Primary glenoid dysplasia

A REVIEW OF 12 PATIENTS

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We reviewed 12 patients with primary glenoid dysplasia. Ten were assessed clinically and two from case notes and radiographs. We identified two groups according to the age at onset of symptoms. The first (seven patients) consisted of boys and younger men, all of whom developed symptoms before the age of 40 years. All four children were free from pain, whereas the three adults in this group had varying degrees of this. Four patients had symptoms of instability. The second group consisted of older men (five patients) all of whom had noted the onset of symptoms, in the form of pain and stiffness, after the age of 40 years. All five had radiological evidence of osteoarthritis.

Although the four children in our study had minimal symptoms, all eight adults had ongoing shoulder pain and dysfunction, despite a specific rehabilitation programme. Four patients required surgery; one had posterior stabilisation for instability and three arthroplasties of the shoulder for osteoarthritis.

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The terms congenital and primary glenoid dysplasia refer to an uncommon condition characterised by incomplete ossification of the lower two-thirds of the cartilaginous glenoid and adjacent neck of the scapula.¹ Since it was first described by Giongo² and Heupke,³ it has been reported in fewer than 80 patients.¹⁻²⁷ Usually a primary isolated lesion, it can be seen in association with other anomalies, or as part of well-described syndromes, and has recently been categorised into five specific groups.¹

Previous reports have observed glenoid dysplasia to present with a variety of symptoms and variable radiological findings, ranging from an asymptomatic incidental finding on a chest radiograph^{11,12} to shoulder pain and dysfunction.^{4,6-9,13-18,20-22}

We report our observations in 12 patients with primary glenoid dysplasia who form two groups based on their age at the onset of symptoms. In some cases the condition is much more disabling than has been previously suggested, and may require surgery.

Patients and Methods

Of 2500 patients with shoulder symptoms referred to the senior author (TDB) over a period of eight years, 12 had a diagnosis of primary glenoid dysplasia. Ten were reviewed clinically and radiologically in a special clinic, and the remaining two were assessed from their case notes and radiographs. All were men and had a mean age of 40 years (12 to 69). All had bilateral radiological involvement.

The clinical review recorded the original timing and mode of presentation, past medical history, details of birth and the family history. A full assessment was carried out according to the method of the American Shoulder and Elbow Surgeons (ASES)²⁸ and the Constant and Murley²⁹ score. The patients were examined for evidence of instability (posterior jerk test, anterior apprehension test, sulcus sign, anterior/posterior translation), posterior dimpling and axillary webs. The index of Beighton, Grahame and Bird³⁰ was used as an indication of generalised ligamentous laxity and both hips were examined. All patients had normal hips on clinical assessment and radiographs were therefore not taken. We felt that a full skeletal survey was not justified in the absence of any other joint problems.

Anteroposterior (AP) and axillary radiographs were reviewed to assess varus angulation of the neck of the humerus, enlargement of the acromion or coracoid, clavicular bossing (hooking), glenoid irregularity (a 'dentate' glenoid), variation in the glenohumeral joint space (superiorly v inferiorly), osteoarthritis and posterior subluxation of the humeral head as seen on axillary views.

Eight patients were managed conservatively, by specific physiotherapy. The symptoms in the other four were severe enough to warrant surgery.

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Table I. Details and grouping of the 12 patients with primary glenoid dysplasia

Case	Age (yr)	Occupation	ASES ADL score /30		Constant score /100		Restriction of movement	Age at onset of symptoms (yrs)	Main symptoms	Treatment
			R	L	R	L				
Group 1										
1	12	Schoolboy	29	29	94	94	No	6	Clicking and instability	Physiotherapy
2	13	Schoolboy	27	27	97	97	Yes	8	Limitation movement	Physiotherapy
3	22	-	-	-	-	-	No	14	Clicking and instability	Physiotherapy
4	24	Office worker	18	6	44	24	Yes	18	Pain, instability and limitation movement	Surgery: left posterior bone block and capsular shift
5	24	Stockbroker	-	-	-	-	No	16	Clicking and instability	Physiotherapy
6	49	Shop manager	19	14	77	69	Yes	29	Pain, instability and limitation movement	Physiotherapy
7	53	Hairdresser	30	9	100	67	R = No L = Yes	33	Pain and limitation movement	Physiotherapy
Group 2										
8	48	Anaesthetist	19	19	77	84	Yes	46	Pain and clicking	Physiotherapy
9	48	Unemployed	13	11	63	54	Yes	41	Pain and limitation movement	Left shoulder hemiarthroplasty
10	60	Lorry driver	21	21	84	80	Yes	50	Pain and limitation movement	Physiotherapy
11	69	Union official	22	5	79	20	Yes	54	Pain and limitation movement	Left shoulder hemiarthroplasty
12	60	Builder	8	24	-	-	Yes	55	Pain and limitation movement	Right shoulder hemiarthroplasty

Results

We divided the patients into two groups according to the age at onset of symptoms (Table I).

Group 1: onset of symptoms at less than 40 years of age (7 patients). In four patients the onset of symptoms occurred before adulthood (at 6, 8, 14 and 16 years). Three of these children presented with clicking and minor symptoms of instability, and one (case 2) simply as a result of problems with swimming backstroke, caused by a previously unnoticed difficulty with forward elevation of the shoulder (Fig. 1). All four children were free from pain and had excellent function at review, after physiotherapy.

A further three patients in this group noted the onset of symptoms between the ages of 18 and 33 years. Two developed symptoms after an injury playing rugby and subsequently had symptoms in the other shoulder, although to a less degree. All three had pain and reduced activities of daily living (ADL) scores and ASES assessment. At review, two had improved after physiotherapy, but remained in pain with some functional disability. One underwent a posterior

glenoid augmentation procedure and capsulorrhaphy for painful instability. Three years after surgery, there was very little change in his subjective assessment of pain, ADL and instability on ASES assessment.

Although two patients (cases 5 and 7) had unilateral symptoms, both shoulders showed radiological evidence of glenoid dysplasia.

Group 2: onset of symptoms at more than 40 years of age (5 patients). All five patients who developed symptoms after the age of 40 years had evidence of osteoarthritis on plain radiographs. In four, there was severe osteoarthritis with subchondral sclerosis, osteophytosis, formation of cysts, and complete loss of joint space; the remaining patient had less marked changes. The mean age at onset of symptoms was 50 years (41 to 55) with a mean duration of symptoms at review of 7.8 years (2 to 15). All five patients had pain and limitation of movement, and the symptoms were bilateral, although the pattern was asymmetrical. This group did not respond well to physiotherapy and at review four required regular daily analgesics. All had a marked functional deficit as recorded by a mean ADL

Fig. 1

Case 2. Radiograph showing primary glenoid dysplasia.

score and ASES assessment of 16/30 (5 to 24) and a mean Constant score of 68 (20 to 84). The progression of one patient (case 9) to severe osteoarthritis over a period of six years is demonstrated in Figure 2, and was bilateral.

Three of these patients required shoulder arthroplasty (Fig. 3). This was technically difficult and the results relatively disappointing compared with standard procedures for primary osteoarthritis, principally because of the lack of glenoid bone stock, which made access and the insertion of a glenoid component difficult. At review, the mean post-operative ADL score on ASES assessment was 6.5/30 with mean pain ratings on the visual analogue score of 5/10.

General. All patients had normal hips on clinical assessment and although four complained of symptoms of shoulder instability, there was no evidence of a sulcus sign, positive posterior jerk test or anterior apprehension. There was no evidence of generalised ligamentous laxity as shown by a normal Beighton's index. One patient had been delivered by caesarean section; the remainder had had normal births. There was no evidence that the glenoid dysplasia was part of an underlying syndrome in any of the patients who were all generally fit and well, although one (case 4) had a history of a cardiac septal defect. Two patients had first-degree relatives who had had previous problems with their shoulder, the nature of which was undetermined. None had evidence of posterior dimpling or axillary webs.

Radiological. The AP and axillary view radiographs were examined for specific features of glenoid dysplasia. Flattening and hypoplasia of the head of the humerus have been described,^{12,15,21,22} but were not seen in our patients. Varus angulation of the head^{7,25} was not a feature. Four of our patients had humeral neck angles of less than 55° (normal male neck angle 60°³¹) and two had angles of more than 65°.

Hooking or 'bossing' of the distal clavicle is well described in the literature^{16,21,25} and was a prominent feature in nine of our patients. None had enlargement of the acro-

Fig. 2

Case 9. Radiographs of a 48-year-old man showing progression to severe osteoarthritis with bilateral involvement.

Fig. 3a

Fig. 3b

Case 11. Radiographs of a 69-year-old man with bilateral involvement showing a) the right shoulder before and b) the left shoulder after arthroplasty.

mion,^{6,12,21,25} but six had a prominent coracoid process.^{6,12,14,21,22} All patients had a 'dentate', irregular appearance of the glenoid^{1,21,25} although the four with marked osteoarthritis were excluded. Retroversion of the glenoid was seen. Although we recognise that measurement of retroversion is relatively inaccurate on plain radiographs, our measured mean angle of retroversion was 58° (40 to 80).

All patients except the four with osteoarthritis had a relative increase in the inferior when compared with the superior joint space.¹ Five patients showed features of osteoarthritis. There was no radiological evidence of posterior subluxation, although this has been previously reported.^{22,24}

Discussion

In 1981 Pettersson¹⁵ suggested that glenoid dysplasia may be more common than had been previously thought and this view has been reiterated since.^{12,16} Our experience would support this. There may be minimal symptoms or the condition may be asymptomatic,^{11,12,15,19} making under-diagnosis inevitable.

The aetiology and inheritance are poorly understood. The pathogenesis appears to be a failure of ossification of the inferior glenoid precartilage.¹ Previous theories concerning a failure of development of the precartilage of the inferior apophysis of the glenoid¹⁴ are not supported by the findings on CT arthrography,^{24,26} plain arthrography^{15,25} and arthroscopy,^{16,21,25} which show that the inferior glenoid precartilage is in fact present, but unossified. The outline of the glenohumeral joint lines on radiographs demonstrating the 'vacuum phenomenon',¹ also supports the view that the radiological glenoid 'deficiency' comprises unossified

cartilage. The underlying cause of this failure of ossification is not established. There appears to be a familial pattern in a small number of cases,^{12,15,27} but in most patients there is no definite family history. A number of conditions may be associated with a similar deformity^{15,32} including Erb's palsy, muscular dystrophy, aseptic osteochondritis, avitaminosis D, avitaminosis C, haemophilic arthropathy and neonatal septic arthritis.¹² Such cases are often unilateral and should easily be excluded on clinical and radiological grounds. Other secondary causes have been described by Currarino et al¹ who divided glenoid dysplasia into five groups according to the underlying condition. The likelihood is that if there is a specific genetic abnormality accounting for primary glenoid dysplasia, it has variable penetrance and may arise as a spontaneous mutation.

The clinical findings in patients with primary glenoid dysplasia may be very varied, from severe pain and disability to no symptoms at all. Most children are asymptomatic^{1,12} and relatively few symptomatic children have been described.^{6,11,12,14,21,27} Children are most likely to be diagnosed incidentally, either from a chest radiograph or after investigation for a minor complaint such as painless clicking or mild instability. Younger adults also present with variable symptoms. Most complain of pain, often with some associated limitation of movement, and symptoms of instability are common.^{12,21,22} True dislocation of the shoulder is unusual and there was a paucity of signs to account for the symptoms of instability, and no evidence of generalised ligamentous laxity. The premature development of osteoarthritis has been described^{15,16,21} and this was the predominant feature in five patients who were older than those with pain and symptoms of instability only. They have similar disability to patients with primary osteo-

arthritis and may require arthroplasty. All our patients had evidence of glenoid dysplasia bilaterally, although the symptoms were not always symmetrical. Only five cases of unilateral glenoid dysplasia have been reported.^{21,22} An association with dysplasia of the hip has been observed,²² but we found no clinical evidence of hip problems.

Previous reports have suggested that in most patients the symptoms resolve after physiotherapy.²¹ This was the case in our younger patients, but with increasing age the symptoms were generally more disabling and the results of physiotherapy less good. One of our patients required surgery for painful, disabling posterior instability and three patients had a shoulder arthroplasty for osteoarthritis. Although we were unable to find previous reports in which shoulder arthroplasty had been undertaken in patients with glenoid dysplasia, Wirth et al²¹ stated that they thought that it may have a role in patients with "unremitting symptoms and moderate or severe degenerative changes of the shoulder joint". Surgery in such patients is technically difficult and the results are less rewarding than those for primary osteoarthritis.

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