Idiopathic Scoliosis: Infantile and Juvenile

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Key Points

- Careful history and physical examination are imperative to rule out other etiologies of scoliosis in growing children.
- Quality AP and lateral spinal radiographs are imperative to the evaluation and management decision making.
- In infantile scoliosis, curves with an RVAD of 20° or more, Cobb angle of 25° or more, or a phase 2 rib head should be followed closely for progression. Curves with an RVAD of less than 20° and a phase 1 rib head almost certainly do not progress.
- All curves 20° or more should be evaluated with advanced imaging (such as MRI) to rule out brain and spine anomalies.
- Our current recommendation for treatment of infantile and juvenile idiopathic scoliosis is dual growing rods placed subfascial using a two-incision technique with a skin bridge. Lengthening should occur every 6 months.

17.1 Introduction

> It "develops rapidly and relentlessly, causing the severest form of orthopaedic cripple with dreadful deformity, marked dwarfing and shortening of life"
>
> J.I. James, MD on infantile scoliosis, 1959

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Management of spine deformity in children 5 years of age or less presents one of the most challenging tasks in spine surgery. It requires intimate knowledge of normal spine development as well as the etiology, natural history, clinical evaluation, and available nonoperative and operative treatments for infantile scoliosis. Early recognition and diagnosis by both parents and pediatricians is essential. Immediate orthopaedic referral is mandatory as early treatment may ultimately affect patient outcome.

Harrenstein [24] in 1936 coined the term infantile idiopathic scoliosis (IIS). He treated 46 children and noted mixed success treating the curve with bracing, and attributed the deformity primarily to rickets [23]. In 1951, James [28] reported on 33 cases of scoliosis in infants aged 3 years and younger. They were predominantly boys with left-sided thoracic curves. Four cases resolved spontaneously, but the remainder progressed very aggressively. In 1954, he first described scoliosis according to chronologic age at a presentation including infantile from birth to 3 years, juvenile with onset up to 8 years, and adolescent with onset from 10 years to maturity [29]. Interestingly, no reference was made for those between 8 and 10 years [29]. Dickson [12] later recommended that scoliosis in children be classified as early (5 years or less) or late (>5 years) onset. The rationale for this is twofold. As Dimeglio and Bonnel [14] have shown, growth velocity in the spine is highest from birth to 5 years, followed by a deceleration between age 6 and 10 years. From 11 to 18 years, there seems to be another peak in growth velocity but not equal to that of early life. Early onset, therefore, more accurately describes this growth. Similarly, this group is at a higher risk for developing significant cardiopulmonary complications if thoracic curves progress, whereas these complications are rare in the late onset group. Complications include pulmonary hypoplasia, restrictive pulmonary

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disease, pulmonary artery hypertension, cor pulmonale, and thoracic insufficiency syndrome.

This chapter aims to equip the spine deformity surgeon with all the relevant knowledge to diagnose, educate, and effectively treat the child with infantile and juvenile scoliosis.

17.2 Natural History

17.2.1 Growth and Development

Dimeglio [15] and Dimeglio and Bonnel [14] very nicely illustrated that spine growth velocity is greatest from birth to 5 years, averaging >2 cm growth per year. From the age of 6 to 10 years, velocity decreases to 0.5 cm per year and increases to 1.3 cm per year from the age of 11 to 18 years. Chest growth is most easily assessed as thoracic volume, which shows a similar trend as spine growth. At birth, it is 5% of adult volume. By 5 years of age, it has reached 30%, a staggering 600% increase in volume [15]. At 10 years of age, lung volume is 50% and reaches adult size at the age of 15, in both males and females. Lung development is best measured by change in alveolar volume and number. It is estimated that 20 million alveoli exist at birth and increase to 250 million by the age of 4 and complete development by 8 years of age. A similar increase in alveolar volume also occurs. Respiratory branches also increase from 20 at birth to 23 by 8 years.

17.2.2 Epidemiology

Several authors have reported the incidence and prevalence of infantile and juvenile idiopathic scoliosis (IIS, JIS) [12, 30, 47]. In the United States, IIS comprises less than 1% of idiopathic cases. A slightly higher incidence has been reported in Europe [30, 47]. Unlike late onset, it is more common in males with a ratio of 3:2, and curves tend to be left sided. It occurs in the mid to lower thoracic spine in 75–90% of cases [12, 30, 60]. Since the initial description by James [29] in 1951, it appears that the incidence has decreased. McMaster [47] most recently reported on a declining prevalence of patients with IIS scoliosis in Edinburgh, a major referral for scoliosis in Scotland. Between 1968 and 1972, they averaged 16.5 new patients per year with a 34% incidence of progressive curves. From 1980 to 1982, there was an average of two referrals per year. On the contrary, referrals for adolescent idiopathic scoliosis increased during this same time period.

JIS accounts for 12–21% of reported idiopathic cases [30, 53]. It is more prevalent in females with a 2:1–4:1 ratio. Between 3 and 6 years of age, the gender difference is neutral, and after 10 years of age, females are affected at a rate of 8:1 [20, 64]. Males are usually diagnosed by 5 years of age and females by 7 years of age. This difference as well as the age of skeletal maturity makes progression more likely in males. Right-sided thoracic and double major curves are the principal curve patterns associated with JIS [20, 42].

17.2.3 Prognosis

James [28] in 1951 reported his initial series of 33 patients, 18 (55%) were progressive, 11 (33%), stationary, and 4 (12%), spontaneously resolved. In 1954, he increased his numbers to include 52 children who were treated with physiotherapy, plaster-of-Paris beds, and orthoses [29]. Curves in 43 patients progressed (83%) with all curves being >70° at the age of 10 and several progressing >100°. In the remaining nine patients (17%), the curves resolved spontaneously without treatment. In 1959, James et al. [30] reported on 212 infantile cases from two separate institutions. Seventy-seven (31%) patients had spontaneous correction and the remainder progressed aggressively (135/212). Of these 135 patients, 47 were between 0 and 5 years, and 23 of these already had a curve >70°. Thirty-seven patients were between 5 and 10 years, and 27 of 37 had a curve >70° and 14, > 100°. Of the 23 children of 11 years and older, 12 had a curve >100°, and two at skeletal maturity had curves in excess of 150°.

Scott and Morgan [60] reported on 28 patients with IIS, of which 14 were followed to skeletal maturity. All had severe scoliosis with a mean of 120°. The remaining 14 were still growing. At 6 years of age, the average Cobb measured 65° with the largest being 112°. Three patients died in the late second and third decades of life from cardiopulmonary complications. All patients in their series had small thoracic cages with reduction in both pulmonary and cardiac function. Younger age at diagnosis and progression were found to be predictors of poorest outcome.

In 1965, Lloyd-Roberts and Pilcher [38] reviewed 100 patients with idiopathic curves who were diagnosed before 12 months of life. Ninety-two of these curves resolved spontaneously. Several other authors have subsequently reported their rates of resolution ranging from 20 to 80% [13, 31, 38]. James [31] followed 90 patients with nonprogressive curves and found that all resolved by the age of 6 years. Diedrich et al. [13] reported 34 patients with resolving curves followed through maturity, and found that none progressed during the adolescent growth spurt. Of the 34, 20 were treated with an orthosis, and no children had significant disabilities related to their spine.

Fernandes and Weinstein [19] reviewed the literature and summarized the data on nonprogressive and progressive infantile idiopathic curves. They identified 573 patients with nonprogressive curves with a male to female ratio close to 3:2. Ninety percent were thoracic curves, 80% apex left with greatest Cobb angle ranging from 20 to 48°. A large majority had associated intrauterine molding features. Perhaps the most significant finding was age at diagnosis that averaged 5.5 months compared to 12 months among the progressive group. Furthermore, the progressive group showed greater variability compared to historic reports. Gender ratio was closer to 1.2:1 (male to female), 81% with thoracic curves and 75% left sided. It is important to recognize that girl infants with right-sided thoracic curves may have a worse prognosis and may not follow the typical rate of spontaneous correction.

Juvenile idiopathic scoliosis differs from IIS in its natural history [36]. The curves progress at a slow to moderate rate [20, 26, 29, 34, 53]. The earlier onset usually leads to more severe deformity than adolescent idiopathic scoliosis. Tolo and Gillespie [64] reported on their series of 59 patients, of which 71% (42) progressed to require surgery. Similarly, Figueiredo and James [20] found that 56% (55) of 98 JIS patients progressed. Mannherz et al. [42] reported on a series of JIS patients who did no progress. All patients presented with curves <25°.

Pulmonary complications are the most morbid results of untreated infantile scoliosis. As previously described, the spine, chest wall, and respiratory system rapidly develop during the first 5 years of life [15]. Alteration in normal development of one of these can have deleterious effects on the others. Scoliosis that presents and progresses during this time period has a higher chance of causing unwanted cardiopulmonary side effects [51]. Infantile scoliosis alters normal development of alveoli and pulmonary vessels resulting in ventilation defects. The severity of pulmonary involvement is directly related to the age of onset of scoliosis. The earlier the onset and progression, the more the disability. Pulmonary dysfunction usually presents as restrictive lung disease with reduced vital capacity (VC), total lung capacity (TLC), and increased residual volume (RV). The loss of compliance of the chest wall and both lungs contributes to the restrictive pattern of disease. Persistence of restrictive lung disease usually results in pulmonary hypertension and cor pulmonale. Hypoxemia is related to reduced tidal volume as gas exchange is normal in these kids. Respiratory failure is a late development as these patients have significant pulmonary reserve. This pattern of disease has been consistently shown in the literature; however, it is a rare finding in curves that present after maturation of the lungs (8 years) [11, 32]. Similarly, it differs from thoracic insufficiency syndrome, which presents with respiratory failure at a very early age [9].

17.2.4 Etiology

Browne [8] in 1956 was the first to suggest that infantile scoliosis was initially attributed to an intrauterine packaging problem. He found in his series that 83% of infants had some form of intrauterine crowding deformity such as plagiocephaly, plagiopelvy, decreased hip abduction, and abnormal rib molding with infantile scoliosis. Mehta [48] later agreed that intrauterine crowding was responsible. In 1965, Lloyd-Roberts and Pilcher [38] termed this association “molded baby syndrome”. Further study would refute this theory as scoliosis was not found to be present at birth and did not explain the gender difference or the variance in geographic regions. The difference in incidence in Europe and the United States gave rise to the thought of an environmental theory. Mau [45] in 1968 proposed that infantile scoliosis was linked to how an infant was positioned for sleeping. In the United States, it was more common to place the infant prone in bed which decompresses the spine. This is in contrast to the Europeans who were placing their infants supine. Children, in this position tend to turn to a slight oblique position with a tendency to lie oblique to the right. He also suggested that the molding deformities noted were caused by constant pressure on the soft bones of infants. He also added four other components to the molding theory: unilateral contracture of neck muscles, associated oblique posture of the head,

calcaneus foot deformity, and the subsequent development of fixed dorsolumbar kyphosis. These findings aim to raise awareness and prompt intervention for earlier diagnosis of infantile scoliosis.

The geographic differences further influenced Wynne-Davies [67] to analyze 180 medical records from the Edinburgh Scoliosis Clinic. She identified 114 eligible patients and studied the prevalence of scoliosis between first, second, and third-degree relatives. She analyzed these patients in two groupings: early (before age 8) and late onset. In the early group, 88% had left thoracic curves with a slight male predilection. She identified a 2.6% prevalence of scoliosis in the infantile group compared to 0.39% of controls, a 30-fold higher risk. The late/adolescent group had an even stronger association at 6.94%. Plagiocephaly was found in 100% of patients compared to 11% among controls. Mental retardation and epilepsy were found in 13% of patients. Advanced maternal age was also commonly associated with progressive curves.

Ward et al. [65] have made recent advances in genetic testing among the adolescent idiopathic group. Several gene locuses have been identified to strongly predict those patients with progressive curves. The future is very promising to expand this technology to infantile and juvenile scoliosis for early detection and treatment.

17.3 Clinical Evaluation

17.3.1 History

A thorough and systematic history prior to physical examination is imperative in the diagnosis of infantile and juvenile scoliosis. Careful attention to detail in the history will lead the spine surgeon to pursue further diagnostic testing. Idiopathic scoliosis is a diagnosis of exclusion, and therefore all measures need to be exhausted for accurate diagnosis. Differential diagnosis includes: neuromuscular scoliosis, syringomyelia, spinal tumor, congenital spinal deformity, intraspinal anomalies, neurofibromatosis, syndromic, and spinal infection. Patients need to be carefully screened for any other associated anomalies including cardiac defects, history of hip dysplasia, cognitive deficits, congenital muscular torticollis, and other molding abnormalities. This information is often overlooked during an interview, and we recommend having history forms that are conducive to eliciting this information.

During history taking, careful attention should be directed to prenatal history of the mother, including any health problems, previous pregnancies, and medications. Birth history should include length of gestation, delivery type (vaginal or cesarean), weight and any complications. Like developmental dysplasia of the hip (DDH), there has been an association between scoliosis and breech presentation. Unlike DDH, however, infantile scoliosis is more common in premature low birth weight males. Careful attention should be given to developmental milestones and cognitive function. This information can be gleaned from conversation with family or from simple observation in the waiting room and during the examination. Wynne-Davies [67] found mental retardation in 13% of males with infantile scoliosis.

17.3.2 Physical Examination

Physical examination should be performed systematically with special attention given to the skin, head, spine, pelvis, extremities, and neurological examination. Findings in this group of patients are often subtle, and workup is largely dependent on examination findings in order not to miss an underlying cause for scoliosis. The skin examination should include careful inspection for café-au-lait spots and axillary freckling seen in neurofibromatosis. A hairy patch along the spine may indicate spinal dysraphism, and bruising may indicate trauma. The head examination aims primarily to identify any plagiocephaly, where the recessed side of the head is often on the left side of patients. Wynne-Davies [67] found a 100% incidence of plagiocephaly among the infantile idiopathic group.

The spine examination should begin with inspection, palpation, and careful evaluation of the child's posture, head, shoulder, trunk, and pelvic symmetry. Owing to the patients age, an Adam's forward bend test (looking for prominence of ribs in the thoracic spine or transverse processes in the lumbar spine) is not possible, but the test can be simulated by lying the child prone over the examiner's knee with the convex side downward. Lateral pressure in this position will illicit curve flexibility. The more rigid the curve, the higher the likelihood of progression. Chest or flank asymmetry and limitation in chest excursion should make the examiner aware of the association with syndromic scoliosis. Abdominal reflex abnormalities should initiate a more thorough neurological examination. Absence of

this reflex has been reported as the only objective finding in patients with Chiari malformations [50]. The abnormal reflex is typically found on the convex side of the curve [69]. Further work-up is appropriate in this setting with total spine magnetic resonance imaging.

Other physical findings that should not be overlooked include plagiopelvy, and developmental hip dysplasia, both with strong associations to idiopathic infantile scoliosis [7, 8, 10, 27, 68]. Hooper [27] found a 6.4% prevalence of congenital hip dislocation among 156 patients with infantile scoliosis. This is approximately ten times higher than the general population. Wynne-Davies [68], similarly reported on four patients among her infantile scoliosis cohort who had DDH. In 1980, Ceballos et al. [10] reported on 113 patients with a 25% prevalence of DDH. Interestingly, the dislocations were found mainly among females and resolving curves. There was no correlation with side of dislocation and direction of curve. Finally, limb length inequality must be ruled out as an etiology for scoliosis. When it is the cause, the lumbar prominence is found on the side of the longer limb. Other means of testing this include a sitting forward bend test or by placing a lift under the short limb to equalize limb lengths.

17.4 Diagnostic Testing

17.4.1 Radiologic Evaluation

Plain radiography is a simple and reliable tool in the work-up of a child with suspected scoliosis. Patients typically are diagnosed in the first 6 months to 1 year of life, and early recognition and treatment are essential for optimal outcomes. Radiographs will help rule out congenital scoliosis as well as establish baseline measurement for future comparisons. Treatment decisions are traditionally based on progression of Cobb angle and rib vertebral angle difference (RVAD) obtained at subsequent visits. Progression has been associated with compensatory curves (including lumbar, double thoracic, and thoracic), greater vertebral rotation, and shorter length of curves.

High-quality radiographs are essential for thorough radiographic analysis. Initial evaluation should include anterior–posterior (AP) and lateral radiographs of the spine (including cervical spine and pelvis). In children too young to stand, films should be obtained supine. Special attention should be paid to the cervical spine for anomalies, as well as to the lumbosacral junction for spinal dysraphism, and the pelvis and hips to ensure a reduced position of the hips. Measurements should include both Cobb angle and RVAD (Fig. 17.1a). Mehta [48] is credited for developing this powerful tool for predicting progression of infantile curves. Out of frustration with the inability to predict progression with Cobb measurements, she evaluated the relationship of the rib attachment to the vertebral body. She noted variability in the takeoff angle of the ribs from the convex vs. the concave side of the curve. The rib vertebral angle measures the angle of a line drawn perpendicular to the apical thoracic vertebra end plate and a line drawn down the center of the concave and convex ribs. The RVAD is calculated by subtracting the convex from the concave angles. An RVAD of less than 20° indicates a curve that is most likely to resolve (85–90%), while an RVAD of 20° or more is frequently associated with progression. She also described a second radiographic parameter to assist in prediction known as the *phase of the rib head* (Fig. 17.1b, c). This radiographic tool uses the relationship of the head and neck of the rib to the vertebral body, at the apex of the convexity of the scoliosis. In phase-1, there is no overlap of the rib head or neck on the apical

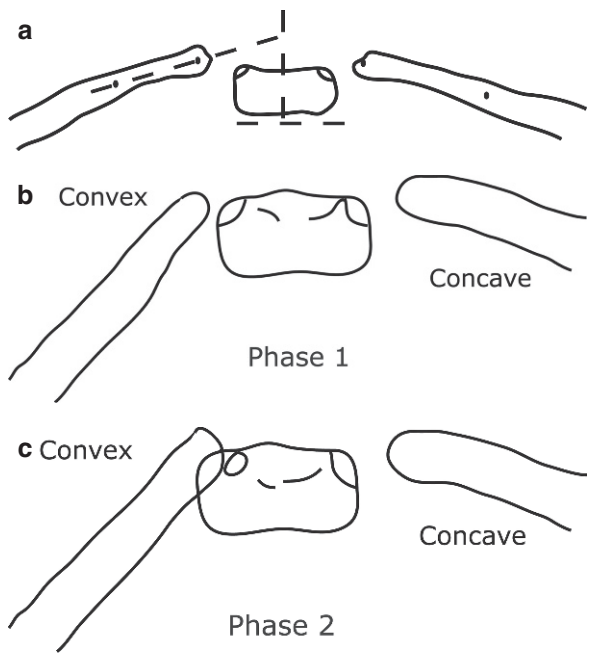

Fig. 17.1 (**a**) Rib vertebral angle difference (RVAD). (**b**) Phase of rib head: phase 1. (**c**) Phase of rib head: phase 2. (Redrawn from reference [48])

vertebra. In this group of patients, the RVAD should be measured to detect progression. In phase-2, the head or neck of the rib is overlapped on the apical vertebra. It has been shown that a phase-2 rib head is a certain predictor for progression and RVAD does not need to be measured. Mehta [48] reported on 46 infantile patients with phase-1 rib heads whose scoliosis resolved. She found that 83% had a RVAD of less than 20°. Of the remaining patients with an RVAD of 20° or more, the angle was found to consistently decrease with follow-up. The decrease in RVAD also preceded the decrease in Cobb angle. Of the group with progressive curves, 84% had an initial RVAD of 20° or more (range 18–30°).

Ceballos et al. [10] corroborated Mehta's findings reporting 92% of their resolving curves having an RVAD of 20° or less. Of the remaining 8% with an RVAD greater than 20° all showed improvement at the 3-month follow-up. Robinson and McMaster [56] in 1996 found that the curves that progressed among their 109 patients had a mean initial RVAD of 31°, while those that resolved had a mean of 9° on initial exam.

Mehta [48] recognized a special radiographic feature among the less common (and more aggressive) double major and lumbar curve patterns. She recognized that the RVAD at the apical thoracic vertebra was frequently less than 20° and found there to be significant asymmetry at the 12th vertebra. Here, she found the rib on the concave side becoming more vertical than the rib on the convex side, making the RVAD negative. The 12th rib is initially part of the upper curve but becomes the apex of a secondary curve developing caudally to the first. Consequently, the rib that is on the concavity of the upper curve drops secondary to the progression of the vertebral rotation and increases in magnitude of the caudal curve.

17.4.2 The Role of Advanced Imaging and Neural Axis Abnormalities

The role of advanced imaging in infantile and juvenile scoliosis is directly related to the presence of neural axis abnormalities. As IIS and JIS are a diagnosis of exclusion, all attempts must be made to identify possible etiologies. The incidence of neurological abnormalities has been reported as high as 20% in patients under the age of 10 [18, 35, 37, 48]. Lewonowski et al. [37] reported an magnetic resonance imaging (MRI) study of 26 consecutive patients with idiopathic scoliosis under the age of 10 years. They found 5 patients (19%) with neuropathology, and only 2 patients with atypical curves. Four of their patients were infantile, and two patients had abnormal findings: a 4-month-old boy with a terminal lipoma and a 3-year-old girl with a syrinx.

Gupta et al. [22] conducted a prospective and retrospective MRI study to evaluate the prevalence of neural axis abnormalities in patients 10 years of age or younger with idiopathic scoliosis and a normal clinical examination. In the prospective arm, he followed 34 patients with a mean age of 9 years and found abnormalities in 6 patients (18%). Within this group, six patients were infantile, and three patients had identifiable neuropathology. Among the 64 retrospective patients, 20% were found to have neural axis pathology.

Most recently, Dobbs et al. [16] in multicenter study identified 11 of 46 infantile scoliosis patients with neural axis abnormalities. All patients were clinically asymptomatic and had curves of 20° or less. Five patients had an Arnold–Chiari type-I malformation, three with syringomyelia, one with a low-lying conus, and one with a brain tumor. Of these 10 patients, 8 required surgical intervention. On the basis of the findings of this paper and other reports, it is our recommendation that all patients with IIS or JIS with a curve of 20° or less have both, a brain and a complete spine magnetic resonance Imaging MRI.

Other imaging modalities exist to aid in management and provide continued relevant information in the care of these children. Computed tomography (CT) scans can be helpful for preoperative evaluation in selected patients where the spine will be instrumented. Pedicular anatomy and bony anomalies are made very clear. CT scans can also be used to assess the three-dimensional lung volumes and can be a marker of treatment.

17.5 Management Themes (Fig. 17.2)

17.5.1 Selecting Surgical Candidates

Management of children with infantile scoliosis is based on anticipated or actual curve progression. Mehta's [48] prognostic criteria, as discussed earlier, are very helpful in identifying curves at risk. Curves with an RVAD of less than 20° and a Cobb angle of less than 25° are at low risk of progression. These patients are safely treated

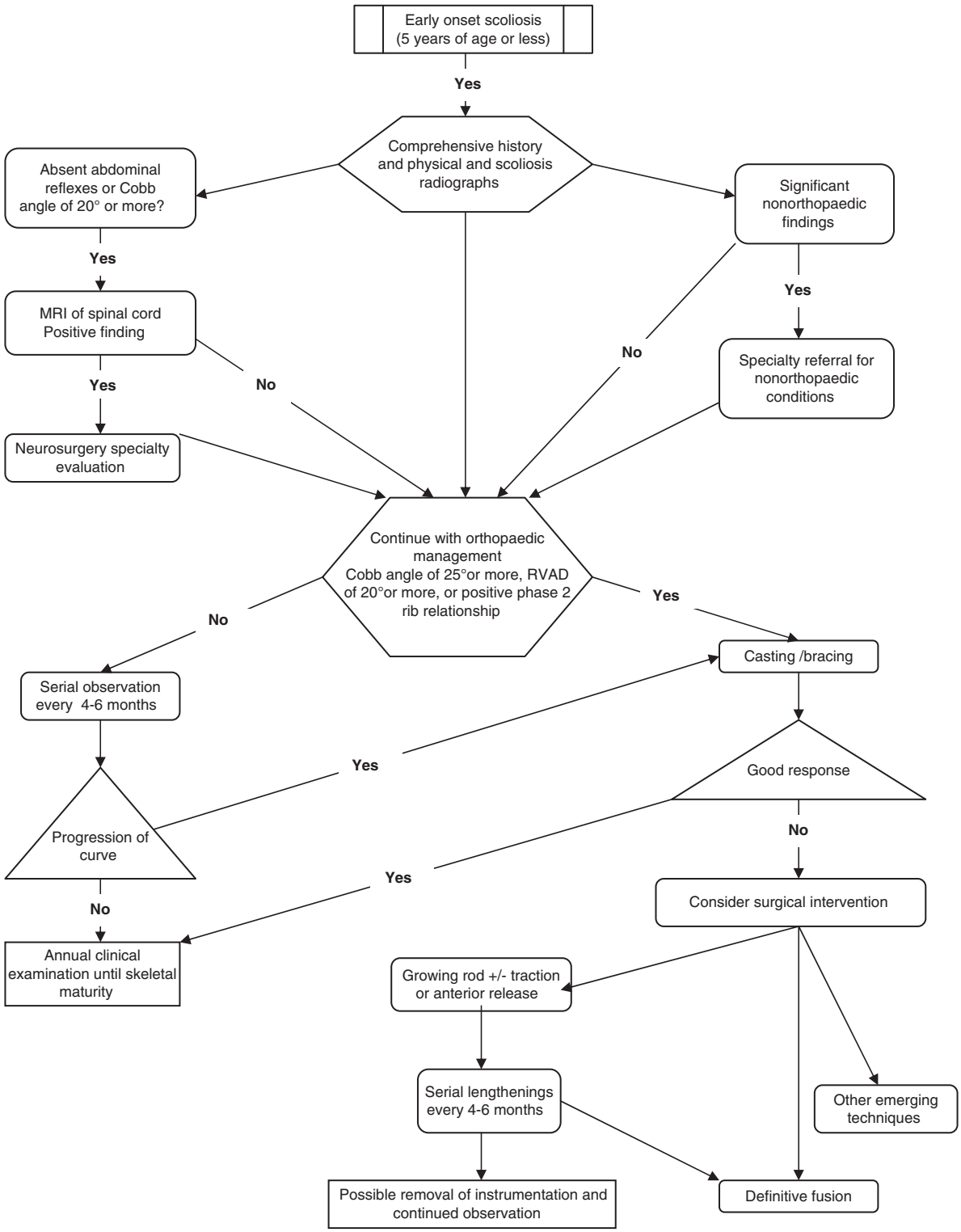

Fig. 17.2 Treatment algorithm for infantile and juvenile idiopathic scoliosis. (Adopted from reference [21])

with observation; however, they should be followed clinically every 4–6 months for progression. Once the curve has resolved, the follow-up interval can be extended to 1–2 years. We recommend following these patients to maturity to ensure that there is no recurrence during the adolescent growth spurt. Diedrich et al. [13] reported on 25-year follow-up of infantile scoliosis, validating the use of RVAD, and demonstrated that there was no advantage to supine plaster bed treatment over physiotherapy, in regard to time to resolution or functional outcome.

Infants with an RVAD of 20° or more or a phase 2 rib–vertebral relationship and a Cobb angle between 20 and 35° have a higher risk of progression. This group of patients should be followed closely at 4–6 month intervals for clinical and radiographic evaluation. Active treatment should be initiated when progression of Cobb angle of 5° or more is documented over 1 year [3]. Active treatment at this point is usually in the form of casting or bracing, which will be discussed thoroughly in separate chapters.

17.5.2 Surgical Treatment: Historic Perspective

The goals of surgical treatment of infantile scoliosis are multifold: To stop curve progression and allow for maximum growth of the spine, lungs, and thoracic cage. Surgery is recommended in children with Cobb angle of 45° or more and documented curve progression. This statement reflects the current trend toward more aggressive operative management since the techniques for fusion-less surgery have become refined, and the natural history of this disease, more clearly understood.

Historically, the goals of surgery were a straight shortened spine rather than a deformed spine of near normal length. Isolated posterior spinal fusion in this age group quickly went out of favor, after Dubousset et al. [17] described the crankshaft phenomenon. This phenomenon seen in skeletally immature patients describes progression of deformity following posterior spinal fusion due to continued anterior growth of the spine. Sanders et al. [58] further correlated open triradiate cartilage and Risser 0 to high risk of crankshaft in light of an isolated posterior spinal fusion. Anterior arthrodesis was therefore recommended, in addition to posterior to prevent crankshaft. Anterior and posterior fusion, however, results in a significant amount of height loss and thoracic development. As discussed earlier, Dimeglio [15] very nicely outlined spinal growth throughout childhood with two noticeable peaks of growth (0–5 years and 10–15 years). Using his formula for calculating normal growth, expected loss of height can be determined in lieu of an anterior–posterior fusion. Winter [66] similarly described a formula for calculating amount of projected height loss. To calculate it in centimeters, you multiply 0.07 by the number of segments fused and the number of growth years remaining. This data is very valuable in educating family and caretaker, of the potential ramifications of fusion in this very young patient population. It should also be noted that the effect of fusion on the spine could have morbid effects on lung and thoracic cage development. This has been a motivating factor over many decades to devise other surgical methods that avoid circumferential fusion.

Over 45 years ago, Roaf [55] attempted to modulate spine growth, much like one would modulate an angular deformity in a pediatric lower extremity with hemiepiphysiodesis. He proposed that the spinal deformity was the result of asymmetric growth between the convex (faster growing) and concave (inhibited) side of the curve. His technique of modulation involved ablation of the convex epiphyseal cartilage and adjacent discs at the vertebrae near the apex of the curve. Only 23% of his treated patients showed improvement of Cobb angle, while 40% showed little or no improvement (Cobb angle <10° change). Marks et al. [44] built upon this idea and used hemiepiphysiodesis and simultaneous Harrington internal fixation. No significant improvement was measured in 13 consecutive patients with 12 demonstrating progression of deformity.

Harrington, [25] in 1962, described a fusion-less technique in 27 idiopathic and postpolio patients, placing a single distraction rod on the concavity of the curve connected to hooks at both ends. The hooks and rods were placed after a subperiosteal approach to the spine. The idea was to instrument the spine without arthrodesis in an attempt to preserve spinal growth, correct deformity, and control the residual deformity. Although no longitudinal results were reported, he believed that children under 10 years could be managed with instrumentation alone, and those, 10 years and older required arthrodesis.

Moe et al. [49] modified the technique described by Harrington and limited subperiosteal exposure to the

site of hook placement and passed the rod subcutaneously. Furthermore, they modified the rod to have a smooth, thicker central portion to prevent scare formation to the threads and allow for sagittal contouring. Patients were lengthened when a loss of Cobb angle >10° occurred. Of the two patients treated with idiopathic infantile scoliosis, both were reported as having a notable decrease in curve magnitude. They furthermore reported a complication rate of 50%, including rod breakage and hook dislodgement from the rod or the lamina.

In 1997, Klemme et al. [33] reported on 20-year experience of the Moe technique. Sixty-seven patients were followed from initial instrumentation to final fusion, with an average of 6.1 procedures per patient. Curve progression was arrested or improved in 44 of 67 patients with an average curve reduction of 30%. Of the remaining 23 patients, 12 were neuromuscular, and the curves progressed on average 33%.

In 1977, Luque and Cardosa [39] described their technique of fusion-less treatment of scoliosis with segmental spinal instrumentation. In 1982, Luque [40] modified this technique by adding sublaminar wires and replacing the Harrington rod with L-shaped rods, later to be known as the Luque trolly. His initial series included 47 paralytic patients who grew by an average of 4.6 cm over the immobilized segment with an average curve correction of 78%. This system became less favored after reports that subperiosteal exposure and sublaminar wire passage created scar tissue and weakened the lamina, which made revision and later definitive fusion difficult. There were also several reports of spontaneous fusion and substantially less growth preservation than predicted. These findings were attributed to the exposure that was required at each level to pass wires.

Patterson et al. [52] combined segmental spinal instrumentation with anterior apical convex growth arrest and fusion in 9/13 patients who had previously undergone surgery at an average age of 5 years and 5 months. Curve correction averaged 46% at 2-year follow-up. Less curve deterioration was identified in those patients who had anterior apical growth arrest compared to those who had segmental instrumentation alone.

In 1999, Pratt et al. [54] performed a retrospective review of patients treated with Luque trolley instrumentation with and without convex epiphysiodesis in 26 patients. Eight were treated with Luque trolley alone and all showed significant curve deterioration. Of those treated with combined convex epiphysiodesis and Luque instrumentation, the Cobb angle worsened in 7 of 13, remained unchanged in 4 and improved for 2. Growth was found to be 49% of predicted in the Luque trolley alone group and 32% among those undergoing combined surgery.

Blakemore et al. [6] further reported periodic lengthening with a submuscular rod with and without apical fusion. Apical fusion was performed on curves 70° or more and in those whose curves were stiff on bending radiographic testing. The rod was placed within the muscle above the spine periosteum, placing the rod closer to the spine for better contour and alignment without inducing spontaneous fusion. He reported on 29 children, 10 idiopathic, all treated in a Milwaukee brace postoperatively. Mean Cobb angle improved from 66 to 38° immediately postoperatively with most recent follow-up showing a slight deterioration to 47°. Complication rate was 24% including hook dislodgement (5), rod breakages (3), and superficial wound infection (1).

17.5.3 Current Approaches to Surgical Management

Once the decision for surgery has been made, several factors have to be considered before choosing the correct surgical approach. The rigidity of the curve plays an important role in decision making, as curves that have little flexibility will not likely be as amenable to a growing construct alone. In this situation, there may be a role for anterior release prior to posterior fusion-less surgery. Marks, [43] in unpublished results, discusses the use of annulectomy vs. nucleotomy as anterior release options. No long-term results exist, however, to make any definitive recommendations.

The next decision to make is which lengthening procedure is ideal for the patient. Salari et al. [53] recently reported on the results of a survey sent to 40 qualified surgeons on ideal treatment of 11 different case scenarios of infantile scoliosis. Seventeen surgeons responded with a wide variation in treatment recommendations for each patient scenario. The most common treatment selected was a dual growing-rod construct (56.7%) followed by nonoperative management (16.6%), SHILLA (15.5%), VEPTR (7%), and fusion or resection, and immediate fusion (4%). This study is important to highlight the lack of standardized

treatments offered to our patients by highly qualified surgeons [57].

The next two sections briefly describe the various fusion-less surgeries. They are subdivided into two categories. Distraction-based growing rods and growth directed surgery. VEPTR, a form of distraction-based growing rod will be discussed in a separate chapter.

17.5.4 Distraction-Based Growing Rods

The unpredictability and high implant associated complication rate associated with single rod distraction techniques led Akbarnia and Marks [1] to develop a dual growing rod technique, building on concepts formulated by Asher (Fig. 17.3a–d). This is the current technique preferred by the authors. Subperiosteal dissection is limited to the proximal and distal foundations (anchor sites). Hooks or pedicle screws are placed on both ends over two or three spinal levels. Foundation sites are fused using local bone graft supplemented with synthetic graft. Upper and lower contoured 3/16 in.-diameter rods are placed submuscularly on both sides of the spine. The rods are joined on each side with extended tandem connectors placed at the thoracolumbar junction to avoid disturbing sagittal balance. The first lengthening is typically performed at the index procedure. A distractor designed to fit within the longitudinal opening in the tandem connector is used at time of lengthening that typically occurs at 6-month intervals starting with the index surgery. The intent of the original lengthening is to obtain modest correction of the scoliotic curve without unduly stressing the foundations. We have found approximately 50% correction of coronal Cobb angles at the original surgery. More aggressive lengthening can be performed starting with the first lengthening after fusion. Somatosensory evoked potential monitoring is performed during each lengthening. Lengthening can be performed as outpatient surgery with appropriate anesthesia and nursing support. Bracing is utilized until fusion is achieved at the foundation sites.

Akbarnia et al. [4] reviewed 13 patients with no previous surgery and noncongenital curves who were followed to final fusion. They found a mean spinal growth of 5.7 cm during a 4.4-year treatment period. The curve improved from 81 to 36° after initial surgery and to 28° at final fusion. T1–S1 length improved from 24 to 29 cm after initial surgery to 35 cm at final fusion. Those patients lengthened at 6-month or less intervals experienced significantly more growth and curve correction than those lengthened less frequently [4].

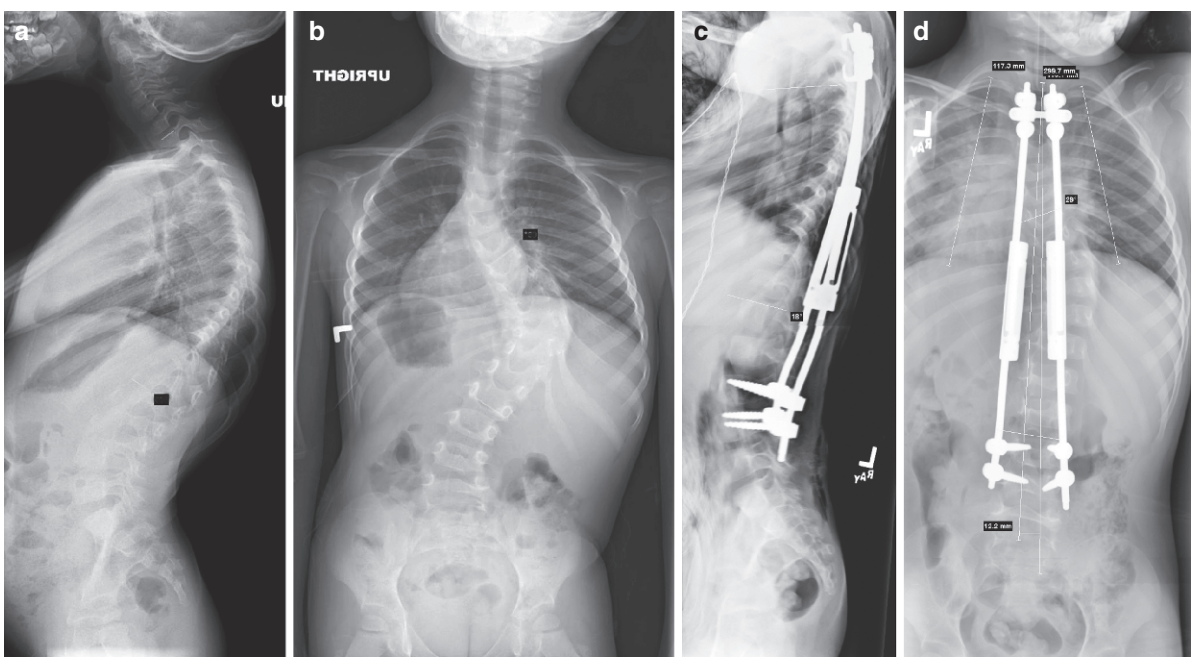

Fig. 17.3 (**a, b**) Severe progressive scoliosis in a 4-year-old patient with idiopathic infantile. (**c, d**) Post initial surgery radiographs

A recent report by Sankar et al. [59] reviewed 782 growing rod surgeries in 252 patients where neuromonitoring was performed. Surgeries included 252 primary rod implantations, 168 implant exchanges, and 362 lengthenings. Neuromonitoring changes occurred in two primary implant surgeries (0.8%), in one implant exchange (0.6%), and one lengthening (0.3%). The change noted in the case of implant exchange also resulted in a clinical deficit, which resolved within 3 months. The monitoring change that occurred in the lengthening was in a child with an intracanal tumor that also had a change during the primary surgery. The final recommendation was that the overall rate of neuromonitoring change seen in primary and implant exchange surgeries justifies its use. No definitive recommendations could be made for lengthenings because of sample size.

Akbarnia et al. [2] reported on a multicenter study with 2-year follow-up (24–111 months) of 23 patients, 7 of which had idiopathic infantile scoliosis. The average age at initial surgery was 5 years and 5 months, with an average of 6.6 lengthenings. Mean Cobb angle improved from 82 to 38° following initial surgery and 36° at latest follow-up. Growth averaged 1.21 cm per year as calculated by T1–S1. Seven patients completed treatment and averaged 11.8 cm of total growth (T1–S1) from preoperative to postfinal fusion (1.66 cm per year). Among 14 patients with thoracic curves, the space available for lung as described by Campbell et al., improved from 0.87 preoperatively to 1.00 at latest follow-up or final fusion. Complications occurred in 11 of 23 patients between initial surgery and final fusion. They included three anchor (hook or screw) displacements, two rod breakages, two deep wound infections, four superficial wound problems, one crankshaft, and one junctional kyphosis requiring an extension of instrumentation. Although the complication rate is high, the authors contested that it is safe and effective, and carried with it a lower complication rate than single rod systems.

Thompson et al. [62] compared the results of single and dual growing rod systems in 28 patients followed to definitive surgery. Five had a single rod construct with anterior and posterior apical fusion, 16 had single rod without apical fusion, and 7 had dual rod without fusion. Mean Cobb angle, respectively, improved from 85 to 65°, 61 to 39°, and 92 to 26°. Spinal growth, respectively, was 0.3 1.0, and 1.7 cm per year. The authors concluded that the improved results seen in dual rod systems are likely attributable to its greater strength and more frequent lengthening.

Mahar et al. [41] recently published results of a biomechanical study investigating the construct of the foundation in a porcine model. They investigated four constructs: (1) hook-hook with cross-link, (2) hook-screw with cross-link, (3) screw-screw with cross-link, and (4) screw–screw without cross-link. They found that a four-screw construct in adjacent vertebral bodies provides the strongest construct in pullout testing. A cross-link did not provide any additional strength to the all screw construct. They also found that the hook construct had significantly higher pullout strength in the lumbar spine compared to the thoracic spine.

In a multicenter study, Bess et al. [5] (Growing Spine Study Group) reported on complications in 910 growing rod surgeries in 143 patients with minimum 2-year follow-up. They divided the group as single (n = 73 patients) or dual rod (n = 70 pts) and subcutaneous (n = 54) or submuscular (n = 89). Complication rate per surgery was <20%. Complication rates were equivalent among single and dual rod constructs. Significance was found in number of implant-related complications requiring unplanned return to the operating room for single rod constructs compared to dual. The subcutaneous group had more complications per patient (1.6 vs. 0.99) and more wound problems (13 vs. 4 patients). Furthermore, subcutaneous placement of dual rods had higher overall complication rate, higher wound problems, prominent implants and patients undergoing implant-related unplanned return to the operating room. The conclusion was that the overall complication rate is comparable to historic reports; dual rods reduce unplanned trips to the operating room and submuscular position of implants is preferred over subcutaneous.

17.5.5 Growth Directed Surgery

Growth directed surgery is the phrase used to describe procedures where reduction of the spinal deformity relies on the remaining growth available. The classic example of this is the Shilla procedure described by McCarthy et al. This surgery involves limited instrumentation and reduction of the apical segment with specialized polyaxial Shilla screws that house two rods and allow those rods to glide within the construct. The concept is to improve the deformity of the spine by naturally directed growth along a new path (the rods that are placed).

McCarthy et al. [46] recently reported on 10 patients with 2-year follow-up. Three of these patients were either infantile or juvenile idiopathic scoliosis. Initial curve correction went from 70.5 (40–86°) to 27° (5–52°) at 6 weeks, and 34° (18–57°) at 2-year follow-up. Two patients had a staged anterior apical release. Complications included rod revision for growth off the end of the rods, rod exchange for a shorter one due to prominence, one broken rod, and two wound infections for a total of five surgeries among all ten patients beyond the index procedure. It was predicted that this same group of patients would have required 49 additional surgeries in a distraction-based growing rod model. For more details regarding this technique, please see Chap. 48.

17.6 On the Horizon and Conclusion

Significant strides have been made in the last decade regarding understanding and management of infantile and juvenile scoliosis. This unique disease entity, however, still leaves many areas undiscovered including genetic etiology, accurate scientific predictability of progression, ideal treatment for individual curves, and refinement in surgical technique.

The ideal surgery would include a minimally invasive approach with a durable and inert implant that rarely requires reoperation. Takaso et al. [61] in 1998, reported on the development of a rod containing a direct-current motor attached to a radio-controlled receiver. They performed successful correction of experimental scoliosis in beagles. The main issues with this device were its size (16 mm) and the placement of the receiver in the abdominal cavity. Akbarnia has recently explored the idea of remote lengthening, and animal studies are under way investigating this promising technology.

Ward et al. [65], as discussed in another chapter, are currently studying this very unique patient population to identify any markers for progression and the genetic basis of IIS and JIS. It is the hope of all treating physicians that success in this arena will be as productive as it has been in identifying these markers in adolescent idiopathic scoliosis.

In conclusion, idiopathic infantile and juvenile scoliosis is a disease entity that if left untreated can result in devastating and life-threatening complications. Early recognition and timely treatment are essential to management and for good outcome. Exciting new technology and improved surgical technique will result in lower complication rates, avoidance of natural history, and ultimately improved patient outcome [62].

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