

Classification of Congenital Scoliosis and Kyphosis: A New Approach to the Three-Dimensional Classification for Progressive Vertebral Anomalies Requiring Operative Treatment

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Study Design. We reviewed three-dimensional (3D) computed tomography (CT) images of congenital spinal deformities and proposed a new classification based on the information obtained.

Objectives. The purposes of this article were to clearly illustrate the limitations of two-dimensional classification, to summarize the clinical significance of 3D analysis of congenital vertebral anomalies, and to propose a new 3D classification of congenital vertebral anomalies.

Summary of Background Data. The classification of congenital scoliosis or kyphosis were based on radiographic findings of plain radiograph images of congenital vertebral anomalies, it is sometimes difficult in classifying the large variety of anomalous vertebrae or severely twisted 3D curves.

Methods. Three-dimensional CT images of more than 150 patients with congenital spinal deformities were analyzed and compared with plain radiograph images. By developing the algorithm for the evaluation of malformed vertebrae in terms of numbers of abnormal vertebrae, type of formation failure, and type of segmentation failure in separate steps, we attempted to revise the classification of congenital spinal deformities.

Results. The images of plain radiograph cannot demonstrate the spatial relationship of each structure of the vertebrae. Three-dimensional findings in congenital-deformed vertebrae included several types of laminae and clearer definitions of each type of anomalous vertebrae. By developing an algorithm for the evaluation of congenital spinal deformity, congenital spinal deformity could be mainly classified into 4 types of congenital vertebral abnormalities: Type 1: solitary simple, Type 2: multiple simple, Type 3: complex, Type 4: segmentation failure.

Conclusion. The large volume of information that can be obtained by evaluating 3D CT images of congenitally deformed vertebrae can be a great help in developing

a strategy for surgical treatment. We need to develop a new classification of congenital scoliosis based on the perspective of 3D imaging to understand the etiology and embryology, as well as to determine an operative strategy.

Key words: congenital spinal deformity, classification, three-dimensional analysis, discordant anomaly. **Spine** 2009;34:1756–1765

There are many factors by which congenital scoliosis and kyphosis can be classified. One of the distinct differences of the vertebral anomalies in congenital spinal deformities is whether or not there are defects of lamina in some of the anomalous vertebrae. Radiographically and clinically, it is very simple and useful to divide them into closed vertebral defects and open spine lesions according to these differences.

Open spine lesions are clearly distinct in morphology from closed defects. This remarkable difference may be due to the differences of initial inductive influences during development of the vertebral bodies and arches, which confer a degree of independence in their initial developmental pattern. This article focuses on the closed vertebral defects similar to those reported in classifications of congenital scoliosis and kyphosis. From the morphologic viewpoint, closed vertebral defects demonstrate great variation in the number of vertebrae involved and patterns. Winter *et al*^{1,2} and McMaster *et al*^{3,4} presented independent classifications of congenital scoliosis and kyphosis. Both classifications, whether they were for scoliosis or kyphosis, were based on the same concept of embryologic maldevelopment of the spine, that is, abnormalities of formation and segmentation. Winter¹ classified congenital scoliosis into failure of formation, failure of segmentation, and the mixed-type. This classifications has been widely used to describe the type of congenital abnormal vertebrae for more than 20 years. However, because all these classifications were based on the radiographic findings of front and lateral, plain x-ray images of congenital vertebral anomalies, they sometimes result in difficulty in classifying them when severe twisted three-dimensional (3D) curves are present, and might result in wrong decision-making during strategic planning of congenital spinal deformities.

We have analyzed the 3D computed tomography (CT) images of more than 150 patients with congenital spinal deformities and have described the results of this analysis

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Figure 1. Complicated case of congenital scoliosis. A unilateral unsegmented bar may be diagnosed with a plain x-ray image (**A,B**). However, 3D CT shows a combination of slight wedged vertebral bodies of the anterior structure and contralateral 3 hemilaminae skipping 1 normal-shaped lamina each on the posterior structure (**C,D**).

in several reports.^{5–7} Based on these 3D CT findings and a review of literature related to the classification of congenital spinal deformity, this review aims to (1) more clearly illustrate the problems of two-dimensional (2D) classification, (2) emphasize the clinical significance of 3D analysis of congenital vertebral anomalies, and (3) propose a new classification system for congenital vertebral anomalies, which might allow us to develop a more logical approach to the surgical strategy as well as more clearly understand the natural history and etiology.

■ The Problems of 2D Classifications

Although the quality of the plain x-ray images has improved significantly through the development of computerized digital radiographic technique, it still has limitations in resolution. Furthermore, plain x-ray images are not 3D and cannot demonstrate the spatial relationship of the structure of each vertebra. In particular, as long as we evaluate congenital spinal deformities with such a variety in curve types and morphologies, it is clearly impossible to understand the 3D structure of each abnormal vertebra using only plain x-ray.

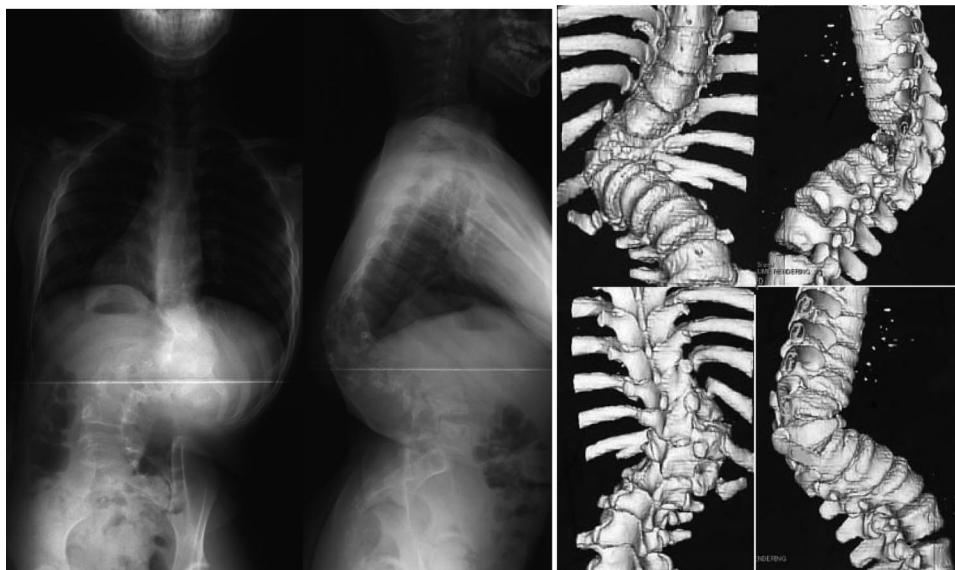
The vertebral body is relatively easier to evaluate in plain x-ray images compared with the posterior components. Thus, it can be said that 2D classifications using plain x-ray images were comprised mainly of vertebral bodies. A solitary hemivertebra can be easily diagnosed by every spine surgeon using only plain x-ray images. By contrast, it is very difficult to judge the type of multiple

complex vertebral anomalies using plain x-ray images (Figure 1). Some may recognize it as a unilateral unsegmented bar. However, 3D images have shown this deformity as a combination of slight wedged vertebral body of the anterior structure and contralateral 3 hemilaminae skipping every normal-shaped lamina on the posterior structure (Figure 1). This vertebral anomaly exhibits a difference in type between anterior and posterior structures. Complicated mixtures of anomalous vertebrae like this case with a combination of the different abnormalities of the anterior structure and the posterior structure would be only classified into mixed type, according to the Winter's classification. It is too simple to put all these types into the mixed type together with the multiple of other possible spine abnormalities.

Severe curvature of the spinal column is another example that is difficult to evaluate by plain x-ray images. It can be very difficult to classify even a single malformed vertebra once the spinal column becomes so twisted so much that it develops into an imbalanced severe 3D kyphoscoliotic deformity (Figure 2).

Failure of segmentation is one of 3 major categories in 2D classification in congenital scoliosis and kyphosis. However, even anomalous vertebrae that are classified as due to failure of formation sometimes exhibit different types of synostosis to the adjacent vertebrae. A semisegmented hemivertebra is 1 example and these anomalous vertebrae have 2 different characters: one is nature of formation failure and the other is of segmentation fail-

Figure 2. Severe curve of congenital kyphoscoliosis. A 12-year-old boy exhibiting 97° of scoliosis and 81° of kyphosis in the thoracolumbar segment. It is almost impossible to determine the type of congenital scoliosis according to Winter's classification. Three-dimensional CT shows a combination of hemivertebra of T11 and a wedge vertebra of T10.



ure. Although this type of anomalous vertebrae has been classified into failure of formation, according to the Winter's classification to date, should this type be classified as the mixed type? A nonsegmented hemivertebra is another example and can also be regarded as a hemivertebra that has a character of segmentation failure both caudally and cranially. From the viewpoint of synostosis with cranial and caudal adjacent vertebrae, both a nonsegmented hemivertebra and a block vertebra should belong to the same group, segmentation failure. However, the former is recognized as a vertebral anomaly of failure of formation and a latter is one of the failure of segmentation, according to the Winter's classification (Figure 3).

When we consider the natural history of each type of congenital vertebral anomaly, it is uncertain without considering the type of anterior and posterior abnormalities, including segmentation abnormalities. The presence of hemivertebrae makes it very difficult to predict progression. Some of hemivertebrae cause severe progression, and others do not progress at all.^{2,3,8} Although McMaster and Ohtsuka³ reported the natural history of each types of congenital vertebral anomalies, it is not

rare that some of them unexpectedly progress or not. Of course, many factors influence the progression. As long as we are evaluating hemivertebrae using plain x-ray images, our abilities to predict the natural history and prognosis are limited.

■ The Clinical Significance of 3D Images of CT in Congenital Spinal Deformity

With the recent improvement of diagnostic imaging, particularly with 3D CT, detailed observation of the anterior and posterior components of the malformed vertebrae and relationships with adjacent vertebrae by means other than macroscopic observation during surgery has become possible. Many reports have described the superiority of 3D CT over plain x-ray from different viewpoints.^{7,9–11} Newton *et al*⁹ mentioned that more than 50% of the cases showed additional abnormalities that were not appreciated on plain radiographs or axial 2D CT images. Hedequist and Emans^{10,11} reported that unexpected anomalies might be encountered posteriorly when plain films alone were used in surgical planning. These morphologic findings might be one of the causative factors that changed the recent strategy

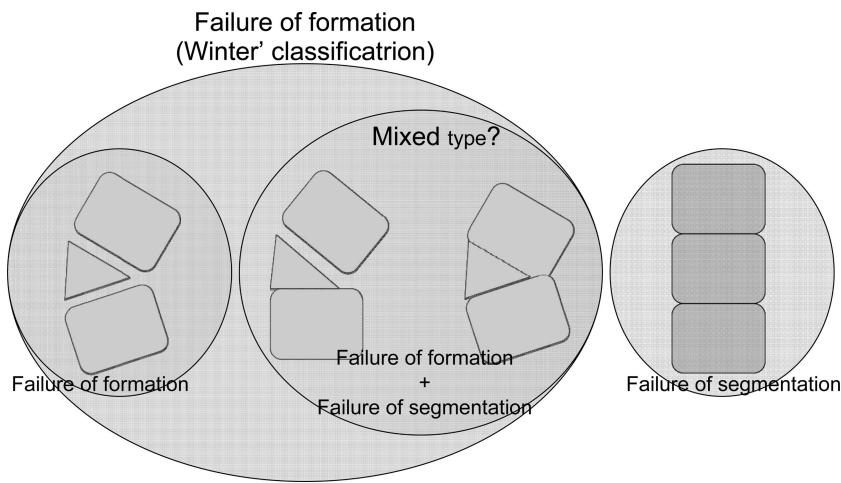


Figure 3. Ambiguous border of segmentation failure. Failure of formation in the Winter's classification includes fully segmented, semisegmented, and nonsegmented hemivertebrae. However, semisegmented and nonsegmented vertebrae have the same characteristics of segmentation failure as a block vertebra that belongs to segmentation failure.

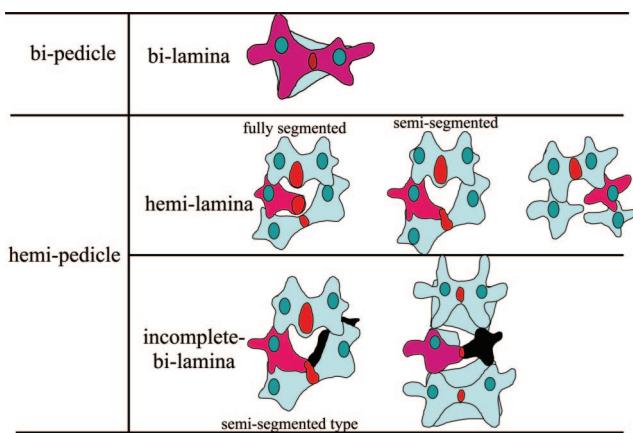


Figure 4. Morphology of the posterior components of malformed vertebrae. These abnormal shapes of laminae are observed not only in the solitary malformation group but also in the multiple group. In the latter, various combinations of abnormal shaped-laminae exist closely together. Reprinted with permission from *Spine*.⁷

of operation from *in situ* fusion or partial correction of the deformity to maximum correction with osteotomy including vertebrectomy.

However, most of the 3D analysis of congenital scoliosis performed in literature reached the conclusion that the 3D CT was superior in demonstrating abnormal vertebrae more clearly and did not describe the detailed characteristics of congenital scoliosis. In 2007, Nakajima *et al*⁷ analyzed the 3D morphology of formation failure type of congenital scoliosis in detail and reported that even the same type of hemivertebrae diagnosed by plain x-ray images exhibited different types of laminae in the same manner as that observed in the vertebral body anomaly. Furthermore, they advocated 2 types of congenital vertebral anomalies, unison anomaly, and discordant anomaly, as the new categories of congenital spinal deformity. Imagama *et al*⁵ attempted to evaluate the type and site of segmentation failure using 3D CT images in congenital spinal deformities and presented the existence of discordant anomaly of segmentation (Figure 4).

■ Anatomic Findings of 3D Morphology in Congenital Spinal Deformities

Each type of anomalous vertebrae, such as hemivertebra, wedge vertebra, butterfly vertebra, and typical type of segmentation failure, each exhibit distinct characteristics in 3D morphology that have been not detected and have indeed been ignored for a long time.

Hemivertebra

In definition, hemivertebra is a wedged-shaped vertebra with 1 pedicle on 1 side. It can be subdivided as fully segmented, semisegmented, and unsegmented, according to the relation with the cranial and caudal adjacent vertebral bodies. There has been no description about the anomalous changes of posterior structure of hemivertebra until 2007. Nakajima *et al*⁷ reported the types of laminae of formation failure and classified them into bilamina and hemilamina, based on the number of pedicles

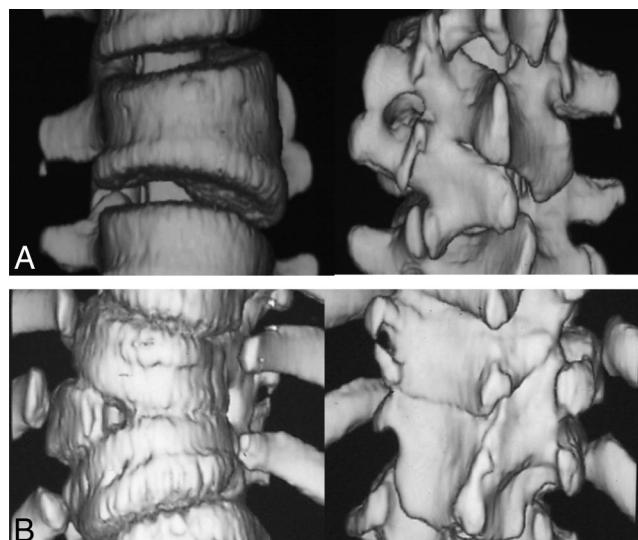


Figure 5. Discordant segmentation in hemivertebrae. A hemivertebra has semisegmented hemibody and fully segmented hemilamina (A). A hemivertebra has fully segmented hemibody and semisegmented hemilamina (B).

of the anomalous vertebra. Laminae with 1 pedicle are subclassified into fully segmented hemilamina, semisegmented hemilamina, spina bifida, and incomplete lamina, which are defined as a nearly normal bilamina with only 1 pedicle (Figure 5). Some hemivertebrae exhibit fully segmented hemibodies with semisegmented hemilaminae and others exhibit semisegmented hemibodies with fully segmented hemilaminae (Figure 4). These findings should be regarded as a discordant segmentation in hemivertebrae and may be one of the reasons why it is difficult to predict the natural history of each hemivertebra.

Wedge Vertebra

Wedge vertebrae are defined as wedge-shaped vertebrae with 1 pedicle on each side. Because wedge vertebrae exhibit 2 pedicles they inevitably exhibit a bilaminar posterior structure. Three-dimensional CT analysis can clearly demonstrate deformation of vertebral body and these findings may contribute to confusion as to whether it is congenital or secondary because wedge-shaped vertebral body can be quite often observed in the apical area of severe scoliosis. These changes of the apical vertebrae are secondarily caused by regression of vertebral growth on the concave side of the apical area. Forty cases of scoliosis (congenital scoliosis is not included) with the Cobb angle more than 80° treated in our hospital were evaluated and identified that the apical vertebra is severely wedged-shaped and that it never exceeded more than 50% of wedging. This study indicated that secondary wedged-deformation of the vertebrae was usually observed along a few consecutive apical vertebrae with an apical vertebra exhibiting the most severe wedged-shaped and never exhibiting a decrease more than 50% of vertebral height on the opposite side. Formation of osteophytes on the edge of the vertebral body or ridge formation on the concave side may be a result of secondary

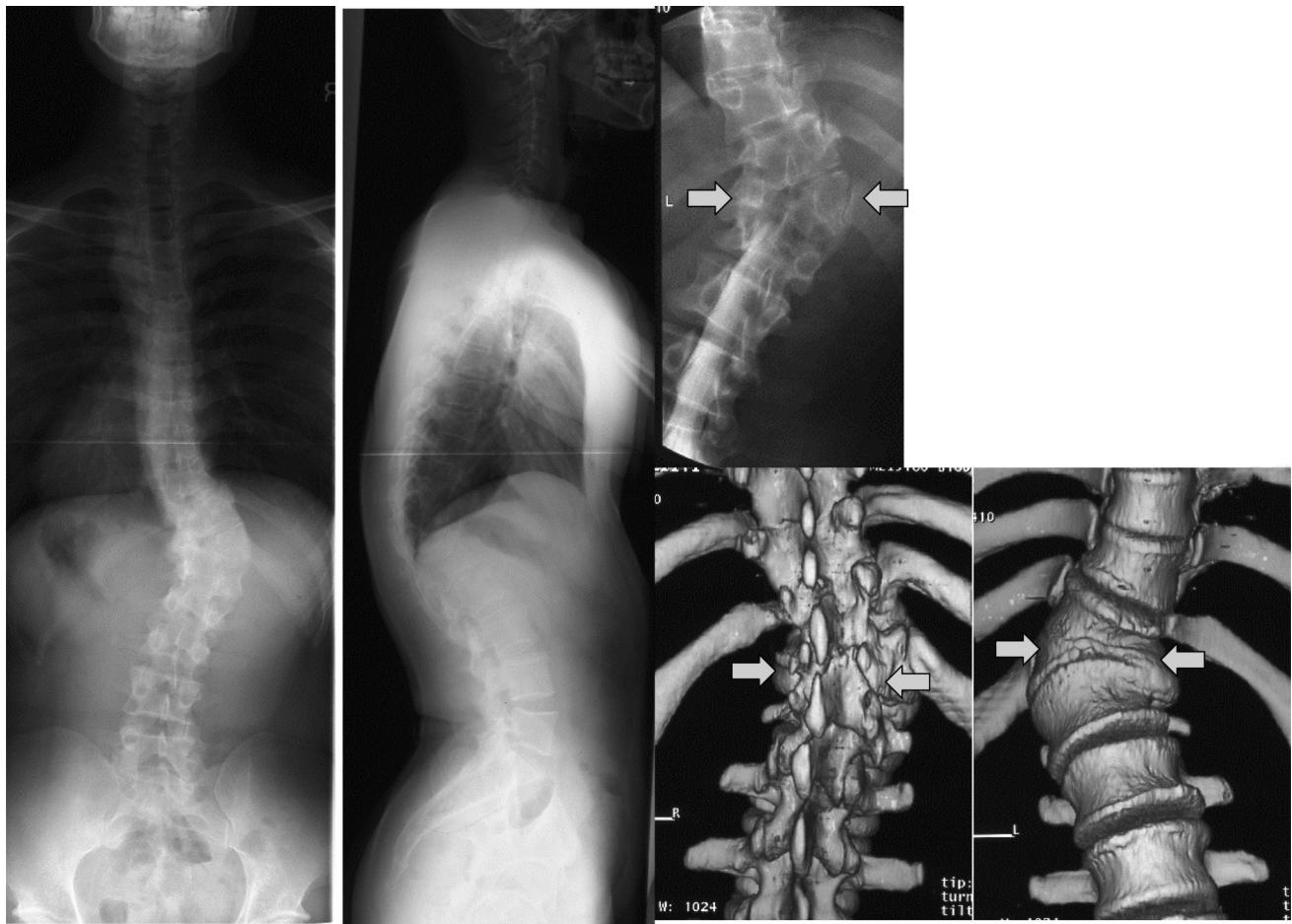


Figure 6. Asymmetrical butterfly vertebra. A 30-year-old man patient complained of lower back pain and numbness of the left lower extremity for more than 5 years. The plain x-ray showed 49° of scoliosis and 50° of kyphosis in the thoracolumbar area. Three-dimensional CT demonstrated an asymmetrical butterfly vertebra of T12 with posterior bilamina (arrow).

deformation of wedge-shaped vertebrae. In this context, wedge vertebra may be defined as a wedged-shaped vertebra in which the height of 1 side decrease by more than 50% of the opposite side, or whose adjacent cranial or caudal vertebra is almost normal shape, not wedged-shaped if the height on the 1 side of the wedged-shaped vertebra decreases less than 50% of the opposite side.

Butterfly Vertebra

The butterfly vertebra belongs to the anterior formation failure in the classification; it is the vertebra with 1 pedicle on each side and partial defect of the vertebral body. This type of formation failure is usually included in the congenital kyphosis because many butterfly vertebrae are clinically observed with symmetrical partial defects of the center of the vertebral body that result in anterior wedged-shaped vertebrae. Since they exhibit 2 pedicles, their posterior components are bilamina despite variations of the asymmetrical shape. An asymmetrical partial defect of the vertebral body often causes scoliosis and is sometimes misidentified as hemivertebra on plain x-ray images (Figure 6). Many butterfly vertebrae might be diagnosed as hemivertebrae or posterior quadrant hemivertebrae before development of 3D CT reconstruction technique. To prevent this misinter-

pretation, we should always check the number of pedicles and the type of posterior components. If an anomalous vertebra exhibits a wedged-shaped body and a bilamina with bilateral pedicles, it is definitely an asymmetrical butterfly vertebra and we should look for a very small hemibody on the opposite side (Figure 6). In some cases, the small opposite hemibody is synostosed to the caudal or

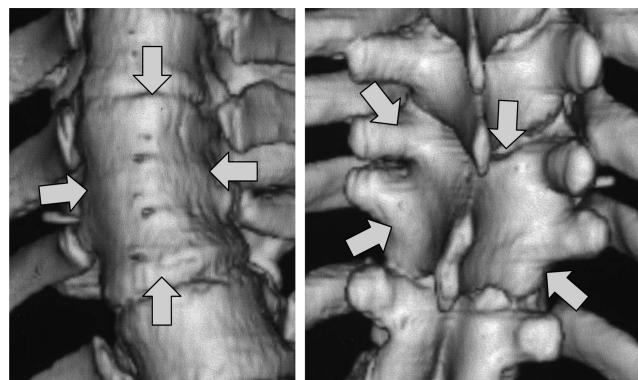


Figure 7. Block vertebra. A 12-year-old girl existing 62° of scoliosis. Front and back images of 3D CT demonstrate no segmentation between T10 and T11 (block vertebra, arrows).

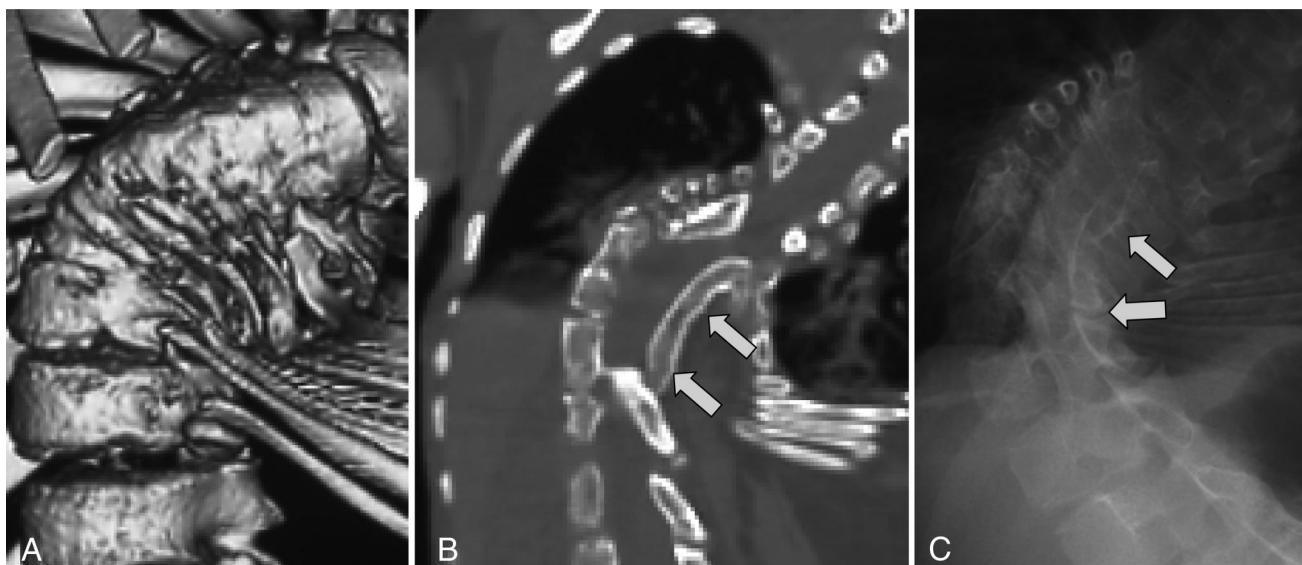


Figure 8. Unilateral unsegmented bar. A 7-year-old girl existing 128° of scoliosis. Three-dimensional CT image of thoracic spine (A) shows a severe scoliotic curvature with severely lateral wedged vertebrae. The long bar can be easily recognized on the coronal reconstructed image (B) and plain radiograph image (C) (arrows).

cranial adjacent vertebra. This mixture of semisegmentation will make it more difficult to make a correct diagnosis.

Segmentation Failure

The circumferentially unsegmented vertebrae (block vertebrae Figure 7) and the unilateral unsegmented bar (Figure 8) typifies segmentation failure in congenital spinal deformity. This latter is considered as one of progressive congenital vertebral abnormalities. According to the location of segmentation, segmentation failure has been reported to be subclassified into anterior, anterolateral, posterolateral, posterior, and circumferential.¹

Although 3D analysis can be used to demonstrate the presence or absence of segmentation in detail, it is very hard to differentiate segmentation from no segmentation

in some patients. In particular, the upper and midthoracic spine is the location where it is almost impossible to judge about the presence or absence of segmentation because of the coverage of the disc space and pedicles with rib heads. Disc space may be only the clue to differentiate segmentation from no segmentation. However, there is also difficulty in evaluation of segmentation for the anomalous vertebrae with disc space narrowing. This may be incomplete segmentation or secondary change influenced by posterior no segmentation observed in the mixed complex type.

Segmentation Abnormalities in Formation Failure

As reported by McMaster *et al*¹² the most progressive type is unilateral unsegmented bar with contralateral

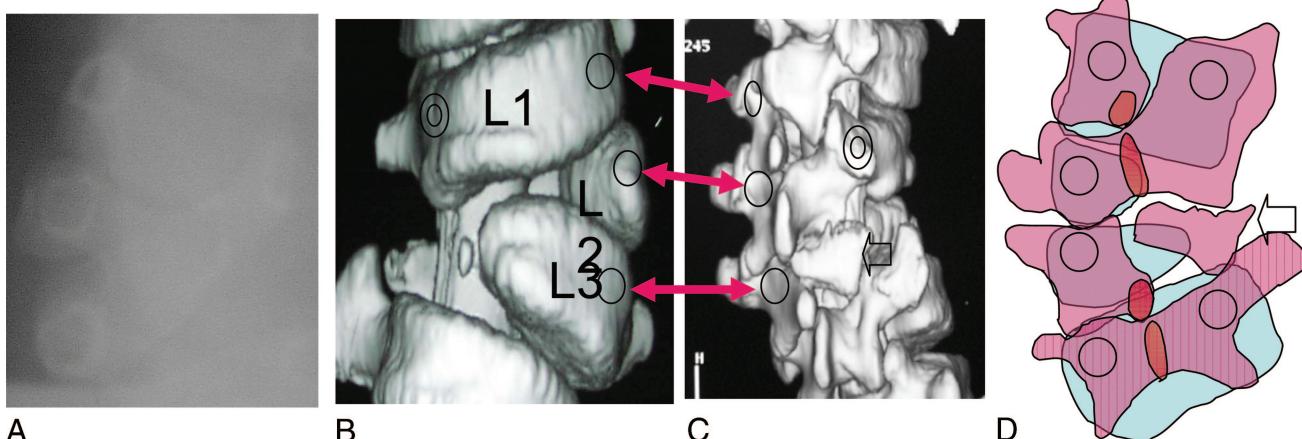
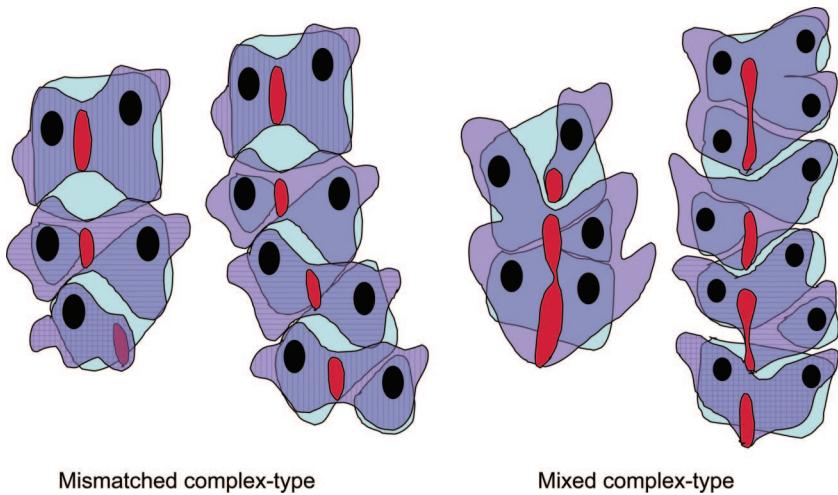


Figure 9. Discordant anomaly in formation failure. The patient was a 6-year-old boy with hemivertebrae at L2 and L3. The front image by 3D CT (B) coincided with the radiographic observation (A). The vertebral arch components connected to the hemivertebra united with the adjacent vertebral arch components of the vertebra in the direction of the head. The circles show the positions of the pedicles of the vertebral arch, and the arrows show the same pedicles of the vertebral arch imaged by 3D CT from the front and back (B, C, D). A vertebral arch was formed by pairing of the vertebral arch components connected to the right pedicle of the vertebral arch at L1 and the vertebral arch components at L2. The area shown by the thick arrow was surgically proved to be isolated hemilamina.

Figure 10. Drawings of the 2 types of complex type. Mismatched complex type: The vertebral arch components of the hemivertebra are united with the opposing vertebral arch components of the adjacent caudal or cranial vertebra. Mixed complex type: The anterior structure of malformed vertebral bodies belonging to formation failure and the posterior structure consists of complex fused vertebral arches (Drawn by A. Nakajima).



hemivertebrae. This type is associated with hemivertebrae on the opposite site, and should belong to the mixed-type. However, if it is hypothesized that it is a combination of some asymmetrical butterfly vertebrae and that 1 side of their smaller hemibodies become synostosized with cranial and caudal adjacent vertebrae, this unilateral unsegmented bar with contralateral hemivertebrae can be regarded as 1 type of segmentation failure in formation failure. In fact, there are so many hemivertebrae and butterfly vertebrae that have synostosis to the adjacent vertebrae. Considering this, it is confusing which type should be named as failure of formation or failure of segmentation.

Three-dimensional CT analysis can demonstrate many different types of failure of segmentation. We need to clarify the location of segmentation failure and formation failure. Imagama *et al*⁶ analyzed the location of synostosis in 40 patients with formation failure in congenital spinal deformities and reported the existence of difference of synostosis of the anterior and posterior structure even in the same type of abnormal vertebrae. We should differentiate these complicated vertebral anomalies from a pure segmentation failure that exhibit no formation failure.

Relationship of the Anterior Structure With the Posterior Structure

Both anterior and posterior structures of anomalous vertebrae correspond with each other in some patients (unison type). The unison type can be also called a simple type from the viewpoint of complexity of the vertebral anomalies. It is not difficult to evaluate these anomalies even using plain x-ray image because malformed vertebrae in the simple type could be explained by formation failure of certain vertebral components one by one. However, other patients exhibit differences between the anterior and posterior structures (discordant type, Figure 9).⁷ Congenital vertebral malformation that shows discordant relationship between the anterior and posterior structures can be called a complex type. It also demonstrates abnormal continuity between the posterior components of the mal-

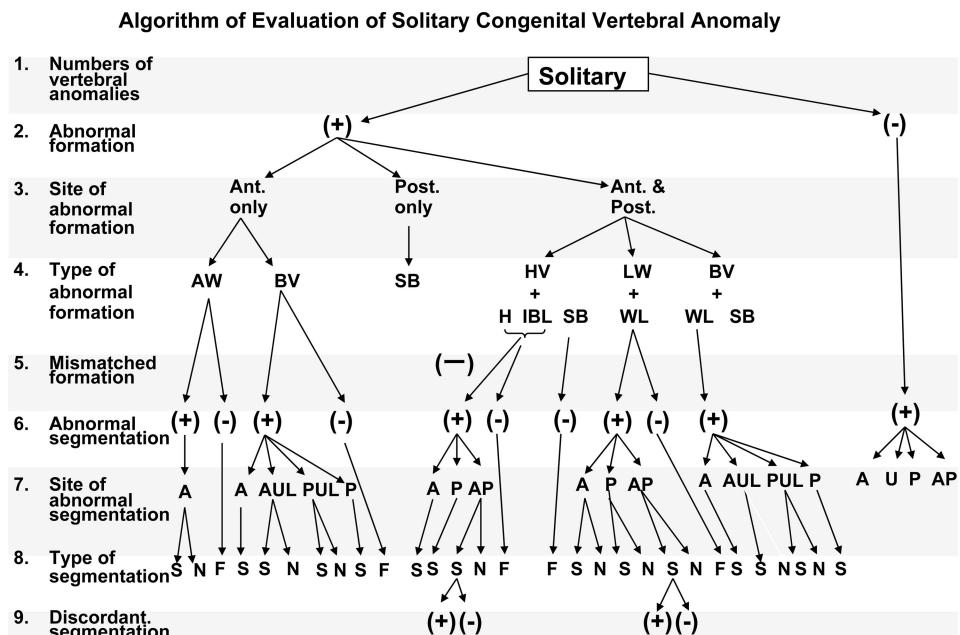
formed vertebrae and those of the neighboring vertebrae. It is very difficult to correlate the anterior structure with the posterior structure of each segment of anomalous vertebrae. Furthermore, it is virtually impossible to judge which vertebrae are normal or abnormal. These characteristics makes independent evaluation of the malformation in each vertebra as one segment impossible, and evaluation of complex malformations, involving multiple vertebrae is necessary. The discordant anomaly can be divided into 2 types, the mismatched complex type and mixed complex type, according to the mode of false fusion between each structure of the abnormal vertebrae (Figure 10). The former is one in which either side of the primordia of vertebral body may couple with an adjacent opposite side of the posterior component. Usually several vertebral segments are involved in this mismatching

Table 1. Algorithm of Evaluating Congenital Spinal Deformities

Step 1	Count the number of vertebral anomalies	Solitary or multiple
Step 2	Detect the abnormal formation	(+), (-), or (\pm)
Step 3	Determine the site of abnormal formation	Anterior, posterior, or both
Step 4	Find out the type of abnormal formation	Anterior: hemivertebra, anterior wedge, lateral wedge, butterfly Posterior: bilamina (wedged), incomplete bi-lamina, hemilamina Spina bifida
Step 5	Detect the mismatched formation	(+) or (-)
Step 6	Detect the abnormal segmentation	(+), (-), or (\pm)
Step 7	Find out the type of segmentation	Anterior: Full, semi, non Posterior: Full, semi, non
Step 8	Determine the site of abnormal segmentation	Anterior, unilateral, posterior, or all
Step 9	Detect the discordant segmentation	(+) or (-)

Full indicates fully segmented; Semi, semi-segmented; Non, nonsegmented.

Figure 11. Algorithm for the solitary type. If only 1 congenitally abnormal vertebra exists in whole spinal column, we should follow the algorithm for the solitary type. Referring the Table 1 the abnormal vertebra should be evaluated step by step. This algorithm helps to clarify the characteristics of abnormal vertebra. A indicates anterior; P, posterior; BV, butterfly vertebra; W, wedged vertebra; HV, hemivertebra; SB, spina bifida; LW, lateral wedged vertebra; H, hemilamina; IBL, incomplete bilamina; AUL, anterior unilateral; PUL, posterior unilateral; S, semisegmented; N, nonsegmented; F, fully segmented; U, unilateral; AP, anteroposterior.



phenomena and we need to evaluate this type of anomaly as an abnormal vertebral complex despite the inclusion of some normal-shaped vertebrae in this complex. The mixed complex type is a group of vertebral anomalies that exhibit differences of the failure type between the anterior and posterior structure.

■ An Approach to a New 3D Classification of Congenital Spinal Deformity

For the last 20 to 30 years, the classification of congenital scoliosis and kyphosis on which we developed

an operative strategy for the treatment of the deformity remains the same. Many spine surgeons have encountered vertebral anomalies whose operative findings did not fit with plain x-ray findings. Once 3D analysis of the deformed vertebrae becomes possible, referring to the findings of their 3D morphology becomes inevitable for the correction of congenital spinal deformity with osteotomy. However, because of the ambiguous definition of synostosis observed in many malformed vertebrae and the detection of discordant anomalies, we need to revise the classification

Algorithm of Evaluation of Multiple Congenital Vertebral Anomalies

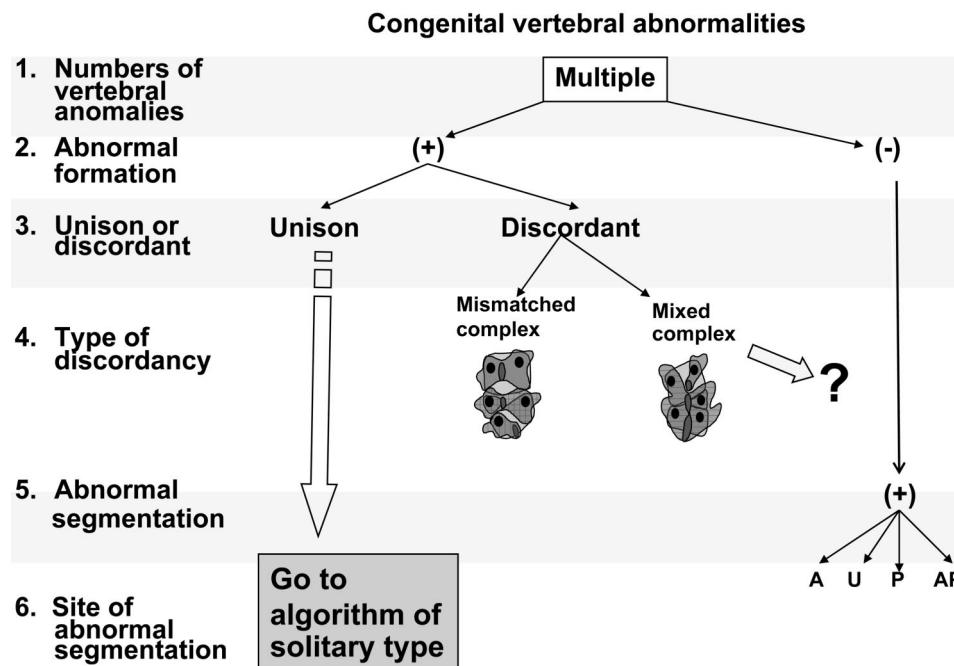


Figure 12. Algorithm for the multiple type. If more than 2 congenitally abnormal vertebrae exist in whole spinal column, we should follow the algorithm for the multiple type. Referring the Table 2 the abnormal vertebrae should be evaluated step by step. The abnormal vertebrae that belong to unison type should be evaluated separately using the algorithm for the solitary type. A indicates anterior; P, posterior; U, unilateral; AP, anteroposterior (circumferential). Unison type of multiple malformed vertebrae should be evaluated using the algorithm for the solitary type.

Table 2. Classification of Congenital Vertebral Abnormalities (Based on Presence or Absence of Abnormal Formation)

Type 1	Solitary simple (unison) type Hemivertebra Wedge vertebra Butterfly vertebra Defect Others
Type 2	Multiple simple (unison) type Combination of hemivertebra, wedge vertebra, or butterfly vertebra Discrete, adjacent, or others
Type 3	Complex (discordant) type Mismatched complex type Mixed complex type
Type 4	No abnormal formation type Pure segmentation failure

of congenital spinal deformity by including new information obtained by 3D analysis.

To make the story much simpler, analysis of congenital spinal deformity should be determined by orders of evaluation. The first step is counting the number of congenitally deformed vertebrae and dividing them into solitary and multiple. The second step is the analysis of formation failure of deformed vertebrae, such as the type of anterior and posterior formation failure, the site, the laterality, and the existence of discordance. The next step is the analysis of segmentation failure of abnormal vertebrae that have been already evaluated, according to the type of formation

failure (Table 1). If deformed vertebrae do not exhibit any signs of formation failure, the type of congenital vertebral anomaly surely belongs to the pure segmentation failure. The fourth step is the analysis of segmentation failure according the site, the type of synostosis, and the existence of discordance. The simple type and the multiple types should be evaluated in a different pathway (Figures 11, 12).

By developing this algorithm for the evaluation of congenital scoliosis in terms of numbers of abnormal vertebrae, type of formation failure, and type of segmentation failure in separate steps, congenital spinal deformity could be mainly classified into 4 types: Type 1, solitary simple; Type 2, multiple simple; Type 3, complex; Type 4, segmentation failure (Table 2). These 4 types of malformed vertebrae could be further modified by several morphologic factors, such as location, laterality, associated rib abnormalities, intracanal abnormalities, etc.

This classification has just been built up and we still have many things to do for getting acceptance in clinical usage. First, the complex type still remains uncertain and should be determined more clearly in this classification. Second, this classification should be analyzed by intra- and interobserver reliability. The aims of building up this 3D classification of congenital scoliosis and kyphosis have been to make the natural history of each congenital vertebral malformation much clearer, to identify which type of vertebral anomaly

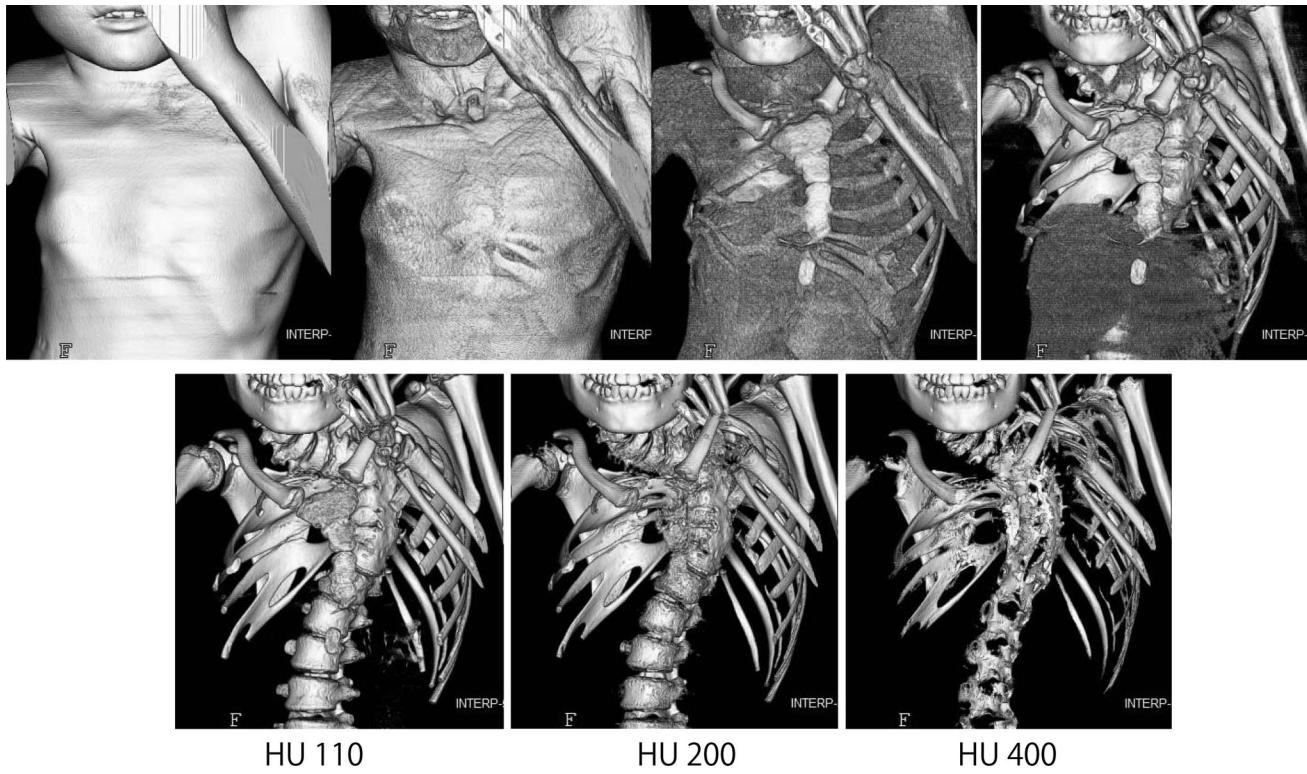


Figure 13. The process of reconstruction of 3D CT images. During processing a reconstruction image of the bony skeleton, not bony structure are erased gradually following masking of the soft tissue. An image demonstrating the vertebrae and ribs without erasing the bony structure can be obtained at step of Hounsfield unit 110.

make the deformity progress and should be operated, and to assist making a strategy of surgical planning. These targets have not been investigated yet. We furthermore have to continue to advance these projects to investigate how it works for the treatment of congenital scoliosis and kyphosis, and how it is superior to the prior classifications in the future.

■ Problems of 3D Analysis

There are many problems related to establishing a new classification of congenital spinal deformity by using 3D CT reconstruction images. First of all, high-resolution CT is absolutely necessary to obtain high quality images. A slice thickness of at least 2 mm is also inevitable although we are always aware of the amount of irradiation patients are exposed. To decrease the irradiation exposure, obtaining 3D CT images of whole spinal column seems to be not always necessary. However, some congenital vertebral malformation such as mismatched complex type may exist several normal-shaped vertebrae between 2 abnormal vertebrae. Scanning on the small area may misunderstand the type of vertebral anomalies. Secondary, the radiologic technicians should be well trained and know the volume-rendering technique because a false image may be constructed during image processing (Figure 13). Third, it is necessary for the spine surgeons evaluating 3D CT reconstructed images to become accustomed with this procedure, because it is still sometimes very difficult to analyze even high resolution images.

We should also understand the limitations of 3D CT reconstruction imaging. First, as mentioned earlier, the disc space and pedicle cannot be imaged because of the existence of rib heads in the thoracic spine. In this case, we need to add an evaluation of multiplanar reconstruction images (coronal and sagittal reconstruction images) besides the 3D CT images. Second, it is very difficult to determine whether the narrowed disc space derives from congenital or secondary means, or whether or not growth plate facing to the narrowed disc still has the potential to grow. Finally, even high-resolution image of 3D CT has limitations in demonstrating the demarcation of 2 structures, which are in contact without any space.

■ Conclusion

The 2D classification of congenital spinal deformity clearly has limitations in clinical usage at present when 3D CT images can be obtained. The information of 3D CT images has contributed greatly not only to the devel-

opment of a strategy for the surgical treatment but also to the proposal of a new classification.

■ Key Points

- Although 3D CT images of congenitally-deformed vertebrae still have many problems, the volume of information that can be obtained by evaluating them has contributed greatly to the development of a strategy for the surgical treatment.
- We concluded that we should revise the 2D classification of congenital spinal deformity through detection of discordant anomalies with 3D imaging.
- By analyzing the 3D images of congenital spinal deformity, they could be mainly classified into 4 types: Type 1, solitary simple; Type 2, multiple simple; Type 3, complex; Type 4, segmentation failure.
- Although this 3D classification of congenital scoliosis may still require revision, we need to develop a new classification of congenital scoliosis, based on the perspective of 3D imaging to understand the etiology and embryology, as well as to determine an operative strategy.

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References

1. Winter RB. *Congenital Deformities of the Spine*. New York, NY: Thieme-Stratton; 1983.
2. Winter RB, Moe JH, Wang JF. Congenital Kyphosis—Its natural history and treatment as observed in a study of one hundred thirty patients. *J Bone Joint Surg Am* 1973;55:223–56.
3. McMaster MJ, Ohtsuka K. The Natural history of congenital scoliosis: a study of two hundred and fifty-one patients. *J Bone Joint Surg Am* 1982;64:1128–47.
4. McMaster MJ, Singh H. The Natural history of congenital kyphosis and kyphoscoliosis. *J Bone Joint Surg Am* 1999;81:1367–83.
5. Imagama S, Kawakami, Matsubara Y, et al. The three-dimensional analysis of congenital scoliosis—segmentation failure. *J Jpn Scoliosis Soc* 2004;19:33–7.
6. Imagama S, Kawakami, Goto M, et al. Spacial relationships between a deformed vertebra and an adjacent vertebra in congenital scoliosis—failure of formation. *J Jpn Scoliosis Soc* 2005;20:20–5.
7. Nakajima A, Kawakami A, Imagama S, et al. Three-Dimensional analysis of formation failure in congenital scoliosis. *Spine* 2007;32:562–7.
8. Nasca RJ, Stelling FH, Steel HH. Progression of congenital scoliosis due to hemivertebrae and hemivertebrae with bars. *J Bone Joint Surg Am* 1975;57:456–66.
9. Newton PO, Hahn GW, Ficka KB, et al. Utility of three dimensional and multiplanar reformatted computed tomography for evaluation of pediatric congenital spine abnormalities. *Spine* 2002;27:844–50.
10. Bush CH, Kalen V. Three dimensional computed tomography in the assessment of congenital scoliosis. *Skeletal Radiol* 1999;28:632–7.
11. Hedequist DJ, Emans JB. The Correlation of preoperative three-dimensional computed tomography reconstructions with operative findings in congenital scoliosis. *Spine* 2003;28:2531–4.
12. McMaster MJ. Congenital scoliosis caused by a unilateral failure of vertebral segmentation with contralateral hemivertebrae. *Spine* 1998;23:998–1005.