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The primary aim of World Journal of Orthopedics (WJO, World J Orthop) is to provide scholars and readers from various fields of orthopedics with a platform to publish high-quality basic and clinical research articles and communicate their research findings online.

WJO mainly publishes articles reporting research results and findings obtained in the field of orthopedics and covering a wide range of topics including arthroscopy, bone trauma, bone tumors, hand and foot surgery, joint surgery, orthopedic trauma, osteoarthropathy, osteoporosis, pediatric orthopedics, spinal diseases, spine surgery, and sports medicine.

INDEXING/ABSTRACTING
The WJO is now abstracted and indexed in PubMed, PubMed Central, Emerging Sources Citation Index (Web of Science), Scopus, Reference Citation Analysis, China National Knowledge Infrastructure, China Science and Technology Journal Database, and Superstar Journals Database. The 2021 edition of Journal Citation Reports® cites the 2020 Journal Citation Indicator (JCI) for WJO as 0.66. The WJO’s CiteScore for 2020 is 3.2 and Scopus CiteScore rank 2020: Orthopedics and Sports Medicine is 87/262.

RESPONSIBLE EDITORS FOR THIS ISSUE
Production Editor: Ying-Yi Yuan; Production Department Director: Xiang Li; Editorial Office Director: Jin-Lei Wang.

NAME OF JOURNAL
World Journal of Orthopedics

ISSN
ISSN 2218-5836 (online)

LAUNCH DATE
November 18, 2010

FREQUENCY
Monthly

EDITORS-IN-CHIEF
Massimiliano Leigheb

EDITORIAL BOARD MEMBERS
http://www.wjgnet.com/2218-5836/editorialboard.htm

PUBLICATION DATE
March 18, 2022

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ONLINE SUBMISSION
https://www.f6publishing.com
Surgical treatment of femoral deformities in polyostotic fibrous dysplasia and McCune-Albright syndrome: A literature review

Giulio Gorgolini, Alessandro Caterini, Lorenzo Nicotra, Fernando De Maio, Kristian Efremov, Pasquale Farsetti

Specialty type: Orthopedics
Provenance and peer review: Invited article; Externally peer reviewed.
Peer-review model: Single blind

BACKGROUND
Surgical correction of femoral deformities in polyostotic fibrous dysplasia (PFD) or McCune-Albright syndrome (MAS), such as coxa vara or shepherd’s crook deformity, is a challenge.

AIM
To evaluate the treatment of patients with femoral deformities caused by PFD or MAS treated by osteotomies and stabilized with different methods, by analyzing the most relevant studies on the topic.

METHODS
A literature search was performed in Medline database (PubMed). Articles were screened for patients affected by PFD or MAS surgically managed by osteotomies and stabilized with different methods.

RESULTS
The initial search produced 184 studies, with 15 fulfilling the eligibility criteria of our study. Selected articles (1987-2019) included 111 patients overall (136 femurs).

CONCLUSION
Based on our results, the preferred method to stabilize corrective osteotomies is intramedullary nailing with neck cross pinning. When the deformity is limited to the proximal part of the femur, a screw or blade plate may be used, although there is a high risk of fracture below the plate. When the femur is entirely involved, a two-stage procedure may be considered.

Key Words: Polyostotic fibrous dysplasia; McCune-Albright syndrome; Coxa vara; Shepherd’s crook deformity; Femoral osteotomy; Intramedullary nailing
INTRODUCTION

Fibrous dysplasia of bone is an uncommon hereditary genetic skeletal disorder, characterized by the replacement of the bone marrow organ with a tissue formed by pre-osteogenic fibroblast-like cells and trabeculae of immature bone. The disease is due to a sporadic, congenital mutation that causes an increased synthesis of the G protein, a factor stimulating the mitosis of pre-osteoblastic cells, with the consequence that only some pre-osteoblastic cells reach a more mature stage. These immature pre-osteoblastic cells form thin bone trabeculae with structural anomalies and poor mineralization, causing bone fragility with possible deformities and fractures[1-3]. The disease was first defined as polyostotic fibrous dysplasia (PFD) by Lichtenstein in 1938, and subsequently Lichtenstein and Jaffe in 1942 described the clinical, radiographic, and histological aspects of the disease[3]. There are monostotic and polyostotic forms (PFD) that may be associated with cafe-au-lait skin spots and hyperfunctioning endocrine disorders in the McCune-Albright syndrome. In MAS, the most frequent endocrinopathies including precocious puberty, hyperthyroidism, growth hormone excess, rickets, and osteomalacia amongst others[4]. PFD and MAS commonly affect the femur and tibia, causing deformities and fractures; however, other bones including the spine and the craniofacial bones may also be affected[4-6]. The proximal third of the femur represents the site where the most difficult deformities that require surgical correction are located, such as coxa vara and shepherd’s crook deformity, sometimes associated with deformities of the diaphysis or of the distal part of the femur. A classification of femoral deformities has recently been proposed[7]. Surgical correction of femoral deformities in patients with PFD or MAS is a challenge, since the fibro dysplastic bone is much weaker and more vascularized compared to normal bone and, in the most severe forms, the medullary canal is absent. To stabilize corrective osteotomies performed in PFD, a cervico-diaphyseal interlocking intramedullary nail may be preferred, because failures are very likely to occur with either screw or blade plates. However, in some deformities, such as isolated coxa vara, screw or blade plate remain the most appropriate devices for stabilizing corrective valgus osteotomy[8-10]. Curettage and bone grafting, both with allograft and autograft, have been commonly used in PFD. However, this treatment usually fails, since no retention of any graft material has been observed over time, as reported in long-term follow-up studies[11,12].

The aim of our study was to analyze a series of papers published from 1987 to 2019, to identify the correct indications for surgical treatment of femoral deformities in patients with PDF, the effectiveness over time of the different corrective osteotomies performed, and finally the best devices to better stabilize the fibro dysplastic bone.

MATERIALS AND METHODS

Inclusion and exclusion criteria were formulated according to the population, intervention, comparator, outcome (PICO) method and are summarized in Table 1[13].

Search strategy and sources of information: authors of this review (GG, AC, LN, FDM, PF) performed a literature search about the topic by querying Medline database, Scopus and Web of Science (WOS). Studies were located by searching the database via Pubmed, Scopus and WOS. The search strategy covers PICO and was performed independently by each author on March 2021. Keywords and Medical Subject Headings (MeSH) terms were identified by a preliminary search and selected by discussion. The
Table 1 Inclusion and exclusion criteria (population, intervention, comparator, outcome)

<table>
<thead>
<tr>
<th>Inclusion criteria</th>
<th>Exclusion criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Population</strong></td>
<td>(1) Patients affected by polyostotic fibrous dysplasia or MAS; and (2) Patients affected by femoral deformities</td>
</tr>
<tr>
<td><strong>Intervention</strong></td>
<td>(1) Osteotomies; and (2) Internal fixation by intramedullary nailing</td>
</tr>
<tr>
<td><strong>Comparison group</strong></td>
<td>Internal fixation by peripheral plate</td>
</tr>
<tr>
<td><strong>Outcome</strong></td>
<td>Studies reporting clinical, radiographic evaluation</td>
</tr>
<tr>
<td><strong>Time</strong></td>
<td>Studies published from any date to 2021</td>
</tr>
<tr>
<td><strong>Study type</strong></td>
<td>(1) Cohort studies; (2) Case-control studies; and (3) Randomized control trials</td>
</tr>
<tr>
<td><strong>Language</strong></td>
<td>English</td>
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</tbody>
</table>

Other languages

<table>
<thead>
<tr>
<th>Inclusion criteria</th>
<th>Exclusion criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Population</strong></td>
<td>(1) Patients affected by monostotic fibrous dysplasia; (2) Patients affected by PFD or MAS originally treated for fractures or impending fractures; (3) Patients affected by other kind of dysplastic pathologies as fibrocartilaginous dysplasia; and (4) Patients treated for deformities caused by PFD but not affecting femur</td>
</tr>
<tr>
<td><strong>Intervention</strong></td>
<td>(1) External fixation only; (2) Bone grafting or transplantations techniques only; and (3) Other surgical techniques</td>
</tr>
<tr>
<td><strong>Comparison group</strong></td>
<td>Not applicable</td>
</tr>
<tr>
<td><strong>Outcome</strong></td>
<td>Not applicable</td>
</tr>
<tr>
<td><strong>Time</strong></td>
<td>Not applicable</td>
</tr>
<tr>
<td><strong>Study type</strong></td>
<td>(1) Letters; and (2) Case reports</td>
</tr>
</tbody>
</table>


No publication date filter was applied to select articles and review articles. Language restriction was applied to identify only English articles. In addition, a manual search was performed of the references cited in the studies included.
The reviewers (GG, AC, LN, FDM, PF) retrieved the data and independently analyzed each selected study; instances of disagreement were resolved by the senior investigator (PF).

The articles were screened for the presence of the following inclusion criteria: patients affected by PFD or MAS; patients affected by femoral deformities (coxa vara, shepherd’s crook deformity, etc.); patients surgically treated by corrective osteotomies and internal fixation; studies providing an adequate level of evidence, including retrospective studies; availability of full text. The studies were excluded if they provided information regarding: patients affected by monostotic fibrous dysplasia or affected by different dysplastic pathologies as fibrocartilaginous dysplasia; patients affected by PFD but originally treated for fractures; patients treated for deformities caused by fibrous dysplasia that did not affect the femur; and patients treated exclusively with external fixation or bone grafting or transplantation techniques.

Figure 1 shows the flowchart for study selection.

RESULTS

The initial search produced 146 studies from the Medline database, 28 studies from Scopus and 10 from WOS, for a total of 184 papers. After a first screening, we eliminated 21 duplicates. Of the remaining 163 studies, after a detailed evaluation based on inclusion and exclusion criteria, articles were screened and only 14 studies fulfilled the eligibility criteria of our study. The other studies were excluded for the following reasons: 4 included monostotic forms, 5 included fractures or impending fractures, 27 included different type of dysplasia or other pathologies, 6 included deformities not affecting femur, one included patients treated by external fixation, 7 included patients treated by curettage and bone grafting, 14 included patients non surgically treated, 20 included patients treated with other surgical techniques, 26 studies were case reports, and 39 articles were published in a different language other than in English. After screening the references by reading the full-text studies included, we added one more article. In conclusion, a total of 15 articles were enrolled in the present review (Table 2).

All of the selected articles were published from 1987 to 2019 and included 111 patients overall (136 femurs). Table 2 presents a list of the studies, summarizing the number of patients and femurs, type of deformity, age at surgery, surgical technique performed, length of follow-up, results and conclusions.

DISCUSSION

The femur is the most common skeletal segment affected in PFD with a high incidence of severe deformities, especially of the proximal part of the bone, which may cause a progressive and disabling condition[2]. The most frequent deformities are represented by coxa vara and shepherd’s crook deformity that, in severe cases, may be associated[3]. Treatment of these deformities is challenging; surgery based on curettage and bone grafting are usually inadequate in symptomatic lesions of the femur, especially in polyostotic form and skeletally immature patients. This treatment generally fails with a high percentage of relapses of the deformity and requires internal fixation in order to achieve satisfactory result[12].

Freeman et al[14] first reported the results obtained in a series of four patients affected by PFD (six femurs) treated by multiple osteotomies and fixation using a Zickel intramedullary nail. The authors concluded that in complex deformities of the femur, Zickel nail applied after multiple corrective osteotomies, provides a good control of the deformity, and allows the patients to return to normal activities. In fact, this cervico-diaphyseal device gives a good stabilization of the entire skeletal segment including the femoral neck through the screw inserted into the femoral head. The same authors stated that internal fixation with peripheral plate avoids prolonged immobilization, but a progression of the deformity often occurs, with a high risk of fracture below the plate. Ten years later, some authors[15] reported a long term-follow-up study on eight patients (7 PFD and 1 MAS) with an average age at diagnosis of 8.4 years and an average follow-up of 19.5 years. Of these patients, only two were operated on at 7 years and 5 years of age respectively, by valgus osteotomy for coxa vara, twice in one case. However, in both children the deformity continued to progress until puberty. Over 80% of patients younger than 18 years, treated by curettage and bone grafting, have an unsatisfactory result[12]. In the subsequent decade, other authors[16-19] reported the results of treatment of four series of patients affected by femoral deformities caused by PFD or MAS, with 24 patients overall (9 PFD and 15 MAS) with 37 femurs involved. The authors performed one or multiple femoral osteotomies stabilized with an intramedullary rigid nail. In some cases, curettage and bone grafting and cryosurgery or medical treatment with bisphosphonates was used in concomitance. Most of the patients were surgically treated in adolescence and followed up at least 2 years after surgery. Better results were obtained using a cervico-diaphyseal nail that Freeman et al[14] had proposed many years earlier, which allows a good stabilization of the femoral neck that in PFD represents an anatomical site where the bone is particularly weak. In fact, some of these authors[18], using an elongating intramedullary rod, without stabilization of the femoral neck in a younger series of patients, observed at follow-up, a progressive coxa vara in
Patients return to normal activities.

Complications

Intraoperative fracture (1), Respiratory distress syndrome (1), Delayed union (1)

Conclusions

Definitive control of deformities and recurrent fracture

Valgus osteotomy in 2 cases + curettage and bone grafting

Deformities continue to progress until puberty

None

The lesions stop progressing after puberty, with the change in activity of pathologic tissue from childhood to adult life

Several corrective osteotomies + curettage, cryosurgery and grafting with definitive I.M. fixation

Progression of the deformities in patients with MAS

None

Functional outcomes of extended lesions are satisfactory although some lesions need multiple procedures

Single or multiple osteotomies and fixation with UFN nail with spiral blade

All patients were painless and able to walk, one with brace and another with crutches

Delayed union (1)

Provide mechanical support to the weak and fragile dysplastic bone through intramedullary nailing

Elongating intramedullary rods + biphosphonates

Improvement of quality of life, decreasing pain and fracture rate and improving walking ability

None

Elongating rod without stabilization of the femoral neck is effective but doesn’t prevent coxa vara

Multiple osteotomies and I.M. nailing with neck cross pinning

All patients were able to return to normal activities of daily living

Loosening of the distal locking screw (1)

Good correction of progressing shepherd’s crook deformity and prevention of recurrences and fractures

Valgus osteotomy, curettage, massive allograft and I.M. nailing with neck cross pinning

Correction of coxa vara from 75° to 120°. No progression of deformity

None

The device represents the first choice of internal fixation, improving limb function and preventing fractures. Good incorporation of allografts

Valgus osteotomy stabilized by DHS plate

Correction of coxa vara from 89° to 129°

Fracture below the plate (1)

Restore the neck-shaft angle and the mechanical alignment of the femur and improve function

Valgus osteotomy stabilized with different devices (plate, I.M. nail, F.E.)

Satisfactory in 3 patients and unsatisfactory in 2 for persistent pain

Loosening of External Fixator (1)

I.M. implants with neck cross pinning are the preferable method of stabilizing osteotomies in shepherd’s crook deformities

Two stages: (1) Valgus osteotomy for correction of coxa vara and hip plate; and (2) Definitive fixation by I.M. nail with spiral blade

Neck-shaft angle and shepherd’s crook deformities were fully corrected

Cut out of the spiral blade (2), Plate’s screw loosening (1), Fracture below the plate (1)

Restore femoral alignment, pain relief and gait improvement, avoiding complications related to peripheral plates

Valgus osteotomy

Correction of

Nail breaking

Proximal humeral nail

## Table 2 Summary of literature data on surgical treatment of femoral deformities in polyostotic fibrous dysplasia and McCune-Albright syndrome

<table>
<thead>
<tr>
<th>Ref.</th>
<th>Patients (femurs)</th>
<th>Type of deformities</th>
<th>Mean age at surgery</th>
<th>Surgical treatment</th>
<th>Length of follow-up</th>
<th>Results</th>
<th>Complications</th>
<th>Conclusions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Freeman et al [14], 1987</td>
<td>4 (6)</td>
<td>Complex</td>
<td>14.5 yr</td>
<td>Multiple osteotomies and fixation with Zickel nail</td>
<td>2.8 yr</td>
<td>Patients return to normal activities</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ozaki et al [15], 1996</td>
<td>8 (11)</td>
<td>Cova vara, Shepherd’s crook deformity</td>
<td>6 yr</td>
<td>Valgus osteotomy in 2 cases + curettage and bone grafting</td>
<td>19.5 yr</td>
<td>Deformities continue to progress until puberty</td>
<td>None</td>
<td>The lesions stop progressing after puberty, with the change in activity of pathologic tissue from childhood to adult life</td>
</tr>
<tr>
<td>Keijser et al [16], 2001</td>
<td>7 (10)</td>
<td>Complex</td>
<td>14.5 yr</td>
<td>Several corrective osteotomies + curettage, cryosurgery and grafting with definitive I.M. fixation</td>
<td>6 yr</td>
<td>Progression of the deformities in patients with MAS</td>
<td>None</td>
<td>Functional outcomes of extended lesions are satisfactory although some lesions need multiple procedures</td>
</tr>
<tr>
<td>Ippolito et al [17], 2002</td>
<td>7 (10)</td>
<td>Complex</td>
<td>17 yr</td>
<td>Single or multiple osteotomies and fixation with UFN nail with spiral blade</td>
<td>2 yr</td>
<td>All patients were painless and able to walk, one with brace and another with crutches</td>
<td>Delayed union (1)</td>
<td>Provide mechanical support to the weak and fragile dysplastic bone through intramedullary nailing</td>
</tr>
<tr>
<td>O’Sullivan et al [18], 2002</td>
<td>5 (10)</td>
<td>Complex</td>
<td>8.6 yr</td>
<td>Elongating intramedullary rods + biphosphonates</td>
<td>&gt; 2 yr</td>
<td>Improvement of quality of life, decreasing pain and fracture rate and improving walking ability</td>
<td>None</td>
<td>Elongating rod without stabilization of the femoral neck is effective but doesn’t prevent coxa vara</td>
</tr>
<tr>
<td>Jung et al [19], 2006</td>
<td>5 (7)</td>
<td>Shepherd’s crook deformity</td>
<td>24 yr</td>
<td>Multiple osteotomies and I.M. nailing with neck cross pinning</td>
<td>2.5 yr</td>
<td>All patients were able to return to normal activities of daily living</td>
<td>Loosening of the distal locking screw (1)</td>
<td>Good correction of progressing shepherd’s crook deformity and prevention of recurrences and fractures</td>
</tr>
<tr>
<td>Yang et al [20], 2010</td>
<td>7 (8)</td>
<td>Cova vara, Shepherd’s crook deformity</td>
<td>22.7 yr</td>
<td>Valgus osteotomy, curettage, massive allograft and I.M. nailing with neck cross pinning</td>
<td>6.2 yr</td>
<td>Correction of coxa vara from 75° to 120°. No progression of deformity</td>
<td>None</td>
<td>The device represents the first choice of internal fixation, improving limb function and preventing fractures. Good incorporation of allografts</td>
</tr>
<tr>
<td>Li et al [21], 2013</td>
<td>12 (12)</td>
<td>Cova vara, Shepherd’s crook deformity</td>
<td>14.3 yr</td>
<td>Valgus osteotomy stabilized by DHS plate</td>
<td>1.5 to 10.6 yr</td>
<td>Correction of coxa vara from 89° to 129°</td>
<td>Fracture below the plate (1)</td>
<td>Restore the neck-shaft angle and the mechanical alignment of the femur and improve function</td>
</tr>
<tr>
<td>Kushare et al [22], 2014</td>
<td>5 (5)</td>
<td>Cova vara, Shepherd’s crook deformity</td>
<td>21.6 yr</td>
<td>Valgus osteotomy stabilized with different devices (plate, I.M. nail, F.E.)</td>
<td>2.2 yr</td>
<td>Satisfactory in 3 patients and unsatisfactory in 2 for persistent pain</td>
<td>Loosening of External Fixator (1)</td>
<td>I.M. implants with neck cross pinning are the preferable method of stabilizing osteotomies in shepherd’s crook deformities</td>
</tr>
<tr>
<td>Ippolito et al [23], 2015</td>
<td>11 (12)</td>
<td>Complex</td>
<td>14 yr</td>
<td>Two stages: (1) Valgus osteotomy for correction of coxa vara and hip plate; and (2) Definitive fixation by I.M. nail with spiral blade</td>
<td>4.5 yr</td>
<td>Neck-shaft angle and shepherd’s crook deformities were fully corrected</td>
<td>Cut out of the spiral blade (2), Plate’s screw loosening (1), Fracture below the plate (1)</td>
<td>Restore femoral alignment, pain relief and gait improvement, avoiding complications related to peripheral plates</td>
</tr>
<tr>
<td>Benedetti et al</td>
<td>5 (8)</td>
<td>Complex</td>
<td>6 yr</td>
<td>Valgus osteotomy</td>
<td>3 yr</td>
<td>Correction of</td>
<td>Nail breaking</td>
<td>Proximal humeral nail</td>
</tr>
<tr>
<td>Reference</td>
<td>Year</td>
<td>Case Details</td>
<td>Surgical Treatment</td>
<td>Complications</td>
<td>Results</td>
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<tr>
<td>[24], 2015</td>
<td></td>
<td>Shepherd’s crook deformity</td>
<td>Corrective osteotomy stabilized by I.M. nail with spiral blade</td>
<td>Most patients were pain free. All patients but one were able to walk, 3 of them with crutches</td>
<td>Corrective osteotomy stabilized by I.M. nail with spiral blade. Loss of coxa vara correction in 2 cases. Nail breakage in one case</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Hefti et al [25], 2017</td>
<td>13 (15)</td>
<td>Shepherd’s crook deformity</td>
<td>Corrective osteotomy stabilized by a custom made retrograde intramedullary nail</td>
<td>Nail breaking (1), Screw penetration into the acetabulum (1), Proximal screw migration (1)</td>
<td>This new operative method corrects and stabilizes severe difficult deformities. The operation is technical demanding requiring a careful ability to manage significant blood loss</td>
<td></td>
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</tr>
<tr>
<td>Majoor et al [27], 2018</td>
<td>6 (6)</td>
<td>Shepherd’s crook deformity</td>
<td>Corrective osteotomy stabilized by angle blade plate or I.M. nail (1 case), plus grafting in 3 cases</td>
<td>Fractures of the distal part of the plate (2)</td>
<td>PDF deformities can be adequately and safety treated with angled blade plates. Based on literature review, they propose an individualized patient-tailored approach</td>
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</tr>
<tr>
<td>Fang et al [26], 2018</td>
<td>6 (6)</td>
<td>Shepherd’s crook or complex deformity</td>
<td>Corrective osteotomy stabilized with I.M. nail (PFNA) plus curettage and grafting</td>
<td>All patients except one had satisfactory functional and radiologic results</td>
<td>This new operative method corrects and stabilizes severe difficult deformities. The operation is technical demanding requiring a careful ability to manage significant blood loss</td>
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</tr>
<tr>
<td>Wan et al [28], 2019</td>
<td>10 (10)</td>
<td>Shepherd’s crook deformity</td>
<td>Corrective osteotomy stabilized by DHS plate or I.M. nail (3 cases) plus PMMA augmentation in 5 cases</td>
<td>Correction of coxa vara from 88.1° to 128.3°. Longer operating times and greater blood loss in I.M. nailing</td>
<td>3D printing osteotomy templates facilitate the correction of shepherd’s crook deformity. DHS plus PMMA yields excellent results</td>
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half of their patients. The main intraoperative technical problems reported in these studies were the difficulty to ream a new medullary canal through the fibrodyplastic bone and the considerable amount of blood loss. Some authors[16] were forced to stop surgery for the massive bleeding observed during exposure of the proximal femur. From 2010 to 2015, we selected five studies on the surgical treatment of PFD or MAS femoral deformities in five corresponding series of patients with coxa vara and shepherd’s crook deformity[20-24]. Some authors[20] suggested correcting the deformity by valgus osteotomy or medial displacement valgus osteotomy and stabilizing it by an intramedullary nail with neck cross pinning associated to curettage and massive implanation allograft. They reported a series of 7 patients with PFD (8 femurs) in adolescent or adult age, followed up 6.2 years after surgery, obtaining a mean correction of the coxa vara from 75° to 120°. By contrast, other authors[21] suggested to stabilize the corrective valgus osteotomy by a dynamic hip screw-plate without grafting. They reported a series of 12 patients with PFD (12 femurs), of average age similar to the previous study, and a length of follow-up from 1.5 years to 10.6 years, with an improvement of the neck-shaft angle from 89° to 129°. One of these patients had a fracture below the plate and he was reoperated, stabilizing the femur by an intramedullary nail with a neck cross screw. To avoid this complication, the remaining authors[22-24] preferred to stabilize the corrective osteotomy by a cervico-diaphyseal intramedullary nail. Other possible devices are not recommended, such as the external fixator used by Kushare et al.[22] that reported a failure of treatment for an early loosening of the hardware which had to be removed. The same authors reported that the additional procedures as curettage and bone grafting using autograft, allograft or calcium sulfate are questionable, because none of their patients had complete radiographic resolution of the fibrodyplastic lesion[22]. Ippolito et al.[23] first proposed to treat these complex femoral lesions by a two-stage surgical treatment: The first stage was performed by correction of the coxa vara and fixation with a hip plate, while the second stage, by correction of a shepherd’s crook deformity and a definitive fixation with a cervico-diaphyseal nail connected to a spiral blade. The second stage procedure was performed as soon as the coxa vara was healed. He reported a series of 11 patients (12 femurs) with a mean age of 14 years, followed up after an average of 4.5 years after the second stage procedure. They concluded that the proposed treatment restored a satisfactory femoral alignment with pain relief and gait improvement, avoiding all the complications related to the peripheral plate. The same authors in another study[24] which involved 5 children (8 femora), aged from 4 years to 7 years, proposed to use intramedullary nailing also in young patients, using a custom-modified adult humeral nail 7-mm thick with a spiral blade. They concluded that this device may represent a useful method of treatment in fixing femoral deformities in young children with PFD.

Regarding the most recent literature, two studies[25,26] recommended stabilizing the corrective osteotomy of the classic shepherd’s crook deformity using an intramedullary nail, while two other studies[27,28], suggested an angle blade plate or a dynamic hip screw plate, adding bone graft or polymethyl methacrylate. Of the first two papers (overall 19 patients, 21 femurs), Hefti et al.[25] introduced a new type of custom made retrograde intramedullary nail, reporting 15 operated femurs followed up 4.5 years after surgery, with satisfactory results, although the surgical technique is demanding with significant blood loss. By contrast, the other two studies reported a total of 16 patients (16 femurs), in which the deformities were stabilized with plates; 10 patients were followed up after more than 10 years. They concluded that all the corrections obtained were stable over time, although in two cases, a fracture of the distal part of the plate occurred. Wan et al.[28], underlined that using the plate instead of the intramedullary nail reduced operation time and blood loss.

According to our review, we believe that isolated coxa vara should be corrected by an osteotomy and stabilized with a peripheral plate, while isolated shepherd’s crook deformity should be treated by multiple osteotomies and stabilized by a cervico-diaphyseal intramedullary nail. Complex deformities in which coxa vara is associated to shepherd’s crook deformity should be treated by two staged procedures.

The main strength of this review is the topic, as PFD and MAS are uncommon disease that, especially when they are presented in severe form, are difficult to manage. The main limitation lies in the papers included in the review, as they are all retrospective studies without a control group. Further studies are needed to address points that remain controversial in the treatment this disease.

**CONCLUSION**

In conclusion, we believe, in accordance with the majority of the authors, that correction of coxa vara and shepherd’s crook deformity as well as the other deformities of the femur when it is entirely involved, remains a demanding procedure and, especially in severe cases, more than one operation is necessary. Intramedullary nailing is often preferred to stabilize osteotomies performed in fibrodyplastic bone, while peripheral plating remains the device of choice to stabilize osteotomies performed for coxa vara. The use of cancellous or cortical bone graft in addition to corrective osteotomy is still controversial. Significant blood loss represents a surgical problem, which must be kept in mind during the operation by the surgeon and the anesthesiologist, especially in patients affected by MAS with complex deformities. High X-ray exposure for both the patient and surgeon must also be considered.
ARTICLE HIGHLIGHTS

Research background
Surgical correction of femoral deformities in polyostotic fibrous dysplasia (PFD) or McCune-Albright syndrome (MAS), such as coxa vara or shepherd’s crook deformity, is a challenge. Different surgical fixation devices have been described in the past.

Research motivation
No common consensus on the optimal surgical treatment for this pathology among orthopedic surgeons is present.

Research objectives
The aim of our study was to identify the correct indications for surgical treatment of femoral deformities in patients with PFD and MAS, the effectiveness over time of the different corrective osteotomies performed and the best devices to better stabilize the fibrodysplastic bone.

Research methods
A review of English language literature from 1987 until now was performed following the population, intervention, comparator, outcome guidelines.

Research results
Fifteen articles were included for qualitative synthesis in the study after the initial screening resulted in 184 papers.

Research conclusions
Correction of coxa vara and shepherd’s crook deformity remains a demanding procedure and, especially in severe cases, more than one operation is necessary. Intramedullary nailing is often preferred to stabilize osteotomies performed in fibrodysplastic bone, while peripheral plating remains the device of choice to stabilize osteotomies performed for coxa vara.

Research perspectives
High-quality prospective randomized clinical trials are needed.

FOOTNOTES

Author contributions: Gorgolini G and Farsetti P designed the study and contributed to manuscript preparation and editing; Caterini A, De Maio F, and Efremov K equally contributed to data analysis and manuscript preparation; Nicotra L contributed to data collection and manuscript preparation.

Conflict-of-interest statement: The authors did not receive any funding or financial support or potential sources of conflict of interest.

PRISMA 2009 Checklist statement: The authors have read the PRISMA 2009 Checklist, and the manuscript was prepared and revised according to the PRISMA 2009 Checklist.

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S-Editor: Fan JR
L-Editor: Filipodia
P-Editor: Fan JR
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