



The Neurofibromatosis Association

Registered Charity No 1078790



SCOLIOSIS

Introduction

The orthopaedic manifestations of Neurofibromatosis Type 1 (Nf1) fall into three main categories:

- scoliosis
- congenital (present at birth) pseudarthrosis
- disorders of bone growth/soft tissue

In this fact sheet we will look at scoliosis and try to answer the most frequently asked questions about spinal problems in Nf1.

What is Scoliosis?

Scoliosis is derived from a Greek word and means a sideways curvature of the spine. It is present in about 6% of people with Nf1 and it can be so mild as not to be noticed or severe enough to be obvious and painful. In the severe form there can be a considerable forward bend as well as a sideways component. If this is predominant in the upper part of the spine the word 'kyphosis' is used. In the lower part of the spine the word 'lordosis' is used to describe this forward bend which often causes the buttocks to appear unduly prominent.

How common is Scoliosis in Nf1 and how do you check for a spinal curvature?

The general consensus is that between 5 — 10% of people with Nf1 have a detectable scoliosis but this is often mild and non-progressive, however, it can be the opposite.

It is important that all growing children with Nf1 should have their backs examined at least annually as part of their regular health check and your GP or paediatrician can do this. To check for spinal curvature, get someone to look along your back when you are bending down towards your toes with your knees straight. If there is an obvious difference on the two sides or any sharp angulation of the spine, or if in you are in doubt, your GP should be consulted. Children with Nf1 can also have this simple test done by their parents, for example every six months or so in between appointments, until they are fully grown at the age of 16 or 17 years. Early detection is preferable as it can result in easier and therefore better treatment if that should be necessary.

When do spinal curvatures first appear?

The most severe spinal curvatures will appear during the first few years of life, typically before the age of 5. As most spinal curvatures are disorders of growth, they will tend to become more severe if there is a long period of growth ahead. The less severe types tend to appear around the age of puberty, at which time they might progress rapidly during the fast growth of adolescence; on the other hand, curvatures, which are only just visible to the naked eye, may not worsen. If by the end of adolescent growth, at around the age of 17 or 18, the curvature is only just visible to the naked eye, it will probably not worsen later in life; but, the more severe and obvious the curvature at this stage, then the more likely it is to increase with age.

Will scoliosis become painful or disabling?

Pain and disability are caused by mechanical failures of the spine. In scoliosis this will occur if the curvature increases over 40-60% of angulation (off the vertical) in adolescence or in later life when the normal stresses and strains produce increasing wear and tear over the curved part of the spine. Pain and disability are, however, very variable in different individuals. For example an individual might have a severe curve with only minor pain whilst someone with a relatively minor curve could experience more severe pain and disability.

How should spinal curvatures be treated?

This is a large and complex subject and impossible to cover here in detail. The most important step is to consult your GP or Consultant and if it is felt that there is a spinal curvature you will be referred to an orthopaedic surgeon with special interest and expertise in this field. Virtually all regions of the country now have such a surgeon but your own doctor will be able to give you more specific information.

Treatment of spinal curvatures usually consists of watching the curvature with X-rays every few months to assess its behaviour. If it is mild and static then nothing else need be done but if it is worsening then your surgeon may prescribe a brace or recommend surgery to stabilise the deformity before it worsens. There is no doubt that surgery is a major procedure but currently with much safer and more effective techniques, surgery can be advised earlier.

It should be remembered that every case of Nf1 is different and the treatment and management of each individual will vary. The timing and type of treatment suggested is a matter of careful judgement and will only be considered after full discussion between the surgeon, yourself and your relatives. Fortunately, treatment is slowly improving all the time.

Is there a risk of paralysis?

The answer is that with early detection of deformities and timely skilled surgery this is exceedingly rare.

Helpful organisations

Scoliosis Association UK (SAUK)

2 Ivebury Court
323-327 Latimer Road
London W10 6RA

Helpline: 020 8964 1166 — Mon-Fri 10am-3pm

Website: www.sauk.org.uk

SUMMARY

The most important priority is to locate the relevant specialist in your area who will provide you with the best possible treatment and your GP will be able to help.

Nf1 is a comparatively common condition but the orthopaedic problems are rare and the pooling of experience is crucial.

Each region will differ in the services they provide e.g. rehabilitation, physiotherapy and occupational therapy. Other support organisations can also be extremely helpful. Please speak to your Specialist Advisor for more information about what is available in your local area.

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