Syndrome Support Group Ireland, I wish to take this opportunity to tell you a little about myself and the vision that I and the Board have for raising awareness of Marfan syndrome. I work in the Health Service Executive and am from County Kildare. I am effected by a connective tissue disorder and so are members of my family. It is very obvious to us all that when we present to our General Practitioners or Hospital Consultants, we are greeted with comments like "I must look that up" or " I have never heard of that condition". This type of response is certainly unnerving and doesn't allow for confidence in our medical personnel, however genuine they seem to be. This is unfortunately because connective tissue disorders are generally extremely rare and are exceptionally difficult to diagnose.

Our vision is therefore to raise as much awareness as possible of Marfan syndrome and similar conditions within the medical profession and general society. I feel we can all play a part in raising awareness, even by doing the simple things like joining our social media sites and signing up as a member of this group. In our situation our biggest tool for awareness raising is word of mouth and social networking. In that regard, I am encouraging as many people as possible to attend our next Annual General Meeting. I really look forward to seeing the new and existing members.

As always , kindest regards and good health

Stephen Toft

The annual Dublin Women's Mini Marathon was sponsored this year by VHI. We are delighted that 10 ladies partook in it, in awful weather, and raised and incredible €2,120 and awareness for Marfan . We very much appreciate their efforts.

In June this year, the Mid-West Vocal Academy and Music School in Co. Limerick, held their end of year Summer concert benefitting both ourselves, the Marfan Syndrome Support Group, and Crumlin Children's Medical Research Foundation. Each charity received €1,175 from the proceeds. We are very grateful to Owen Gilhooly, Director of the school for choosing us as part beneficiaries. Great awareness was raised as a result.

As a result of the above fundraiser, we also were later presented with a cheque by the Limerick Lions Club for €500!

We have silicone bracelets for sale for €1.50 each. All money goes directly back to support families and publish materials. It's vital that we raise awareness, therefore, hopefully, saving lives. Please let us know if you wish to order any.

We have a Page on Facebook, give us a 'like'!
www.facebook.com/marfanireland

We also have a private group - search Marfan Support Group Ireland (Private Group)

Clinical Trial Results and Recommendations

Following on from a clinical trial in the USA on the use of the beta blocker Atenolol and angiotensin receptor blocker (ARB) Losartan in slowing down aortic growth, The Marfan Foundation have released the following guidelines.

Medications

Medications can help treat many types of problems with the heart and blood vessels. It is recommended that either a beta blocker or angiotensin receptor blocker (ARB) be started at the time of diagnosis of Marfan syndrome with the goal of reducing the rate aortic root enlargement. Doses suggested are those given in the 2014 Atenolol vs. Losartan randomized trial.

- **Atenolol** should be increased to a maximum dose of **4 mg/kg/day (not to exceed 250 mg/day)** with a goal of a 20% or greater decrease in average heart rate measured on a 24-hr recording. This high dose of atenolol is generally well tolerated.
- Alternatively, in patients who cannot tolerate beta blockers, **angiotension receptor blockers (ARBs) such as losartan**, provide a similar amount of protection against aortic enlargement. Losartan should be started at an **initial dose of 0.4 mg/kg/day and increased based on weight to a maximum dose up to 1.4 mg/kg/day, not to exceed 100 mg.**
- Since the trial data provides evidence that in younger patients both drugs are associated with a greater decrease in aortic-root z-score over time, beta blockers or angiotension receptor blockers should be prescribed at the time of diagnosis even in the youngest children. Therefore, it is recommended that once a diagnosis is made, with or without aortic dilation, medical therapy should be started, maintained and continued after surgery indefinitely.
- Based on the patient's history, individualized treatment plans must be developed when deciding on which medical therapy to use (beta blocker [atenolol] or ARB [losartan]). However, since the trial only investigated use of either atenolol or losartan, that is the only hard evidence- based recommendation that can made at this time. There are other ongoing which may additional information about combination therapy with both ARB drug and beta blocker
- There has been a small study of ACE-inhibitor therapy in Marfan syndrome; more information is needed before recommending the use of this class of agents for the prevention of aortic disease in Marfan syndrome.

Credit: THE MARFAN FOUNDATION