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Mechanism for Superior Subluxation of the Glenohumeral Joint in Fibrodysplasia Ossificans Progressiva

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Progressive heterotopic ossification leads to ankylosis of the major joints in patients who have fibrodysplasia ossificans progressiva. Joint subluxation has not been recognized widely in patients with this disease. The clinical records and radiographs of 79 patients with fibrodysplasia ossificans progressiva were reviewed and, it was found that humeral to chest wall synostosis and subluxation of the glenohumeral joint had occurred in 21% of skeletally immature patients and in 74% of skeletally mature patients. In fibrodysplasia ossificans progressiva, synostosis of the humeral shaft to the chest wall commonly occurs by 7 years of age, well before the age of proximal physal closure. The continued growth of the proximal humeral physis in the presence of a humeral to chest wall synostosis causes the humeral head to migrate superiorly, thus promoting growth related subluxation. The clinical significance of this finding for patients who have fibrodysplasia ossificans progressiva is unknown, but this unique model

will be useful in the study of shoulder biomechanics and growth plate physiology.

The major clinical features of fibrodysplasia ossificans progressiva have been described.^{2-4,8,9} A colleague of the authors (verbal communication, Frederick S. Kaplan, MD, 1996) observed that many patients with fibrodysplasia ossificans progressiva who had humeral to chest wall synostosis also had superior subluxation of the glenohumeral joint. Based on this observation, the authors evaluated the prevalence of humeral to chest wall synostosis and superior subluxation of the glenohumeral joint in a large group of skeletally immature and skeletally mature patients who had fibrodysplasia ossificans progressiva.

METHODS AND MATERIALS

From 1990 to 1996, 110 patients with the diagnosis of fibrodysplasia ossificans progressiva were observed by the fibrodysplasia ossificans progressiva working group at The University of Pennsylvania Medical Center. The diagnosis of fibrodysplasia ossificans progressiva was based on the presence of congenital malformation of the great toes and progressive postnatal heterotopic ossification. Patients were divided into two groups: patients who were 18 years of age or

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younger (skeletally immature), and patients who were older than 18 years of age (skeletally mature). Available radiographs were evaluated for the presence or absence of humeral to chest wall synostosis and for the presence or absence of superior subluxation of the glenohumeral joint. The correlation between humeral to chest wall synostosis and subluxation of the glenohumeral joint was assessed using Fisher's exact test.⁶

RESULTS

Seventy-nine (72%) of the 110 patients had appropriate radiographs for review. Forty-eight (61%) of the 79 patients were skeletally immature and 31 (39%) of the 79 patients were skeletally mature. In the skeletally immature group, there were 17 boys and 31 girls. In the skeletally mature group, there were 13 males and 18 females. The mean age of the skeletally immature and skeletally mature patients was 9 years old and 33 years old, respectively.

A 3-year-old girl with fibrodysplasia ossificans progressiva had undergone a right forequarter amputation after aggressive juvenile fibromatosis was misdiagnosed. The operating surgeon had not recognized the characteristic malformation of the great toes, a cardinal clinical finding that distinguishes early fibrodysplasia ossificans progressiva from aggressive juvenile fibromatosis. The child was excluded from the study.

The condition of the left and right shoulders was symmetric in 76 (97%) of the 78 patients. Of the 47 patients who were skeletally immature, 10 (21%) patients had humeral to chest wall synostosis and superior subluxation of the glenohumeral joint, whereas 34 (72%) patients had neither humeral to chest wall synostosis nor superior subluxation of the glenohumeral joint (Fig 1; Table 1). Three (7%) of the 47 patients were discordant for the presence of humeral to chest wall synostosis and subluxation (Table 1). Humeral to chest wall synostosis developed in all three patients, but subluxation of the glenohumeral joints had not yet developed. The correlation be-

tween humeral to chest wall synostosis and superior subluxation of the glenohumeral joint in skeletally immature patients with fibrodysplasia ossificans progressiva was highly significant ($p = 6 \times 10^{-8}$).

Of the 31 patients who were skeletally mature, 23 (74%) patients had humeral to chest wall synostosis and superior subluxation of the glenohumeral joint, whereas six (19%) patients had neither humeral to chest wall synostosis nor superior subluxation of the glenohumeral joint (Fig 1; Table 1). Two (7%) of the 31 patients were discordant for the presence of humeral to chest wall synostosis and subluxation. Both patients had evidence of humeral to chest wall synostosis without evidence of subluxation of the glenohumeral joints. In both cases, humeral to chest wall synostosis occurred during late adolescence when little skeletal growth remained. The correlation between humeral to chest wall synostosis and superior subluxation of the glenohumeral joint in skeletally mature patients with fibrodysplasia ossificans progressiva was highly significant ($p = 4 \times 10^{-5}$).

Among all patients, the correlation between humeral to chest wall synostosis and superior subluxations of the glenohumeral joint was highly significant ($p = 5 \times 10^{-17}$). There was a significantly higher prevalence of humeral to chest wall synostosis ($p = 7 \times 10^{-6}$) and glenohumeral subluxation ($p = 5 \times 10^{-6}$) in patients who were skeletally mature than in those who were skeletally immature.

DISCUSSION

The major finding of this study is that superior subluxation of the glenohumeral joint is extremely common in patients who have fibrodysplasia ossificans progressiva and is highly correlated with the presence of humeral to chest wall synostosis in an age related manner. In a recent study on the natural history of fibrodysplasia ossificans progressiva, Cohen et al² showed that the



	A	B	C	D
Gender	F	F	F	F
Age (years)	12	8	42	25
Skeletal Maturity	IM	IM	MA	MA
Synostosis	+	+	+	-
Subluxation	-	+	+	-

Fig 1. Representative radiographs of the shoulder in patients who have fibrodysplasia ossificans progressiva. Panels A and B: skeletally immature (IM); Panels C and D: skeletally mature (MA). Note humeral to chest wall synostosis (+) in Panels A, B, and C; and subluxation (+) in Panels B and C. Note absence of humeral to chest wall synostosis (-) in Panel D, and absence of superior subluxation of the humeral head (-) in Panels A and D. F = female.

mean age of onset of heterotopic ossification at the glenohumeral joint was 7 years of age, well before the time of proximal humeral physal closure which generally occurs at 14 to 17 years of age in girls and 15 to 18 years of age in boys. Once synostosis has occurred between the chest wall and the proximal humerus, superior migration of the humeral head likely progresses secondary to continued growth at the proximal humeral physis until the time of skeletal maturity.⁴ The significant difference in the prevalence rate of humeral to chest wall synostosis and superior subluxation of the glenohumeral joint between the skeletally mature and skeletally immature patients most likely reflects the fact that many of the patients in the skeletally immature group have not yet formed the osseous bridge re-

quired for subsequent growth related subluxation.

There are several major limitations of this study: First, radiographs were not available on 31 of the 110 patients. Second, radiographs were not specifically obtained to quantify the extent of subluxation. The presence of subluxation was noted incidentally on radiographs obtained for other reasons, most commonly chest radiographs or scoliosis studies. Third, only a few patients had a series of radiographs in which to follow the temporal evolution of glenohumeral subluxation in a longitudinal fashion. Despite these limitations, the data suggest that a growth related mechanism likely exists for superior subluxation of the glenohumeral joint in patients who have fibrodysplasia ossificans progressiva.

TABLE 1. Humeral to Chest Wall Synostosis and Superior Subluxation of the Glenohumeral Joint in Patients With Fibrodysplasia Ossificans Progressiva

Diagnosis	Immature	Mature	Total
+ Synostosis + subluxation	10 (21%)	23 (74%)	33 (42%)
- Synostosis - subluxation	34 (72%)	6 (19%)	40 (51%)
+ Synostosis - subluxation	3 (7%)	2 (7%)	5 (7%)
- Synostosis + subluxation	0 (0%)	0 (0%)	0 (0%)
Total	47	31	78

Glenohumeral subluxation in patients with fibrodysplasia ossificans progressiva provides unique insight into the competence of soft tissue restraints about the shoulder joint. The Hueter-Volkman law states that pressures parallel to the axis of physal growth affect the rate of growth.¹ More specifically, compressive forces across the physis, if large enough, can inhibit growth of the physis. Although there are numerous examples of the Hueter-Volkman law, the exact amount of compressive force required to inhibit physal growth remains unknown. The observation of superior subluxation of the glenohumeral joint in patients with fibrodysplasia ossificans progressiva shows that the soft tissue structures about the shoulder do not provide sufficient resistance to inhibit the growth of the proximal humeral physis once ankylosis of the humeral shaft to the chest wall has occurred.

Superior subluxation of the glenohumeral joint, although rare in the general population,^{5,7} is a common finding in patients who have fibrodysplasia ossificans progressiva. This phenomenon occurs in patients who have synostosis of the humeral shaft to the chest wall develop in the presence of an open proximal humeral physis. Continued growth of the proximal humeral physis causes superior migration of the humeral head relative to the glenoid fossa, resulting in superior glenohumeral subluxation. Although there is currently no effective treatment available for patients with fibrodysplasia ossificans progressiva, this novel growth related mecha-

nism for superior glenohumeral subluxation provides useful information on shoulder biomechanics and growth plate physiology that may be beneficial to patients who have more common afflictions of the musculoskeletal system.

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