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Unicameral bone cysts

Natural history and the risk of fracture

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Summary. *Unicameral bone cysts in 57 patients, seen at the Alfred I. duPont Institute between 1949 and 1982, were studied retrospectively. We present the natural history in several patients and report a "cyst index" which allows the progress of the cyst to be followed and makes it possible to determine whether there is a significant risk of fracture which would warrant treatment. We have found that measuring the cyst index in patients treated by injections of methylprednisolone acetate helps to assess the response to treatment.*

Résumé. *Nous avons réalisé une étude rétrospective des 57 cas de kyste essentiel des os, observés à l'Institut Alfred I. duPont de 1949 à 1982. Nous relatons l'histoire naturelle de cette affection et nous présentons un "index kystique" qui permet de suivre l'évolution du kyste et rend possible de prévoir la survenue d'un risque important de fracture, pouvant justifier un traitement. Nous avons trouvé que la mesure de cet index, chez les malades traités par injections d'acétate de méthyl-prednisolone, aide à évaluer la réponse à cette thérapeutique.*

Introduction

Unicameral cyst is the commonest benign bone lesion in childhood [12]. It has been a well recognised for at least 80 years [23], and has been known by a number of names such as a benign bone cyst, a juvenile unicameral bone cyst, and a simple bone cyst. Several aetiological theories have been proposed including intraosseous haemorrhage, involution of a giant cell tumour, growth disturbance [11, 23], inflammation [6], blockage of interstitial fluid circulation [6, 18], and disturbance of venous drainage [5]. None of these theories has been proved and the precise cause of these cysts is not known.

In 1942, Jaffe and Lichtenstein described the radiological, pathological and histological appearances, and defined active and latent phases of evolution which were dependent on the proximity of the cyst to the growth plate [11]. The results of treatment were better in latent cysts, i.e. those not located next to the physis. Garceau and Gregory related results to the age of the patient, and indicated that recurrence was more common under the age of ten years [9]. Neer et al. stressed that the size and site of the cyst were the most important factors [17].

The classical treatment is curettage of the lining membrane and grafting with homogenous or autogenous bone, but this has been associated with a high risk of recurrence [2, 7, 9, 17, 21]. Many attempts have been made to reduce this risk by improving operative methods. Spence et al. emphasised the importance of the graft material and surgical technique [21]. They showed that packing the cyst completely with freeze-dried, crushed cortical bone allograft, or with fresh allograft, decreased the risk of recurrence. Others have demonstrated that the more extensive the resection, the lower the risk [7, 10, 13, 15].

Radical resection, or diaphysectomy, has a recurrence rate of from 0% to 10%. This method is seldom used for femoral lesions since there is a risk of subsequent growth disorder because the cyst is close to the epiphyseal plate, and there are

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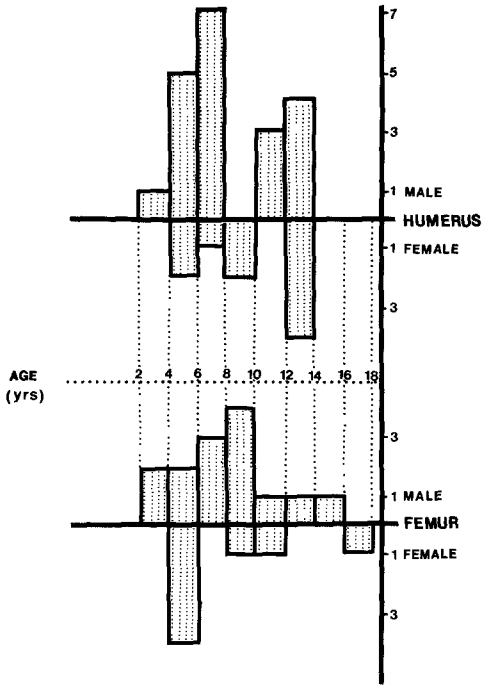

Fig. 1. Histograms showing the distribution of patients by age at diagnosis, indicating the child's sex and the site of the cyst

difficulties in stabilising the proximal fragment in a growing child.

In 1974, Scaglietti et al. first described the treatment of unicameral bone cysts by injection with methylprednisolone acetate, and since that time both curettage and bone grafting, and injection, have been used with varying degrees of success [4, 7, 19].

In spite of numerous reports of the results of treatment, little is known about the natural history of these cysts. Most authors have failed to find evidence of spontaneous healing and so have chosen active treatment [3, 11, 19]. Since the cysts are benign, the only problem is the risk of pathological fracture, but so far no reliable means has been devised for predicting the risk of this complication, or for assessing the indications for treatment.

Neer's [17] and Baker's [2] classification do not allow full assessment of the cyst because these methods were developed only for determining the results of operative treatment.

Based on a retrospective study, we have devised a "cyst index" which makes it possible to follow the progress of a cyst and determine whether there is a significant risk of fracture warranting treatment. We also report on the natural history in several patients who were observed for some time before the cyst filled in spontaneously.

Table 1. Treatment according to site of cyst

Curettage and graft			
	Site		
No. of procedures	Humerus	Femur	Other
1	4	7	3
2	4	2	0
3	1	3	0
No. of patients	9	12	3
Total no. of patients 24			
Total no. of procedures 38 = 1.58 procedures/patient			

Methylprednisolone acetate injection			
	Site		
No. of injections	Humerus	Femur	Other
1	3	2	2
2	1	1	0
No. of patients	4	3	2
Total no. of patients 9			
Total no. of injections 11 = 1.2 injections/patient			

Operation and injections			
	Site		
	Humerus	Femur	Other
No. of operations/injections	1/2	1/2	
No. of patients	1	2	
Total no. of patients	3		

Observation (no treatment)			
	Site		
	Humerus	Femur	Other
	17	3	0

Summary			
	Site		
	Humerus	Femur	Other
	31	21	5
Total no. of patients 57			
Total no. of procedures 61 = 1.07 procedures/patient			

Clinical material

Between 1949 and 1982, 57 patients, with 58 bone cysts, were treated at the Alfred I. duPont Institute. The number and site of the cysts was proximal humerus 31, proximal femur 19, distal femur 2, proximal tibia 2, distal tibia 2, proximal fibula 1, and distal fibula 1.

Boys were affected more commonly than girls (ratio 2:1). The average age at diagnosis was 8.3 years (range 3.2–17 years), but the distribution in relation to sex and site varied (Fig. 1).

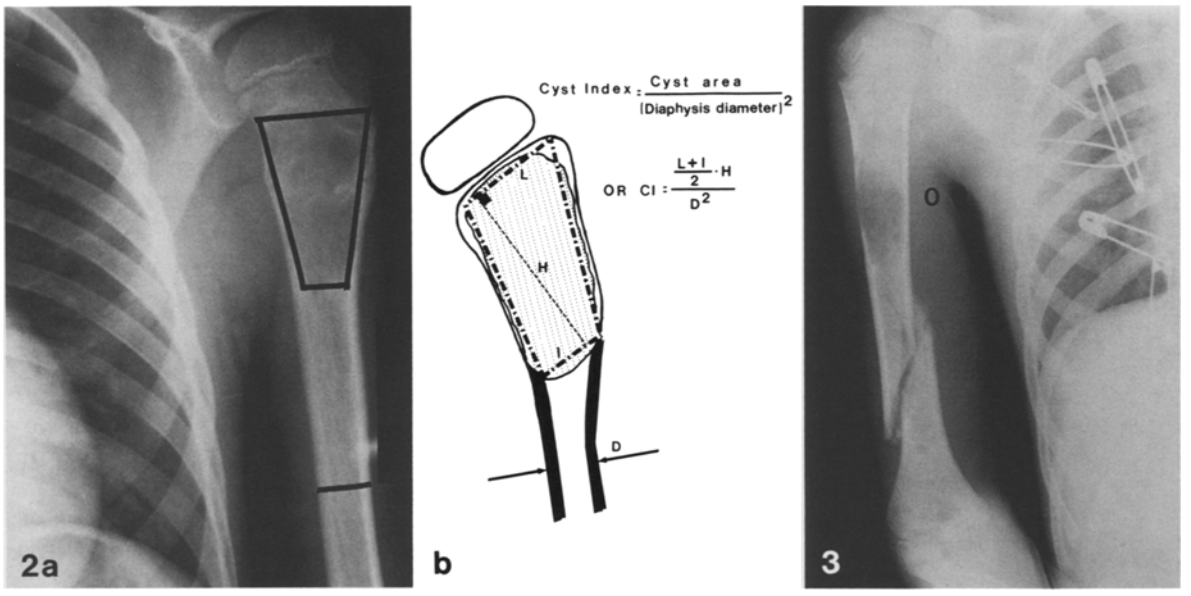

Fig. 2. a A trapezoid is drawn round the cyst in the radiograph and the diameter of the diaphysis is measured at its tubular part; **b** diagram showing the way the area is measured

Fig. 3. Radiograph of a spiral fracture distal to a unicameral bone cyst in the humerus. Index: 4.62

The diagnosis was made after fracture in 71% of patients, but the rate of fracture at diagnosis varied with the site. In 94% of humeral cyst the first manifestation was a fracture; the corresponding rate in the femur was 47% and in the tibia and fibula 33%. Cysts without a fracture were diagnosed when the child complained of a persistent pain or a limp. A positive diagnosis was made by biopsy or analysis of the cystic fluid in 34 cases; the other 24 cysts did not have any surgical procedure, but presented with the typical radiological appearance and healed subsequently. The 31 children with humeral cysts sustained a total of 50 fractures (average 1.6 fractures a child; range 0–5). The 21 children with femoral cysts sustained a total of 14 fractures (average 0.7 fractures a child; range 0–2).

The methods of treatment were curettage and bone graft, methylprednisolone acetate injection, and observation alone (Table 1).

Children with humeral cysts who were not treated surgically were immobilised for an average of 2.4 months, whereas those who were operated on were immobilised for an average 2.8 months. Thus the length of immobilisation was not affected by the type of treatment.

The cyst index

A cyst developing in the metaphyseal area expands centrifugally and, although the cortex is resorbed, a thin bony border remains around the cyst without any periosteal reaction. The inner borders are either smooth or scalloped. During its developmental phase, the cyst stays in the physis because its growth is more rapid than bone growth. The cyst moves away from the physeal line either because it is in a stable or healing phase, or because the patient has entered a rapid adolescent growth phase when bone growth becomes more rapid than cyst growth. The diameter of the cyst may equal the width of the physis, but rarely becomes greater. The shape of the metaphysis is disturbed by lack of resorption.

Internal crests or septa often appear after fracture and may be transient. Spontaneous healing occurs by bone formation in the cyst's cavity, the cortex progressively thickens and the cyst fills in centripetally. The periosteum is not involved in the healing process, but periosteal apposition appears as a result of the fracture healing. After complete healing, the site is permanently marked by a sclerotic scar.

A constant radiological finding was that the larger the cyst, the more cortex was destroyed and the bone weakened. To quantify the strength of the remaining cortex, which is related to the size of the cyst and the size of the involved bone, we devised the cyst index. This gives the proportion between the radiographic area of the cyst and the size of the involved bone, measured as the diameter of the diaphysis squared:

$$\text{Cyst Index} = \frac{\text{Area of the cyst}}{\text{Diaphysis diameter}^2}$$

The areas involved are shown in Fig. 2 and the cyst index is the relationship between the two areas. The size of the bone, the distance to the physeal line, the length of the cyst, and the radio-

graphic density which depends on the technique, are not important features in this evaluation.

The area of the cyst is measured by its largest radiological dimension. Often the index is the same on the anteroposterior and lateral radiographs. The shape may be oval or trapezoidal, and the area is easily calculated by drawing one or two trapezoids around the borders of the cyst. The diameter of the diaphysis is measured in its tubular part (Fig. 2).

A low cyst index indicates a small cyst area in relation to the bone and a stronger cortex; conversely, a high index indicates a large cyst and a weaker cortex. The smallest cyst measured 0.1, the largest 12.78.

Results

The cyst index in pathological fractures

Recent pathological fractures were found in 53 radiographs and the cyst index was measured in each. Forty-nine of these fractures followed minor trauma and this is reflected by the minimal displacement seen. The average index in humeral fractures was 6.12 (SD = 1.8) and in femoral fractures 4.74 (SD = 1.19). The humeral index is higher because these fractures occurred later than those in the lower limb.

One distal tibial fracture followed a significant injury and had an index of only 2.21. On three occasions torsion had produced a spiral fracture distal to a humeral cyst through normal cortex (Fig. 3). In these cases, the average index was 3.33 (range 1.62–4.62) indicating that the cyst wall was strong enough to resist relatively high energy trauma. None of the observed fractures involved the physeal line.

Before the first fracture, the children are normally active, but once the diagnosis is made activity is reduced, and for a subsequent fracture the trauma is likely to be less than in the first instance. The index would then be larger, and our results confirmed this hypothesis. In 21 initial fractures through humeral cysts, the average index was 5.38 (SD = 1.02), and in 14 recurrent fractures the index was 7.38 (SD = 2.4) which is a highly significant difference ($P < 0.005$) Student's *t*-test). The increased index is also an indication of progression of the cyst in some patients after the first fracture.

In femoral fractures the average index of the first fractures was 4.4 (SD = 0.75) and that of recurrent fractures 7.17.

The lowest indices for humeral and femoral fractures were 3.9 and 3.6, respectively. Although we cannot predict the fracture which leads to the diagnosis of a cyst, we now can predict recurrent fractures. There were no fractures in 40 patients who had an index lower than 3.5 and who did not curtail their activities.

The differences between humeral and femoral fractures can be explained by their biomechanical pattern. The humerus is under axial tension and shearing stress, whereas the femur is under compression shearing forces. Microlesions in compression through a thinner cortex explain the incidence of pain and limp, and indicate why a femoral cyst can be diagnosed before fracture.

The level of confidence can be estimated by Gaussian distribution of the cyst index for the first humeral or femoral fracture. The standard deviation is a statistical means of assessing the spread of the results. In the initial humeral fractures the average index is 5.38 (SD = 1.02). It is logical to suppose that in a weight-bearing bone like the femur a smaller cyst will allow a pathological fracture; this is confirmed by an average index of 4.4 (SD = 0.75) for initial femoral fractures.

For the recurrent humeral fractures, the comparison of the index of the initial and subsequent fractures indicates that the increased index is a sign of further growth of the cyst. Thus fractures occur despite restricted sports and physical activity. These observations support the reliability of the index for predicting pathological fractures through cysts in a long bone with a tubular diaphysis.

The cyst index and natural history

The natural history of unicameral bone cysts was observed in 11 untreated humeral cysts that were followed for more than one year. Spontaneous healing occurred in five cases (Fig. 4), who had sustained a total of nine fractures. One patient had no fractures, 2 patients had two fractures each, and one had four fractures. All fractures were immobilised and healed within one month; no patient lost movement of the shoulder or elbow, or had any shortening of the arm.

The index does not show any tendency towards regression of the cyst after a fracture. The cyst often continues to grow in parallel to the healing process. After three months, periosteal new bone has been incorporated and partially resorbed. The cyst is then strong enough to transmit load without further stimulation of the periosteal reaction. All humeral fractures healed within 2 or 3 weeks and femoral fractures in 10 weeks.

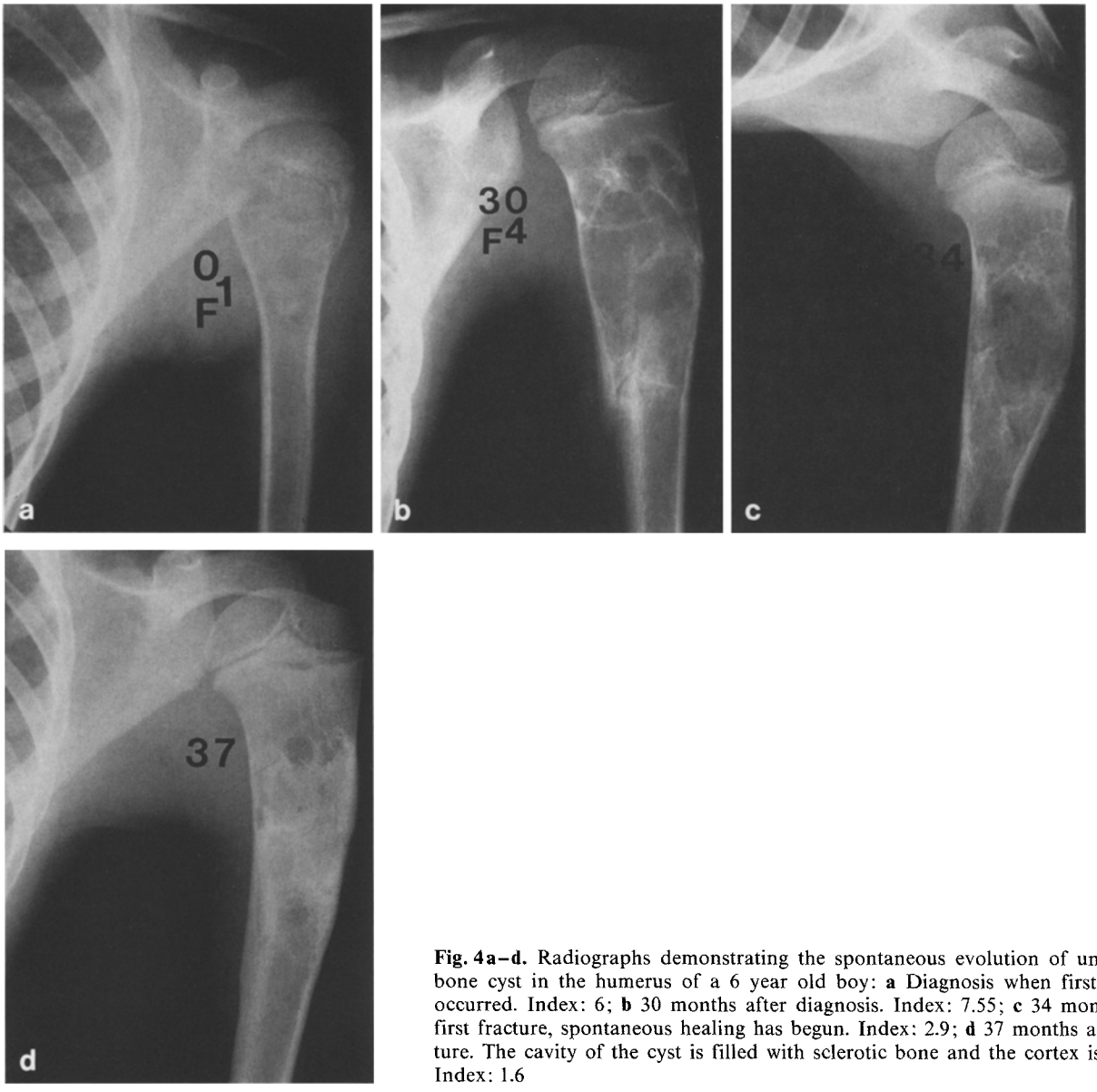

Fig. 4a–d. Radiographs demonstrating the spontaneous evolution of unicameral bone cyst in the humerus of a 6 year old boy: **a** Diagnosis when first fracture occurred. Index: 6; **b** 30 months after diagnosis. Index: 7.55; **c** 34 months after first fracture, spontaneous healing has begun. Index: 2.9; **d** 37 months after fracture. The cavity of the cyst is filled with sclerotic bone and the cortex is thicker. Index: 1.6

In 4 cysts, each of which had several fractures, the first fracture occurred with an average index of 5.8 and the subsequent fracture with an average of 7.0; this increase is proportional to the period between the first and second fractures. In one, the index increased by 0.65 after 4 months, in another by 1.10 after 7 months, and in two by 2.0 and 3.9 after a year. In these cases the slope of the increase remained in the same range as before fracture, showing a rise of about 2 points a year. Thus a fracture does not change the natural course of the development of a cyst.

In our 57 patients, no further increase in the index occurred after its spontaneous decrease, which means that all cysts which are healing spontaneously will ultimately heal completely. Cysts with at least two consecutive decreased indices of less than 3 and a cortical wall thicker than 2 mm are considered to be healed. When these conditions are present, fractures never occur and the result is always favourable.

Cyst index and treatment

Twenty-six healed femoral and humeral cysts were studied. Of the 15 proximal humeral cysts, 8 were treated by curettage and grafting, 5 of whom had a second procedure for recurrence. Two others benefited from injection of methylprednisolone acetate, and 5 were treated conservatively for a fracture and the cyst was observed.

Table 2. Humeral cysts

	Treatment	
	Failure	Success
No. of operations	8	7
No. of previous fractures/patient	1.4	1.3
% of latent cysts	40	78
Previous operations	1	6
Age at operation (years)	7.3	12.7
Duration of disease (months)	7	47
Cyst index	5.58 ± 2.35	3.38 ± 1.53

Table 3. Femoral cysts

	Treatment	
	Failure	Success
No. of operations	13	7
No. of previous fractures/patient	0.46	0.15
% of latent cysts	26	73
Previous operations	4	9
Age at operation (years)	7.2	10.2
Duration of disease (months)	7	30
Cyst index	4.66 ± 3.23	1.86 ± 0.82

Table 4. Cyst index: points of reference

	Cyst Index	
	Average	Lowest measured
First humeral fracture (n = 21) (normal activity)	5.38 ± 1.02	3.9
Recurrent humeral fracture (n = 14) (restricted activity)	7.38 ± 2.4	4.8
First femoral fracture (n = 10)	4.40 ± 0.75	3.6

Of the 11 patients with cysts of the proximal femur, 7 had curettage and grafting; 2 had one procedure each, 3 had 2 and 2 had 3. One patient had two operations for curettage and grafting and two injections of methylprednisolone acetate, and 3 cysts healed after a single injection each.

In order to assess whether the cyst index is related to the prognosis after curettage and grafting, we compared humeral and femoral cysts in two series of patients with success or failure of treatment (Tables 2 and 3). In humeral cysts the number of fractures was similar whether or not treatment was successful, and the latency of a cyst is not necessarily an indication of success. The age at operation and the duration of the lesion are significant factors. In older patients with a longer history, the success rate is higher.

The index was 5.58 in the failures and 3.38 in the successful group which is significant ($P > 0.05$). The cysts which responded well had already sustained fractures (index >4) and were in a phase where the index was diminishing. After a long evolution and in adolescence, cysts were beginning to heal spontaneously before they were treated. In the failures, the children were younger and the cysts had been present for a shorter period. The index was high and at the level where there was a risk of fracture; treatment failed because the cysts were in a developing phase.

Comparison between the successes and failures in femoral cysts leads to similar conclusions. Cysts already healing respond well to operation, and the difference between indices (4.66 in the failures and 1.86 in the successes) is significant ($P < 0.025$). The shorter time between diagnosis and healing results from more aggressive surgical treatment and a better response to treatment at this site.

It is paradoxical that cysts in the lower limb with an index of >4 need treatment because of the higher risk of fracture, and in this group the failure rate was higher. Otherwise, when treatment is not necessary and the index is <4, the conditions are favourable for success.

Healing occurs in two ways after curettage and grafting. There is rapid incorporation of the graft which becomes sclerotic and the cortex thickens after two or three months; microcysts my appear in the new cortex. Healing occurred in 12 of our patients. In three cases, incorporation of the graft was followed by a recurrence, with an index up to 3; spontaneous healing occurred after two or three years.

Injections of methylprednisolone acetate were given to 9 patients (11 injections). These numbers were too few for an accurate analysis to be made. Scaglietti et al. found that three or four injections were necessary to produce healing. In our cases, ten injections were followed by little or no response; further injections were required in four cases, operation in one case and observation only in the remainder. The average index was 4.49. Four cysts healed after injections (average index 3.73), but a larger study is needed to see whether this is statistically significant. In these cases the cyst filled with sclerotic bone after about four months, but the lining membrane remained radiolucent; after six months the cortex thickened and healing occurred.

Table 5. Indications for treatment or observation

Cyst index	Increasing	Decreasing
High fracture risk Humerus >4 Femur >3.5	Fracture prevention Injection[a] If no response, consider operation in the femur	Fracture prevention Injection can accelerate healing
Low/no fracture risk Humerus <4 Femur <3.5	Observation Consider injection[a]	No treatment

[a] Injection of methylprednisolone acetate

Discussion

McLachlin reviewed the natural history of bone cysts [14] and Cohen analysed more than 1000 cases [7]. Morton reported that in his series of 76 cysts most ran their own course and were not greatly affected by treatment [16].

It is necessary to anticipate the risk of pathological fracture before deciding whether treatment is necessary. No correlation was found between the length or area of the cyst [4] and the risk of fracture, but there was a significant correlation when the area of the cyst was related to the size of the bone expressed as the square of the diameter of the diaphysis.

A review of the English literature [1, 2, 14, 16, 20, 22] revealed 57 cases where the cyst healed spontaneously, representing 15% of all the cysts reported in these papers. The reasons for deciding on conservative treatment were not explained.

The effectiveness of any treatment can only be evaluated by comparison with the natural history of the cysts, and a high rate of good results in the spontaneous healing phase is no proof of effectiveness.

There is no doubt that unicameral bone cyst is a self-limiting benign condition and that the only indication for treatment is the potential risk of fracture. The cyst index, which we have described, is an easily reproducible method of assessing the mechanical resistance of the cyst wall. The distribution of results correlate closely with the anatomical site. Its use makes it possible to plan treatment for each patient related to the risk of fracture. Table 4 summarises the risk in relation to the index. Assuming that normal bone can be broken by severe trauma, we give the lowest limit for a pathological fracture through the proximal humerus as 4, and as 3.5 for the proximal femur.

We have shown that treatment is successful when the cyst is already healing. A high recurrence rate is related to a rising cyst index which leads to a higher risk of fracture.

The index is also valuable in evaluating the response to injection of methylprednisolone acetate.

The scheme of treatment depends on the phase of evolution as measured by an increasing or decreasing index, and on the risk of fracture as indicated by the instant value of the index. Table 5 reviews the indications for treatment of cysts in tubular bones.

After a radiographic diagnosis is made, the limb should be protected if there is a high or moderate risk of fracture. If the cyst is in an active growth phase, an attempt should be made to stop its development. Injection of methylprednisolone acetate should be considered first because it is innocuous. Surgical treatment is reserved for cysts which do not respond to injection and which have a high risk of pathological fracture and subsequent deformity. This particularly applies to cysts in the femoral neck.

References

1. Adams AW (1926) Report of a case of solitary fibrocystic disease of the humerus exhibiting spontaneous resolution: with a review of the literature and a consideration of the etiology and treatment. Br J Surg 13: 734–741
2. Baker DM (1970) Benign unilateral bone cyst: a study of forty-five cases with long-term follow-up. Clin Orthop 71: 140–151
3. Boseker EH, Bickel WH, Dahlin DC (1968) A clinicopathologic study of simple unicameral bone cysts. Surg Gynecol Obstet 127: 550–560
4. Capanna R, DalMonte A, Gitelis S, Campanacci M (1982) The natural history of unicameral bone cyst after steroid injection. Clin Orthop 166: 204–211
5. Chaves D (1980) Treatment of solitary cysts of the humerus. Treatment by diaphyseal resection and bone grafting. Int Orthop 3: 253–256
6. Chigira M, Maehara S, Arita S, Udagawa E (1983) The aetiology and treatment of simple bone cysts. J Bone Joint Surg [Br] 65: 633–637
7. Cohen J (1977) Unicameral bone cysts: a current synthesis of reported cases. Symposium on Tumors of the Musculoskeletal System. Orthop Clin North Am 8: 715–736
8. Fahey JJ, O'Brien ET (1973) Subtotal resection and grafting in selected cases of solitary unicameral bone cyst. J Bone Joint Surg [Am] 55: 59–68

9. Garceau GJ, Gregory CF (1954) Solitary unicameral bone cyst. J Bone Joint Surg [Am] 36: 267–280
10. Gartland JJ, Cole FL (1975) Modern concepts in the treatment of unicameral bone cysts of the proximal humerus. Orthop Clin North Am 6: 487–498
11. Jaffe HL, Lichtenstein L (1942) Solitary unicameral bone cyst: with emphasis on the Roentgen picture, the pathologic appearance and the pathogenesis. Arch Surg 44: 1004–1025
12. Marotexau P (1979) Bone diseases of children. Lippincott, Philadelphia, pp 388–390
13. McKay DW, Nason SS (1977) Treatment of unicameral bone cysts by subtotal resection without grafts. J Bone Joint Surg [Am] 59: 515–518
14. McLachlin AD (1943) Treatment and results in localized osteitis fibrosa cystica (the solitary bone cyst). J Bone Joint Surg 25: 777–790
15. McNamee WB, Gartland JJ, Irani RN (1981) Diaphysectomy for unicameral bone cyst. Orthop Rev 10: 97–101
16. Morton KS (1982) Unicameral bone cyst. Canadian J Surg 25: 330–332
17. Neer CS, Francis KC, Marcove RC, Terz J, Carbonara PN (1966) Treatment of unicameral bone cyst: a follow-up study of one-hundred-seventy-five cases. J Bone Joint Surg [Am] 48: 731–745
18. Neer CS, Francis KC, Johnston AD, Kiernan HA (1973) Current concepts on the treatment of solitary unicameral bone cyst. Clin Orthop 97: 40–51
19. Scaglietti O, Marchetti PG, Bartolozzi P (1982) Final results obtained in the treatment of bone cysts with methylprednisolone acetate (Depo-Medrol) and a discussion of results achieved in other bone lesions. Clin Orthop 165: 33–42
20. Siegel IM (1966) Brisement force with controlled collapse in treatment of solitary unicameral bone cyst. Arch Surg 92: 109–114
21. Spence KF, Bright RW, Fitzgerald SP, Sell KW (1976) Solitary unicameral bone cyst: treatment with freeze-dried crushed cortical-bone allograft. A review of one-hundred-and-forty-four cases. J Bone Joint Surg [Am] 58: 636–641
22. Stewart MJ, Hamel HA (1950) Solitary bone cyst. Southern Med J 43: 927–934
23. von Mikulicz J (1906) Über cystische Degeneration der Knochen. Verh Ges Dtsch Naturforsch Ärzte 76: 107