Notes from the FDSSUK Annual conference in Edinburgh

Saturday 1st June 2019

Dr. Michael Collins – Overview of Fibrous Dysplasia/McCune Albright Syndrome

A brief overview of FD/MAS was given with an explanation of how bones and hormones are affected by the disease.

We were shown an article dated back from 1936, which was written by Donovan McCune and Fuller Albright. There have been many different names for the disease of the years.

There is not much literature available so, NIH started a study in 1998 of patients of all different ages with FD/MAS. The data has been collected on each patient from all different types of Dr's involved within their care plans. Working internationally with other Dr's and sharing information has helped improve the literature in more recent times.

We were shown graphs with percentages from the study and the skull, pelvis and femur are the most commonly affected bones. We were shown examples of x-rays showing the spine and femur. It has recently been found that the pancreas can be affected.

There were also some images of skull-based FD and Dr. Collins explained the types of obstructions that the disease can cause to the optic nerve and ear canal. Early diagnosis, careful observation and watchful waiting is highly recommended over surgery.

We were then shown some slides with information on how the gene mutation happens. The disease establishes itself in the cells very early on during the pregnancy. It will not spread to other parts of the body or appear in unaffected bones later on in life. It is not inherited.

Dr. Collins explained the symptoms of precocious puberty for both girls and boys, including growth issues and he described the drugs that are available for this. Early diagnosis is best. He also went on to give a description of hyperthyroidism and phosphate issues and the treatments currently available.

Dr. Faisel Ahmed & Dr. Zilla Huma – Endocrine Issues in Children with Fibrous Dysplasia/MAS and Pain Management

Dr. Huma gave examples of how FD/MAS can be discovered in a patient – by bone pain, a minor injury, café au lait spots or early puberty. She showed us examples of two children one who had pain after injuring their arm in a fall, and the other who had café au lait spots. Both children were sent away with no further treatment by their hospitals. She explained that some patients have never been seen by a Paediatric Dr. or an Endocrinologist and have not been referred on by their Orthopaedic surgeons. Lots of patients have never been screened by their local care services.

Dr. Huma works at The Royal National Orthopaedic Hospital in Stanmore.

She showed us graphs of four of her patients who have been screened for growth, calcium, vitamin D, alkaline/phosphate, thyroid, parathyroid and puberty. She explained that phosphate medication is very difficult for children to take.

She made the following recommendations:

Thyroid tests should be carried out throughout childhood and puberty.

Height and weight checked regularly.

Phosphate levels checked regularly.

Pubertal progress should be documented.

Stephen McMurray – Hypnotherapy for Pain Management

Stephen gave a brief overview of his journey with FD/MAS. He is a trained Hypnotherapist.

He gave a brief history of hypnotherapy and described the difference between hypnotherapy and hypnosis, which is commonly seen on television. Hypnotherapy works at a sub conscious level which triggers the body's natural ability to reduce and distract attention away from the pain. Pain is not just physical, it affects emotions as well. Hypnotherapy is not available on the NHS.

Stephen showed us four good evidence examples from the BBC and various medical journals.

Dr. Robert Stanton – What you need to know before Surgery in Children

In order to understand the extent of the disease, a bone scan should be carried out at around the age of 5 years old. Dr. Stanton said that the orthopaedic literature on FD is limited.

He explained the difference between Monostotic FD and Polyostotic FD. Monostotic is not aggressive, only affects one bone and there is little deformity. Polyostotic FD is more aggressive, causes more deformity and thin and weak bones. It usually requires different surgical techniques.

Things that you must not do when treating Polyostotic FD:

Do not use the patients own bone for grafting or use cancellous graft.

Do not try to remove the diseased area of bone, as it will grow back.

Do not allow the deformity to get severe.

Things to remember:

The use of plates and screws is only temporary.

Rods are not always available for small children.

A single operation will not solve the problem.

Dr Stanton explained that it is challenging to reverse deformity especially in the femur. There is more fracture frequency in the femur as it is a weight bearing bone, but not so much in the upper limb bones. In the weight bearing bones, no surgery until necessary.

Goals after surgery:

Early weight bearing.

Avoid casts.

Minimize bed rest.

More intramedullary rods and less bone grafting.

He explained the different types of Orthopaedic Dr's and what questions to ask them:

Do you know the disease?

Do you have any patients with FD?

Have you done surgery to other patients with FD?

What can you tell me about MAS?

Dr. Stanton showed us some x-ray examples of an adult with severe pelvic disease and a child with a femur/hip screw.

Dr. Robert Stanton – Scoliosis in Fibrous Dysplasia

Once scoliosis has been noted it should be radiographed over time to determine the progression. Each patient should be examined. Bracing should be discouraged to those patients who have FD in their ribs. It should be followed up, especially in growing children. Bisphosphonates don't have an effect on Scoliosis.

We were shown x-rays of patients who have had surgery on their spine.

Sunday 2nd June 2019.

Professor Hamish Simpson – What you need to know before surgery in Adults

Reasons for operating:

To reduce pain and improve function.

To prevent pain and preserve function.

FD is not an easy disease to treat as there is no literature available to look at. More data is needed in a registry.

Considerations for surgery:

Chances of improving symptoms?

How big is the procedure?

Risks of alternatives?

Complications?

Types of Surgery:

Joint replacement

Straightening bone

Correction rotation

Leg lengthening

Professor Simpson talked us through types of material used in hip replacements in FD patients and we were shown slides with examples and images of both internal and external fixations that can be used in the weight bearing bones.

He has advised that your surgeon should have a surgical plan and a back-up plan in-case something can't be done.

Blood loss can be a big issue for FD patients and surgeons should use a tourniquet or limb elevation to prevent this.

We were shown some x-rays of patients who have had a leg lengthening procedure and Professor Simpson talked us through the process.

Robert Cameron – Citizens Advice Bureau - An Overview of Welfare Benefits

Mr. Cameron gave a brief overview of his career and showed us a short old black and white video of how CAB started. The video showed different conversations between different sets of people showcasing the types of advice people needed when there was no help available. Once the CAB was formed in 1939, people could visit the office and get the advice they needed.

We were shown a graph of the types of enquiries that are received, and the most common area was benefits at 42% followed by debt at 21%. The different types of benefit groups were explained.

Mr. Cameron went through each benefit for disabled people in detail and explained how the DWP assess it and the rates of payment that are available for each one. He also gave tips on how to fill in the form and what to ask for. As well as benefits he told us about the blue badge scheme, the taxi card scheme, disabled persons rail card and the motability scheme. The benefits covered within the presentation were:

Disability Living Allowance

Personal Independence Payment

Attendance Allowance

Carers Allowance

Universal Credit which is slowly replacing Income Support, income-based Job Seekers Allowance and Employment and Support Allowance, Child Tax Credit, Working Tax Credits and Housing Benefit.

Financial health checks are also popular topics and help is given to people on how to manage their bills.

Dr. Line Caes – The Journey of How Families Deal with a Paediatric Illness

Emotional responses to diagnosis were explained. Once a condition is diagnosed there are feelings of anxiety, fear, depression, denial and disorganisation for the parents. For the child there can be an impact on social development, resilience and self-esteem.

Dr. Caes explained the different types of support that are given to the family including coping strategies, relaxation, respite options, positive thinking and problem solving. She also told us what to do when your child is in pain.

When the child reaches adolescent age, there will be separate sessions with the child to help prepare them for making their own decisions with regard their medical conditions.

We were shown a video called the magic glove, hypnotic pain management for children.

Dr. Kassim Javaid – New Drugs, Bisphosphonates and their Side Effects and Pain Results from the Rudy Study

The pain in Fibrous Dysplasia is confusing, Dr's are not sure why it hurts. Some of the different types of pain were explained:

Scar pain – some people who have had multiple operations have numbness around their scar. Oil and massage can help with this.

Joint pain caused by Osteoarthritis – some patients may have reduced movement, stiffness and pain on walking.

Referred pain – this pain can be coming from nearby joins such as the spine and not FD.

Sudden Severe pain – this can be a fracture or an aneurysmal bone cyst.

Dr. Javaid explained the types of pain and issues that can be caused by craniofacial FD, including:

Psychological impact, hearing, vision and dental/maxillofacial problems.

The different types of Bisphosphonates - Pamidronate, Zoledronate and Risedronate were explained and how they work on the bones. We were shown scan examples of how they stick to the bones. It is advised that if one type of Bisphosphonate doesn't work for a patient, they may try switching to another, but if there is no response then the treatment should be stopped. The side effects of the drugs were explained including the flu like symptoms and very rarely problems with the jaw and dental complications.

We were shown some of the results from The Rudy study, where FD patients can record their pain and keep a diary of their medications and fractures. Surveys are filled out every 6 months by patients and the data is recorded for the study.

It has been noted that abnormal nerve pain is common in patients with FD. We were told that there are some new drugs which are being used at specialist centres only and some research undertaken.

Dr. Jason Malone – Surgical Challenges in Fibrous Dysplasia

A brief overview of FD was given and we were shown x-rays of normal and FD pelvis, femurs and tibias.

We were shown examples of different materials that have been used for bone grafting over the years and their success rates were explained. It is not recommended to use the patients own bone for this procedure.

We were shown examples of nails, rods, pins and plates that have been used in FD and an explanation of the pro's and cons of each one was given. An intramedullary nail is the preferred option.