Spine

Nonoperative treatment of infantile spinal deformity

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ABSTRACT

Progressive infantile scoliosis is one of the more challenging problems faced by pediatric spinal deformity surgeons. These curves can be rapidly progressive and lead to significant respiratory compromise. Moreover, surgical options are fraught with complications. There has been a widening role for casting in this population over the past 10-15 yr with encouraging outcomes. The literature of the last few years has reinforced evidence that casting works best in younger patients (< 2 yr old) with smaller, idiopathic curve types. There also is expanded evidence for the use of casting to delay surgery when curve resolution does not seem to be a realistic expectation. Studies have also demonstrated the role of increased body mass index in predicting better outcomes in idiopathic infantile patients and have examined how reduced vertebral body wedging improves the likelihood of maintaining curve correction after casting. In sum, these findings confirm that casting is an effective intervention and outline some new variables that can predict successful outcomes both before and after cast treatment.

Key Words
early onset scoliosis, infantile scoliosis, casting, conservative

INTRODUCTION

Conservative management of the infantile form of early onset scoliosis (EOS), defined as those under 3 yr of age, essentially includes casting or observation. Bracing, which is a mainstay of care in older children, is challenging in this rapidly growing younger age group with little to no literature on its effectiveness, except for maintenance of correction after casting.[1,2] The latest literature on casting continues to provide us with new insights, and reinforces existing data from the earlier literature.

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IMAGING IN INFANTILE SCOLIOSIS

Intraobserver and Interobserver Measurement Variability of the Rib-Vertebral Angle Difference

Nearly 50 yr ago Dr. Mehta taught us that the rib-vertebral angle (RVAD) was crucial to predicting progression of infantile idiopathic scoliosis, with a value of 20 degrees generally differentiating those who will and those who will not progress,[3] a finding that has been reproduced in multiple studies.[4–6•] There is some literature on reliability of measuring the RVAD as well as the Cobb angle in infants,[7] which demonstrated good reliability. However, a more recent project by Tysklind *et al.*[6•] looked at the magnitude of the variability, something not done in earlier work, and found large differences between measurements. They demonstrated remarkably high values for both intraobserver (24 degrees) and interobserver (23 degrees) error. They recommended caution relying solely on the RVAD for counseling and medical decision making.[6•]

The Prevalence of Intraspinal Anomalies in Infantile and Juvenile Patients with "Presumed Idiopathic" Scoliosis: MRI-based Analysis of 504 Patients

Evaluating the neuroaxis with an MRI is generally recommended in patients with early onset scoliosis, especially when there is no underlying syndrome or neuromuscular disease to explain the presence of a spinal deformity.[8] The incidence of abnormal findings on MRI in presumed idiopathic EOS has been reported to be between 11% and 26% in generally small studies.[9]

Zhang *et al.*[9] looked to better define the incidence of abnormal MRI findings in a much larger population of infantile and juvenile patients (all under the age of 10 yr). They identified 504 patients with presumed idiopathic early onset scoliosis, which they defined as otherwise normal children no older than 10 yr of age with normal neurological examinations, and no other diagnoses. All patients also had a full spinal MRI. In their study, 18.7% of patients had a documented intraspinal neural axis abnormality, solidly in the middle of the range previously reported in the literature.

The most common finding was an isolated Arnold-Chiari malformation, which was found in 65% of patients, but they did not differentiate between Chiari I and Chiari II malformations. A syrinx was found in 33% of patients, with most of these also having a Chiari malformation. Other less common findings included diastematomyelia (13%), tethered cord (10%), and spinal cord tumor (3%). A number of patients had multiple findings which is why the total number is over 100%.

The authors did not report on the number of patients who had surgical treatment for the finding or if the findings altered treatment of outcome in any way. They did find that abnormal MRI findings were more common in males as well as in patients with left thoracic or right lumbar/thoracolumbar curves. Age, curve magnitude, apex location, thoracic kyphosis, and curve span were not found to be associated with increased abnormal MRI findings. The incidence was statistically equivalent in the infantile and juvenile age groups.

In summary, this study was the largest of its kind to evaluate the incidence of MRI abnormalities in early onset scoliosis, with findings in line with other studies. The recommendation continues to be to obtain an MRI in cases of apparent idiopathic EOS as approximately one in five patients will have an abnormal finding that could impact management.

PREDICTORS OF SUCCESSFUL CAST MANAGEMENT

There were several articles in the past few years that reinforced earlier findings that younger children with smaller curves do better, and idiopathic curves do better than nonidiopathic curves. There has also been some more novel research looking at other clinical and radiographic predictors including body mass index (BMI), in cast correction, and the ratio of the convex and concave heights of the periapical vertebrae.

Initial Cast Correction as a Predictor of Outcome Success for Infantile Idiopathic Scoliosis

Gomez *et al.*[10•] utilized data from the Children's Spine Study Group database to evaluate clinical and radiographic factors associated with successful outcomes after elongation-derotation casting for idiopathic infantile scoliosis. They defined success as a curve that was under 15 degrees at last follow-up, a rather narrow, but useful, definition.

The authors identified 68 patients from the study group's database with an average follow-up of 2.5 yr. Twenty-five patients (37%) met their definition of success. Across the entire population, the major coronal Cobb angle improved 50%, from 46 to 23 degrees in the first cast, and averaged 25 degrees at last follow-up. Comparing their successes and failures, the success group was significantly younger at initial casting (1.4 vs. 2.1 yr), had smaller pre-casting major Cobb angles (40 vs. 50 degrees), and had more in cast curve correction (57% vs. 48%). Additionally, successful patients spent less time in treatment and had fewer total casts than the failure group (median 9 mo in five casts vs. median 18 mo in seven casts). Multivariable logistic analysis demonstrated that for each year younger, the odds of successful cast treatment increased 2.5 times, and for each additional cast the odds of success decreased by 23%. It is not clear, however, if an increase in the number of casts represents a surrogate for severity or nonresponse to treatment so this last finding must be interpreted with some caution.

This study did not find that lower pre-cast RVAD correlated with success. They also did not find BMI to be a significant predictor of outcome.

Overall, these results greatly confirm findings from previous studies, namely that casting is more successful in younger patients (under 2 yr of age) with smaller curves (under 50 degrees).[3,11] This is a message that has been reproduced repeatedly in the literature, and cannot be overstressed.

Serial Mehta Cast Utilization in Infantile Idiopathic Scoliosis: Evaluation of Radiographic Predictors

A 2017 study by Hassanzadeh *et al.*[4] reported post-casting radiographic predictors of sustained curve correction in infantile idiopathic scoliosis. Their study population was 45 consecutive patients who were casted at a mean age of 19 mo. These patients had an average major Cobb angle of 52 degrees and an average follow-up of over 3 yr *after* their last cast. They looked at multiple radiographic variables including Cobb angle, RVAD, focal deformity (Cobb angle from 1 level above the apex to one level below the apex) and the ratio of the coronal height of the concave edge to the convex edge of the periapical vertebrae (Figure 1). This last variable is an indication of vertebral wedging with smaller ratios indicating more wedging. Their population had 50% improvement in Cobb angle after the last cast (52 to 26 degrees) and this further improved to 16 degrees at the last follow-up. The majority of their patients were transitioned to bracing after casting (62%), while 29% experienced curve resolution and 9% progressed to surgery. The radiographic criteria associated with success, defined as sustained curve correction at final follow-up, included the concave to convex ratio and focal deformity correction. Clearly these two findings are related, although the author's analysis did not say if these predictors were independent of each other or not. The concave to convex ratios that were associated with sustained correction were at least 0.86 at the apex +1 vertebra, and at least 0.63 at the apex -1 vertebra. They also found that

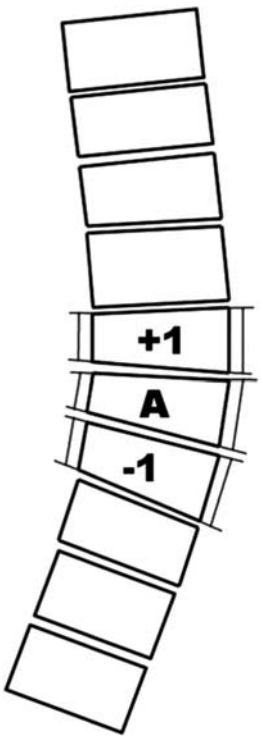

FIGURE 1. Image depicting measurements of concave and convex aspects of the 3 apical vertebrae. "A" indicates the apex vertebral segment.

an RVAD at final casting of less than 20 degrees was associated with successful outcomes.

This study had excellent results with rather large curves, greater than 50 degrees on average, likely because the age at casting was young, 18.8 mo, lending further support for early casting in this population. The concept of convex to concave height ratios, essentially a surrogate for vertebral wedging, shows that if treatment is able to create more rectangular peri-apical vertebrae through growth, then sustained improvement in Cobb angle can be expected.

Serial Casting for Infantile Idiopathic Scoliosis: Radiographic Outcomes and Factors Associated with Response to Treatment

This relatively small study of 21 patients with infantile idiopathic scoliosis looked at pre-casting radiographic and clinical predictors of successful outcome in casting for this condition.[1] They defined success as sustained improvement in the major Cobb angle of at least 10 degrees from baseline (pre-casting) at last follow-up. They reported a 71% success rate by this definition in their group, with the majority of the improvement coming after the first cast (14 degrees). The only pre-casting factor found to be correlated with successful outcome was BMI, with a lower BMI found in the failure group (17.6 vs. 14.8). They calculated a 2.4-fold increase in successful treatment for each unit increase in BMI. This is in contrast to most other studies looking at this factor that have found no relationship,[10•] although Mehta described the "sturdy" phenotype which had better outcomes.[3]

Pre-casting RVAD, Cobb angle, age at first cast, rib phase, and curve flexibility (in contrast to the study by Gomez *et al.*[10•]) were *not* found to be pre-casting predictors of successful treatment as defined in this study. The small study population likely contributed to this. However, over time the influence of these factors did evolve, and by 2 yr after casting was initiated, the successful group had significantly lower RVADs, smaller Cobb angles, and fewer patients with phase II ribs. Additionally, all five curves that resolved had phase I ribs pre-casting.

Another important finding was that even though four patients in this study were candidates for open surgery, casting allowed a delay in surgery by an average or 4.4 yr, a very positive outcome of casting as this likely reduces surgical complications in that group.

This study's major finding was demonstrating that higher BMIs were associated with better outcomes, although it is important to remember that none of these patients had a BMI that went into the obese range, so there likely is an upper limit to this effect.

Serial Derotation Casting in Idiopathic and Nonidiopathic Progressive Early-Onset Scoliosis

Gussous *et al.*[5] reported their consecutive series of 74 patients with EOS treated with casting, comparing the outcomes of those with idiopathic scoliosis (41 patients) to those with nonidiopathic scoliosis (33 patients).[5] The groups were similar with respect to pre-casting Cobb angle and RVAD, but the response to casting was dramatically better in the idiopathic group. For patients completing casting in the idiopathic group, Cobb angles improved 28 degrees (from 46 to 18 degrees), while in the nonidiopathic group Cobb angles only improved 10 degrees (from 42 to 32 degrees).

They also reported on other factors associated with outcomes. Patients who were candidates for open surgery were older at first casting (29 mo vs. 20 mo) and had larger curves (62 vs. 42 degrees). Within the idiopathic subgroup, initiating casting prior to 24 mo of age was associated with better outcomes, but within the nonidiopathic group, no such age cutoff could be found. There was no difference in the precasting RVAD between these groups.

Results of Casting in Severe Curves in Infantile Scoliosis

Stasikelis *et al.*[12•] looked at patients with infantile scoliosis and at least 50-degree curves to study the impact of casting in this more severely affected group. They had 44 total patients, and they split them into those with (18) and without (26) comorbidities (abnormal MRI or genetic syndromes). Overall, 27% of all patients had resolution of their curves with casting, but similar to the findings of Gussous *et al.*[5] the idiopathic group did much better (35% resolution vs. 17%). For the patients in whom the deformity did not resolve, 32% maintained their curve to within 15 degrees of their pre-cast curve and 30% improved at least 15 degrees. The remaining five patients (11%) had progression of more than 15 degrees and at the time of writing, four of them had undergone growth friendly open surgery after an average delay of over 6 yr.

Regarding age at casting, they found that the likelihood of curve resolution diminished with age, although they did document curve resolution in children as old as 3 yr. Overall, this study showed positive results with casting larger curves as patients experienced curve resolution, curve stabilization, or a significant delay of surgical intervention.

CONCLUSION

The last few years of research on infantile scoliosis has reaffirmed earlier findings: progressive curves in this age group respond best with early casting (less than 2 yr of age) in smaller, idiopathic curves (under 40 to 50 degrees). This is a message that cannot be overstated, as it has been reproduced in every study. However, when these criteria are not met, there is still a role for casting as a certain percentage of patients with even large idiopathic curves can have their curves resolve, and at worst growth friendly surgery can be delayed, often by several years. Future studies should help resolve the role for casting in nonidiopathic scoliosis as well as further delineate the cognitive and psychosocial impact of multiple anesthetics on this population.

REFERENCES AND RECOMMENDED READING

Papers of particular interest published within the annual period of review are highlighted as:

- •of special interest
- ••of outstanding interest

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This study demonstrated a very wide range in the magnitude of inter- and intra-observer reliability of the RVAD, over 20 degrees, and recommended not making decisions based solely on this variable.

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10. • Gomez JA, Grzywna A, Miller PE, *et al*. Initial cast correction as a predictor of treatment outcome success for infantile idiopathic scoliosis. *J Pediatr Orthop*. 2017; 37:e625–e630.

This is one of the larger studies on casting in the literature, derived from the largest EOS registry. Their findings reinforced the need to initiate early casting in this population (under 2 years of age).

11. Sanders JO, D'Astous JL, Fitzgerald M, *et al*. Derotational casting for progressive infantile scoliosis. *J Pediatr Orthop*. 2009; 29:581–587.
12. • Stasikelis PJ, Carpenter AM. Results of casting in severe curves in infantile scoliosis. *J Pediatr Orthop*. 2018; 38:e186–e189.

This is the first study specifically looking at larger (>50-degree) curves in infantile scoliosis undergoing casting. They reported encouraging outcomes and demonstrated that casting was able to resolve or stabilize 90% of these curves.