Fibrodysplasia Ossificans Progressiva (FOP)

What is Fibrodysplasia Ossificans Progressiva (FOP)?
Fibrodysplasia ossificans progressiva, also known as myositis ossificans progressiva or MOP, is a disabling condition, which is caused by the formation of bony bars within the muscles of the body. This bone formation is usually first noticed in early childhood as a series of hard lumps in the neck or along the spine. These lumps, which may be tender, gradually shrink in size as the affected muscles are replaced by bone. The appearance of bony lumps in muscles is usually spontaneous but can also be provoked by any injury to the muscles. Disability in FOP is physical and very variable in extent; intelligence in unaffected.

How does the condition progress?
FOP is progressive in that more muscles become involved with increasing age, but the rate of progression is very variable from one person to another. Furthermore the condition tends to show long periods of inactivity (of up to several years in length).

Can any muscles be affected?
Certain muscles are never involved in this disease. These include the muscles of the eyes, the face, the tongue, the gullet, the intestines and the muscles of continence (bowel and bladder control). The heart is never involved in this condition. Chest expansion may be reduced in FOP but the diaphragm, which is the single most important breathing muscle, is never involved.

What is the life-span of people who have FOP?
General health remains good and several people with FOP in Britain are over 60 years old. In view of the reduced chest expansion respiratory tract infections need to be treated vigorously.

Are there any other features of FOP?
Most people affected by FOP have some abnormalities of the fingers or toes. The big toes are most commonly involved and are usually shortened and deviated. These changes to the big toes are usually apparent at birth.

How common is FOP?
This disorder occurs in all countries in the world and affects about one person in a million. Men and women seem to be equally affected.
Is FOP hereditary?
The basic genetic error is a mutation in the activin receptor IA (ACVR1) gene. In most affected patients worldwide an identical mutation is found and this mutation is not present in the parents and has occurred at the moment of conception. In this situation the chances of recurrence in the family are negligible. For a person with FOP, however, there will be a one in two chance of handing on the condition to any children. Further information on this point may be obtained from your local genetic counselling clinic.

Is there a treatment?
Many types of treatment have been tried in the past and none of these has been shown to be of benefit. A number of new medications are being evaluated at the present time and further information about these trials is available on the IFOPA website, see below.

Does anything make FOP worse?
Injury to the muscles can often provoke local bone formation. For this reason intramuscular injections should be avoided if possible. Similarly operations on the muscles to remove pieces of bone almost invariably result in increased bone formation. Also, some forms of dental treatment may result in bone formation in the jaw muscles and you should therefore warn your dentist that you have FOP.

We’re here for you at the point of diagnosis and at every stage thereafter, and can:
- give you accurate and up-to-date information about your or your child’s muscle-wasting condition, and let you know of progress in research
- give you tips and advice about day-to-day life, written by people who know exactly what it’s like to live with a muscle-wasting condition
- put you in touch with other families living with the same muscle-wasting condition, who can tell you about their experiences
- tell you about – and help you get – the services, equipment and support you’re entitled to.

If you would like your GP or other health professional to have more information about Fibrodysplasia Ossificans Progressiva, we have some relevant materials. We’ve developed an online training module for GPs, as well as one for physiotherapists working with adults with muscle-wasting conditions. Contact our helpline or email us to find out more.

If you have feedback about this factsheet please email info@musculardystrophyuk.org.
Here for you
The friendly staff in the care and support team at the Muscular Dystrophy UK’s London office are available on 0800 652 6352 or info@musculardystrophyuk.org from 8.30am to 6pm Monday to Friday to offer free information and emotional support.

If they can’t help you, they are more than happy to signpost you to specialist services close to you, or to other people who can help.

www.musculardystrophyuk.org

This factsheet is under review, due for updating later in 2017. If you have any queries, please contact us.

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