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Pediatric Lower Limb Deformities

Principles and Techniques
of Management

 Springer

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General

Deficiency in the shape, strength and structure of bone tissue due to altered bone mineral homeostasis is called *metabolic bone disease* (1). The major factors affecting this homeostasis can be thought of as a three 3's: the intracellular and extracellular levels of *three* ions (calcium, phosphorus, and magnesium), which are controlled by *three* hormones (parathyroid hormone, calcitonin, and 1, 25-dihydroxyvitamin D) and act upon *three* tissues (bone, gut, and kidney) [1, 2]. Common clinical manifestations of metabolic bone disease in children include electrolyte disturbances, fractures, bone deformity, abnormal gait, and short stature.

The most commonly encountered forms of metabolic bone disease in children are the various types of rickets and renal osteodystrophy. Other less common but important pediatric metabolic conditions include osteoporosis, malabsorption syndromes, inherited diseases such as hypophosphatasia, X-linked hypophosphatemia, and various forms of vitamin D-dependent rickets [1, 3] (Box 15.1).

Pathophysiology

Although there are several types of *rickets*, the basic pathogenesis is a relative decline in calcium or phosphorus (or both) of large enough amount so that it interferes with phy-

Box 15.1

- Metabolic problems related to vitamin D must be addressed prior, during, and after the surgical treatment.
- There are various forms of metabolic bone diseases. Thus, collaboration with the pediatric/internal medicine department is essential.

seal growth and mineralization of the bone matrix in the growing child [1, 3, 4]. This decrease in the mentioned serum ion levels may result from inadequate intake or diminished absorption of phosphorus, calcium or vitamin D, reduced conversion of vitamin D to its active form, end-organ insensitivity, impaired release of calcium from bone, or phosphate wasting. In addition, there is evidence that insufficient vitamin D may interfere with mineralization independent of calcium or phosphate levels [1, 3].

In renal osteodystrophy, glomerular damage leads to phosphate retention, and tubular damage causes decreased production of the active form of vitamin D (i.e., 1,25-dihydroxyvitamin D) due to absence of 1-hydroxylase activity. These two factors severely impede intestinal calcium absorption and reduce plasma ionized calcium. The subsequent hypocalcemia generates secondary hyperparathyroidism, which remains ineffective in increasing intestinal absorption of calcium. Consequently, the body's only means of increasing serum calcium levels is by bone resorption. Metabolic acidemia may further deteriorate this condition (Figs. 15.1 and 15.2) [1, 3, 4].

In patients with *osteoporosis*, the bone is structurally normal but is reduced in overall amount. In children, osteoporosis may be idiopathic, as in juvenile osteoporosis, or may be due to disuse or chronic corticosteroid administration. The mechanism is uncertain, but numerous theories include increased bone resorption versus decreased bone formation, possibly due to deficient 1,25-dihydroxyvitamin D or calcitonin

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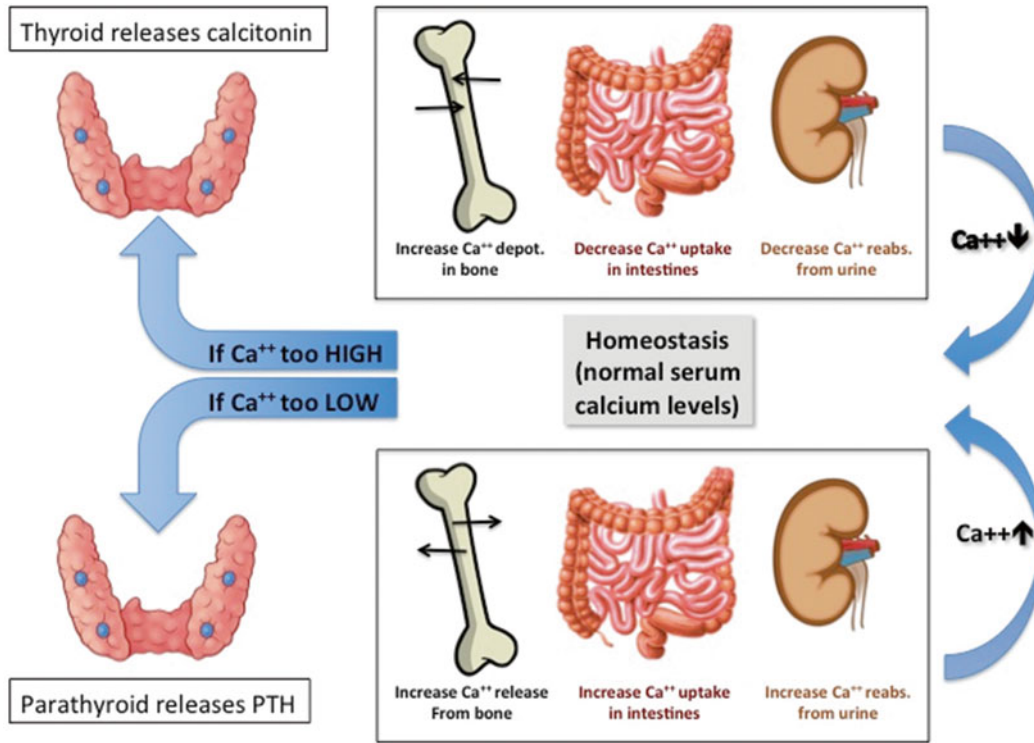
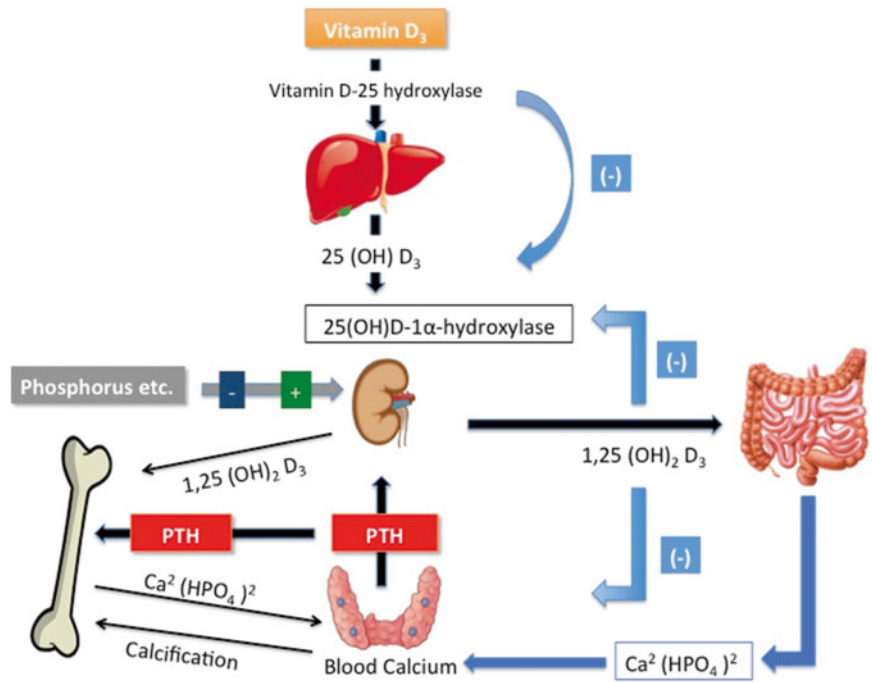


Fig. 15.1 Metabolic control of calcium levels

Fig. 15.2 Vitamin D metabolism



or due to a major interruption in transduction of mechanical forces that stimulate new bone formation [1, 3].

Hypophosphatasia results from a genetic error in the synthesis of alkaline phosphatase, the enzyme necessary for the

maturation of the primary spongiosa in the physis. This condition leads to normal production of osteoid tissue but inadequate mineralization, with resultant skeletal deformities that resemble rickets [1, 3, 4].

Nutritional Rickets

General

The primary etiology of this condition is vitamin D deficiency in the diet. This deficiency leads to failure of calcification of cartilage and osteoid tissue. Vitamin D deficiency results in an inability to absorb calcium and phosphorus. Parathormone (PTH) is released in response to hypocalcemia. Subsequently, serum calcium levels become normal or slightly decreased, but phosphate and vitamin D levels remain low (Table 15.1). Thus, radiologically, the physes display an elongation and a hazy appearance related to alterations in the provisional zone of calcification [4, 5]. The widened growth plate differentiates rickets from more common physiologic angular deformities of the lower extremity [6]. With treatment, calcification occurs and radiographic appearance gradually normalizes.

Treatment

The routine treatment for nutritional rickets include the supplemental administration of vitamin D. Radiographs typically show improved mineralization within 2–4 weeks of initiating medical treatment [4, 6]. If the child does not respond to vitamin D therapy, vitamin D-resistant rickets should be suspected. Because residual deformity is very rarely observed after adequate medical treatment of nutritional rickets, there is no specific orthopedic treatment besides follow-up to ensure acceptable lower limb alignment.

Rickets of Prematurity

Premature infants with comorbidities, followed up in intensive care units, sometimes present with pathologic fractures, probably caused by passive motion exercises. With treatment of the underlying rickets, the fractures consolidate with orthopedic immobilization techniques [4].

Drug-Induced Rickets

Certain antiepileptic medications, such as valproic acid and phenytoin, have been known to create rachitic changes in children [7] (Box 15.2). These drugs lower vitamin D levels through a P-450 microsomal enzyme system mechanism in the liver. Patients mostly present with pathologic fractures while on treatment for seizures. Medical therapy with vitamin D supplementation and consultation with the treating neurologist is helpful.

Box 15.2

- Metabolic bone diseases can be caused by certain medications as well.
- Remember that certain antiepileptics such as valproic acid and phenytoin may lower vitamin D levels in the body.

Vitamin D-Resistant Rickets (Familial Hypophosphatemic Rickets)

General

Vitamin D-resistant rickets involves a group of disorders in which normal dietary ingestion of vitamin D is insufficient to accomplish normal mineralization of bone [2, 6]. There are four major forms of vitamin D-resistant rickets, the most common of which is inherited as an X-linked dominant trait, followed in occurrence by an autosomal dominant type [2, 4, 6]. The inherent pathology is the renal tubule's incapability to retain phosphate, which causes hypophosphatemia. The third group is characterized by failure of the kidney to accomplish the second hydroxylation of vitamin D. This condition can simply be treated medically, therefore orthopedic treatment is infrequently needed. In the fourth group, also known as renal tubular acidosis, the kidney excretes fixed base and wastes bicarbonate resulting in wasting of calcium and sodium [3, 6]. Laboratory findings are listed in Table 15.1.

Vitamin D-resistant rickets typically becomes evident between the ages 1 and 2 years, slightly older than nutritional rickets. The major complaints are delayed walking and angular deformities of the lower extremities (Figs. 15.3 and 15.4). Systemic manifestations are generally absent. The deformities are much more severe when compared to nutritional rickets. Once affected children begin to walk, genu varum develops, although genu valgum may occur in some children [4, 6]. Short stature is also a feature of hypophosphatemic rickets, with their standing height often being 2 standard deviations below the mean for their peers [8]. Radiologically, the physes are widened, there is genu varum and coxa valga. Furthermore, a varus deformity of the distal tibia often leads to varus malalignment of the ankle joint (Fig. 15.5). The upper extremities are involved, to a lesser degree, as well [4].

Treatment

Medical Treatment

The standard treatment consists of oral supplementation of large doses of phosphorus along with administration of vitamin D. Studies have revealed that longitudinal growth is superior in children who receive vitamin D treatment [4].

Table 15.1 Biochemical abnormalities in rickets

| Biochemical abnormality | | | | | | |
|--|---------|-----------|-----|-----|---------------|------------------------------|
| Type of metabolic bone disease | Calcium | Phosphate | ALP | PTH | 25 (OH) Vit D | 1,25 (OH) ₂ Vit D |
| Nutritional | N | N/↓ | ↑ | ↑ | ↓↓ | ↓ |
| Vitamin D resistant | N | ↓ | ↑ | N | N | N |
| Vitamin D dependent I (inability to hydroxylate) | ↓ | ↓ | ↑ | ↑ | ↑↑ | ↓↓ |
| Vitamin D dependent II (receptor insensitivity) | ↓ | ↓ | ↑ | ↑ | N/↑↑ | ↑↑↑↑ |
| Renal osteodystrophy | N/↓ | ↑ | ↑ | ↑↑ | N | ↓↓ |

**Fig. 15.3** Photograph of a patient with severe genu valgum deformity**Fig. 15.4** Photograph of a patient with severe genu varum deformity

Furthermore, treatment by growth hormone administration also increases height and has positive influence on bone density [9]. Recently, analogs of vitamin D₃ (1,25-dihydroxyvitamin D₃) have been proved to be more effective than the previously used supplements [10].

Orthopedic Treatment

The orthotic management of skeletal deformities in patients with vitamin D-resistant rickets has not been successful. If patients complain of increasing pain and difficulty walking, angular deformities should be surgically corrected [4, 11]. The postoperative management should be done in close collaboration with the attending nephrologist or endocrinologist, since calcium levels tend to suddenly increase with postoperative immobilization period. The most common deformity seen in this patient group is a gradual anterolateral bowing of the femur accompanied by tibia vara (Figs. 15.6 and 15.7). In order to reach a physiologic lower extremity

alignment, multilevel osteotomies are often necessary [4, 11, 12]. The mechanical axis can be slightly overcorrected during surgery. The suggested fixation modality varies among reports. While external fixation allows fine-tuning of the alignment postoperatively [13] (Fig. 15.8), intramedullary fixation and plate fixation have also been reported [12, 14–16] (Fig. 15.9; Table 15.2). Regardless of type of implants utilized, careful preoperative planning of the surgery for these multiplanar deformities is obligatory to successfully reestablish the alignment of the lower extremity.

Recurrent deformity is a common sequela of osteotomies in patients with hypophosphatemic rickets [4, 12, 14] (Fig. 15.10). As expected, younger patients have a higher risk of recurrence. Therefore, milder deformities should not be operated early in the infancy. At this stage guided growth by hemiepiphyseodesis is an advisable alternative [17]. Only when gait is compromised by a thrust, or symptoms such as pain start, a corrective osteotomy should be done.



Fig. 15.5 X-ray showing distal tibial varus deformity



Fig. 15.7 Photograph of a patient with severe anterior bowing of the femur



Fig. 15.6 X-ray of a patient with a anterior bowing of the femur



Fig. 15.8 X-ray of external fixation for the tibial deformity



Fig. 15.9 Intramedullary fixation of a tibial deformity correction

Short stature is also noted amongst children with hypophosphatemic rickets. The common indication for long bone lengthening is a shortening of the entire or segment of limb in one or both legs [11]. The procedure can be performed by monolateral or circular external fixators or by lengthening over an intramedullary nail (LON) (Fig. 15.11) [12]. The application of flexible intramedullary nails in limb lengthening for children is also an alternative with multiple advantages [18]. This technique also respects the bone biology that is essential during the limb lengthening.

Author's Preferred Method, Tips and Tricks

The pathological change caused by hypophosphatemic rickets occurs very close to the growth plate, often leading to juxta-articular multiplanar deformities and severe malalignment. Preoperative assessment usually reveals multiple centers of rotation of angulation (CORA) (Fig. 15.12); thus several osteotomies may be required to fully correct the multiplanar deformities. Many methods of treatment have been described. The most common involve acute or gradual correction, using either circular or monolateral external fixators, which can secure accurate correction of the deformity and address the limb-length discrepancy. However, these methods are uncomfortable for the patient, especially when both legs are involved. There are further disadvantages, such as the need for daily adjustments, weekly follow-ups, a high rate of pin-track infection, and a long duration of external fixation.

We use the fixator-assisted nailing technique described by Paley and Herzenberg [19], with osteotomies created at the center(s) of the deformity(ies), followed by correction using a monolateral fixator and stabilization with locked intramedullary nailing (Figs. 15.13, 15.14, and 15.15). Removal of the external fixator at the end of the operation reduces postoperative discomfort and avoids pin-track infections, except where patients require additional postoperative lengthening over the nails. However, this technique does not allow residual correction or adjustment postoperatively. Thus, careful analysis of the deformity and preoperative planning are crucial. We recommend using this technique for the patient, who has essentially reached skeletal maturity. Flexible intramedullary nails (elastic nails) can be used in the pediatric age group [18].

Osteotomies in the long bones can be executed through limited incisions percutaneously either by the Gigli saw technique (Fig. 15.16) or by the multiple drill hole technique (Fig. 15.17). This technique combines the accuracy, minimal invasiveness, and safety of external fixation with the patient convenience of internal fixation. The intramedullary nail prevents the recurrence of the deformity, which is especially important in patients with metabolic bone diseases who are prone to recurrence of the deformity as the metabolic problem continues. In addition, the surgeon must be familiar with both external fixation and intramedullary nailing techniques, as well as combined techniques, which can be technically demanding.

Surgical Technique

The patient is placed supine on a radiolucent table and the lower limb is checked for imaging on the radiography table from the hip joint to the ankle joint on both frontal and side views (Fig. 15.18). In patients with genu valgum, resulting both from the femur and/or the tibia, mini-open release of the peroneal nerve is performed prior to correction of the deformity to avoid neurapraxia due to traction. Two 6-mm conical Schanz screws are placed into the proximal and distal segments above and below the osteotomy to maintain the stability, perpendicular to the anatomical axis, taking care to leave enough space for the inserted intramedullary nail without making contact with the screws (Figs. 15.19, 15.20, and 15.21). The level of osteotomy is chosen based on preoperative planning and performed percutaneously using the multiple drill hole technique for the femur or a Gigli saw for the tibia. Following the osteotomies, the deformity is corrected using a monolateral external fixator (Fig. 15.22). The correction is confirmed by obtaining frontal and side view long radiographs. If the desired correction is not achieved, it is adjusted and confirmed with new X-rays. Once the surgeon is satisfied with the correction, the intramedullary nailing is performed through a 2 cm transverse incision over the ligamentum patellae (Fig. 15.23).

Table 15.2 Review of literature

| Authors | Journal (year) | Title | Number of patients | Results |
|--------------------|------------------------------|--|---------------------|---|
| Fucentese et al. | J Child Orthop (2008) | Metabolic and orthopedic management of X-linked vitamin D-resistant hypophosphatemic rickets | 12 | Single bilateral surgical correction was performed in six patients; one patient each had three and five corrections. Bone lengthening was performed in three patients. At last follow-up, the height of seven operated patients was within normal range. In addition, leg axis was normalized in six patients with mild genua vara in two. Bone healing was excellent: surgical complications were rare. In case of bone deformity, surgery can safely be performed, independent of age or bone maturation |
| Kocaoglu et al. | J Bone Joint Surg Br (2011) | Combined technique for the correction of lower-limb deformities resulting from metabolic bone disease | 17 (43 segments) | 43 segments in 17 patients with metabolic bone disease underwent surgical treatment by the fixator assisted nailing technique. The deformity correction was achieved with a low complication rate. The use of intramedullary nail prevented recurrence of deformity and refracture |
| Kanel et al. | J Pediatr Orthop (1995) | Unilateral external fixation for corrective osteotomies in patients with hypophosphatemic rickets | (29 segments) | Corrective osteotomies were performed on 29 bones in nine children with hypophosphatemic rickets. Use of the Orthofix external fixator enabled precise correction of the deformities without interruption of medical management |
| Rubinovitch et al. | Clin Orthop Relat Res (1988) | Principles and results of corrective lower limb osteotomies for patients with vitamin D-resistant hypophosphatemic rickets | 10 (44 osteotomies) | Osteotomies were combined with shortening and compression plating. Recurrence of deformity occurred in 27 % of the cases. While osteotomies were safe and provided dramatic improvement to limb deformity, postoperative control of vitamin D metabolism was the one constant factor for maintenance of correction |
| Petje et al. | Clin Orthop Relat Res (2008) | Deformity correction in children with hereditary hypophosphatemic rickets | 10 (53 segments) | 37 corrective operations were performed on ten children. Depending on the patient's age, external fixation was used in 53 segments: Kirschner wires in 18, DynaFix in three, the Taylor patial Frame device in 13, and the Ilizarov device in 19. Internal fixation with intramedullary nailing was performed in 12. Deviation of the mechanical axis and knee orientation lines was increased at the follow-ups conducted during a period of 5–12 months. Additional follow-ups revealed a recurrence rate of 90 % after the first corrective procedure and 60 % after a second procedure |
| Song et al. | Acta Orthopedica (2006) | Deformity correction by external fixation and/or intramedullary nailing in hypophosphatemic rickets | 20 (55 segments) | 55 segmental deformities (20 femora, 35 tibiae) from 20 patients were examined retrospectively. Distraction osteogenesis was used in 28 segments and acute deformity correction in 27. External fixation was applied in 24 segments, intramedullary nailing in six, and external fixation and intramedullary nailing in 25. Recurrent deformity or refracture occurred in 10 of 21 segments with distraction osteogenesis by external fixation only, 4 of 6 with acute correction by intramedullary nailing, and 1 of 25 with distraction osteogenesis or acute correction by external fixation and intramedullary nailing. External fixation and intramedullary nailing can be recommended to prevent complications during or after deformity correction in hypophosphatemic rickets |
| Eralp et al. | J Bone Joint Br (2004) | A correction of windswept deformity by fixator assisted nailing | 2 (7 segments) | Seven segments in two patients with vitamin D related metabolic bone disease were treated by fixator-assisted nailing technique. All deformities were accurately corrected and there was no consolidation problem at the osteotomy sites. The total treatment time was found less than with other techniques |
| Stanitski | Clin Orthop Relat Res (1994) | Treatment of deformity secondary to metabolic bone disease with the Ilizarov technique | 8 (18 segments) | Complications were limited to several pin-tract infections and mild translational deformity in two patients. Healing index averaged approximately twice that seen in pediatric femoral lengthening and was 25 % greater than for patients undergoing tibial lengthening. The lack of implants requiring removal, modularity, and reasonable treatment time make this technique an attractive alternative to conventional osteotomy for management of limb-length deformity associated with metabolic bone disease |



Fig. 15.10 Note the recurrence of the deformities in a patient despite the intramedullary nails

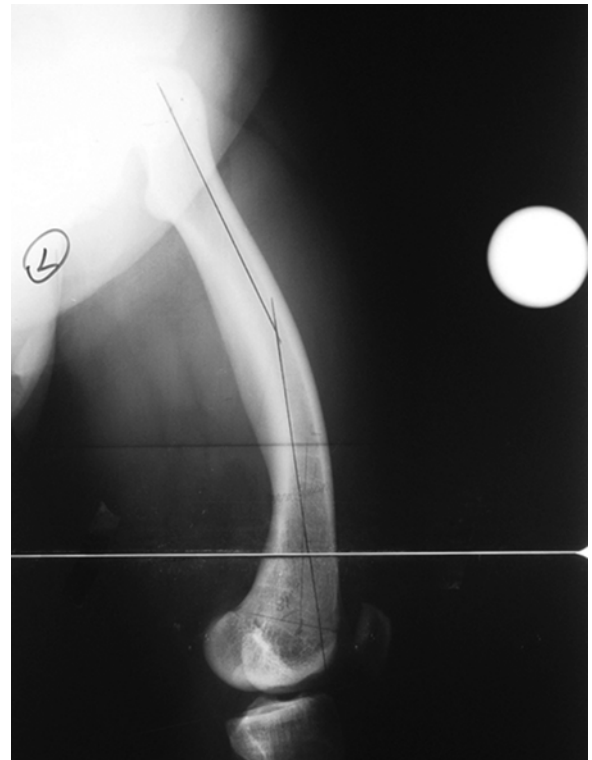


Fig. 15.12 Long sagittal bowing deformity at the femur: note there are multiple centers of rotation in these type of deformities



Fig. 15.11 Following deformity correction (fixator assisted nailing) lengthening over nail was performed at the femur



Fig. 15.13 X-ray showing a varus bowing deformity at the femur

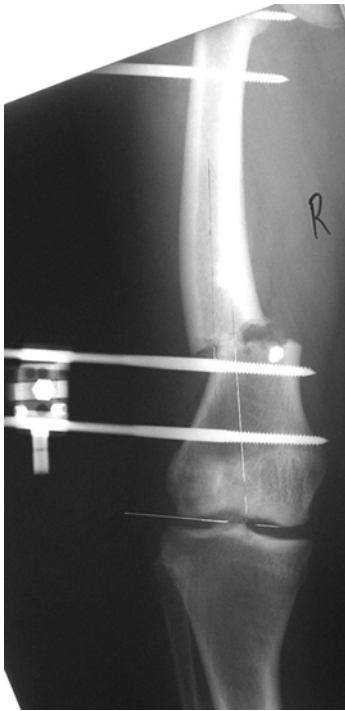


Fig. 15.14 The deformity has been acutely corrected with use of an unilateral external fixator



Fig. 15.15 Once the satisfactory correction was achieved, intramedullary static fixation was performed (FAN procedure)

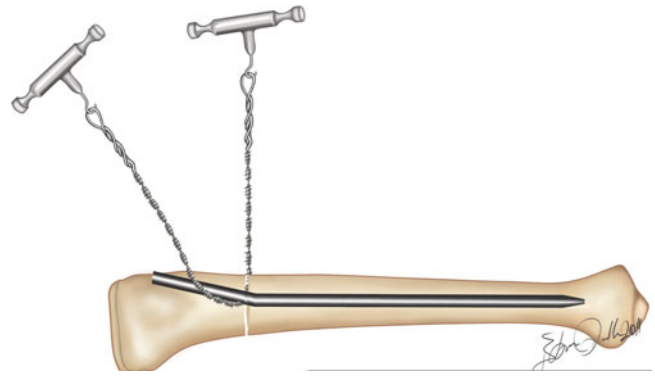


Fig. 15.16 Gigli saw osteotomy technique

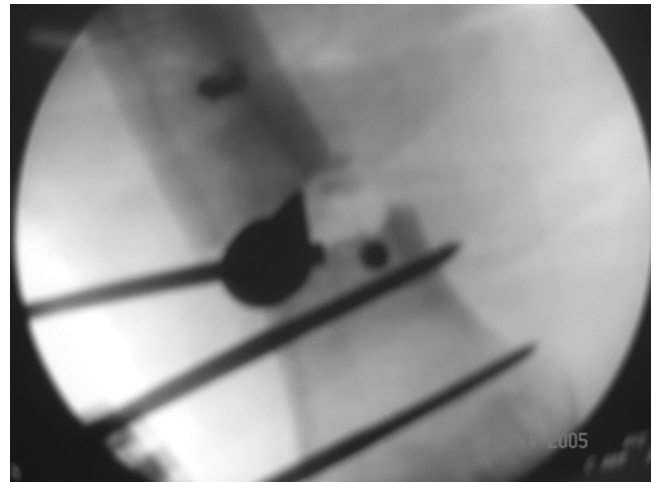


Fig. 15.17 Drill hole technique which is completed by an osteotome

The standard ligament split approach is used to open the tibial or femoral canals under fluoroscopic control. Over a guide wire, the medullary canal is over-reamed 1 mm larger than the diameter of the nail to be used (Fig. 15.24). Interference blocking screws are placed before or after nail insertion to reduce the larger diameter of the medullary canal at the metaphyseal level to prevent motion of the nail (Fig. 15.25). The nail is inserted and locked both proximally and distally if no lengthening is planned. The positions of the nail and the Schanz screws and the deformity correction are once again confirmed under fluoroscopic control, and then the fixator is removed (Figs. 15.26 and 15.27) (Box 15.3).

Follow-Up

Isometric exercises are begun postoperatively and partial weight-bearing is allowed with crutches as tolerated. Full weight-bearing is allowed only after achieving the consolidation of three of four cortices on AP and lateral radiography (approximately 3 months).

Fig. 15.18 Long X-rays for deformity analysis provided by a radiolucent table

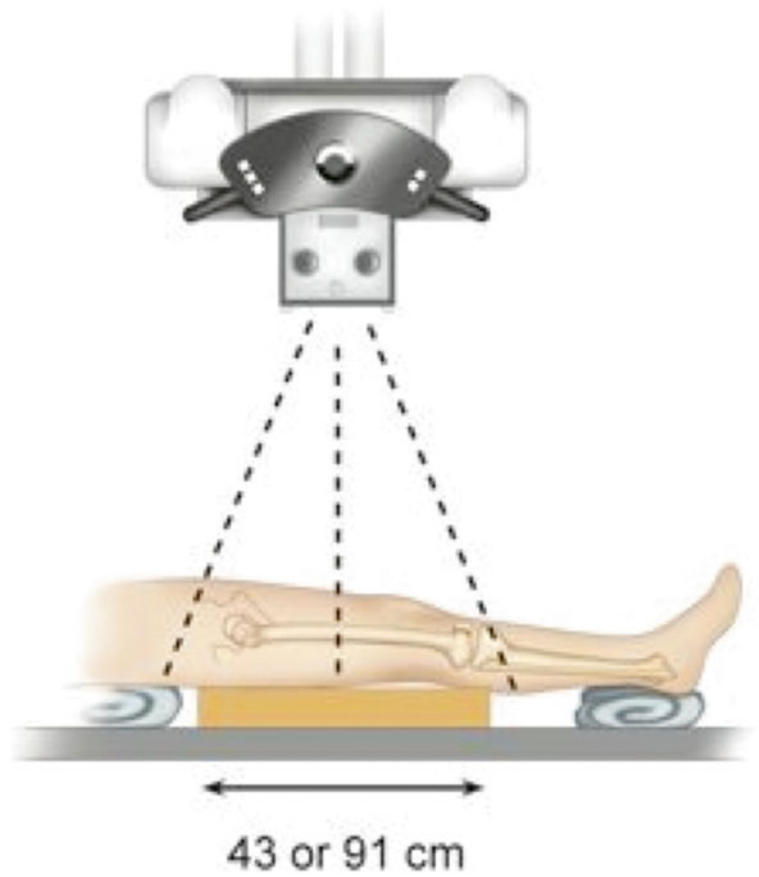


Fig. 15.19 Two parallel Schanz screws distally at the femur



Fig. 15.20 Two parallel Schanz screws proximally at the femur

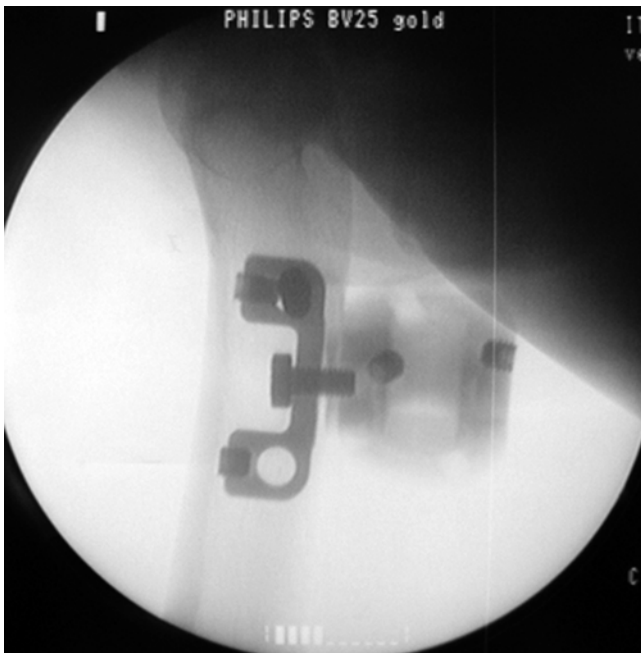


Fig. 15.21 Schanz screws must leave enough space for intramedullary nailing

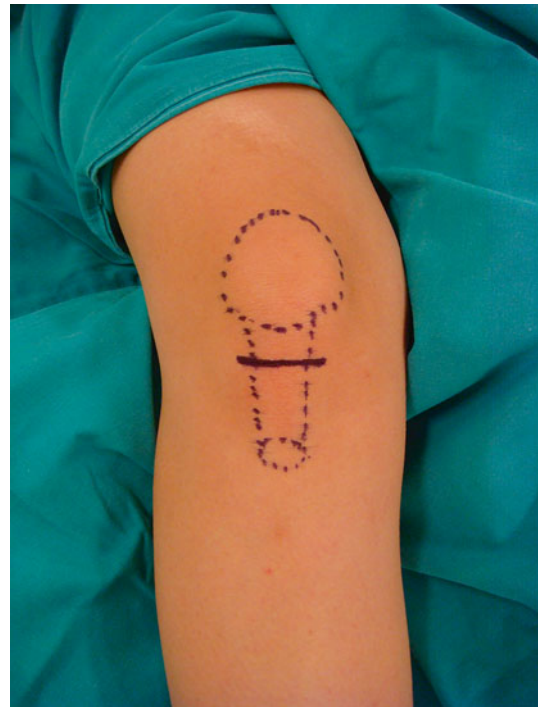


Fig. 15.23 Transverse skin incision over the ligamentum patella



Fig. 15.22 Following osteotomy the correction is performed and maintained by a monolateral external fixator



Fig. 15.24 The medulla is overreamed by 1 mm

Tumor-Related Hypophosphatemic Rickets (Oncogenic Hypophosphatemic Osteomalacia)

Certain tumors have been found to secrete phosphatonins that result in phosphaturia. Neurofibromatosis, fibrous dysplasia, less commonly osteoblastoma and hemangiopericy-

toma tend to produce rachitic changes in bone. The condition generally resolves with excision of the tumor [20].

Renal Osteodystrophy

General

As the use of renal transplantation for treating renal failure in children has increased, the prevalence of renal osteodystrophy has also climbed. Manifestations of renal osteodystrophy are present in 66–79 % of children with renal failure [21].



Fig. 15.25 Interference screws are used to help maintain the correction

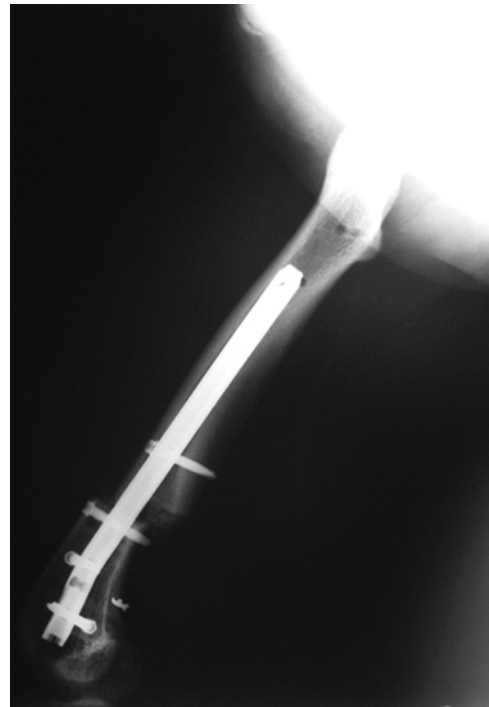


Fig. 15.27 Lateral view following intramedullary static fixation

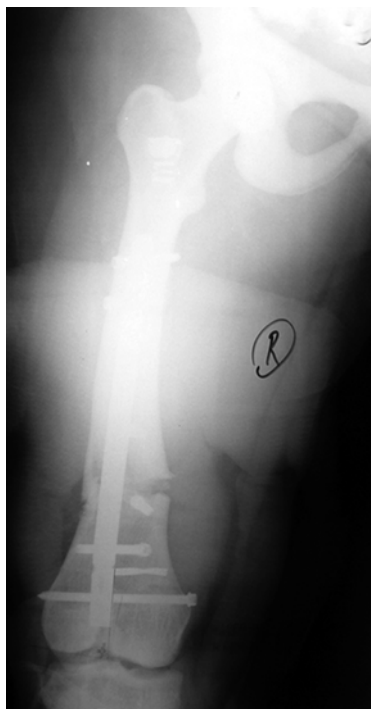


Fig. 15.26 AP view following intramedullary static fixation

Box 15.3

- The deformities are multiapical (long bowing) and tend to recur.
- Intramedullary nailing should be performed always if possible to prevent recurrence.
- Preoperative planning is of paramount importance, and should contain paper-tracing to simulate the surgical procedure. This will increase the precision of the procedure while shortening the surgical time.
- Fixator-assisted nailing is very effective for accurate deformity correction and for prevention of recurrences.

Renal osteodystrophy is noticeably different from either nutritional or hypophosphatemic rickets. It is often driven by the presence of secondary hyperparathyroidism, which leads to activation of osteoclasts and resorption of bone [2, 4].

Features of both rickets and hyperparathyroidism are present in children with renal osteodystrophy. Rachitic changes include lack of calcification in the zone of provisional

calcification of the physis while secondary hyperparathyroidism induces osteoclastic resorption of bone.

Serum vitamin D and calcium levels are usually low, accompanied by increased levels of blood urea nitrogen and creatinine in the blood along with acidosis (see Table 15.1).

Children affected by renal osteodystrophy are often short-statured and have fragile bones. These patients often have bone pain and fractures occur easily. The most common orthopedic manifestations are skeletal deformities, usually genu valgum, periarticular enlargement of long bones, slipped capital femoral epiphysis (SCFE), muscle weakness, and Trendelenburg gait especially if SCFE is present [4]. On radiographs, widespread osteopenia with thin cortices and unclear trabeculae are present. The underlying bone has a ground glass appearance. The physes are increased in thickness, with an indistinct zone of calcification. In severe and persistent renal failure, aggressive lytic areas in long bones may develop (*brown tumor*). Since many patients with renal failure, especially those who undergo renal transplant, are treated with steroids, osteonecrosis can also develop.

Treatment

Medical Treatment

Treatment of the causal renal disease is of crucial importance. Dialysis and renal transplantation are prolonging the survival of these patients. Medical therapy is initiated with the use of 1,25 dihydroxy form of vitamin D. The use of calcitriol significantly decreases serum PTH levels and delays secondary bone changes [22]. The treatment of acidosis with sodium bicarbonate also improves metabolic bone disease.

Decreased skeletal growth and short stature is a significant problem, probably due to disturbances in the growth hormone—insulin like growth factor axis. Recombinant human growth hormone (rHGH) restores growth in these children. However, rHGH, also weakens the physes and may predispose the child to develop SCFE [23]. In patients with renal osteodystrophy, who are refractory renal osteodystrophy to medical treatment, parathyroidectomy may be indicated.

Orthopedic Treatment

Patients with renal osteodystrophy are generally referred to the orthopedic surgeon for the treatment of three pathologies: angular deformity of lower extremity long bones, SCFE, and avascular necrosis [4]. Any surgical intervention in this patient population should be carefully considered, as the perioperative risks are amplified due to associated anemia, hypertension, bleeding tendencies, and electrolyte imbalances. The risk of infection is additionally increased in patients with a renal transplant who are under immunosuppressive therapy.

Angular Deformity

Angular deformity occurs in renal osteodystrophy because the bone is soft, undermineralized, and prone to bend with weight bearing. Genu valgum is the most common deformity, but genu varum or a windswept deformity may also occur [4, 24]. If the beginning of renal osteodystrophy occurs before 4 years of age, varus deformity may develop, because the normal alignment of the leg is in slight varus, which then is accentuated when the bone becomes weak. Similarly, older children are prone to the development of genu valgum because of the physiologic valgus alignment of the lower extremity. Valgus at the ankle may also accompany the genu valgum [25].

Some milder deformities will correct with medical treatment of the renal osteodystrophy [4, 25]. Deformities do not respond well to bracing. If the patient becomes symptomatic, and has had optimum medical treatment without resolution of the deformity, corrective osteotomy should be accomplished [4, 17, 25]. Usually, the greatest deformity is in the distal femoral metaphysis, but sometimes, a supplementary proximal tibial osteotomy is also needed. Internal or external fixation may be used. While external fixators have been successfully applied, taking care of achieving stable constructs and utilize of hydroxyapatite coated Schanz pins, but bone healing may be delayed [26]. Recurrence of the skeletal deformity is common in patients with continuing metabolic pathology, so medical treatment should be adjusted before and continued after corrective osteotomy. Elevation of serum alkaline phosphatase concentration above 500 U/L is a worthy marker of ongoing metabolic bone disease [4, 25]. Milder deformities may respond to hemiepiphyseodesis [17].

Slipped Capital Femoral Epiphysis

The clinical picture of a child with SCFE secondary to renal osteodystrophy differs from the usual cases of SCFE. Often, these patients are younger, and obesity is not a part of the clinical picture. Bilaterality is also very common. Radiologic pathology in the physis is more pronounced, accompanied by physeal widening and generalized osteopenia [27]. The goal of standard management of SCFE is to prevent further deformity and promote early closure of the proximal femoral physeal growth plate. However, cessation of proximal femoral growth may not be desirable in a young child with renal osteodystrophy. Moreover, physeal healing is compromised due to the underlying metabolic pathologic condition. Fortunately, pain and physeal widening resolves with initiation of appropriate medical treatment. If the slip is displaced or symptoms continue despite of medical treatment, fixation with one screw, accomplishing stability across the physis without closing it, should be performed.

Avascular Necrosis

Prolonged use of corticosteroids is the likely cause. Avascular necrosis occurs frequently bilateral, affecting the hip. Treatment is usually symptomatic [4].

Hypophosphatasia

General

Hypophosphatasia is a rare, genetic defect of alkaline phosphatase production, resulting in pathologic mineralization of bone. There is a wide variation in the severity of the disease and the prognosis depends on the age of onset; perinatal, infantile, childhood and adult hypophosphatasia can occur [4, 28]. The genetic defect for this disorder is determined to be in the tissue-nonspecific alkaline phosphatase gene (*TNSALP*), and many different mutations have been described [29].

The pathology detected in hypophosphatasia closely resembles that seen in patients with rickets. Osteoid production remains undisturbed, but deprived of effective *ALP*, mineralization of osteoid is compromised [4, 28, 29]. As a result, the physes are widened with persistence of the zone of calcification and metaphyseal remnants of cartilage islands. If hypercalcemia is added to the clinical picture, heterotopic calcification *foci* can be formed. In laboratory tests, *ALP* levels are decreased not only in the serum but also in tissues like kidney, bone, etc. [1, 4]. Serum phosphorus, vitamin D, and PTH levels may remain within normal limits, but hypercalcemia is sometimes present. Characteristically, urine pyrophosphate levels are increased [1, 4].

In the severe perinatal form, the infants may be stillborn. If they survive delivery, they frequently die from respiratory infection in early infancy. All bones display severe demineralization on radiographs. The infantile form starts later, usually around 6 months of age. These babies have demineralized bones with severe rachitic changes. Fractures and bowing of the extremities are frequent. If these children can survive infancy, they tend to improve clinically, but they are short statured throughout childhood due to absence of normal endochondral bone growth [1, 4].

Treatment

Although there is no conventional medical treatment of hypophosphatasia, successful bone marrow cell transplantation with improvement of the disease has been described [30].

Fractures and deformities need orthopedic management. Fracture healing is commonly delayed. Multiple osteotomies with intramedullary fixation are often required to correct the

bowing and provide structural support to the long bones [4]. Thus, when possible it is advised to utilize an intramedullary nail in all corrected bone segments.

Medical control of the underlying disease is of paramount importance as the deformities tend to recur in various metabolic bone diseases. Therefore, consultation with the endocrinologist should be done preoperatively and continued after the surgical intervention.

Example Cases

Case 1: Figures 15.28, 15.29, 15.30, 15.31, 15.32, 15.33, and 15.34 show the treatment sequences of a patient (hypophosphatemic rickets) with a profound bilateral femoral varus deformities.

Case 2: Figures 15.35, 15.36, 15.37, 15.38, 15.39, 15.40, 15.41, 15.42, and 15.43 show the treatment sequences of a patient (hypophosphatemic rickets) with a profound genu valgum deformity.

Case 3: Figures 15.44, 15.45, 15.46, 15.47, 15.48, 15.49, 15.50, 15.51, and 15.52 show the treatment sequences of a patient (hypophosphatemic rickets) with a profound genu varum deformity.

Case 4: Figures 15.53, 15.54, 15.55, and 15.56 show the treatment sequences of a patient (X-linked rickets) by guided growth technique.



Fig. 15.28 A patient's orthoroentgenogram showing severe varus deformities

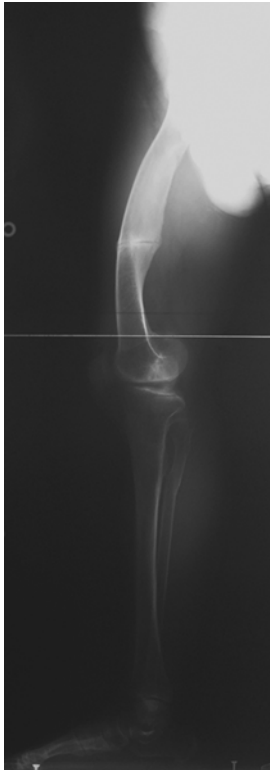


Fig. 15.29 Right lateral orthoroentgenogram shows anterior femoral bowing deformity. Note there is pending stress fracture at the femoral diaphyseal level

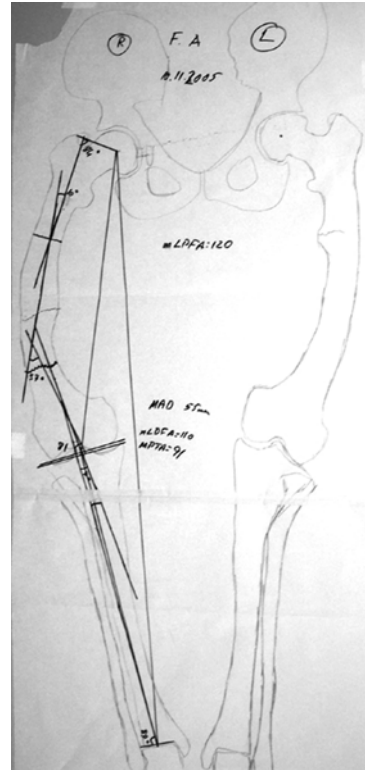


Fig. 15.31 Paper-tracing for deformity analysis

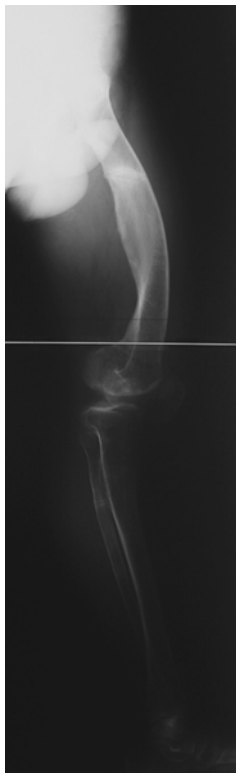


Fig. 15.30 Left lateral orthoroentgenogram shows anterior femoral bowing deformity

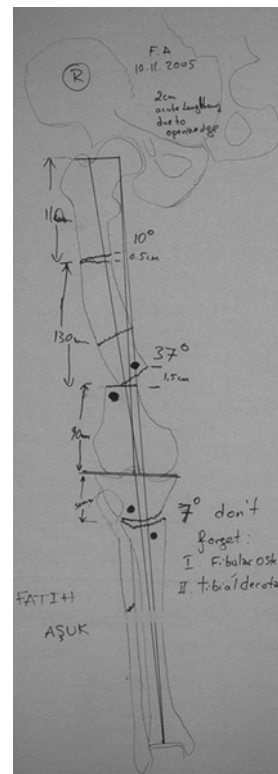


Fig. 15.32 Paper-tracing for surgical simulation



Fig. 15.33 Orthoroentgenogram following FAN procedure at the femur and tibia on the right



Fig. 15.35 Orthoroentgenogram of a patient with severe genu valgum deformity

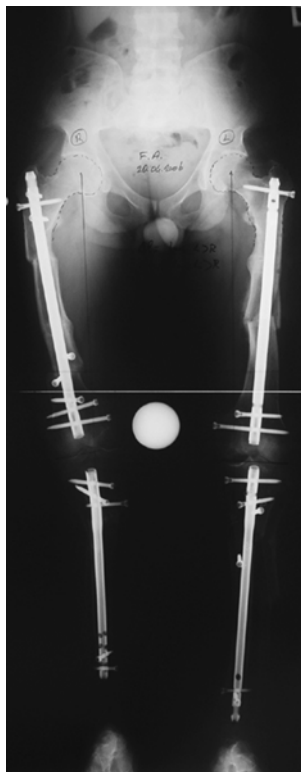


Fig. 15.34 Orthoroentgenogram following final FAN procedure at the femur and tibia on the left



Fig. 15.36 Right lateral orthoroentgenogram of the same patient



Fig. 15.37 Left lateral orthoroentgenogram of the same patient



Fig. 15.39 X-ray of the right femur following FAN procedure

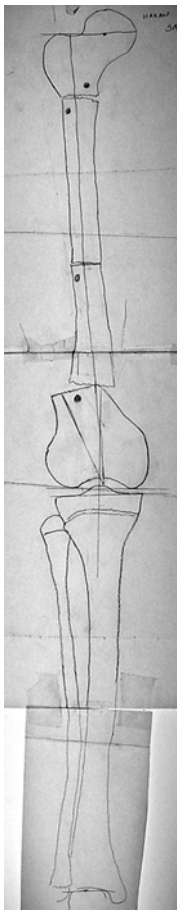


Fig. 15.38 Paper-tracing for surgical planning



Fig. 15.40 X-ray of the left femur following FAN procedure



Fig. 15.41 Orthoroentgenogram at the end of the treatment



Fig. 15.43 Photographic documentation of the same patient after the treatment



Fig. 15.42 Photographic documentation of the same patient prior to surgery

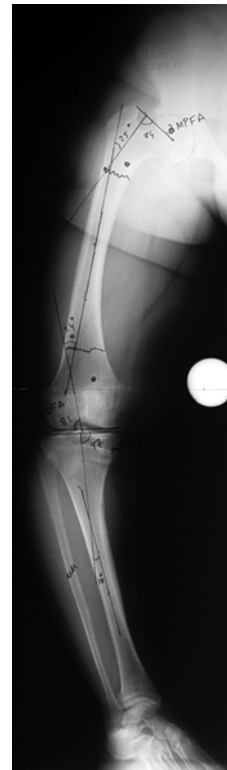


Fig. 15.44 Right frontal orthoroentgenogram showing severe genu varum

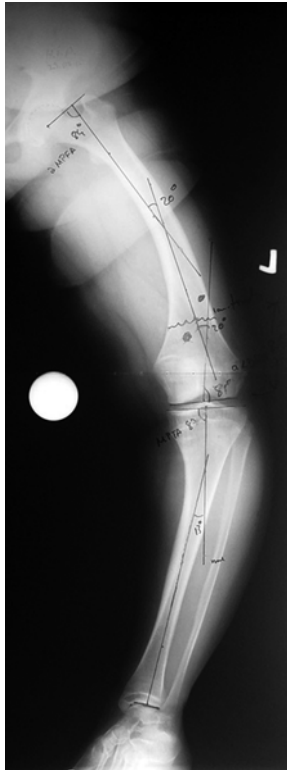


Fig. 15.45 Left frontal orthoroentgenogram of the same patient showing severe genu varum



Fig. 15.47 Left lateral orthoroentgenogram showing anterior bowing (same patient)



Fig. 15.46 Right lateral orthoroentgenogram showing anterior bowing (same patient)

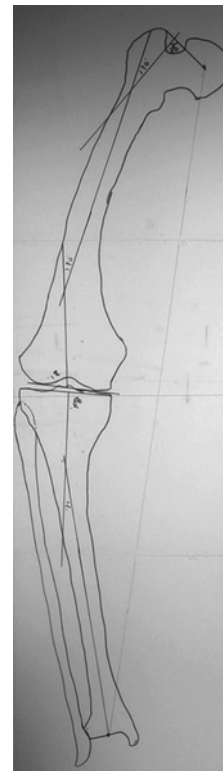


Fig. 15.48 Paper-tracing for deformity analysis on the right

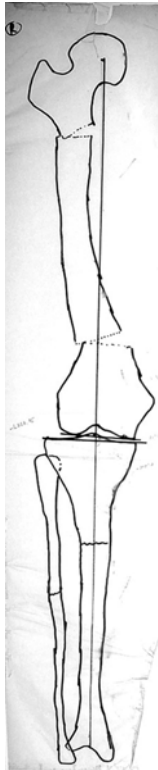


Fig. 15.49 Paper-tracing for surgical simulation



Fig. 15.51 Photographic documentation of the same patient prior to surgery



Fig. 15.50 Orthoroentgenogram at the end of the treatment



Fig. 15.52 Photographic documentation of the same patient after the treatment



Fig. 15.53 Orthoroentgenogram of a patient with X-linked rickets prior to treatment



Fig. 15.55 Orthoroentgenogram of the same patient after 3 months later



Fig. 15.54 Orthoroentgenogram of the same patient after application of hemiepiphysodesis

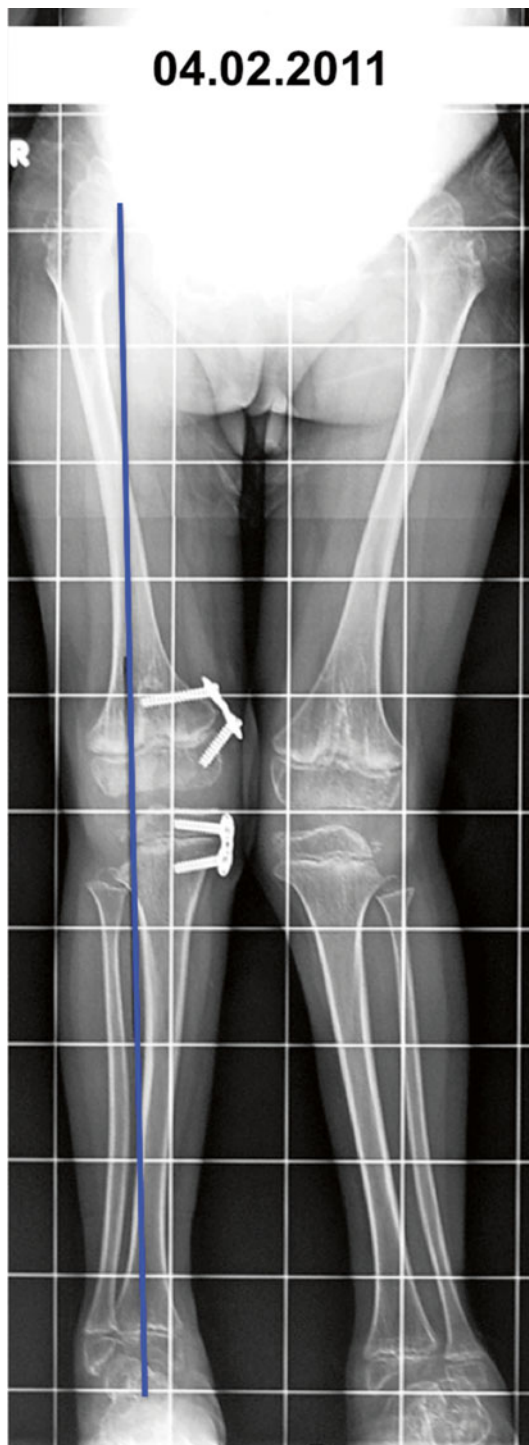


Fig. 15.56 Orthoroentgenogram of the same patient after 7 months later. Please note the correction of the mechanical axis

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