Prevalence of Asymptomatic Cardiac Valvular Anomalies in Adolescent Idiopathic Scoliosis- A Descriptive Study from Retrospective Data Analysis

Dhanya Vijayan[1], Ashok Ramakrishnan[2], Jayakumar Christudas[3]

[1]Assistant Professor, Department of Anaesthesia, Government Medical College, Thiruvananthapuram, Kerala, India. [2]Assistant Professor, Department of Orthopaedics, Government Medical College, Thiruvananthapuram, Kerala, India. [3]Associate Professor, Department of Anaesthesia, Government Medical College, Thiruvananthapuram, Kerala, India.

ABSTRACT

BACKGROUND

This study aims to measure the prevalence of asymptomatic cardiac valvular anomalies in females with adolescent idiopathic scoliosis. The association between severity of scoliosis and prevalence of valvular anomalies was also looked into. The secondary objective of this study was to enumerate the other systemic co-morbidities in adolescent onset scoliosis. We also attempted to establish the association between these co-morbidities and valvular anomalies in our study population.

METHODS

Pre anaesthesia assessment records of 84 females, with adolescent-onset idiopathic scoliosis who presented to our hospital for surgical correction, were analysed after obtaining permission from the human ethics committee.

RESULTS

The mean age at the time of diagnosis was 14.4 ± 2.03 years. Using echocardiograph and colour Doppler, 29 valvular lesions were detected in 20 patients, totalling a prevalence rate of 23.8%. The most common valvular anomaly was Mitral valve prolapse with a prevalence of 44.83%. The severity of scoliosis did not have a significant association with the presence of valvular anomalies. (chi-square value 2.487 with a p-value of 0.478 for thoracic curve, chi-square value 1.044 with a p-value of 0.903 for lumbar scoliosis). Thirteen patients in the study group presented with systemic diseases. The association between systemic diseases and valvular anomalies was found to be significant. In 8 out of 13 patients with comorbidities, valvular anomalies were detected (61.5%) whereas valvular anomalies were diagnosed in 12 out of 71 patients without systemic diseases (16.9%).

CONCLUSIONS

From this study, it was concluded that patients with adolescent idiopathic scoliosis exhibited a spectrum of asymptomatic valvular anomalies irrespective of the severity of the disease. Systemic diseases, when present in adolescent scoliosis patients, had a greater prevalence in those with valvular anomalies.

KEYWORDS

Adolescent Idiopathic Scoliosis, Cardiac Valvular Anomalies, Echocardiography, Systemic Comorbidities

Corresponding Author:
Dr. Ashok Ramakrishnan,
Assistant Professor,
Department of Orthopaedics,
Government Medical College,
Thiruvananthapuram, Kerala, India.
E-mail: ashrampillai@gmail.com

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BACKGROUND

Scoliosis is a complex deformity of the spine characterized by lateral curvature and rotation of vertebrae as well as deformity of rib cage.[1] Over time, secondary involvement of respiratory, cardiovascular and neurological systems occurs. Prevalence is more in adolescents and has a male to female ratio of 1:3. Idiopathic scoliosis, comprising 70% of total cases, is the most common aetiology of scoliosis. Of these, the adolescent origin is the most frequently encountered. Congenital, neuromuscular and mesenchymal disorders associated are other, less frequently encountered types. Adolescent idiopathic scoliosis is a variant occurring in late childhood or adolescence. Genetic and environmental factors are implicated in the aetiology of adolescent scoliosis.[2] Treatments include observation, bracing and/or surgery.[3] The severity of scoliosis is measured by the Cobb's angle. A Cobb's angle more than 80° with flexibility less than 25 per cent on bending films are classified as severe scoliosis and a Cobb's angle less than 80° is called non-severe scoliosis. A thoracic Cobb's angle of greater than 50° or lumbar curve greater than 40° is an indication for surgical correction.

Prevalence of scoliosis varies between 0.3-15.3%. However, a Cobb's angle of more than 30° exists in only 0.3% of affected patients.[4]

Medical literature provides ample evidence of the association between scoliosis and congenital heart disease.[5] Currently the association between asymptomatic valvular anomalies and scoliosis is being studied in detail.[2,6,7] Various researchers have established the association of mitral valve prolapse and mitral regurgitation with idiopathic scoliosis.[8,9,10,11,12,13,14] Tricuspid regurgitation is another association of idiopathic scoliosis.[6]

The primary objective of our study is to estimate the prevalence of asymptomatic valvular heart disease in adolescent females with idiopathic onset of scoliosis. Also, we strive to detect any possible association between the severity of scoliosis and the presence of cardiac valvular anomalies. Another objective of this study is to estimate the prevalence of systemic diseases in idiopathic scoliosis and their association with valvular anomalies in these patients with idiopathic scoliosis.

METHODS

This retrospective study consisted of 84 female patients (10-19 years) with adolescent-onset idiopathic scoliosis who presented to our hospital between March 2016 and March 2019 for surgical correction. Patients with congenital and neuromuscular scoliosis were excluded. Patients with symptoms of cardiac dysfunction or previously diagnosed heart diseases were also excluded from our study. After obtaining approval from the hospital ethics committee, the sample size was calculated using the reference study.[8]

Sample Size and Sampling

$$n = \frac{Z_a^2 * (p * (1 - p))}{d^2}$$

α at 5% level of significant, $Z_{0.05} = 1.96$
As observed by Lang C, Wang R et al. (2019)[8] the values for p and d can be assigned as:
p =0.24
d=precision =10% = 0.10
n = 70.07
By considering 20% drop out, the minimum required sample size.
n=84

The following data were accessed from the patients' medical records - age at diagnosis, weight, height, medical and family history. Cobb's angle was measured from MRI. Respiratory assessment was done using pulmonary function tests using Vitalograph Pneumotrac 6800. Standardization done using accuracy check button and a 3L precision syringe.

Cardiac function was evaluated by 12-lead ECG; and 2-D echo (M mode) and colour Doppler examination using Philips EPIQ 7 series ultrasound machine.

Statistical Analysis

The descriptive analysis included mean, range and standard deviation of the quantitative variable. The student's t-test or the Mann Whitney non-parametric test were used to compare the means. The Chi-squared test was used to compare proportions. Statistical significance was set at a p-value of less than or equal to 0.05. Data were processed with the SPSS software package, version 16.0.

RESULTS

Comorbidities	Cardiac Valvular Anomaly Present		Absent		Total	
	No.	%	No.	%	No.	%
Present	8	61.5%	5	38.5%	13	100%
Absent	12	16.9%	59	83.1%	71	100%
Total	20	23.8%	64	76.2%	84	100%

Table 1. Association of Cardiac Valvular Anomaly and Co-Morbidities

Chi square = 12.069 p value - 0.001

Co-morbidity	Count
Family history of scoliosis	2
Marfanoid habitus	2
Congenital lobar emphysema	2
Childhood onset seizures	2
Hypothyroidism	2
Pierre Robin syndrome	1
Congenital hearing loss	1
Umbilical hernia	1
Bronchial asthma	1
Gilbert's syndrome	1

Table 2

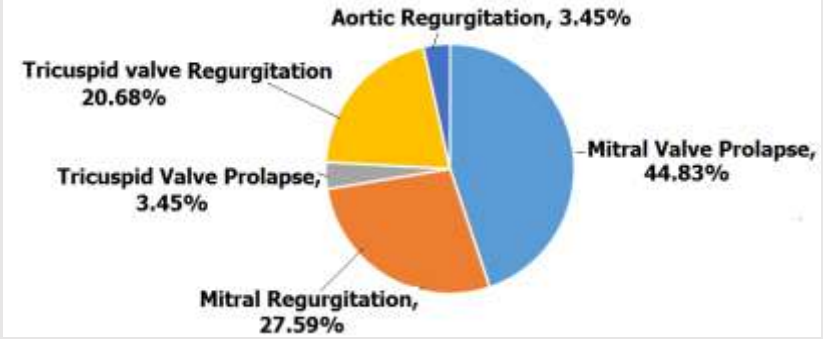

Figure 1. Distribution of the Study Subjects based on the Type of Cardiac Valvular Anomalies Present

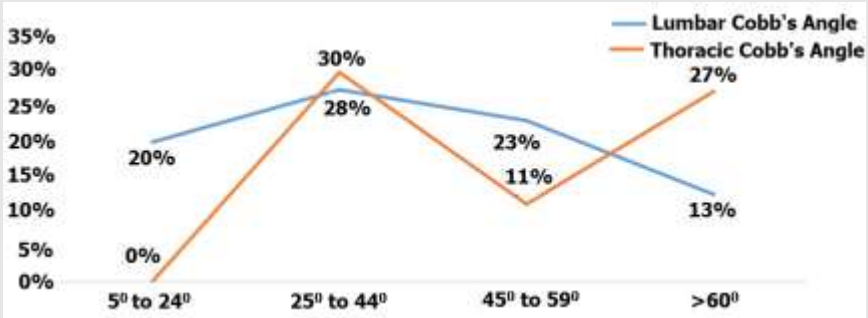

Figure 2. Prevalence of Cardiac Valvular Anomalies Present for Various Cobb's Angles

The mean age of patients at diagnosis was 14.44 with a standard deviation of 2.03 years. The age of the study subjects ranged from 10 years to 19 years. Of the 84 study subjects, twenty patients (23.8%) had cardiac valvular anomalies. Some subjects had more than one cardiac lesion, so total cardiac valvular lesions found was 29.

Cobb's angle was grouped as non-severe (less than 80°) and severe (more than 80°). 31 (36.94%) out of 84 patients had thoracic Cobb's angle greater than or equal to 80°. The mean thoracic Cobb's angle was 68.97. For thoracic Cobb's angle, the Chi-square value was found to be 2.487 and p value - 0.478. This difference was not found to be significant with a chi square value of 2.487 at a p-value of 0.478 Of the 84 patients, 6 patients did not have an abnormal lumbar Cobb's angle. For the other 78 patients, the mean lumbar Cobb's angle was 42.38°. All of these patients had a lumbar Cobb's angle of less than 80°. For lumbar Cobb's angle, the Chi-square was found to be 1.044 and p-value was 0.903. This difference was not found to be significant. 61.5% of those with co-morbidities had cardiac valvular anomalies, whereas 16.7% without co-morbidities had cardiac valvular anomalies. This difference was found to be significant with a chi square value of 12.069 at a p value of 0.001. There were 13 patients who had 15 co-morbidities.

DISCUSSION

Association between spine deformities and congenital cardiac diseases has been studied in detail.[5] Incidence of scoliosis in patients with congenital heart disease ranges from 11 to 34%. Cyanosis in congenital heart disease is postulated to cause scoliosis.[4,15] Post thoracotomy and sternotomy patients operated for congenital heart diseases are also found to have an increased incidence of scoliosis.[16,17] Rather than the surgical procedure, the coexisting cardiac condition is thought to be a significant causative factor in the development of scoliosis. Idiopathic scoliosis is 10 times more common in children with congenital heart disease compared to unaffected children.[18] In recent years, researchers have focused on the prevalence of asymptomatic valvular anomalies in idiopathic and congenital scoliosis. Several reports revealed an increase in the incidence of congenital heart abnormalities, namely mitral valve prolapse (MVP), atrial septal defect (ASD), ventricular septal defect (VSD), tricuspid regurgitation and mitral valve dysplasia, among adolescent patients with idiopathic scoliosis[13,14,19] Since both mitral vasculogenesis and vertebral formation occur during 5th to 7th week of gestation, it is postulated that any insult to the foetal growth pattern during intrauterine differentiation at this time could be the common precipitating factor for cardiac valvular anomalies and skeletal deformities.[10,12,20]

Mitral valve prolapse co-exists with various skeletal conditions, such as Pectus Excavatum and various spine anomalies.[9,10,11,12] The diagnosis of MVP is sometimes considered in patients who have thoracic skeletal abnormalities; the most common being scoliosis. Hirschfield et al[13] described a 28% prevalence of MVP in patients with idiopathic scoliosis. However, they reported a 41% prevalence in patients with a positive family history of scoliosis. The prevalence further rose to 53% if the patient had a first degree relative with scoliosis. Dhuper et al[14] reported a 13.6% prevalence of MVP in a series of patients with idiopathic scoliosis. They concluded that MVP is four times more common in patients with severe severe idiopathic scoliosis than in the normal adolescent population,

and is associated with body weight in idiopathic scoliosis patients with MVP than in idiopathic scoliosis patients without MVP. In another study by Stephen et al, MVP was present in 48% of their subjects with hereditary scoliosis and kyphoscoliosis. Prevalence increased to 53% in patients with a positive family history of skeletal abnormality. The cardiac and skeletal manifestations may be attributed to a common generalized soft-tissue defect.[21] M J Colomina et al[7] also concluded that mitral valve prolapse was the commonest (75%) asymptomatic valvular anomaly, in their study population consisting of adolescent scoliosis patients.

Salomon J et al[22] studied the prevalence of thoracic skeletal abnormalities in a series of subjects with mitral valve prolapse. They found that 75 per cent of their study subjects had a definite thoracic skeletal deformity, including pectus excavatum (62%) and severe scoliosis (8%). They proposed that the association of idiopathic mitral valve prolapse with skeletal deformities may represent an attenuated manifestation of Marfan's syndrome.

Bozcali E et al[22] found that mitral valve prolapse was the most common cardiac abnormality in their patients with idiopathic and congenital scoliosis. Atrial septal aneurysm, pulmonary regurgitation, aortic regurgitation, atrial septal defect patent foramen ovale, dextrocardia, bicuspid aortic valve, aortic stenosis, ventricular septal defect, and cardiomyopathy were other, less frequently detected cardiac lesions in their study subjects.[22] Zhu et.al reported that tricuspid regurgitation was the commonest valvular anomaly in their research subjects with idiopathic scoliosis.[6] They observed that regurgitation with tricuspid valve involvement was statistically significant in patients with thoracic curve. They found no significant associations were found between the severity of scoliosis or pulmonary function and cardiac abnormalities

Our study results were analogous to those of Hirschfield, Dhuper and Colomina. In this study 20 out of 84 patients had 29 valvular lesions with a prevalence of 23.8% Mitral valve prolapse was the commonest observed valvular anomaly with 44.83% prevalence. Mitral regurgitation had a prevalence of 27.6% Tricuspid regurgitation came next with 20.68%. Other less common anomalies were aortic regurgitation (3.45%) and Tricuspid valve prolapse. (3.45%). Consistent with the findings of Zhu et al, it was found that there was no significant association between the severity of scoliosis and cardiac valvular anomalies. It must be noted that none of our study subjects with valvular anomalies suffered perioperative cardiovascular complications.

Congenital and secondary scoliosis have many systemic associations such as Osteogenesis imperfecta[23] Marfan syndrome[24,25] Stickler syndrome,[26] Ehlers-Danlos syndrome[27] and the muscular dystrophies,[28] are associated with scoliosis. But no such definite association has been found to occurs in idiopathic scoliosis. However, M J Columina et al,[18] in their study describing the prevalence of asymptomatic cardiac valvular anomalies in idiopathic scoliosis noted a series of systemic associations in these patients. They also observed that valvular anomalies in patients with idiopathic scoliosis exhibited a greater prevalence in those patients with coexisting systemic associations than those without.

From this study, it appears that a positive relationship exists between scoliosis, valvular anomalies and systemic comorbidities. 13 patients in this study presented with 15 systemic co-morbidities. Prevalence of valvular anomalies in the group with co-morbidities was 61.5% and in the group without co-morbidities was 16.9%. The co-morbidity group consisted of two-second degree relatives with adolescent-onset scoliosis, two subjects with marfanoid habitus, Pierre Robin's syndrome, cleft palate, congenital hearing loss, umbilical hernia, bronchial asthma, congenital lobar emphysema, Gilbert's syndrome, childhood-onset seizures and hypothyroidism. The analysis of these co-morbid conditions show that some of these had an obvious genetic component (as in the patients with a family history of scoliosis) while others seem to have originated in the embryonic period.

CONCLUSIONS

Adolescent-onset scoliosis is associated with asymptomatic valvular anomalies. A common insult during the embryonic development could be the cause of coexisting valvular and vertebral anomalies in adolescent patients with idiopathic scoliosis. Therefore we are justified in having a high index of suspicion of valvular lesions, especially mitral valve prolapse, in patients with idiopathic scoliosis. Age of diagnosis of scoliosis or severity of scoliosis had no relation to the prevalence of valvular anomalies. Adolescent scoliosis was associated with several systemic co-morbidities which in turn is significantly related to the occurrence of cardiac valvular abnormalities in these patients. A screening echo in patients with adolescent-onset idiopathic scoliosis would help in early diagnosis of these cardiac valvular lesions.

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REFERENCES

[1] Kulkarni AH, Ambareesha M. Scoliosis and anaesthetic considerations. Indian J Anaesth 2007;51(6):486-495.

[2] Adolescent idiopathic scoliosis. Genetics Home Reference (GHR). September 2013. http://ghr.nlm.nih.gov/condition/adolescent-idiopathic-scoliosis. Accessed 11/10/2014.

[3] Idiopathic Scoliosis: Adolescents: treatment. Scoliosis Research Society 2014. http://www.srs.org/patient_and_family/scoliosis/idiopathic/adolescents/treatment.htm. Accessed 11/10/2014.

[4] Kawakami N, Mimatsu K, Deguchi M, et al. Scoliosis and congenital heart disease. Spine (Phila Pa 1976) 1995;20(11):1252-1255.
[5] Ogilvie J. Congenital heart disease and scoliosis. In: Bradford D, Lonstein J, Moe J, et al, eds. Moe's textbook of scoliosis and other spinal deformities. Philadelphia: W.B. Saunders Co 1995:564-565.
[6] Zhu XD, Wang JL, Li M. Prevalence of valvular regurgitation in adolescent idiopathic scoliosis patients. J Pediatr Orthop B 2011;20(1):33-36.
[7] Colomina MJ, Puig L, Godet C, et al. Prevalence of asymptomatic cardiac valve anomalies in idiopathic scoliosis. Pediatr Cardiol 2002;23(4):426-429.
[8] Lang C, Wang R, Chen Z, et al. Incidence and risk factors of cardiac abnormalities in patients with idiopathic scoliosis. World Neurosurg 2019;125:e824-e828.
[9] deLeon AC, Ronan JA. Thoracic bony abnormalities with the click and late systolic murmur syndrome. Circulation 1971;44(Suppl 2):157.
[10] Kumar UK, Sahasranam KV. Mitral valve prolapse syndrome and associated thoracic skeletal abnormalities. J Assoc Physicians India 1991;39(7):536-539.
[11] Salomon J, Shah PM, Heinle RA. Thoracic skeletal abnormalities in idiopathic mitral valve prolapse. Am J Cardiol 1975;36(1):32-36.
[12] Udoshi MB, Shah A, Fisher VJ, et al. Incidence of mitral valve prolapse in subjects with thoracic skeletal abnormalities--a prospective study. Am Heart J 1979;97(3):303-311.
[13] Hirschfeld S, Rudner C, Nash CL, et al. Incidence of mitral valve prolapse in adolescent scoliosis and thoracic hypokyphosis. Pediatrics 1982;70(3):451-454.
[14] Dhuper S, Ehlers KH, Fatica NS, et al. Incidence and risk factors for mitral valve prolapse in severe adolescent idiopathic scoliosis. Pediatr Cardiol 1997;18(6):425-428.
[15] Roth A, Rosenthal A, Hall JE, et al. Scoliosis and congenital heart disease. Clin Orthop Relat Res 1973;(93):95-102.
[16] Durning RP, Scoles PV, Fox OD. Scoliosis after thoracotomy in tracheoesophageal fistula patients. A follow-up study. J Bone Joint Surg Am 1980;62(7):1156-1159.
[17] Bisgard JD. Thoracogenic scoliosis influence of thoracic disease and thoracic operations on the spine. Arch Surg 1934;29(3):417-445.
[18] Herrera-Soto JA, Vander Have KL, Barry-Lane P, et al. Retrospective study on the development of spinal deformities following sternotomy for congenital heart disease. Spine (Phila Pa 976) 2007;32(18):1998-2004.
[19] Liu L, Xiu P, Li Q, et al. Prevalence of cardiac dysfunction and abnormalities in patients with adolescent idiopathic scoliosis requiring surgery. Orthopedics 2010;33(12):882.
[20] Read R, Tahl AP, Wendt VE. Symptomatic valvular myxomatous transformation (the floppy valve syndrome). A possible forme fruste of the Marfan syndrome. Circulation 1965;32(6):897-910.
[21] Hirschfeld SS, Rudner C, Nash CL, et al. Incidence of mitral valve prolapse in adolescent scoliosis and thoracic hypokyphosis. Pediatrics 1982;70(3):451-454.
[22] Bozcali E, Ucpunar H, Sevencan A, et al. A retrospective study of congenital cardiac abnormality associated with scoliosis. Asian Spine J 2016;10(2):226-230.
[23] Sillence DO, Barlow KK, Cole WG, et al. Osteogenesis imperfecta type III. Delineation of the phenotype with reference to genetic heterogeneity. Am J Med Genet 1986;23(3):821-832.
[24] Winter RB. Posterior spinal fusion in scoliosis: indications, techniques, and results. Orthop Clin North Am 1979;10(4):787-800.
[25] Pyeritz RE, McKusick VA. The Marfan syndrome: diagnosis and management. N Engl J Med 1979;300(14):772-777.
[26] Ahmad NN, Ala-Kokko L, Knowlton RG, et al. Stop codon in the procollagen II gene (COL2A1) in a family with the Stickler syndrome (arthro-ophthalmopathy) Proc Natl Acad Sci USA 1991;88(15):6624-6627.
[27] Steinmann BU, Royce PM, Superti-Furga A. The Ehlers-Danlos syndrome. In: Royce PM, Steinmann BU, eds. Connective tissue and its heritable disorders: Molecular, genetic, and medical aspects. 5th edn. New York: Wiley-Liss 1993:351-407.
[28] Emery AE. X-linked muscular dystrophy with early contractures and cardiomyopathy (Emery-Dreifuss type). Clin Genet 1987;32(5):360-367.