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CHINESE SECTION

# A retrospective study of congenital scoliosis and associated cardiac and intraspinal abnormities in a Chinese population

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**Abstract** Our objective is to assess the incidence of cardiac and intraspinal abnormities in Chinese congenital scoliosis (CS) patients and to study the relationship between the associated abnormities and the different CS types. Five-hundred and thirty-nine consecutive Chinese patients with CS were retrospectively studied, and the records of echocardiography, plain radiograph of the entire spine, magnetic resonance imaging of the entire spine and/or myelogram were reviewed. The results indicated that the incidence of cardiac and intraspinal abnormities in CS patients was 14.1 and 24.5%, respectively. There was no difference in the incidence of associated cardiac and intraspinal abnormities in different CS types ($P > 0.05$). The most common cardiac abnormities in CS patients was mitral valve prolapse, which was followed by congenital heart diseases, including atrial septal defect, ventricular septal defect, bicuspid aortic valve and patent ductus ateriosus. The cardiac abnormities were not likely to be concurrent with intraspinal abnormities in CS patients ($P = 0.04$). The intraspinal abnormities were more common in female and older patients (all $P < 0.05$). One or more abnormities mentioned above could be found in 36.8% CS patients and were more likely to be found in female patients ($P < 0.01$). We concluded that CS is not a simple abnormity, due to the high incidence of associated deformities of other organs, comprehensive assessment was strongly recommended before the surgical correction for CS patients.

**Keywords** Congenital scoliosis · Associated abnormities · Chinese population

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## Introduction

Congenital scoliosis (CS) is a curvature of the spine that results from anomalies or abnormally developed vertebrae, the building blocks of the spinal column. These anomalies occur in utero at 4–6 weeks of gestation. Specific abnormalities include hemivertebra, which is a wedge-shaped or half vertebra (failure of formation), unsegmented bar, which is a failure of the normal separation of the individual building blocks of the spine (failure of segmentation), and mixed abnormalities.

CS is often associated with the defects of other organs, cardiac and intraspinal abnormities have been shown to be commonly associated with CS, which maybe the result of the closely related embryonic development of these organs from the mesoderm [1–5]. The concurrent abnormities may increase the risk of surgical correction in the CS patients.

Reckles et al. [6] found congenital heart disease was present in 10% CS patients, echocardiography was suggested in patients who were about to undergo CS surgery. Bradford et al. [1] evaluated 42 congenital scoliosis patients with magnetic resonance imaging(MRI) and found intraspinal abnormalities in 38% patients, and due to such a high incidence of intraspinal abnormity, routine MRI screen in CS patients was suggested. Rajasekaran et al. [7] found the incidence of intraspinal abnormities was 35% in 60 Indian CS patients.

In this retrospective study, 539 consecutive Chinese patients with congenital scoliosis were reviewed. The aims of the study were to assess the overall incidence of intraspinal and cardiac abnormities and to study the relationship between the associated abnormities and the different CS types in Chinese CS patients.

## Materials and methods

From January 2004 to October 2009, 539 CS patients (214 male and 325 female patients) were admitted to Peking Union Hospital for surgical correction of the CS. All patients underwent a full clinical examination, plain radiograph of the entire spine, neural axis MRI from brain stem to sacrum and/or mylogram to screen the intraspinal abnormities. All the records of MRI and/or mylogram were available. Part of these CS patients underwent echocardiography because of the cardiac murmur or just for routine screen of cardiac abnormities, and echocardiographic records of 475 patients were available. All these clinical data and imaging records were reviewed.

The congenital scoliosis was classified as a resulting from the failure of formation, failure of segmentation, and mixed abnormalities (mixed and complex lesions) [8]. The $t$ test and the $\chi^2$ test (as appropriate for the data type) were used for the statistical analysis. A $P$ value $<0.05$ was considered statistically significant.

## Results

A total of 539 patients were involved in the study, the mean age was 12.8 ± 5.8 years and 39.7% (214/539) was male. Among them, the failures of formation were noticed in 361 (70.0%) patients, failure of segmentation in 168 (31.2%) patients and the mixed abnormalities in 70 (13.0%) patients.

Surgical correction of the spinal abnormities was performed in 493 CS patients, 46 CS patients with the spinal abnormities did not receive the correction surgery because of very mild abnormities or high operation risk.

Of 475 patients whose echocardiographic records were available, cardiac abnormities were found in 67 (14.1%) patients and the specific types of cardiac abnormities are shown in Table 1. The most common cardiac abnormity was mitral valve prolapsed (MVP), which was followed by congenital heart diseases, including atrial septal defect (ASD), ventricular septal defect (VSD), bicuspid aortic valve and patent ductus ateriosus (PDA). The diagnosis of associated heart diseases had been made in nine patients before their referral to our hospital. There were no differences in the age, gender, body height and CS types between CS patients with or without cardiac abnormities (all $P > 0.05$) (Table 2), but the incidence of intraspinal abnormities and body weight was lower in the CS patients with cardiac abnormities ($P = 0.04$ and $P = 0.02$, respectively) (Table 2).

**Table 1** Associated Cardiac abnormities in CS patients

| Cardiac abnormities | Number (%)[a] |
|---|---|
| Mitral valve prolapsed | 34 (7.2) |
| Atrial septal defects (ASD) | 12 (2.5) |
| Ventricular septal defects (VSD) | 8 (1.7) |
| Bicuspid aortic valve | 5 (1.1) |
| Patent ductus arteriosus (PDA) | 4 (0.8) |
| Tetralogy of fallot | 3 (0.6) |
| Dextrocardia | 1 (0.2) |
| Cor triatriatum | 1 (0.2) |
| Hypertrophic cardiomyopathy | 1 (0.2) |
| Aortic valve prolapsed | 1 (0.2) |

[a] Multiple cardiac abnormities were found in three patients: 1 with ASD and VSD, 1 with ASD and PDA, 1 with VSD and PDA

Myelogram and/or magnetic resonance imaging was performed in all 539 patients. The intraspinal abnormalities were detected in 24.5% (132/539) patients, the main manifestations were diastematomyelia (95/539, 17.6%), syringomyelia (27/539, 5.0%), tethered cord (22/539, 4.1%), intraspinal mass (8/539, 1.5%), spinal meningocele (5/539, 0.9%), and syringomyelus (4/539, 0.7%). Combination of two or more intraspinal abnormities existed in 27 (5.0%) patients. The intraspinal abnormities were more likely to be found in female or older CS patients (all $P < 0.05$) (Table 2). There were no differences in the body height, body weight and CS types between CS patients with or without the intraspinal abnormities (all $P > 0.05$) (Table 2).

In the 475 CS patients whose imaging records including plain radiograph of the entire spine, MRI of the entire spine and/or myelogram, and echocardiography were all available, 175 (36.8%) patients were found to have at least one abnormity (cardiac or intraspinal abnormities), and no differences were found in most clinical data between the patients with or without associated intraspinal and cardiac abnormities, except the associated abnormities were more likely to be found in female CS patients ($P < 0.01$) (Table 2).

## Discussion

To our knowledge, this is the largest study of CS and associated abnormities of other organs in Chinese population. The incidence of intraspinal, cardiac abnormities and

**Table 2** Clinical data of CS patients with or without cardiac or intraspinal abnormities

| | With cardiac abnormities ($n = 67$) | Without cardiac abnormities ($n = 408$) | $P$ value | With intraspinal abnormities ($n = 132$) | Without intraspinal abnormities ($n = 407$) | $P$ value | With intraspinal or cardiac abnormities ($n = 175$) | Without intraspinal or cardiac abnormities ($n = 300$) | $P$ value |
|---|---|---|---|---|---|---|---|---|---|
| Age (years) | 11.7 ± 4.4 | 12.9 ± 5.7 | 0.11 | 13.7 ± 5.1 | 12.5 ± 5.9 | 0.046 | 12.9 ± 4.8 | 12.6 ± 6.0 | 0.65 |
| Gender (male), *n* % | 23 (34.3%) | 166 (40.7%) | 0.32 | 37 (28.0%) | 177 (43.5%) | <0.01 | 53 (30.3%) | 136 (45.3%) | <0.01 |
| Body height when admitted (cm) | 136.1 ± 22.4 | 139.1 ± 22.0 | 0.33 | 140.0 ± 16.7 | 138.0 ± 23.3 | 0.22 | 138.9 ± 19.0 | 138.6 ± 23.6 | 0.89 |
| Body weight when admitted (kg) | 33.6 ± 14.4 | 38.0 ± 14.1 | 0.02 | 37.9 ± 12.0 | 37.4 ± 15.3 | 0.72 | 38.1 ± 14.9 | 36.1 ± 12.9 | 0.16 |
| CS types, *n* % | | | | | | | | | |
| Failure of formation | 40 (60.0%) | 227 (55.6%) | | 71 (53.8%) | 230 (56.5%) | | 96 (54.9%) | 171 (57.0%) | |
| Failure of segmentation | 22 (32.8%) | 125 (30.6%) | | 45 (34.1%) | 123 (30.2%) | | 62 (35.4%) | 85 (28.5%) | |
| Mixed defects | 5 (7.5%) | 56 (13.7%) | 0.37 | 16 (12.1%) | 54 (13.3%) | 0.70 | 17 (9.7%) | 44 (14.7%) | 0.14 |
| Intraspinal abnormities, *n* % | 10 (14.9%) | 108 (26.5%) | 0.04 | | | | | | |

the relationship between the different CS types and associated abnormities were retrospectively studied.

The incidence of congenital heart diseases was 22.7% in a series of 126 congenital spine abnormities which included 110 CS patients [4] and the authors did not mention the incidence of valvular abnormities such as valvular prolapse. This study showed that valvular disease, mainly MVP, was the most frequent abnormity in Chinese CS patients, which had not been described previously. Dhuper et al. [9] suggested that the thorax abnormity in idiopathic scoliosis patients may cause the change of the heart shape and subsequently may lead to the mitral valve prolapse. We supported this assumption in CS patients. It is important that echocardiography should be recommended in all CS patients because before admission to our hospital, the heart abnormities were found only in nine CS patients.

Basu et al. [4] found that the proportion of congenital heart diseases was higher in patients with CS types of mixed defects and the incidence of intraspinal anomalies was higher in CS type of segmentation and mixed defects. But this study did not draw the same conclusion. No difference was noticed in the incidence of cardiac and intraspinal abnormities among the different CS types. Interestingly, we found although both cardiac and intraspinal abnormities were not uncommon in CS patients, these two abnormities were seldom present in the same patient ($P = 0.04$), this was not described previously.

The high incidence of associate with cardiac or intraspinal abnormities in this study reminds us of the importance to assess CS patients carefully and comprehensively before the surgical correction. This was a retrospective study and not all patients underwent echocardiographic examination, so bias may happen in the statistical analysis.

## Conclusion

CS is not a simple abnormity and it is often associated with multiple abnormities of other organs. Due to the high incidence of associated abnormities, comprehensive assessment was strongly recommended before the surgical correction.

**Conflict of interest** None.

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