Demographic Findings and Associated Anomalies in Patients with Congenital Kyphosis

Abstract

Background: Congenital kyphosis is a spinal deformity resulting from disruption in the formation of the anterior part of the vertebrae on the sagittal plane during early fetal development. Understanding demographic trends and associated anomalies in affected individuals within specific regions and countries is pivotal for tailoring more suitable treatment approaches within these patients.

Methods: This retrospective cross-sectional study was conducted on 108 patients referred to Shafa Yahyaian Hospital in Tehran between 2010 and 2019. Demographic and radiological criteria were assessed in all patients who underwent surgery for congenital kyphosis at this hospital. Parameters such as age, gender, BMI, presence of cardiac, urogenital system, and spinal anomalies, kyphosis type, as well as Cobb's angle before and after surgery, were gathered, reviewed, and analyzed from patients' records.

Results: Of the cohort, 74 patients were categorized as congenital kyphosis type 1, 21 as type 2, and 13 as type 3, encompassing 69 female (63.9%) and 39 male patients (36.1%). The mean age at the time of examination and symptom onset was 6.5 years, while the mean age at the time of surgery was 16.5 years. Notably, 38 patients exhibited a cardiac anomaly, 14 had genitourinary system anomalies, and 39 presented with spinal anomalies. A statistically significant correlation existed between the presence of a cardiac anomaly and the type of congenital kyphosis, with type one exhibiting a markedly higher incidence of cardiac anomalies compared to the other two types.

Conclusion: The findings revealed a higher frequency of type 1 congenital kyphosis. Moreover, patients with type 3 congenital kyphosis underwent surgery at a significantly younger age than those with the other two types, potentially attributed to its higher progression rate. Spinal cord, cardiac, and genitourinary tract anomalies were respectively observed as the most prevalent anomalies among the patients.

Keywords: Kyphosis, Demographic, Congenital abnormalities

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Mohammadreza Chehrassan, MD[1]; Mohammadreza Shakeri, MD[1]; Farshad Nikouei, MD[1]

[1]Shafa Orthopaedic Hospital, Iran University of Medical Sciences, Tehran, Iran.

Corresponding Author:
M Shakeri ; MD
Email Address:
mshakeri3116@gmail.com

Introduction

Congenital kyphosis arises from a disruption in the formation of the anterior part of the vertebrae in the sagittal plane during early fetal development [1-4]. It represents a common spinal deformity that can impact various bodily systems, including the nervous, respiratory, and cardiac systems [3]. A comprehensive understanding of this condition is crucial for devising improved surgical strategies and halting disease progression [2, 3]. Notably, congenital kyphosis is more prevalent in females [5], and when symptomatic before the age of 10, it typically carries a poorer prognosis [6].

The condition is broadly categorized into three types: type 1 results from a disruption in the proper formation of the vertebral body; type 2 stems from a disturbance in vertebral body segmentation; type 3 involves a simultaneous disorder in both the formation and segmentation of the vertebral column. This classification aids in predicting disease progression speed, thereby facilitating the development of more effective treatment plans for patients [5, 7].

Type 1 congenital kyphosis, commonly observed in the thoracolumbar regions,

exhibits a faster development rate compared to other subtypes and is associated with a higher incidence of paraplegia [3, 8, 9].

Type 2 congenital kyphosis progresses less rapidly than type 1 and is predominantly found in the cervical region and the thoracolumbar junction of the spine [3]. Conversely, type 3 congenital kyphosis progresses more swiftly than type 2 and is more frequently observed in the thoracolumbar junction zone [2, 5].

This study aims to assess the epidemiological and radiological characteristics of patients who underwent surgery for congenital kyphosis. The goal is to enhance the planning and implementation of treatment strategies for these patients.

Methods

A retrospective cross-sectional study was designed and conducted, involving 108 patients referred to Shafa Yahyaian Hospital in Tehran between 2010 and 2019. The study focused on evaluating demographic and radiological criteria of all patients who underwent surgery for congenital kyphosis at this hospital. Parameters examined and recorded included age, gender, body mass index (BMI), presence of cardiac anomalies, urogenital system anomalies, spinal anomalies, kyphosis type, and Cobbs angle before and after surgery.

Data from patients' medical records were collected and analyzed utilizing SPSS 26 software. The normality of quantitative data was assessed using the Kolmogorov- Smirnov test. Parametric tests were applied for normally distributed data, while corresponding non- parametric tests were employed for skewed distributions. Throughout this study, a P-value < 0.05 was deemed statistically significant for all conducted tests.

Results

In this research, 108 patients who underwent surgery between 2010 and 2019 were included. Among these individuals, 74 patients (68.5%) were diagnosed with congenital kyphosis type 1, 21 patients (19.5%) were identified as type 2, and 13 patients (12%) were categorized as type 3. Of the total, 69 patients were female (63.9%), and 39 patients were male (36.1%). Specifically, there were 52 female patients in group one, only 7 female patients in group two, and 10 female patients in group three. However, no significant relationship was found between gender and the type of kyphosis.

The mean age of patients at the onset of symptoms and during examination was 6.5±2.4 years. Additionally, the mean age at the time of surgery was 16.5±5.8 years. A statistically significant relationship was identified between the patients' age at the time of surgery and the type of congenital kyphosis (correlation coefficient = - 0.162 with a P-value=0.039). Notably, the average age of patients at the time of surgery was notably lower in type three congenital kyphosis (5.8±12.2 years) compared to the other two types (Table 1). However, no statistically significant relationship was observed between the age of patients at the onset of symptoms and the type of congenital kyphosis.

The average Body Mass Index (BMI) among patients was 22.6±2.3 kg/m². Interestingly, there was no statistically significant relationship between BMI and the type of congenital kyphosis (correlation coefficient of 0.29 with a P- value of 0.706) (Table 2).

Associated anomalies of other organs involving the cardiac, urogenital, and spinal cord systems. Among the patients, 38 (35.2%) exhibited cardiac anomalies, 14 (13%) presented urogenital anomalies, and 39 (36.1%) displayed spinal cord anomalies, encompassing conditions such as syrinx, tethered cord, and diastematomyelia.

A statistically significant relationship was found between cardiac anomalies and the type of congenital kyphosis (P- value = 2.0). Notably, the incidence of cardiac anomaly was notably higher in type 1 (9.45%) compared to the other two types (7.7% in type 2 and 3.14% in type three). However, the presence of a genitourinary system anomaly did not exhibit

a statistically significant relationship with the type of congenital kyphosis. Conversely, there was a statistically significant relationship observed between the presence of spinal cord anomalies and the type of congenital kyphosis. Specifically, in type one congenital kyphosis, 43.2% of patients exhibited spinal cord anomalies, while in type 2 and 3, the percentages were 7.7% and 6.28%, respectively.

In evaluating concurrent anomalies, 45 patients (41.7%) did not display any of the mentioned anomalies. Among the remaining patients, 17 (15.7%) exclusively presented with cardiac anomalies, six (5.6%) solely had urinary tract anomalies, and 14 (13%) were solely affected by spinal cord anomalies. Additionally, one patient (9.0%) displayed anomalies in both the heart and genitourinary system, while 18 patients (16.7%) exhibited simultaneous anomalies in the heart and spinal cord. Furthermore, five patients (4.6%) had anomalies in both the genitourinary and spinal systems, and two patients (1.9%) experienced anomalies in the cardiac, genitourinary, and spinal tracts concurrently. Importantly, the presence of any of these anomalies did not demonstrate an association with the presence of other types of anomalies (P value > 0.05).

The pre-surgery average Cobbs angle measured 88.4°±25.5, which significantly reduced to 2.26°±7.9 post-surgery. On average, there was a substantial change in the Cobbs angle by 62.2±20.7 degrees. A statistically significant relationship was observed between the Cobbs angle before surgery and the type of congenital kyphosis (correlation coefficient = 0.225 with a P value = 0.003). Particularly, the Cobbs angle before surgery in type three congenital kyphosis was notably higher than in the other two types (Table 3). However, no statistically significant relationship was identified between the Cobbs angle after surgery and the type of congenital kyphosis.

Table 1: Mean of the patients age at the time of diagnosis and surgery based on type of congenital kyphosis

Type of congenital kyphosis	Mean age at the time of diagnosis	Mean age at the time of surgery
Type 1	6.7 ± 2.3	17.1±5.1
Type 2	8.8±1.5	19.8±5.9
Type 3	4.5±1.9	12.2±5.8

Table 2: Mean BMI based on congenital kyphosis type

Type of congenital kyphosis	BMI
Type 1	22.6±2.2
Type 2	23.1±1.9
Type 3	22.2±2.9

Table 3: Cobbs angle changes based on congenital kyphosis type

Type of congenital kyphosis	Cobbs angle before surgery	Cobbs angle after surgery	Cobbs angle change
Type 1	84.2±22.3	25.1±9.5	59.1±17.9
Type 2	76.8±27.9	26.7±5.1	50.1±25.3
Type 3	110.5±23.3	29.7±11.8	80.8±15.9

Discussion

Congenital kyphosis represents a spinal disorder where inadequate or untreated management may lead to significant impairments in a patient's life. Non-surgical interventions often have limited efficacy, with early posterior and anterior fusion being the most effective treatment option thus far [10]. The primary surgical objective for kyphosis treatment is to rectify the sagittal curvature and restore spinal balance [11]. A comprehensive understanding of congenital kyphosis features aids in selecting the appropriate treatment for each patient.

Regarding gender distribution, among the patients, 69 (63.9%) were female, and 39 (36.1%) were male. While previous studies have reported a higher prevalence of congenital kyphosis in females compared to males [4], this present study did not establish a statistically significant relationship between gender and the type of congenital kyphosis (P-value=0.117).

The average age of patients at symptom onset was 6.5±2.4 years, whereas the mean age at surgery was 16.5±5.8 years. Notably, the average age at surgery for type 3 congenital kyphosis was significantly higher than for the other types. However, the age of symptom onset had no significant association with the type of kyphosis [12, 13].

Studies conducted earlier by Winter et al. [10] and McMaster et al. revealed that clinical manifestation of congenital kyphosis before the age of 10 years could lead to a poorer prognosis. They reported an annual progression of 5 to 6 degrees in kyphosis curve in patients manifesting kyphosis before 10 years and puberty [3].

Analyzing the frequency of anomalies, this study found cardiac anomalies in 38 patients, urinary-genital system anomalies in 14 patients, and spinal anomalies in 39 patients. This prevalence differs from earlier studies like Basu et al.'s [9], where cardiac anomalies among individuals with congenital spine anomalies were reported as 26%, while it was 35.2% in this study. Similarly, the presence of spinal anomalies in individuals with spine anomalies was 56% in Bliko et al.'s study [14, 15] but stood at 36.1% in this current investigation. Notably, the variation in patient cohorts across studies may account for these differences. Rai et al. noted that patients with congenital spine abnormalities and genitourinary involvement tend to exhibit more skeletal- muscular, digestive, and cardiac abnormalities [11]. However, in this study, while anomalies in the genitourinary system did not show a significant relationship with the type of congenital kyphosis, spinal anomalies did.

Additionally, a statistically significant relationship was observed between the Cobbs angle before surgery and the type of congenital kyphosis. Specifically, the pre-surgery Cobbs angle in type three congenital kyphosis was significantly higher than the other types. Audat et al.'s study in 2020 on severe thoracolumbar kyphosis patients reported a range of Cobbs angles before surgery from 35–180 (81.48±39.1) degrees, which improved to 0–45 (21.72±13.47) degrees after surgery [16].

Conclusion

Based on the findings, the most prevalent form of congenital kyphosis observed was type 1. Notably, the average age of patients undergoing surgery for type 3 congenital kyphosis was significantly lower compared to the other types, potentially associated with its higher rate of progression.

The most frequently observed anomalies among patients were spinal cord, cardiac and genitourinary tract anomalies, respectively. Particularly, cardiac and spinal anomalies were significantly more prevalent in type 1 congenital kyphosis compared to the other types. Additionally, both the pre- surgery Cobbs angle and the post- surgery changes in the Cobbs angle were notably higher in type 3 congenital kyphosis than in the other two types.

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