Original Article

Klippel–Feil: A constellation of diagnoses, a contemporary presentation, and recent national trends

ABSTRACT

Background: Klippel–Feil syndrome (KFS) includes craniocervical anomalies, low posterior hairline, and brevicollis, with limited cervical range of motion; however, there remains no consensus on inheritance pattern. This study defines incidence, characterizes concurrent diagnoses, and examines trends in the presentation and management of KFS.

Methods: This was a retrospective review of the Kid's Inpatient Database (KID) for KFSpatients aged 0–20 years from 2003 to 2012. Incidence was established using KID-supplied year and hospital-trend weights. Demographics and secondary diagnoses associated with KFS were evaluated. Comorbidities, anomalies, and procedure type trends from 2003 to 2012 were assessed for likelihood to increase among the years studied using ANOVA tests.

Results: Eight hundred and fifty-eight KFS diagnoses (age: 9.49 years; 51.1% females) and 475 patients with congenital fusion (CF) (age: 8.33 years; 50.3% females) were analyzed. We identified an incidence rate of 1/21,587 discharges. Only 6.36% of KFS patients were diagnosed with Sprengel's deformity; 1.44% with congenital fusion. About 19.1% of KFS patients presented with another spinal abnormality and 34.0% presented with another neuromuscular anomaly. About 36.51% of KFS patients were diagnosed with a nonspinal or nonmusculoskeletal anomaly, with the most prevalent anomalies being of cardiac origin (12.95%). About 7.34% of KFS patients underwent anterior fusions, whereas 6.64% of KFS patients underwent posterior fusions. The average number of levels operated on was 4.99 with 8.28% receiving decompressions. Interbody devices were used in 2.45% of cases. The rate of fusions with <3 levels (7.46%) was comparable to that of 3 levels or greater (7.81%).

Conclusions: KFS patients were more likely to have other spinal abnormalities (19.1%) and nonnervous system abnormalities (13.63%). Compared to congenital fusions, KFS patients were more likely to have congenital abnormalities such as Sprengel's deformity. KFS patients are increasingly being treated with spinal fusion.

Level of Evidence: III

Keywords: Congenital anomaly, congenital scoliosis, Kid's Inpatient Database, Klippel–Feil

INTRODUCTION

Klippel–Feil syndrome (KFS) is classically defined as having the clinical triad of a lower posterior hairline, brevicollis, and limited cervical range of motion, which is due to the presence of fused cervical vertebrae.[1,2] However, studies have found that up to half of patients with KFS may not present with such physical findings.[3] Conversely, the development of adjacent segment disease has been associated with KFS due to improper vertebral segmentation, leading to altered spinal mechanics. This results in segmental hypermobility and instability as well as neurological compromise.[4-9] Due to the inconsistencies of these associated pathologies, congenital

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Access this article online	
Website: www.jcvjs.com	Quick Response Code
DOI: 10.4103/jcvjs.JCVJS_65_19	

How to cite this article: Zhou PL, Poorman GW, Wang C, Pierce KE, Bortz CA, Alas H, *et al.* Klippel–Feil: A constellation of diagnoses, a contemporary presentation, and recent national trends. J Craniovert Jun Spine 2019;10:133-8.

fusion of the cervical spine remains the most constant trademark of KFS.

Restricted mobility secondary to osseous and soft-tissue restrictions, often associated with KFS, has been shown to result in excessive motion and shear stress in the nonfused segments, which has the further potential to clinically manifest as degenerative disc disease later in life.[9] As such, when indicated, operative management strategies have included arthrodesis and stabilization of the cervical spine.[10] Other surgical treatment options have been attempted, including cervical disc replacement, in order to restore motion as well as to potentially addressing concerns over further loss of motion and development of adjacent segment disease.[11-13]

While KFS is limited to the cervical spine, congenital fusions of the spine can occur elsewhere in the spine and can oftentimes co-occur with hemivertebrae in the case of incomplete formation or block vertebrae or unilateral bars in the case of failure of segmentation.[14,15] Frequently, these congenital vertebral anomalies can result in the development of coronal or sagittal malalignment due to asymmetrical growth. Contrarily, these anomalies may also remain clinically undetected as minimal to no deformity may develop, especially in the case of block vertebrae.[14]

There is much in the literature that explores associated risk factors and the prognosis of KFS; however, none have investigated the long-term trends in operative management and complications in such patients.[4,6,8] Furthermore, the true incidence of KFS syndrome has yet to be properly assessed and may be due to variability between patient populations.[16] Accordingly, the purpose of this study is to determine define the incidence, characterize concurrent diagnoses, and examine trends in the presentation and management of pediatric patients given a diagnosis of KFS. Similarly, we aimed to assess the same trends for those diagnosed with congenital fusions in relation to those with KFS.

METHODS

Data source

The Kid's Inpatient Database (KID) is the largest publicly available all-payer pediatric (age <21 at admission) health-care database in the United States. This database is created under the Agency for Healthcare Research and Quality's Healthcare Cost and Utilization Project (HCUP). KID sampling includes complicated and uncomplicated births as well as other pediatric inpatient procedures from community and nonrehabilitation hospitals. The KID database contains 107 data elements, with diagnoses and procedures in the International Classification of Disease, Ninth Revision, Clinical Modification (ICD-9) format. With over 12 million patients from 2003 to 2012, the database is designed to allow accurate calculation of medical condition incidences using HCUP-provided trend weights. A detailed overview of the KID design is available at (https://www.hcup-us.ahrq.gov/kidoverview.jsp). Given the deidentified nature of HCUP's KID, International Review Board approval is not needed for this study.

Study design

This is a retrospective review of charge-based KID from the years 2003, 2006, 2009, to 2012. KID supplied hospital- and year-adjusted weights allowed for accurate assessment of the national incidence of both KFS and congenital fusions. Patients aged 0–20 years were identified by ICD-9 coding and placed into two groups: Klippel–Feil patients (KFS; ICD-9 code 756.16) and congenital fusion patients (ICD-9 code 756.15). Incidences of both KFS and congenital fusion were established using KID-supplied year- and hospital-trend weights. Demographics and secondary diagnoses commonly associated with KFS were evaluated in both KFS and congenital fusion patients to assess for the prevalence. Comorbidities, anomalies, and procedure type from the years 2003 to 2012 were identified and assessed to determine trends.

Statistical analysis

Statistical analysis was performed using SPSS version 23.0 (IBM, Armonk, NY, USA). The prevalence of concurrent comorbidities and secondary diagnoses in both KFS and congenital fusion patients were analyzed by calculating the weighted incidence of concurrent diagnoses based on corresponding ICD-9 codes. These were later compared between groups KFS and congenital fusion groups using Pearson's Chi-square test of independence. Trends for surgical factors including approach, use of osteotomies, use of decompression, and other surgical details were plotted and assessed by mapping out their incidence over the years.

RESULTS

Overview of study population

A total of 12,718,381 pediatric patients were included, which represented 29,013,399 discharges after applying discharge level weighting. We identified an incidence rate for KFS of 1/21,587 discharges and 1/39,799 for congenital fusions affecting the cervical spine. Eight hundred and fifty-eight discharges with any diagnosis of KFS (mean age: 9.49 years; 51.1% females) and 475 patients diagnosed congenital fusions (mean age: 8.33 years; 50.3% females) were identified. Demographics are shown in Table 1. The rate of diagnosis for KFS increased every year from 1/28,279 discharges in 2003 to 1/16,942 in 2012 (66.9% increase). The diagnosis for congenital

fusion remained relatively stable from 2003 to 2009 (1/46,599 discharges and 1/41,174, respectively) but saw a drastic increase in 2012 (1/31,939 discharges). KFS patients had significantly shorter hospital lengths of stay (6.05 days) as compared to congenital fusion patients (7.56 days; $P = 0.003$). However, KFS patients had significantly higher Charlson Comorbidity Index scores (KFS: 0.43 vs. congenital fusion: 0.30; $P < 0.001$).

Concurrent spinal abnormalities

Overall, KFS patients were significantly less likely to be diagnosed with all congenital anomalies as compared to congenital fusion patients (36.5% vs. 48.2%; $P < 0.001$). Both KFS and congenital fusions had similar incidences of spina bifida (1.30% and 0.88%, respectively; $P = 0.417$), tethered cord (6.05% and 4.91%, respectively; $P = 0.296$), and diastematomyelia (0.44% and 0.25%, respectively; $P = 0.532$). KFS patients had a significantly higher rate of concomitant Arnold–Chiari malformations (6.22% vs. 2.79%; $P < 0.001$). Overall, KFS patients presented with significantly lower additional spinal abnormalities (hemivertebra, spina bifida, congenital spondylolisthesis, tethered cord, Arnold–Chiari malformation, diastematomyelia, and missing vertebra) than congenital fusion patients (19.06% vs. 24.79%; $P = 0.002$).

Muscular abnormalities

Both KFS and congenital fusion patients had similar incidences of any concurrent neuromuscular abnormality (34.0% and 33.7%, respectively; $P = 0.624$). However, 6.36% of KFS were also diagnosed with Sprengel's deformity, which was significantly greater than the 0.41% diagnosed in those with congenital fusions ($P < 0.001$). Congenital torticollis was found to be similar in both groups (1.50% and 1.27%, respectively; $P = 0.609$). Muscular dystrophy was found to be very rare in both KFS (0.59%) and congenital fusion (0.22%) patients, but incidences remained similar ($P = 0.249$) [Table 2].

Neurological abnormalities

Overall, KFS and congenital fusion patients had similar incidences of a co-occurring neurological diagnosis (5.68% vs. 5.31%, respectively; $P = 0.395$). KFS patients had higher incidence rates of Stilling–Türk–Duane syndrome (1.63% vs. 0.00%; $P < 0.001$), which is a congenital strabismus of the eyes that is often found in those with KFS.[17] The incidence of cerebral palsy was found to be similar between both groups of patients (KFS 2.81% vs. congenital fusion 2.76%; $P = 0.492$).

Other abnormalities

About 36.51% of KFS patients were diagnosed with a nonspinal or nonmusculoskeletal anomaly, with the most prevalent anomalies being of cardiac origin (12.95%) followed by urinary anomalies (10.37%) and those of the nervous system (5.68%). Those diagnosed with congenital fusions had significantly greater rates or cardiac (23.95%; $P < 0.001$), gastrointestinal

Table 1: Demographics of those with Klippel-Feil syndrome and congenital fusions

	KFS (%)	Congenital fusion (%)	*P*
CCI	0.4257	0.2998	<0.001
Any other congenital anomaly	36.51	48.89	<0.001
Any neuromuscular anomaly	2.93	3.37	0.624
Age in years at admission	9.46	8.17	<0.001
Female	51.15	49.76	0.52
Male	48.75	50.05	0.544
Caucasian	46.84	48.43	0.495
African American	9.72	6.10	0.003
Asian	16.42	17.14	0.654
Other	27.02	28.34	0.555

KPS – Klippel–Feil syndrome; CCI - Charlson comorbidity index

Table 2: Percentage of patients with concomitant congenital anomalies in Klippel-Feil syndrome and congenital fusion patients

Congenital anomaly	KFS (%)	Congenital fusion (%)	*P*
Sprengel's deformity	6.36	0.41	<0.001
Tetraparesis	0.53	0.00	0.046
Spina bifida	1.30	0.88	0.284
Hirschsprung's disease	0.23	0.18	0.554
Stilling-Türk-duane	1.63	0.00	<0.001
Multiple hemangiomas	0.45	0.18	0.225
Torticollis	1.50	1.27	0.383
Cervical dislocation	0.76	0.69	0.547
Congenital spondylolisthesis	0.21	1.75	<0.001
Tethered cord	6.05	4.91	0.173
Arnold-Chiari malformation	6.22	2.79	<0.001
Diastematomyelia	0.44	0.25	0.415
Other orthopedic	0.12	1.23	0.002
Cardiac	12.95	23.95	<0.001
Gastrointestinal	3.60	15.63	<0.001
Genital	1.00	6.20	<0.001
Urinary	10.37	15.13	0.001
Nervous system	5.68	5.31	0.395
Ophthalmalogical	1.04	2.12	0.053
Ear	2.33	4.88	<0.001
Neck	4.30	7.26	0.003
Pulmonary	1.72	3.31	0.014
Endocrine system	0.12	0.00	0.415
Any other congenital anomaly	36.51	48.89	<0.001
Muscular dystrophy	0.59	0.22	0.249
Cerebral palsy	2.81	2.76	0.492
Hemivertebra	3.33	6.42	<0.001
Klippel-Feil syndrome	100.00	4.49	<0.001
Congenital fusion	1.60	100.00	<0.001

KPS – Klippel–Feil syndrome

(15.63%; $P < 0.001$), genital (6.20%; $P < 0.001$), and urinary anomalies (15.13%; $P = 0.021$).

Surgical management

From 2003 to 2012, 7.34% of patients with KFS underwent anterior fusions, whereas 6.64% of KFS patients underwent posterior fusions [Figure 1]. The average number of levels operated upon was 4.99 with 8.28% receiving decompressions. Interbody devices were used in 2.45% of cases [Figure 2]. The rate of fusions with <3 levels (7.46%) was found to be comparable to that of 3 levels or greater (7.81%).

In congenital fusion patients, the rate of anterior and posterior fusions was marginally increased as compared to KFS patients with 8.00% of patients undergoing anterior fusion and 8.63% undergoing posterior fusion [Figure 3]. Both the number of levels operated upon and the percentage of those receiving decompressions were similar to KFS patients at 4.56% and 8.21%, respectively [Figure 4]. Bone morphogenetic protein was used more prevalently in congenital fusion patients (3.58%) than in KFS patients (1.52%).

DISCUSSION

While KFS is often described as the clinical triad of brevicollis, low posterior hairline, and limited range of motion in the neck secondary to congenitally fused cervical segments, diagnoses are often made without the presence of these featuring characteristics, as there is a spectrum of anomalies that can concomitantly present.[3,18] As a result, it remains unclear, whether KFS is a single pathological process or one of many in a spectrum of congenital spinal abnormalities.[18] This uncertainty may present as disparities in the diagnosis of patients with fused vertebral segments. This study aims to elucidate the discrepancy in those diagnosed with KFS and those with congenital fusions as well as determine trends in both presentation and management of patients with KFS.

We found an incidence rate for KFS of approximately 1/21,500 discharges, whereas congenital fusions were found to occur in approximately 1/40,000. Previous reports have estimated the incidence rate of KFS to be approximately 1 in 40,000–42,000 births.[18] The true prevalence of KFS is believed to be higher than previous reports due heterogeneity in clinical presentation leading to missed diagnoses.[16,18,19] In addition, some patients who have stable fusion patterns may never become symptomatic and may live their lives unaware. The discrepancy between our reported rate and the literature is due in part to previous literature detailing incidences per birth, while our numbers describe those diagnosed after hospital discharges. Our reported numbers may overestimate

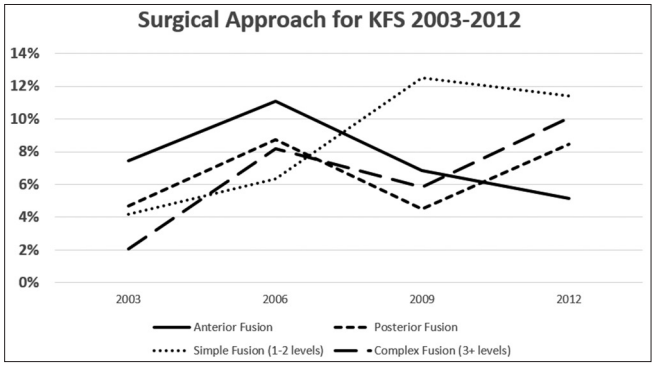

Figure 1: Surgical approach for KFS from 2003 to 2012

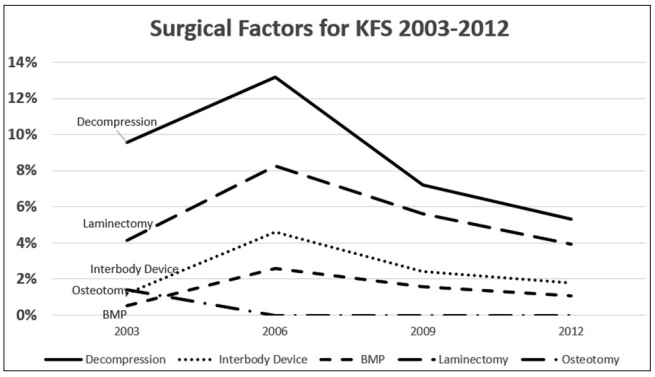

Figure 2: Surgical factors for KFS from 2003 to 2012

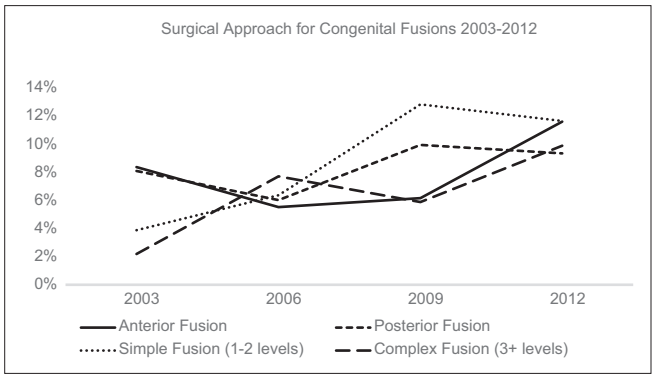

Figure 3: Surgical approach for congenital fusions from 2003-2012

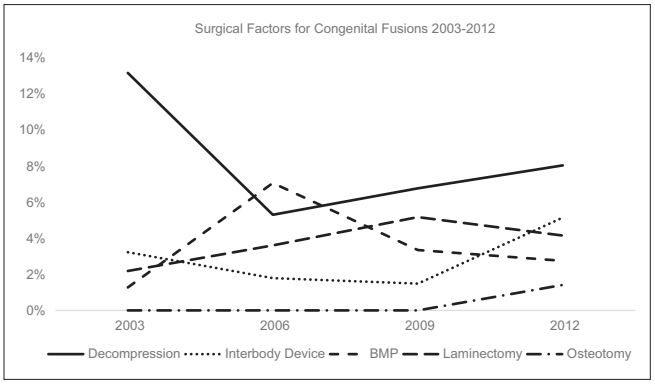

Figure 4: Surgical factors for congenital fusions from 2003-2012

the true incidence of KFS, however, due to the nature of KFS and the possibility of going undiagnosed at birth, our study accounts for those undiagnosed at that time period and may reflect a more reasonable estimate of the true incidence in a nonneonatal population.

Besides cervical pathologies, patients with KFS often have osseous and soft-tissue anomalies located in other parts of the spine as well as other musculoskeletal deformities.[7,20,21] Previous studies have reported a high association of Sprengel's deformity in KFS patients, with rates ranging from 7% to 42% of cases of KFS.[3,7,22] We identified a concomitant rate of 6.18%, which is in line with the lower range of incidences reported in the literature.[22] Aside from Sprengel's deformity and Stilling–Türk–Duane Syndrome, a rare congenital pathology characterized by the lack of eye abduction that is commonly associated with KFS, both KFS and congenital fusion patients had similar rates of other neuromuscular deformities.[17] Overall, KFS patients had a high incidence rate of having another concurrent spinal anomaly at 18.88%. A number of genetic mutations implicated in the formation of bones have been identified in the development of KFS, which may in part explain the overall high rate of coinciding spinal anomalies observed.[18,23] In comparison, both KFS and congenital fusion patients had similar rates of additional spinal anomalies. Furthermore, given the similar pathology of both diagnoses and the wide spectrum in which both congenital anomalies may present, it is possible that KFS is a single manifestation in a wide spectrum of associated congenital spinal deformities.[18,23]

While musculoskeletal anomalies were similar between KFS and congenital fusion patients, anomalies of the other organ systems were significantly greater in congenital fusion patients. Hensinger *et al*. identified a 14% incidence rate of congenital cardiac anomalies in KFS patients, similar to our finding of 12.95%.[3] Overall, our study population had a high rate of anomalies of other organ systems, which may be attributed to the vertebral anomalies, anal atresia, cardiac malformations, tracheoesophageal fistula, renal abnormalities, and limb deformities and other congenital anomaly associations.[24] While it remains unknown why these congenital anomalies often appear together, it is possible that the mesodermal origin of all these tissues may play a role.[25,26]

In both KFS and congenital fusion patients, we saw an overall increase in the percentage of patients who underwent surgical fusion as a treatment option from 2003 to 2012. Interestingly, we saw a decrease in the amount of decompressions performed for KFS in the same time period. Indications for surgical management of KFS include persistent pain refractory to medical therapy as well as neurological deficits, with the overall goal being increased stability of the cervical spine. Theiss *et al*. followed 32 patients over the course of >10 years and found that only 2 (6.25%) required surgery for cervical instability in pain.[27] These numbers are comparable to ours for 3 or greater levels but are less than our findings for 1–2 level fusions, which have tripled over the study period from 3.87% to 11.63%. In a more recent study by Samartzis *et al*., 11% of patients required operative management for neurological symptoms.[8] It should be noted that the majority of patients who require surgical intervention may not be captured in our pediatric cohort as oftentimes, patients may undergo surgery in the second or third decade of life or even later.[28]

We appreciate certain limitations to this study including its retrospective nature and the use of a nationwide database. While providing data on a large scale, the use of CCS coding often lacks granularity. ICD-9 codes used can have general diagnosis codes that encompass several pathologies. Furthermore, as a discharge-level database, KID which may exaggerate the incidence of some pathologies as some patients may be accounted for multiple times due to multiple discharges.

CONCLUSIONS

KF is a relatively uncommon disorder associated with a plethora of congenital anomalies. KF patients were more likely to have other spinal abnormalities (19.1%) and nonnervous system abnormalities (13.63%). Compared to congenital fusions, patients with KFS were more likely to have certain congenital abnormalities such as Sprengel's deformity. Recent years show that KF patients are increasingly being treated with spinal fusion.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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