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SURGERY FOR SCOLIOSIS

Dr Alison Kerr: March 2002, updated March 2004

Scoliosis is common in Rett syndrome. Most adults have some degree and by 15-20 years about half have a moderate or severe curve. About 10% of the British cohort have required surgical correction. The back clearly requires careful monitoring in a department with specialist skills in scoliosis management.

There are several theories of causation and in Rett it seems likely that defective control of muscle tone – from hypotonia to hypertonia, always with an element of dystonia – and distorted body image play a part. Work on scoliosis in chickens has intriguingly suggested a link between the development of scoliosis and melatonin deficiency and this deserves investigation, particularly as melatonin levels have been reported low in Rett.

Prevention better than cure

Physiotherapy, satisfactory positioning at rest and encouragement to be active are of value in preventing or minimising the deformity. Riding and swimming with support and hydrotherapy seem to facilitate spontaneous correction of posture in Rett. Light splinting has a place with care taken that this does not restrict activity.

If rapid progression of scoliosis is making surgery inevitable:

- make sure that the procedure is well discussed with the family and is acceptable.
- toilet use should be encouraged. Most people manage habit training and this reduces problems with constipation post-operatively.
- time the operation so that walking skills are not lost in an effort to avoid surgery. Once lost they are difficult to recover, although they do tend to survive operation.
- introduce the patient and family to the surgical, nursing and therapy team so that the abnormal breathing, epilepsy and other features of the individual 'baseline' are understood.
- make sure that nutritional state is optimal, by PEG if necessary (tube directly into the stomach through the skin).
- admit the parent as well as the patient and encourage and facilitate their care as far as possible.
- surgical correction needs to be robust in order to allow for seizures and to permit early mobilisation.
- give adequate analgesia and mobilise as quickly as possible.
- maintain satisfactory nutrition and fluid intake using PRG if necessary, continuing after discharge.
- ensure adequate physiotherapy, dietary advice and family support on return home.

Following the immediate post operative period

The first year after operation tends to be difficult for the family and patient. Following that period reports are almost always favourable. Families report better posture, digestion, less chest infections and continue mobility provided that this had not already been lost. In general, people with operated scoliosis enjoy better health than those at the same age with a comparable scoliosis who have not been operated. Undoubtedly the level of family and therapy support is an important factor as well as the actual operation.
Well-supported riding and swimming should be possible.