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SCOLIOSIS PREVALENCE IN CONGENITAL HEART DISEASE PATIENTS

KONJENİTAL KALP HASTALIĞI OLAN HASTALARDA SKOLYOZ PREVALANSI

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SUMMARY*:*

The association between scoliosis and congenital heart disease has been well known for many years. The incidence of scoliosis in patients with congenital heart disease has been reported as 4.2%. This rate is higher for patients who are indicated for surgical corrections. There are theories advocating that thoracotomy also leads to developmental scoliosis. In this study, in order to test the accuracy of these views for both surgical and non-surgical cases of patients with congenital heart disease (both from the ages of 0–10 and adult patients), we aimed to investigate the prevalence of scoliosis.

356 patients with a diagnosis of congenital heart disease were enrolled in this study. 226 of them were between the ages of 0 and 10, with a mean age of 3.6 ± 5.1 years and a male/female ratio of 111/115. The remaining 130 patients were 26 to 76 years of age, with a mean age of 48.9 ± 22.8 years and a female/male ratio of 49/81. The patients were retrospectively evaluated with chest X-rays, and the cervicothoracic and thoracic regions included in the X-rays were examined for spinal deformities. The prevalence of scoliosis in both groups was calculated separately.

In addition, the effect of receiving surgery or not on the prevalence was also studied.

In this study, the prevalence of scoliosis was found to be 7.30% when all patients were included. This rate was found to be 6.64% for children between 0–10 years of age, and 8.46% for adults.

This study shows that patients who received surgery for congenital heart disease in childhood had an increased prevalence of scoliosis as adults. This result suggests that thoracotomy can have an effect on the development of scoliosis, especially after the age of 10. In the light of these results, it is important to obtain scoliosis X-rays for patients who are diagnosed with any congenital heart disease, and to follow up these patients. Also, for patients with scoliosis, if there are any surgical indications, a detailed cardiac examination and all necessary investigations must be performed before surgery.

Key words: Congenital heart disease, scoliosis, prevalence

Level of evidence: Retrospective clinical study, Level III

ÖZET:

Skolyoz ile konjenital kalp hastalığı birlikteliği uzun yıllardır çok iyi bilinmektedir. Konjenital kalp hastalığı olanlarda skolyoz görülme insidansı % 4.2 olarak bildirilmiştir. Cerrahiye gidenlerde bu oran yükselmektedir ve torakotominin gelişimsel skolyoza yol açtığı konusunda görüşler mevcuttur. Bu çalışmada, bu görüşlerin doğruluğunu test etmek üzere cerrahiye gitmiş ve gitmemiş 0-10 yaş arası ve erişkin yaştaki konjenital kalp hastalığı olan hastalarda skolyoz prevalansı araştırılması amaçlanmıştır.

Konjenital kalp hastalığı tanısı almış 356 hasta bu çalışmaya dâhil edilmiştir. Bu hastalardan 226 adedi 0-10 yaş arasında olup ortalama yaşları 3.6 ± 5.1, kız/erkek oranı 115/111' dir. Geri kalan 130 hasta, 20 ile 76 yaş arasında olup yaş ortalaması *48.9 ± 22.8 ve kadın/erkek oranı 49/81' dır.*

Hastaların geriye dönük olarak, PA akciğer grafileri değerlendirilmiş, radyolojik alana giren servikotorasik, torasik bölgelerinde omurga deformitelerinin olup olmadığı incelenmiştir. Her iki grupta ayrı ayrı skolyoz prevalansı hesaplanmıştır. Ayrıca operasyon olup olmamasına göre prevalans farkı olup olmadığı araştırılmıştır.

Bu çalışmada konjenital kalp hastalığı tanısı alan 356 hastada, tüm hastalar dâhil edildiğinde skolyoz prevalansı % 7.30, 0-10 yaş arası çocuklarda % 6.64, erişkinlerde % 8.46 olarak bulunmuştur.

Çocukluklarında konjenital kalp hastalığı nedeniyle opere edilen hastalarda prevalansın sadece erişkinlerde skolyoz prevalansının artışına sebep olmaktadır. Bu durum torakotominin özellikle 10 yaş sonrası skolyoz gelişimine *etkide bulunduğunu* düşündürmektedir. Bu çalışmanın verileri ışığı altında, konjenital kalp hastalığı olan hastaların skolyoz grafilerini çekilmesi ve bu yönde takip edilmelerinin, skolyozu olan hastaların ise özellikle opere edileceklerse ayrıntılı kardiyak muayene ve incelemelerden geçirilmelerinin çok önemli olduğu fikri elde edilmiştir.

Anahtar Kelimeler: Konjenital kalp hastalığı, skolyoz, prevalans

Kanıt Düzeyi: Retrospektif klinik çalışma, Düzey III

INTRODUCTION:

The coexistence of congenital heart disease and scoliosis has been known for a long time[6,9,14,17,22,25]. The incidence of congenital heart disease in cases of idiopathic scoliosis has been reported to be 12%[24,29]. In recent decades, due to advances in medical and surgical treatments for congenital heart disease, patients' life expectancy has increased, and so musculoskeletal deformities such as scoliosis become more apparent in this population[14].

Studies on congenital heart disease patients have revealed the scoliosis incidence to be 4.2%[32]. For patients who have received surgical intervention, this increases to 11%[18]. In recent studies, it has been suggested that thoracotomy may lead to the development of scoliosis[2-3,12-13,28,30,33].

In this study, in order to test this hypothesis, the prevalence of scoliosis was investigated in congenital heart disease patients who were aged 0–10 or adult, who either received surgery or did not.

MATERIALS AND METHODS:

356 patients in the hospital information system (HIS) who applied to the pediatric, cardiology or cardiovascular surgery departments between 1st January 2005 and 30th August 2012 with a definitive diagnosis of congenital heart disease were included in this study. 226 of these patients were aged 0–10 (3.6 ± 5.1) years with a female/male ratio of 111/115. The rest of the 130 patients were aged 20–76 (48.9 ± 22.8) years with a female/male ratio of 49/81.

Of the 226 patients in the child age group, 12 (5.3%) had mitral or aortic valve malformations, 18 (7.9%) had Tetralogy of Fallot, 33 (14.6%) had coarctation of aorta, 54 (23.59%) had ventricular septal defects (VSD), and 109 (41%) had atrial septal defects (ASD). 46 (20.4%) of those patients had never received any surgery, while 180 (76.9%) were collected from HIS who had had some cardiological intervention, surgically or invasively, in another center, and were followed up in our hospital.

Of the 130 patients in the adult age group, two (1.5%) had dextrocardia, six (4.6%) had pulmonary artery stenosis, 22 (16.9%) had mitral or aortic valve malformations, ten (7.7%) had Tetralogy of Fallot, 13 (10.0%) had coarctation of aorta, 24 (18.5%) had VSD, and 53 (40.8%) had ASD. 29 (22.3%) of the patients received surgery in our hospital for primary cardiac reasons, 26 (20.0%) received surgery in other centers during their childhood, 75 (57.7%) applied to the hospital with coronary artery disease or cardiac insufficiency, and 65 (5.0%) had had surgical or any other invasive intervention (53 patients) in their childhood. 35 (26.9%) of these patients had coronary angiography in our hospital, 25 (19.2%) had coronary bypass surgery, and the remaining 70 patients had medical treatment.

Patients were investigated retrospectively using their postero–anterior chest X-rays, and the cervicothoracic and thoracic regions that fell into the X-ray region were investigated for vertebral pathologies. The investigation was performed by a radiology specialist (S.Ç) and two orthopedists (T.B and B.Ç), who are the authors of this paper, and the data analysis was controlled with a joint evaluation.

The scoliosis prevalence was separately calculated for children and adults. The difference in prevalence according to the presence or

absence of surgery was also investigated. The prevalence was compared for the children and adult age groups.

SPSS 16.0 for Windows was used for statistical analysis. The significance for comparing two percentiles was 0.05.

RESULTS:

When all patients were included, the prevalence of vertebral deformities was 7.30% (26 patients). Of the 356 patients, scoliosis of more than 10° was seen in 25 (7.02%) patients, and congenital scoliosis was found in one (0.28%) patient. The prevalence of vertebral deformities in children was 6.64% (15 patients), and in adults this was 8.46% (11 patients).

For the 46 patients who never received surgery in childhood or were newly diagnosed, scoliosis of 10° or more was found in three patients and the prevalence was calculated to be 6.52%. Of the three patients with juvenile scoliosis, all three had less than 20° of deformity (14°, 18°, and 20°) and did not require treatment.

Of the 180 children who received surgery for congenital heart diseases, 12 (6.67%) had vertebral deformity. 11 of those 12 children (6.11%) had idiopathic scoliosis between 10° and 32.5°, with an average of 18.9 ± 9.1°, and one child (0.56%) had T6–7 and T8–9 hemivertebrae without deformity.

The scoliosis prevalence of the adult patients was 8.46% (11 patients).

All of those 11 patients had idiopathic scoliosis of greater than 10°, and the average Cobb's angle was 22.4 ± 12.2° (10–34°). Eight of those 11 patients received surgery in childhood, and only three did not require surgery or were neglected. The 82 patients who had had thoracotomy during childhood had a scoliosis prevalence of 9.56% (8 patients), and 48 patients who did not receive surgery had a scoliosis prevalence of 6.25% (3 patients).

When the prevalence in children was compared to the prevalence for adult patients who had surgery in childhood (Table-1), and when all the patients in all groups were included, from the group of children (6.64%) and adults (8.46%), the prevalence of vertebral deformation was statistically significant ($t=4.41$, $p<0.05$). However, in both groups, the prevalence for patients who did not receive surgery was not statistically significant (children: 6.52%, adults: 6.25%, $t=0.34$, $p>0.05$). Also, in the adult group comparing patients who received surgery (9.56%) and those who did not (6.25%), the prevalence was significant ($t=7.24$, $p<0.05$) (Table-1).

DISCUSSION:

The co-occurrence of congenital heart disease and vertebral deformities has been known in the literature for a long time, but no etiopathological connections have yet been found[6,9,14,17,22,24,25,29]. Shneerson et al. suggested that sudden deaths observed during vertebral surgery for idiopathic scoliosis could be due to undiagnosed congenital heart disease[32].

Table-1. Distribution of scoliosis prevalence in congenital heart disease patients in children (0–10 years of age) and adults (20–76 years of age), according to the presence of surgery.

	Surgery	No surgery	p	Total
Children (226 patients)	6.52% (3 patients)	6.67% (12 patients)	> 0.05	6.64% (15 patients)
Adult (130 patients)	6.25% (3 patients)	9.56% (8 patients)	< 0.05	8.46% (11 patients)
p	> 0.05	< 0.05	-	< 0.05
Total (356 patients)	1.68% (6 patients)	5.62% (20 patients)	< 0.05	7.30% (26 patients)

Beals et al. reported the incidence of congenital heart disease in vertebral malformations as 20%, and the incidence of vertebral malformations in congenital heart disease as 12% [4,5]. Bitan et al. reported no correlation between the severity of congenital heart disease and the severity of scoliosis[6]. In 1995, Kawakami et al. investigated chest X-rays of 680 patients who had undergone surgery, and found 10° or more of scoliosis in 10.9% of the patients[18]. Colomina et al., in a study in 2002, reported valve malformations in 11 out of 64 idiopathic scoliosis patients, without any comorbidities[7]. Liu et al., in 2010, reported high rates of cardiological disorder in echocardiograms of idiopathic scoliosis patients who required surgery, and suggested that patients who experience postoperative cardiac decompensation may fall into this category[20]. Ipp et al. published postoperative cardiac surveys of 212 idiopathic scoliosis patients, and showed that 13% had abnormal ECG or echocardiogram findings, and 3.3% had congenital valve abnormalities[16].

Our study is the first study in the literature to show the prevalence of scoliosis for both patients in childhood (0–10 years) and adulthood (20–76 years) with congenital heart diseases. In this study, including 356 patients with congenital heart diseases, the general scoliosis prevalence was 7.3%. The scoliosis prevalence was 6.64% in childhood and 8.46% in adulthood.

Recently, due to advances in the treatment of congenital heart disease, the life expectancy of patients with these diseases has increased, and scoliosis deformities have became more prevalent[14]. Conversely, there has also been the suggestion that thoracotomy results in scoliosis[2,12,13,18,28,30,33]. Bal et al., in a study that included 49 children with an average age of 10.2 with indications for cardiac surgery, reported the scoliosis prevalence as 31% [2]. Ruiz-Iban et al. retrospectively investigated 128 patients with median sternotomy with chest X-rays, and reported scoliosis of 10° or more in 34.4% of patients[30]. Herrera-Soto et al. published a study in 2006 that included 68 patients who had undergone a thoracotomy procedure, and reported scoliosis in 26% of patients, giving an incidence of scoliosis ten times higher than that of the normal population[12].

The same group published another study in 2007 in which the scoliosis prevalence of 108 median sternotomy patients was 28% [13]. It has been suggested that the development of scoliosis after thoracotomy is observed around the age of 11[14]. Based on this, in our study we included children aged 0–10 and adults aged 20–76 who had had congenital heart disease operations in childhood, and the scoliosis prevalence was studied in these two groups. For the group who had no surgery in childhood, the scoliosis prevalence was 6.67%, which is not statistically significant ($p>0.05$). The group of adult patients who had had surgery in the childhood showed a statistically significant increase in the scoliosis prevalence (9.56%). This ratio is lower than that found in the literature, and it suggests that surgical interventions increase the prevalence of scoliosis.

Ruiz-Iban et al. found the prevalence of 10–20° of scoliosis in patients who received surgery for congenital heart disease as 12.5%, and the prevalence of more than 20° of scoliosis as 25.8%[30]. Herrera-Soto et al. studied the Cobb angle of patients who received surgery for congenital heart disease and found an average scoliosis of 25° (11–88°), and seven patients with more than 30° of deformation[12]. In our study, we found that the deformity in children who received surgery was between 10° and 32.5°, with an average of 18.9 ± 9.1°, and for the adults who received surgery in childhood, the average Cobb angle was 22.4 ± 12.2° (10–34°).

We observed the scoliotic deformity of more than 30° in only one patient in the group of patients in childhood, and in five patients in the adult group.

Gillingham et al. reported a high rate of accompanying congenital heart disease in early onset scoliosis patients. The deformities were generally lower than 20° and did not require treatment[11]. In our study, we found the scoliosis in the group of childhood patients was generally less than 20°.

Congenital syndromes from mesenchymal tissues and the coexistence of congenital heart diseases and scoliosis are reported in many publications[14,25]. Hou et al., in a study on rats in 2006, showed that mutant rats with kyphoscoliosis had a comorbidity risk for congenital heart diseases[15].

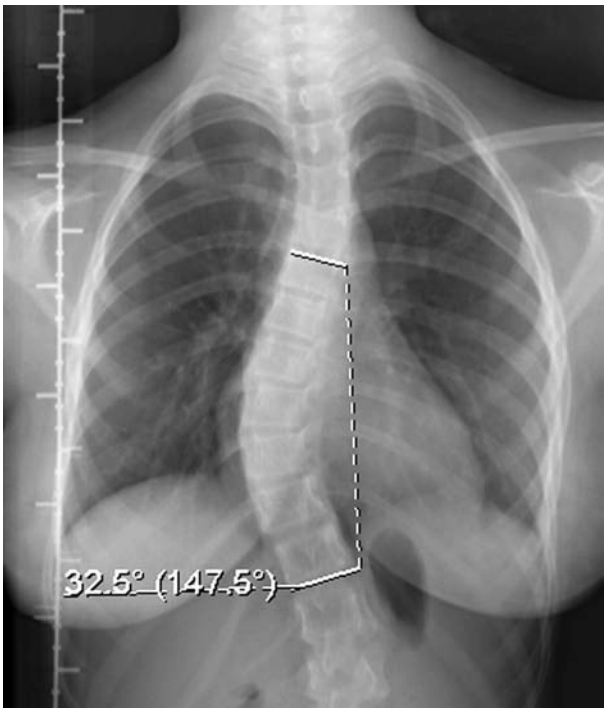

Figure-1. 32.5° of scoliosis visible by thoracic X-ray in a 10-year-old female patient with ASD.

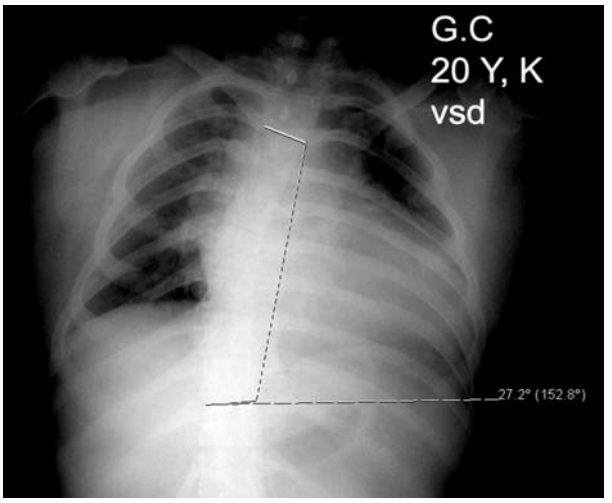

Figure-2. 27.7° of scoliosis visible by thoracic X-ray in a 20-year-old female patient with ASD.

Massin et al. followed up 1058 congenital heart disease patients for ten years, and found coexisting problems such as scoliosis in 21.2% of the patients, 11.2% of which were part of a syndrome[23]. In a study by Liu et al. published in 2011, they reported high rates of coexistence of congenital heart disease and scoliosis[21]. In particular, the incidence of congenital scoliosis has been reported to be higher for patients with syndromes such as trisomy 13, Pilotto syndrome, Mayer-Rokitansky-Küster-Hauser syndrome, and dextrocardia,[1,8,10,19,26-27,31]. In our study, only one patient had congenital scoliosis. This has been attributed to the early death due to cardiac reasons of patients with syndromes and serious congenital heart malformations, or the follow-up of those patients being conducted in pediatric cardiovascular surgery centers.

In conclusion, in this study that included 356 patients with a diagnosis of congenital heart disease, the scoliosis prevalence was 7.30% when all the patients were included, in the 0–10 year age group it was 6.64%, and in adults it was 8.46%.

For patients who received surgery in childhood for congenital heart diseases, the scoliosis prevalence was only increased for adults. This suggests that thoracotomy results in scoliosis after the age of 10. In the light of these findings, congenital heart disease patients should have scoliosis X-rays and should be followed up, and patients with scoliosis who require surgical intervention should be evaluated for cardiac pathologies.

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