



Novel Classification in Congenital Spinal Deformity

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Introduction

Congenital spinal deformity (CSD) including scoliosis and kyphosis is defined as spinal deformity caused by solitary or multiple congenital vertebral anomalies. We should NOT include any spinal deformity that exist at birth without any congenital vertebral anomalies.

At present, mainly two classifications of CSD exist, one is based on the plain X-ray findings, which has been established by Winter^{1,2}), McMaster³) and another that is based on 3DCT findings reported by Kawakami, et al⁵).

The former classified CSD into three categories: Formation failure (FF), Segmentation failure (SF), and Mixed type (MT). While this classification has been widely used for analysis of congenital scoliosis and kyphosis, there are some known cases of CSDs with atypical vertebral anomalies, which do not belong to any of the aforementioned three categories. This is because it is based on radiographic findings recognized on plain X-ray images.

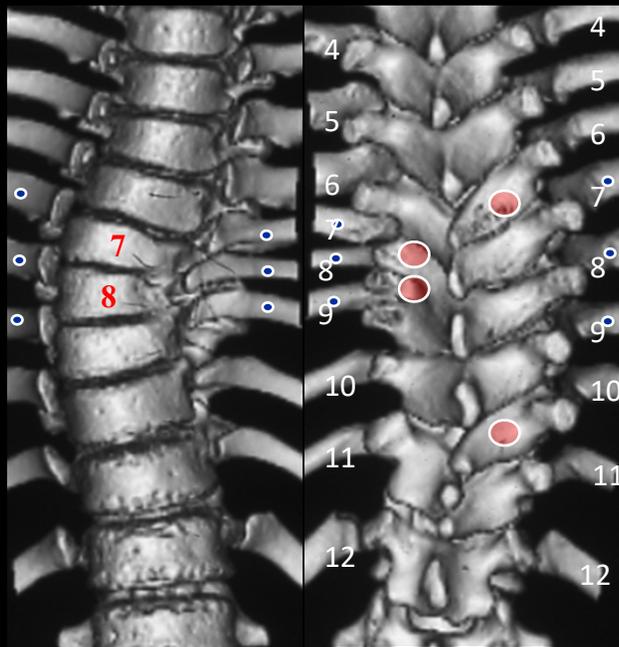


Confusion and Issues in Winter's Classification (1)

1) How should you classify these two CSD?



This first case may be diagnosed as Contralateral Hemivertebrae (Hemimetamelic shift) and classified into formation failure. Those are a fully segmented hemivertebra on the L2 left side and a semisegmented hemivertebra on the L4 right side. However, there are almost normal lamina from T12-L5. Can this pair of hemivertebrae be classified into formation failure?



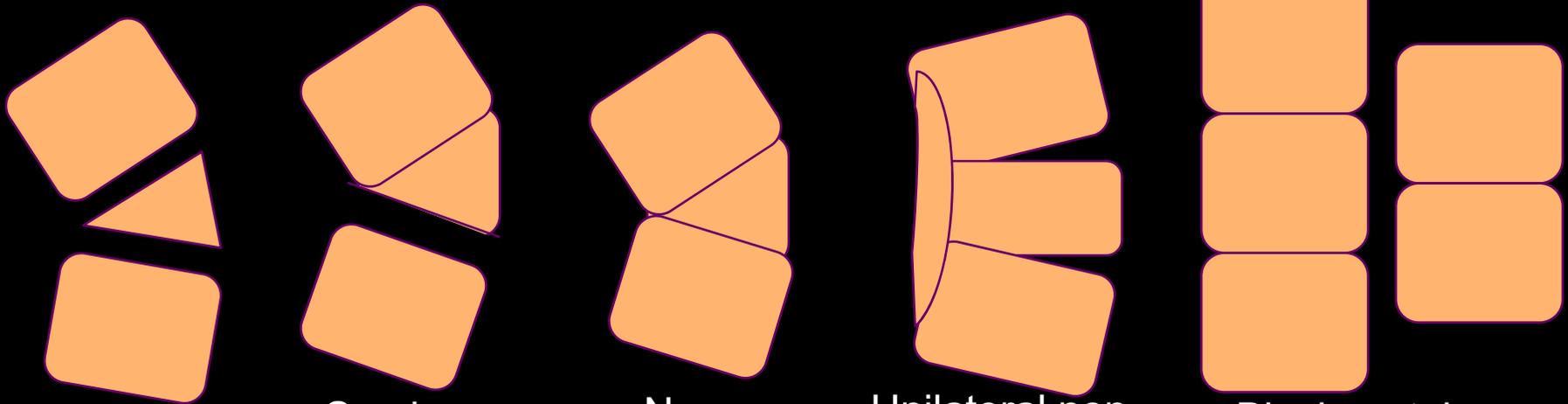
This second case is very difficult to classify using Winter's classification. There were no typical vertebral anomalies on the anterior structure except slight wedging on the T8 vertebral body, however, with multiple hemilaminae and one of them being semisegmented in the posterior structure. Winter's classification does not consider posterior structure anomalies.

Confusion and Issues in Winter's Classification (2)

2) Which vertebra is "congenitally abnormal"?

Formation failure

Segmentation failure



Fully- segmented

Semi-
segmented

Non-
segmented

Unilateral non-
segmented bar

Block vertebrae

It is easy to identify an abnormal vertebra if a fully-segmented vertebral anomaly exists. However, if a vertebral anomaly is combined with segmentation anomaly, it is difficult to identify whether it is normal or abnormal.

Outstanding issues of Winter's classification are

- 1) Evaluation of only vertebral body using plain X-ray images.
- 2) All formation failures as well as those that occur in conjunction with segmentation failure are included in formation failure.
- 3) The mixed type category includes almost all unclassified vertebral anomalies and in its current form, is a "waste bin."

Kawakami's Classification⁵⁾

On the other hand, the latter introduces discordancy, which may be a clue to solve the puzzle of CSD. One of problems in this 3D classification is its complexity and its inherent difficulty to understand.

◆ Type 1 Solitary Simple (Unison)

Hemivertebra
Wedge vertebra
Butterfly vertebra
Defect
Others

◆ Type 2 Multiple Simple (Unison)

Combination of Hemiv.
Wedge v.
Butterfly v.

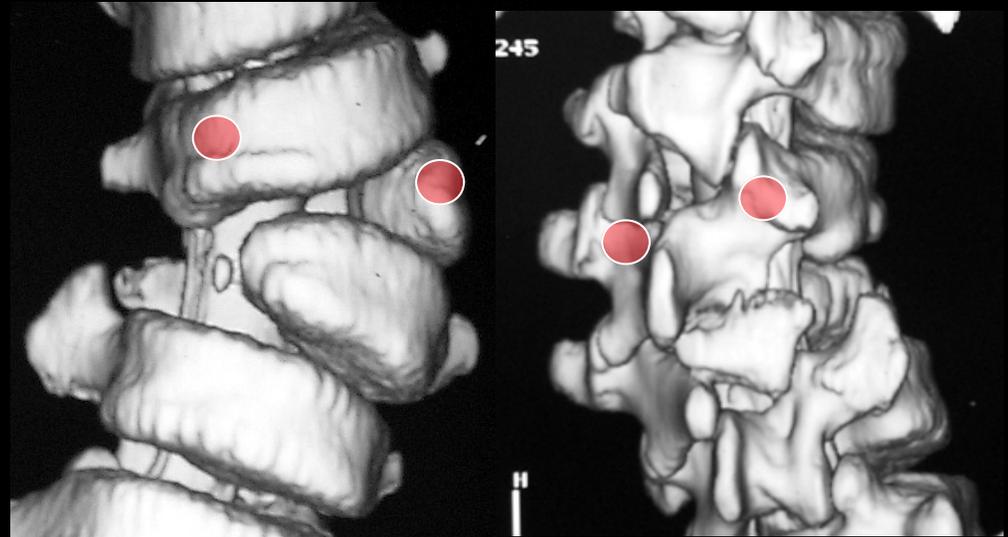
Discrete, adjacent, or others

◆ Type 3 Complex (Discordant)

Mismatched complex type
Mixed complex type

◆ Type 4 No abnormal formation pure segmentation failure

Discordancy in CSD



Nakajima, Kawakami, et al. 2007⁴⁾
Kawakami, et al. 2009⁵⁾

Purpose of This Study

The purpose of this study was to analyze 3D anatomical relationship of CSD in terms of discordancy and to introduce a novel classification of CSD by dividing them into four categories.

Materials

From 2001 to 2013, 566 patients with CVA visited Meijo Hospital complaining back deformity. Spinal deformity in 566 patients varied from none or slight to very severe. Of them, 332 patients with CSDs were evaluated using 3D-CT images to investigate whether a) they should be surgically treated or b) to determine surgical strategy for vertebrectomy. These 332 patients were materials of this study. Open spine lesions including spina bifida except those existed in only the sacrum were excluded in this study.

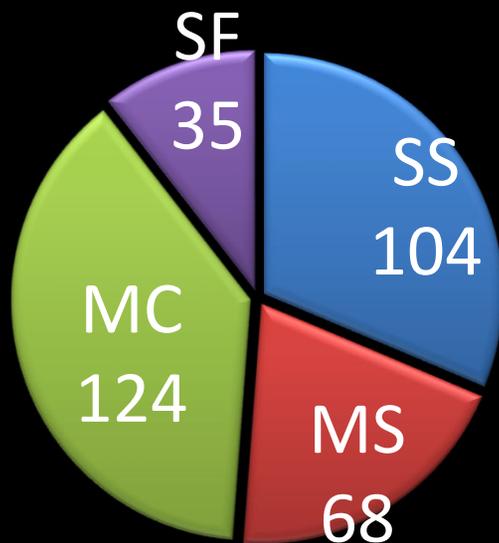
Methods

According to 3D-Classification presented by Kawakami, et al, 331 were classified into four types such as SS, MS, MC, and SF. Anterior, posterior structure, and the relationship between them in each patient were evaluated according to the algorithm of evaluating CSD by two well-experienced spine surgeons (NK, TS) who were familiar with 3D-CT images of CSD. 3D-CT images were mainly taken using the TOSHIBA Aquilion 64, and the slice thickness was 2 mm. Three-dimensional images were expressed by volume rendering.



Types of Vertebral Anomalies in 332 patients.

Mean age of 332 patients at the time of CT: 8.9 years (2-50)
Sex of the patients: male in 144, female in 187.



Discordant Anomaly

	All patients	Patients with MC	Patients with mismatch (+) in MC
Number of patients	332	124	38

Evaluation of discordancy in 38 patients with the MMC type clarified that 12 of 38 patients exhibited very unusual vertebral anomalies in addition to mismatch phenomena. They could be subclassified into three types, anterior, posterior, and anteroposterior.

Twelve Patients with Vertebral Anomalies with Sole Mismatch Phenomena

Case	Sex	Type	Vertebral anomalies	scoliosis
1	M	Anteroposterior	Rt.T3 (FSHV+discordant SSL), Rt.T7 & Rt.T11(SSHV+SSL), Lt. L5(FSHV+SSL)	60
2	F	Anteroposterior	Lt.T8, Rt.L1 (FSHV+discordant FSHL)	55
3	M	Anteroposterior	Lt. T10 (FSHV+FSHL) 、 Rt.T12(SSHV、 discordant FSHL)	19
4	F	Anteroposterior	Rt.T3 (FSHV+discordant FSHL), Lt.T6(FSHV+SSHV), Rt.T8(FSHV+SSL),Lt.T10(SSHV+discordant SSL)	32
5	F	Anteroposterior	Lt.T7 (FSHV+discordant FSHL), Rt.T10 (SSHV+discordant SSL), Lt.L1 (SSHV+discordant SSL)	35
6	M	Anteroposterior	Lt C7 (SSHV+NL) 、 Rt T7(SSHV+NL) , T8-T10 Hemilamina	51
7	M	Anteroposterior	Lt.T10 (FSHV+discordant FSHL), T12(BV), Rt. L2(FSHV+SSL), Lt.L6 (pedicle defect)	45
8	F	Anteroposterior	Rt T1(SSHV+FSHL), Lt.T8 (FSHV+NL) , Rt.T10 (SSHV+NL) 、 Rt.T13 (SSHV+NL)	32
9	F	Anterior	Lt. T1 (FSHV+NL) 、 Rt. T8 (FSHV+NL) 、 Lt. T13 (FSHV+NL)	65
10	F	Anterior	Lt. L2 (FSHV+NL), Rt. L4 (SSHV+NL)	36
11	F	Posterior	Lt. T5 (NVB+FSHL), Rt.T6 (NVB+FSHL)	33
12	M	Posterior	Rt.T6(NVB+FSHL), Lt.T8 (NVB+SSL), Lt.T9 (NVB+SSL), Rt. T10 (NVB+FSHL)	40

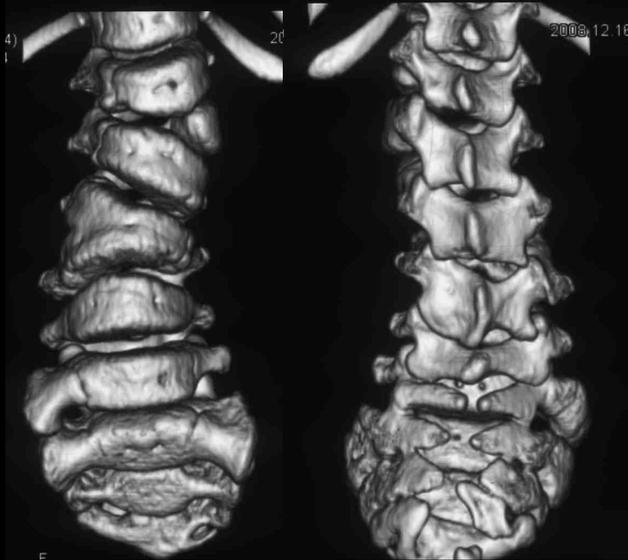
FSHV: fully-segmented hemivertebral body, SSHV: semisegmented hemivertebral body, BV: butterfly vertebra

FSHL: fully-segmented hemilamina, SSL: semisegmented hemilamina, NVB: normal vertebral body, NL: normal lamina

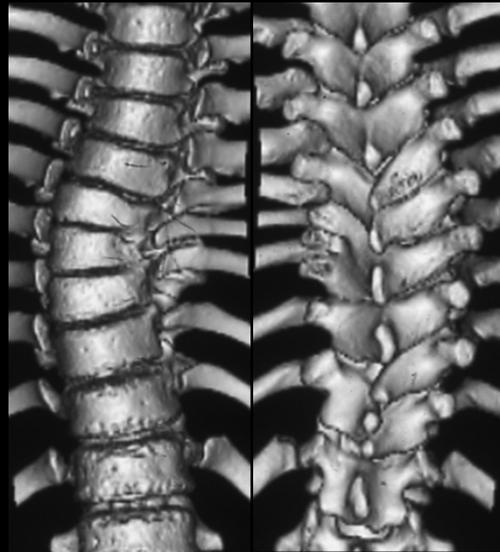


Three Types of Unusual Vertebral Anomalies with Mismatch Phenomena

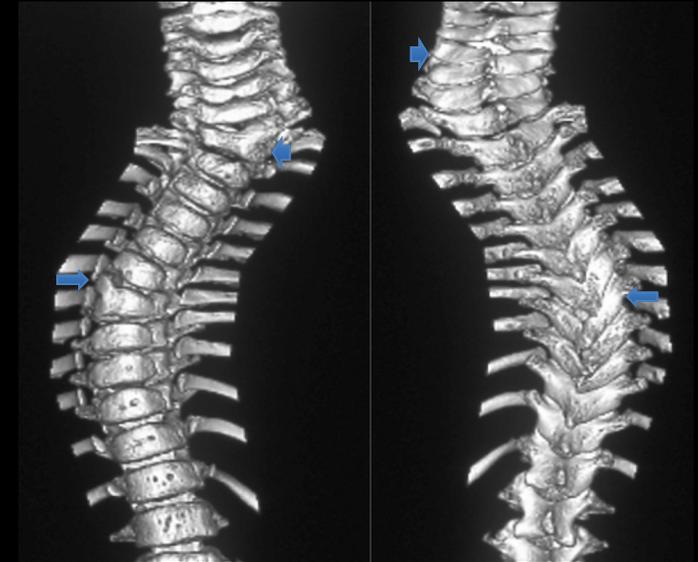
The Anterior type



The Posterior type



The Anteroposterior type



Left: almost normal laminae (L1-L5) with contralateral hemilvertebrae.

Middle: multiple contralateral hemilaminae with almost normal vertebral bodies

Right: discordant combination with multiple contralateral hemivertebral bodies and multiple contralateral hemilaminae.

Common features of these 3 types:

- 1) Discordant anomaly with a discordant combination of normal vertebral bodies and/or laminae
- 2) It was hard to identify the normal vertebrae from the abnormal.

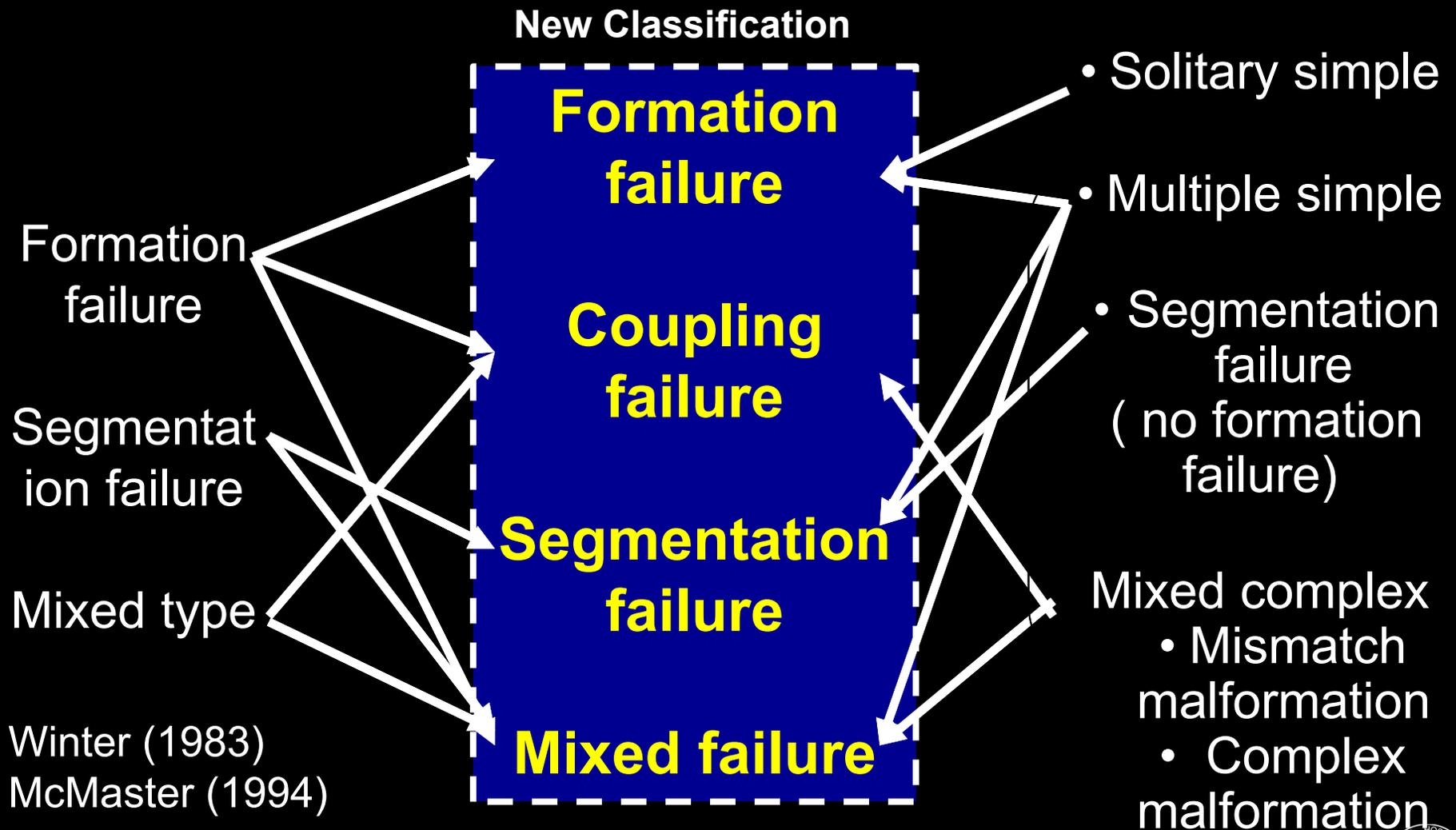
Discussion

- 1) While these three types may look like some other type of formation failure due to the existence of discordancy, they should not be grouped in neither formation failure or segmentation failure.
- 2) Tsou⁶⁾ reported the development mechanism of hemivertebrae; even a single hemivertebra is formed by hemimetameric asynchronous development, which is thought as false fusion of primordia during this period.
- 3) Lehman reported that contralateral hemivertebrae were formed by false coupling of somites⁷⁾. Based on these developmental considerations of the mismatch phenomena; it should be separated from formation failure, which can be explained by the concept of partial formation failure of the vertebrae. We have grouped these three types that are caused by the mismatch phenomena together and named it as “**Coupling failure**”, the new fourth category of CSD.
- 4) The Mixed type in classification presented by Winter and McMaster and the Mixed complex type in classification by Kawakami, et al. could be regarded as an assembly of multiple vertebral anomalies due to combination of formation, segmentation, and/or coupling failure. In other words, this type can be expressed as a “**Mixed failure.**”



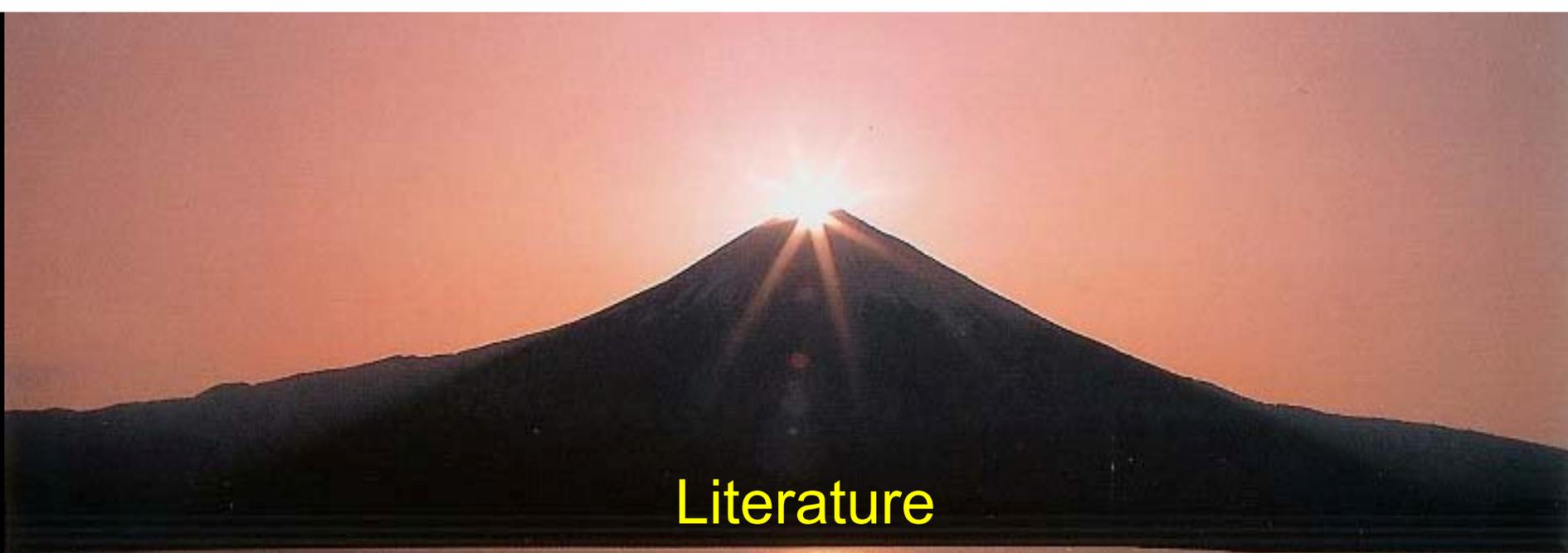
A Proposal of a New Classification of Congenital Spinal Deformity

- We propose a new classification of congenital spinal deformity adding the fourth category “coupling failure”



Winter (1983)
McMaster (1994)





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