**Focus On**

Recent advances in the treatment of scoliosis in children

**Genetics**

Adolescent idiopathic scoliosis (AIS) has been shown in both clinical and genetic studies to have a genetic component to its aetiology.\(^1,^2,^3,^7\) although the pattern of inheritance still remains elusive; X-linked dominant,\(^3\) autosomal dominant or multifactorial patterns\(^1,^2,^7\) have all been suggested. Genealogy work with a cohort of 145 patients with AIS demonstrated a 97% connectedness to other families with AIS, with differences in both expressivity and penetrance, suggesting that there is more than one gene responsible for AIS.\(^9\) Work using genome-wide scanning continues to support this hypothesis by identifying specific areas on the human genome which are potentially significant in the aetiology.\(^7\) Miller et al.\(^5\) in one study of 1198 individuals in 202 families, reported linkage to loci on chromosomes 6p, 6q, 9q, 16q, 17p and 17q. This followed work by Wise et al.\(^10\) who identified loci on chromosomes 6p, 10q and 18q in one polygenerational family and Chan et al.\(^11\) who reported loci on chromosomes 19p and 2q.

Predicting the risk of curve progression has also been the focus of genome studies. Braun et al.\(^12\) identified 12 DNA markers as having diagnostic utility in AIS. When compared with standard radiological methods, these markers provided a superior assessment of risk of progression. Further development of this work has led to the ‘Scoliscore AIS prognostic test’ becoming available to spine specialists in the US in 2009.\(^13\) This saliva-based genetic test uses 53 DNA markers which have been linked to the progressive form of scoliosis. Validation work by the development company, Axial Biotech, quotes a 99% negative predictive value (95% confidence interval (CI) 96% to 100%) but independent validation is awaited.

**Bracing**

The use of orthotics to treat AIS is a time-honoured tradition in countries around the world.\(^14\) Despite bracing being the mainstay of conservative treatment, its efficacy has not been demonstrated definitively in prospective or randomised clinical studies.\(^15\) It is possible to find support in the literature on the effectiveness of braces in preventing curve progression.\(^16-20\) This support includes the Scoliosis Research Society brace study, which is the only prospective, multicentre controlled study so far reported, that compares bracing with observation and concluded that bracing was more effective than observation alone.\(^21,^22\) but it must be noted that this study ‘was non-randomised, non-blinded, the baseline differences between the groups were not statistically adjusted for and the results did not include the surgical rates’.\(^14\) A meta-analysis performed in 1997\(^15\) also supported the use of brace treatment and advocated a 23-hour regimen. This was shown to be significantly more effective than any other form of conservative treatment, including regimens that involved fewer hours per day in the brace. Conversely, there are multiple studies which have concluded that bracing is ineffective in preventing curve progression,\(^23-25\) including a review article in 1999\(^26\) which critically deconstructs the results and conclusions of previous papers which support bracing, while advocating a watchful waiting policy. A more recent systematic review of the relevant clinical studies aimed to identify a pooled estimate of the incidence of surgery in patients who had been treated by observation only (three studies, 139 patients) and for those treated in a brace (15 studies, 1814 patients). The pooled surgical rate was 23% (95% CI 20% to 24%) after bracing and 22% (95% CI 16% to 29%) after observation leading to the conclusion that there was no evidence to recommend bracing rather than observation.\(^27\)

Compliance with brace use has always been a major problem, with most regimens being intensive in terms of time, social acceptability and limitations in physical activity. By designing a discrete choice experiment\(^28\) it was shown that patients with scoliosis would be prepared to undergo brace treatment only if it were to provide a sizeable reduction in the risk of surgery. Effectiveness and discomfort in wearing a brace were the most important determinants of choice. The issue of brace efficacy is thus far from resolved. In an attempt to improve brace comfort and, therefore, compliance, there has been work undertaken on flexible spinal orthoses.\(^29\) However, as yet these have not proven to be better than rigid orthoses in either efficacy or patient acceptance.\(^30\)

It appears that there are many issues in respect of the brace treatment of AIS that have yet to be resolved. In order to help clinicians, patients and parents make a more informed choice about brace treatment, better quality prospective studies are needed. Some key issues may be answered in the Bracing in Adolescent Idiopathic Scoliosis Trial (BrAIST), a 27-centre randomised, controlled trial currently undergoing its second year of recruitment.\(^31\) This trial aims to randomise patients at high risk of curve progression into either a group treated with a thoracolumbar spinal orthosis or a group treated by observation alone, and will compare clinical, radiographic and psychosocial outcomes.
**Instrumentation and fusion**

The instrumentation technique used to supplement fusion in surgery for AIS is constantly evolving. Harrington’s spinal instrumentation system\(^3\) gave improved curve correction and allowed early mobilisation in a brace rather than a bulky cast. Luque introduced the principles of segmental instrumentation and correction via translation and derotation by using sublaminar wires.\(^3\) With the advent of Cotrel-Dubousset implants\(^4\) these concepts have been expanded to include hook, hybrid and most recently, all-screw constructs.\(^5\) In the last decade in particular, increased attention has been paid to re-establish normal sagittal balance during the surgical correction of scoliosis.

All-screw constructs have become the gold standard instrumentation technique for thoracic AIS. A pedicle screw achieves three-column fixation of the vertebral body\(^6\) and displays biomechanical properties that are superior to other forms of construct.\(^6\) This allows the surgeon to achieve greater correction of the curve in all three planes, including rotation, with relatively simple rod derotation manoeuvres; this was a failing of techniques that used hooks. There is also less canal intrusion and risk of dislodgement or migration into the canal when compared with hook fixation.\(^7\) Coronal correction of the curve by up to 70% has been reported with all-screw constructs, hook fixation achieving less than 50%.\(^7\) Better correction of the reduced thoracic kyphosis seen in AIS has also been reported with the use of an all-screw technique,\(^7\) although some have noted a tendency for all-screw constructs to cause flattening of the thoracic spine during the correction,\(^7\) which occurred when rods of inadequate thickness were used.

Before the introduction of all-screw constructs, the more severe thoracic AIS curves (Cobb angle > 60°) required a combined anterior and posterior fusion in order to improve the coronal correction and fusion rates. The deterioration in pulmonary function with a thoracotomy in these cases is well documented.\(^5\) A posterior spinal fusion using an all-screw construct has been shown to provide an equivalent degree of curve correction compared with the combined approach in patients with a curve of 70° to 100°, negating the need for thoracotomy in these patients.\(^5\)

Pedicle screws are not only effective but have also been shown to be safe in all types of curve. This was an initial concern for many surgeons with their use in the thoracic spine. Three recent, large studies have demonstrated the complication rates of using pedicle screws in AIS. Neurological injuries have been reported in 0% to 1.5% of patients, with all deficits being transient.\(^5\) Screw malposition is said to be 1.5% when assessed by plain radiography\(^5\) and 6% when assessed by fine-cut CT scans; however, these patients were asymptomatic and did not require screw revision anyway.\(^5\) A pedicle fracture rate of 0.24% was seen, with one patient out of 462 requiring revision surgery; intraoperative screw loosening was noted in 0.76% of patients.\(^5\) Peri-operative pulmonary function has also been shown to improve in patients after all-screw constructs when compared with hook systems. This is thought to be because of improved alignment of the chest wall with all-screw constructs.\(^5\)

The use of all-screw constructs has been extended from AIS to adult idiopathic scoliosis. Adult curves are stiffer and have a higher rate of pseudarthrosis when compared with their adolescent counterparts.\(^5\) Hook-based constructs are less likely to provide adequate curve correction and this can lead to an increased need for combined anterior and posterior surgery. The improved biomechanical profile of all-screw constructs results in improved curve correction while avoiding the morbidity of anterior surgery; in one study this conferred a significantly lower complication rate.\(^5\) Re-establishment of normal sagittal balance is particularly important in the adult and may necessitate spinal osteotomies such as the chevron or Ponte, pedicle subtraction osteotomy or vertebral column resection.

**Surgical treatment of early onset scoliosis**

Patients with AIS typically present at a stage in their development where the thorax has achieved most of its adult volume and has near-normal vital capacity. Surgical treatment involves definitive spinal fusion and has a negligible effect on thoracic growth or long-term pulmonary outcome. Early-onset scoliosis, irrespective of cause (congenital, neuromuscular, etc.) presents the surgeon with a different set of challenges. Spinal lordosis and curve rotation leaves the thorax with a volume-depletion deformity and growth inhibition ultimately leading to Thoracic Insufficiency Syndrome (TIS). This is defined as “the inability of the thorax to support normal respiration or lung growth.”\(^5\) Associated abnormalities of the rib cage can add to the thoracic disability. Standard surgical treatment of scoliosis is often not possible in these cases because of the detrimental effect that spinal fusion might have on spinal and thoracic growth.\(^5\)

The goal of surgical treatment in early-onset scoliosis is “to stop the progression of the curve whilst allowing maximum growth of the spine, lungs and thoracic cage”.\(^5\) In order to achieve this a number of different surgical approaches can be employed.

Posterior instrumentation without fusion, using dual growing rods, is used when the primary problem is at the vertebral column. The dual rod technique has been shown to give significantly better correction of the scoliosis when compared with single rod techniques.\(^5\) Subperiosteal dissection and fusion is performed only at the upper and lower anchor sites of the construct. Each rod is then measured, contoured and cut into an appropriate length that is approximately 8% greater than the vertebral length. The goal is to achieve the greatest possible correction of the curve while avoiding the morbidity of anterior instrumentation. The Cobb angle is reduced from a mean of 82° to 40°, improving thoracic kyphosis from 72° to 36°. The mean spinal growth was 1.21 cm per year, which approaches that associated with normal spinal growth.\(^5\)
When the primary problem involves the thoracic cage, for example TIS has developed or the patient has rib fusions, reconstruction of the thoracic cage needs to be considered. Expansion thoracoplasty and insertion of a vertical expandable prosthetic titanium rib (VEPTR) has been developed to treat such cases. This technique addresses all related components of the thoracic deformity in the growing child, including the spine, rib cage and diaphragm, without compromising thoracic growth. Rather than directly correcting the spinal deformity with instrumentation, this technique involves correcting the thoracic deformity with rib osteotomies or intercostal muscle lysis. This enlarges the constricted thorax which is then maintained by insertion of the VEPTR device. As with the growing rods, lengthening is required approximately every six months.

Growth of the spine in cases where the VEPTR technique has been used is nearly normal. Improvement in thoracic constriction and lung function has been difficult to confirm because of the problems in measuring pulmonary function in young children; however, some studies have shown a decreased ventilator dependence and increases in thoracic and lung volumes. The interpretation of these results must be guarded as all of the level data remains as the interval between lengthening procedures increased. With repetitive lengthening the expansion portion of the device can be outrun and requires exchanging, which is a more expensive procedure compared with simple lengthening.

References