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Review Article:

Early Onset Scoliosis: Definition, Etiology, Physical Examinations, Classification, and Methods of Treatment

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Abstract

Background: Early Onset Scoliosis (EOS) is defined as scoliosis occurring before the age of 5 years. It affects not only the growing spine but also has a direct influence on the cardiopulmonary development and function of the child. Cardiopulmonary function is compromised as a result of decreased size of the thoracic cavity, which has a negative effect on lung alveolar development.

The goal in the management of EOS is to control the deformity and allow continued growth of the spine and thoracic cavity.

Non-fusion instrumentation facilitates this and allows ongoing respiratory development and optimal cardiopulmonary functions. However some authors describe that early onset scoliosis is that scoliosis starts before the age of 10 years. The debate in these definitions is made because of the difference in spine growth and cardiopulmonary compromise before and after age of five.

Key Words: *Early onset scoliosis – Growing rods – VEPTR – Magnetically controlled rods.*

Review Article

Etiology of EOS:

As mentioned before EOS is that scoliosis starting before the age of 5/10 years **[1-4]** it has a special etiological factors that could be (A) Idiopathic (infantile and juvenile idiopathic scoliosis) (B) Congenital (failure of formation, failure of segmentation, combined or unclassified) (C) Neuromuscular (cerebral palsy, myelodysplasia, muscle diseases ...etc) and syndromic **[5,6]**.

Natural history:

The natural history of EOS is bleak. Pehrsson et al., reported a significantly increased mortality rate in patients with EOS, likely resulting from respiratory failure **[7]**. Several studies have shown that humans cease growing new alveoli after age 7 years **[8-10]**. Thus, deformity leading to decreased chest volume before this age has a permanent, negative effect on pulmonary function regardless of restoration of alignment later in life. Early-onset scoliosis is a serious condition that requires early intervention.

Physical examination:

The physical examination of a young child with a spinal deformity begins with a thorough medical history documenting the onset of the spinal deformity and its changes over time as well as associated medical conditions such as skin, pulmonary, and syndromic abnormalities. The overall health of the child can be assessed with a routine patient evaluation with a special consideration of the child's neurologic status and musculoskeletal features. The information obtained from the patient's history is paramount in conducting an appropriate physical examination and formulating an effective treatment plan **[11]**.

Classification:

Classification systems can be useful tools for clinical care and research but must be proven as reliable. The C-EOS scheme can be used as a tool for classifying EOS patients for clinical communication and research **[12]**. Fig. (1).

Treatment of EOS:

A- *Non surgical option:*

Bracing has a very little role in EOS (patients older than 20 months).

Serial casting, casting correction can benefit patients with EOS due to congenital or non-

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congenital causes. Although the therapeutic efficacy of casting on congenital patients is not as good as that of non-congenital patients, the importance of casting for those with congenital EOS is to delay the need for initial surgery [13]. Fig. (2).

B- *Surgical option:*

1- *Distraction based:* These implants correct and maintain the spinal deformity via distraction which may be spine to spine, spine to rib, pelvis to spine and pelvis to rib [3].

Growing rods:

These rods provide structural support, correct and maintain the deformity and allow axial spine growth. Serial lengthening of the rods is done every 6-12 months [14,15]. In early trials, a single growing rod was frequently used and applied on the concavity but as a frequent complication was seen most of the treating surgeon shifted to using the dual growing rod technique [16]. Another important issue was discussed in using growing rod which is the anchoring points and applying the rods in various methods being spine to spine, spine to rib, pelvis to spine or pelvis to rib. Some surgeons prefer using the iliac crest as the distal anchoring points being stronger than the vertebra [17]. Fig. (3).

Vertically Expandable Prosthetic Titanium Rib (VEPTR):

It is a surgical technique that does not involve spinal fusion but instead uses an expansion thoracoplasty via an opening wedge thoracostomy and implantation of a thoracic VEPTR. This procedure directly treats thoracic insufficiency syndrome by lengthening and expanding the constricted hemithorax and allowing growth of the thoracic spine and rib cage, and indirectly corrects EOS scoliosis without the need for spine fusion [18]. To accommodate growth and maintain correction of the deformity, the device is lengthened every 6 months. The results of this procedure have been favorable to date. As reported by Campbell et al., the longitudinal growth of the thoracic spine after expansion thoracoplasty was 7.1mm per year, compared with a normal 6mm/y for children 5 to 9 years old [19]. In addition, the Space Available for the Lung (SAL) ratio and the mean interpedicular ratio was increased [19]. Fig. (4).

Magnetically controlled rods:

Magnetically controlled growing rods was introduced as an alternative to tradional growing rods with similar to better result and having the advantages of avoiding extra surgeries for lengthening and skin complications these rods are inserted in the index surgery in similar way to traditional rod but lengthening procedures are done in the outpatient clinic using a special remote control [20]. Fig. (5).

2- Tension based (growth inhibition): The concept of using staples for growth modulation of long bones has been repeatedly tested and is fundamentally sound. Staples for epiphysiodesis of long bones in angular deformity have been used for over 50 years. Similarly, the potential benefits in the spine were noted around the same time. In 1951, Nachlas and Bordenwere initially succeeded in their ability to create and correct lumbar scoliosis in a canine model using a staple that spanned several vertebral levels [21]. This technique is used now to control EOS deformity with great success [22]. Fig. (6).

3- Growth guidance: This technique allows for continuation of natural spinal growth while correcting the deformity should be the goal of treating this complex condition. Both SHILLA and Luque trolly systems are used to allow for continued growth of the pediatric spine while correcting and guiding the apex and guiding the future growth of the curvature, Fig. (7). The system involves selective fusion across the apex of the curvature, and minimally invasive instrumentation is then used above and below the apex to allow for continued growth of the spine [23,24].

Etiology	Cobb angle (Major Curve)	Maximum Total Kyphosis	Progression Modifier (Optional)
Congential/ structural	1:≤20º	(–):≤20º	P0: <10º/yr
Neuromuscular	2:21-50º	N:21-50º	P1: 10-20º/yr
Syndromic	3:51-90º		
Idiopathic	4:>90º	(+):>50º	P2: >20º/yr

Fig. (1): C-EOS (classification of early onset scoliosis).

Fig. (2): Casting for EOS [13].

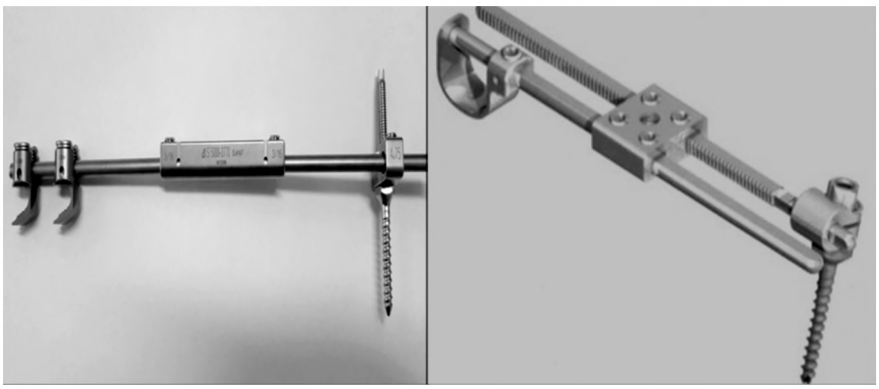

Fig. (3): ISOLA type of growing rods (Left) and GSP type (Right).

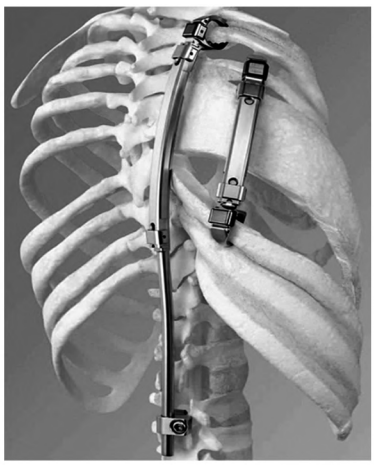

Fig. (4): VEPTR instrumentation **[18]**.

Fig. (5): Magnetically controlled growing rods **[20]**.

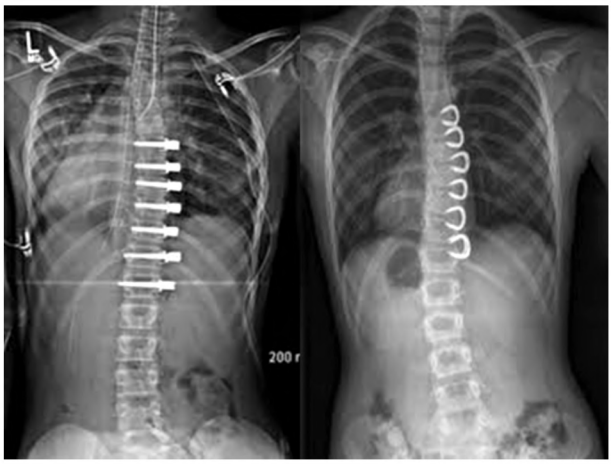

Fig. (6): Tethering techniques in EOS.

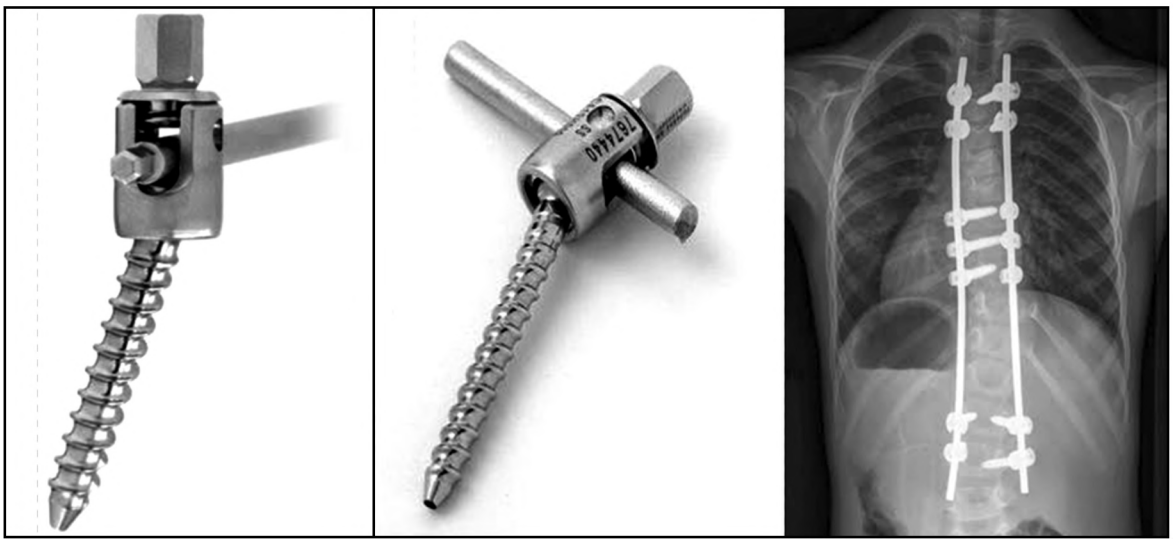

Fig. (7): Shilla technique for EOS **[23]**.

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الجنف المبكر فى الآطفال
(تعريفه، آسبابه، طرق التشخيص، تقسيمه وآنواعه وطرق العلاج)

يبدآ إعوجاج العمود الفقرى (الجنف) المبكر قبل سن الخامسة من عمر الطفل ويشمل كل آنواع الإعوجاج سواء كانت ناتجة عن عيوب خلقية آو نتيجة خلل فى الجهاز العصبى والعضلى آو نتيجة لوجود بعض المتلازمات المصحوبة بإعوجاج العمود الفقرى آو مجهولة السبب. كما يرى بعض الآطباء آن الجنف المبكر هو ذلك النوع من الجنف الذى يبدآ قبل سن العاشره من عمر الطفل.

كما يعانى المختصون فى علاج حالات إعوجاج العمود الفقرى من صعوبة فى علاج الجنف المبكر سواء جراحيا آو تحفظيا حيث تفشل آغلب طرق العلاج التحفظى سواء بإستخدام آنواع مختلفة من الآحزمة آو بإستخدام العلاج الطبيعى. كما آن التدخل الجراحى التقليدى لعمل تثبيت وإلتحام للفقرات فى الآطفال دون الخامسة يؤدى إلى وجود تقصير فى طول جسم الطفل حوالى ١٢.٥سم مقسمة إلى ٧.٨سم فى الفقرات الظهرية و٤.٧ فى الفقرات القطنية.