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Fibrous Dysplasia

AN ANALYSIS OF OPTIONS FOR TREATMENT†*

BY ROBERT B. STEPHENSON, M.D., MICHAEL D. LONDON, M.D., FRED M. HANKIN, M.D., AND HERBERT KAUFER, M.D., ANN ARBOR, MICHIGAN

From the Section of Orthopaedic Surgery, University of Michigan Medical School, Ann Arbor

ABSTRACT: The results of treatment of sixty-five symptomatic lesions in forty-three patients who had fibrous dysplasia were reviewed. For fourteen (93 per cent) of the fifteen times that a lesion in the upper extremity was treated non-operatively, the result was satisfactory. These results were independent of the patient's age at the time of the initial presentation of symptoms. In patients who were eighteen years old or older, eight (88 per cent) of the nine times that a lesion involving the lower extremity had closed treatment and both times that a lesion was treated with curettage and bone-grafting, the result was satisfactory. The results of these modalities of treatment in patients who were less than eighteen years old were discouraging. Twenty-eight (88 per cent) of the thirty-two times that closed treatment was used and twenty-five (81 per cent) of the thirty-one times that curettage and bone-grafting was used, the result was unsatisfactory. However, after eighteen (86 per cent) of the twenty-one times that a lesion in the lower extremity was treated by internal fixation in a patient who was less than eighteen years old, there was a satisfactory outcome.

We concluded that closed treatment of a symptomatic lesion in the upper extremity generally provides satisfactory results. In patients who are less than eighteen years old, neither closed treatment nor curettage and bone-grafting is adequate treatment for a symptomatic lesion in the lower extremity. Internal fixation should be strongly considered in these young patients.

Fibrous dysplasia is a benign pathological condition that affects skeletal development. The lesion and its association with pigmentation of the skin and dysfunction of the endocrine system were described initially by Albright et al. in 1937. Lichtenstein and Jaffe classified fibrous dysplasia as a congenital anomaly that is manifested by a malfunction of bone-forming mesenchyme. The histological characteristics of this fibro-osseous tissue, with its poorly oriented osseous trabeculae and islands of cartilage, have been studied by numerous authors. Fibrous dysplasia can compromise the structural integrity of affected bones, and patients are often initially seen during the first two decades of life with progressive deformities and pathological fractures.

Fractures that result from fibrous dysplasia are often managed non-operatively, as it is thought that these fractures usually heal without difficulty. Harris et al., in a classic study of fibrous dysplasia, recommended surgical treatment only under very limited circumstances: severe or progressive deformity of an extremity, non-union of a fracture, fracture of the femoral shaft in an adult, and persistent pain in a site of fibrous dysplasia that is unresponsive to non-operative treatment. Grabias and Campbell recommended non-operative treatment of fractures that are associated with fibrous dysplasia. They advocated traction for patients who have a fracture of the femoral shaft, and they suggested that traction followed by the use of a cast-brace could be used to treat fractures of the upper and lower extremities in children.

In most reviews concerning fibrous dysplasia, the authors have paid little attention to the results of different methods of treatment. In order to assess the effect of treatment on outcome, we retrospectively reviewed our experience with patients who had symptomatic fibrous dysplasia of the extremities. We sought to correlate the method of treatment, the patient's age at the time of the initial symptoms, the location of the lesion, and the type of disease (monostotic or polyostotic) with the outcome of treatment. Each symptomatic lesion, rather than each patient, was followed over time in order to document the results of the treatment or treatments that were required for that lesion.

Materials and Methods

Between 1954 and 1984, fifty-nine patients who had fibrous dysplasia were treated at our affiliated institutions. In sixteen patients the disease involved only the skull or mandible, and they were excluded from additional analysis. Of the remaining patients, forty had a total of sixty-one symptomatic skeletal lesions of an appendage, and three patients had four symptomatic lesions of the spine or pelvis; they formed the basis of this prospective study.

The lesions were categorized according to whether they had a monostotic or polyostotic pattern, to their location in the upper or lower extremity, and to the skeletal maturity or immaturity of the patient at the time of the initial examination. A patient was considered to be skeletally mature if he or she was eighteen years old or older. Twenty-four patients had monostotic and nineteen had polyostotic fibrous
Fibrous Dysplasia

The ages of the patients at the time that they were initially seen with symptomatic fibrous dysplasia. Twenty-three patients were male and twenty were female. The average duration of follow-up was 10.4 years (range, two to fifty-five years).

The clinical history, radiographs, and histological characteristics of the treated lesions in each patient were reviewed. In all twenty-four patients who had a monostotic lesion, that was confirmed histologically. All of the patients who had polyostotic lesions had characteristic radiographic features, and in seventeen of the nineteen the lesions were confirmed histologically. Two patients had Albright syndrome.

Fifty-one per cent of the patients had pain alone when they were initially seen, while 49 per cent had pain that was secondary to a pathological fracture. Lesions that involved the spine or pelvis originally caused only pain. The age at which the initial symptoms occurred ranged from one and a half to sixty-eight years. The average age at which the symptoms occurred in the patients who had monostotic disease was fifteen and a half years (range, one and a half to sixty years), and in the patients who had polyostotic disease it was thirteen years (range, two to sixty-eight years) (Fig. 1).

When a patient was first seen with a symptomatic lesion, an independent plan for treatment was prescribed by the attending physician. A number of orthopaedic surgeons (including H. K.) at our affiliated hospitals were involved in these decisions. No specific protocol for treatment was used; it was the choice of the individual physician. A patient was considered to have completed the course of treatment if he or she returned to the pre-morbid level of activity and an attending physician thought that osseous healing and rehabilitation were complete, or when the method of treatment had failed and a new plan was initiated.

Each time the treatment for a symptomatic lesion was completed, the lesion was retrospectively assigned to one of four groups: (1) closed methods (cast, splint, or traction), (2) curettage and autogenous bone-grafting, (3) internal fixation (intramedullary rods or bone-plates), or (4) amputation or excision. Subsequent treatment of a lesion resulted in its reassignment to the appropriate treatment group. Therefore, a single lesion that was treated several times required multiple entries into the data base. The sixty-five symptomatic

<table>
<thead>
<tr>
<th>Location of Lesion</th>
<th>No. of Lesions (Monostotic/Polyostotic)</th>
<th>No. of Times Treatment Employed* (Monostotic/Polyostotic)</th>
<th>Total No. of Times Treatment Employed</th>
</tr>
</thead>
<tbody>
<tr>
<td>Upper extremity</td>
<td>21</td>
<td>Closed: 15, CBG: 8, ORIF: 1, Amputat. or Excis: 0</td>
<td>24</td>
</tr>
<tr>
<td>Humerus</td>
<td>3/7</td>
<td></td>
<td>12</td>
</tr>
<tr>
<td>Radius</td>
<td>1.4</td>
<td></td>
<td>5</td>
</tr>
<tr>
<td>Ulna</td>
<td>1.3</td>
<td></td>
<td>4</td>
</tr>
<tr>
<td>Hand</td>
<td>0.2</td>
<td></td>
<td>3</td>
</tr>
<tr>
<td>Spine or pelvis</td>
<td>2.2</td>
<td></td>
<td>4</td>
</tr>
<tr>
<td>Lower extremity</td>
<td>40</td>
<td></td>
<td>102</td>
</tr>
<tr>
<td>Femur — proximal part</td>
<td>10.13</td>
<td>Closed: 6, CBG: 10, ORIF: 4, Amputat. or Excis: 12</td>
<td>68</td>
</tr>
<tr>
<td>Femur — mid-part of shaft and distal part</td>
<td>2/1</td>
<td>Closed: 0, CBG: 2, ORIF: 0, Amputat. or Excis: 0</td>
<td>3</td>
</tr>
<tr>
<td>Tibia</td>
<td>4.7</td>
<td></td>
<td>28</td>
</tr>
<tr>
<td>Fibula</td>
<td>2.1</td>
<td></td>
<td>3</td>
</tr>
</tbody>
</table>

* CBG = curettage and bone-grafting and ORIF = open reduction and internal fixation.
lesions in the forty-three patients required 130 separate evaluations.

The clinical result of each treatment was classified as satisfactory or unsatisfactory. A satisfactory result was defined as: normal use of the affected extremity, no symptoms or only occasional pain that could be relieved by non-steroidal anti-inflammatory medication, and a limb-length discrepancy of less than two centimeters. An unsatisfactory result was defined by the presence of one or more of the following characteristics: recurrent pathological fracture, chronic pain necessitating additional intervention, limb-length discrepancy of two centimeters or more, progressive skeletal deformity, symptomatic non-union, or postoperative infection.

The locations and methods of treatment of the symptomatic lesions are listed in Table I. Both the monostotic and the polyostotic lesions predominantly involved the long bones, with the femur being the most common site. The proximal part of the femur was symptomatic much more often than the remainder of the femur. Closed treatment and curettage with bone-grafting were the most frequently employed methods.

**Results**

*Upper Extremity*

Twenty-one symptomatic lesions were treated a total of twenty-four times (Table II). Closed treatment was used fifteen times; curettage and bone-grafting, eight; and internal fixation, once. No excisions or amputations were performed. Twenty-one (88 per cent) of the twenty-four times...
TABLE II
RESULTS OF TREATMENT OF MONOSTOTIC AND POLYOSTOTIC LESIONS OF THE UPPER EXTREMITY ACCORDING TO AGE-GROUP

<table>
<thead>
<tr>
<th>Type of Treatment</th>
<th>Result</th>
<th>Total in Age-Group</th>
<th>Total in Series</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Satisfactory</td>
<td>Unsatisfactory</td>
<td></td>
</tr>
<tr>
<td>Patients who were &lt;18 yrs. old</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Closed</td>
<td>7</td>
<td>1*</td>
<td>8</td>
</tr>
<tr>
<td>Curettage and bone-grafting</td>
<td>5</td>
<td>1*</td>
<td>6</td>
</tr>
<tr>
<td>Open reduction and internal fixation</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Patients who were ≥18 yrs. old</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Closed</td>
<td>7</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>Curettage and bone-grafting</td>
<td>1</td>
<td>1*</td>
<td>2</td>
</tr>
<tr>
<td>Open reduction and internal fixation</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

* Pathological fracture.

that a lesion of the upper extremity was treated, there was a satisfactory clinical result (Figs. 2-A, 2-B, and 2-C). There were only three unsatisfactory results, and each was due to recurrent pathological fracture. These failures occurred in two patients; one of them was skeletally immature and had polyostotic disease, and the other patient was skeletally immature and had polyostotic disease.
mature and had monostotic disease.

Spine and Pelvis

There were four lesions in the spine or pelvis in three patients. They were treated non-operatively with bed rest and non-specific measures for the symptoms. A satisfactory result was achieved after the treatment of each lesion.

Lower Extremity

The data on the lesions of the lower extremity were analyzed relative to the pattern of the disease (monostotic or polyostotic) and the skeletal maturity of the patient (younger than eighteen years or eighteen years or older) at the time of the initial examination (Tables III-A and III-B). Forty symptomatic lesions were treated a total of 102 times.

Closed treatment: Thirty-four lesions were treated closed a total of forty-one times. The skeletally immature patients who had polyostotic disease were treated closed a total of twenty-seven times; in twenty-three (85 per cent) instances the result was unsatisfactory. Nineteen of these unsatisfactory results were due to recurrent pathological fracture. All five times that a skeletally immature patient who had monostotic disease was given closed treatment, the result was unsatisfactory, because of a recurrent pathological fracture in four instances. Eight of the nine skeletally mature patients who were managed by closed treatment had a satisfactory result, regardless of whether the disease was monostotic or polyostotic. The one failure was a recurrent pathological fracture in the femur of a patient who had polyostotic disease.

Curettage and bone-grafting: Curettage and bone-grafting was performed thirty-three times for eighteen le-
sions in the femur, tibia, and fibula (Fig. 3). There were twenty-five (74 per cent) unsatisfactory and eight (26 per cent) satisfactory results. Thirteen (93 per cent) of the fourteen times that such treatment was performed in a skeletally immature patient who had polyostotic disease, there was an unsatisfactory result. All of the unsatisfactory results were due to recurrent pathological fracture. In addition, twelve (71 per cent) of the seventeen times that curettage and bone-grafting was done in a skeletally immature patient who had a monostotic lesion, there was an unsatisfactory result. The results in the two skeletally mature patients were satisfactory.

Open reduction and internal fixation: Open reduction and internal fixation was performed twenty-two times for nineteen symptomatic lesions of the lower extremity. There were nineteen (86 per cent) satisfactory results (Figs. 4-A and 4-B). The devices for fixation included fifteen intramedullary rods of various types, three sliding hip-screws, and four compression plates. There were no recurrent fractures after internal fixation. The treatment was performed six times in skeletally immature patients who had monostotic disease, and there were five satisfactory results. The only unsatisfactory result was in a seven-year-old child in whom a non-union developed after intramedullary fixation of the tibia with a Rush rod. Internal fixation was performed fifteen times in skeletally immature patients who had polyostotic disease. There were thirteen (87 per cent) satisfactory and two (13 per cent) unsatisfactory results. One failure was due to infection after the use of a Badgley hip-nail for a proximal fracture of the femur and the other was due to a tibial non-union after the insertion of a Lottes nail.

Excision or amputation: Excision or amputation was
performed six times for five symptomatic lesions. Amputation was done in two patients who had polyostotic disease: a Syme amputation in an eleven-year-old boy who had severe deformity of the ipsilateral femur and tibia that resulted in a ten-centimeter limb-length discrepancy, and a below-the-knee amputation as the definitive treatment in a twenty-year-old man whose tibia remained deformed despite multiple previous corrective osteotomies (Figs. 5-A and 5-B).

Excision of a lesion was performed four times. The involved segment of bone was removed for the treatment of one fibular lesion, and the outcome was satisfactory. Excisional arthroplasty of the hip was performed three times and two satisfactory results were obtained, both following a Girdlestone arthroplasty. The unsatisfactory result involved re-fracture distal to the site of resection in a Colonna trochanteric arthroplasty.

Discussion

In 1942, Lichtenstein and Jaffe first recognized and differentiated the monostotic and polyostotic patterns of fibrous dysplasia. Early efforts were directed at describing the pathological and radiographic appearance of the disease, and it was demonstrated that monostotic and polyostotic fibrous dysplasias are histologically indistinguishable. The radiographic appearance of a lesion can range from purely lytic, to a classic ground-glass appearance, to sclerotic and densely calcified. Therefore, it may be very difficult to diagnose monostotic fibrous dysplasia solely by radiographic criteria, and a biopsy is usually necessary for confirmation. In contrast, polyostotic fibrous dysplasia can frequently be diagnosed radiographically due to the presence of multiple lesions that may also lead to characteristic deformities (Fig. 7).

In the majority of patients, the lesions of fibrous dysplasia become clinically manifest in the first two decades of life. The appearance of the disease this early is consistent with a congenital origin, followed by a functional deficit as the patient grows and his or her level of activity increases. Whether the lesions of fibrous dysplasia resolve or become inactive with the onset of puberty or skeletal maturation is unclear. Reed noted no change in biopsy specimens that were obtained over periods as long as ten years. Similarly, Harris et al. reported that the examination of serial biopsy specimens demonstrated only subtle histological differences over time. They observed decreased cellularity and a slightly decreased quantity of bone relative to fibrous tissue, but no conversion of fiber-bone trabeculae to lamellar bone. Malignant sarcomatous transformation has been reported in fewer than 1 per cent of patients, and it was not seen in our patients.

In contrast to the relatively stable histological appearance of fibrous dysplasia, radiographic progression of the disease has been documented. In the study by Harris et al., twenty-six patients had a complete radiographic skeletal survey, and thirteen (50 per cent) were noted to have extension of the osseous lesions over time. This was observed both before and after puberty. Henry documented radiographic progression of the disease with pregnancy.

Although patients usually seek medical attention because of pain, with or without a pathological fracture, there have been few reports on treatment and functional outcome. Harris et al. reviewed the cases of fifty patients who had fibrous dysplasia, thirteen of whom had monostotic and thirty-seven, polyostotic disease. They did not evaluate the results of treatment, but they noted that fractures were seldom displaced and healing was usually not delayed. Consequently, closed treatment of these pathological fractures, even in areas of polyostotic involvement, was advocated. Henry, in a study of the cases of fifty-six patients who had monostotic disease, noted a rate of success of 61 per cent after treatment with curettage and cancellous bone-grafting. These results improved with extraperiosteal excision. However, Henry's data are difficult to interpret because he included the radiographic appearance as an integral part of a
successful outcome. Stewart et al. reported the results of treatment in twenty patients. Thirteen of the sixteen patients who had monostotic disease had involvement of a lower extremity. Seven good, three poor, and three uncertain results were recorded after curettage and bone-grafting. They recommended early curettage and bone-grafting for weight-

Fig. 6-A

Figs. 6-A, 6-B, and 6-C: Spectrum of the radiographic presentation of fibrous dysplasia.
Fig. 6-A: Lytic lesion.
Fig. 6-B: Classic ground-glass appearance.

Fig. 6-C: Sclerotic lesion.
Fig. 7: An eighteen-year-old man who had polyostotic disease and a characteristic shepherd’s crook deformity of the proximal part of the right femur.
bearing bones but asserted that good results were unusual in skeletally immature patients. Internal fixation was used in three patients, but no conclusions were drawn due to inadequate follow-up. Strassburger et al. reported on the cases of nine patients, of whom five had curettage and bone-grafting. By our criteria, only two of the five had a satisfactory result. They concluded that only small, solitary lesions responded well to curettage and bone-grafting.

Fibrous dysplasia of the hip and proximal part of the femur has been the subject of relatively recent reports. The loss of structural integrity in the proximal part of the femur, coupled with the stresses of weight-bearing, can result in pathological fractures that are difficult to treat. Nakashima et al. reviewed the cases of eight patients who had monostotic disease that involved the femoral neck. Curettage and bone-grafting was considered curative in six patients, but the authors failed to elaborate on the functional outcome. Funk and Wells contended that monostotic disease of the hip is more predictable and easier to control than polyostotic disease. The two patients in their series who had monostotic disease were treated with curettage and bone-grafting, followed by a period of non-weight-bearing. Nevertheless, each patient subsequently sustained two fractures each, and ultimately required osteotomy with internal fixation to correct a deformity of the proximal part of the femur. Four of their patients who had polyostotic disease (in seven hips) required nineteen operations. Funk and Wells stated that repeated bone-grafting and osteotomy may not control the deformities that are associated with severe polyostotic disease.

Fig. 8: A twenty-two-year-old man who had polyostotic disease engaged in normal activities with the dominant upper extremity and had only occasional pain that could be relieved by aspirin.

Fig. 9: A twenty-one-year-old woman who had monostotic fibrous dysplasia with deformity of the proximal part of the femur and pain. After osteotomy and internal fixation, she became asymptomatic.
Our retrospective review of the cases of patients who had fibrous dysplasia dealt specifically with the results of four different methods of treatment. Each symptomatic lesion, rather than each patient, was followed to document the clinical outcome fully. In this way, we thought that the true morbidity of the disease could be assessed.

All patients in whom a symptomatic lesion of the spine or pelvis was treated by closed means had a satisfactory result. Although the number of symptomatic lesions of the spine and pelvis in our series was small, our data suggest that non-operative management may be sufficient.

Eight patients had twenty-one symptomatic lesions of the upper extremity. Twenty-one (88 per cent) of the twenty-four times that such a lesion was treated, the result was satisfactory. There was no correlation between the clinical outcome and the size or location of the lesion (Fig. 8). Closed treatment was usually sufficient, and the long-term functional morbidity that was caused by these lesions was minimal. Open reduction and internal fixation was available as a final option, but our data suggested that it is rarely, if ever, indicated as the primary treatment.

While minimum functional morbidity was associated with symptomatic fibrous dysplasia of the upper extremity in our patients, regardless of age, this was not the case for the lesions of the lower extremity. In the lower extremity, the results were highly dependent on the patient’s age at time of the initial presentation of symptoms. In patients who were eighteen years old or more, eight (88 per cent) of the nine times that a lesion involving the lower extremity had closed treatment, and both times that a lesion was treated with curettage and bone-grafting, the result was satisfactory. In contrast, patients who were less than eighteen years old usually had an unsatisfactory outcome. Eighty-eight per cent of the patients had twenty-one symptomatic lesions of the lower extremity. In the lower extremity, closed treatment was usually sufficient, and the long-term functional morbidity of the patient, and neither closed treatment nor curettage and bone-grafting was adequate for symptomatic lesions that were treated by internal fixation and bone-grafting, the result was satisfactory. Conclusively, patients who were more than eighteen years old and who had a lesion of the lower extremity generally had a satisfactory outcome with closed treatment. In this particular group of patients non-operative management may be sufficient.

Conclusions

Our review of symptomatic lesions of fibrous dysplasia revealed that significant long-term morbidity is associated with this disease. Closed treatment of symptomatic lesions in the upper extremity provided a satisfactory functional outcome regardless of the pattern of the disease or the age of the patient, and neither closed treatment nor curettage and bone-grafting were adequate for symptomatic lesions of the lower extremity in skeletally immature patients. These lesions frequently require internal fixation in order to achieve a satisfactory result.

References