CONGENITAL PSEUDO-ARTHROSIS OF THE TIBIA

The Findings in One Case and a Suggestion as to Possible Etiology and Treatment

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INTERMITTENT pressure of sufficient force, when applied to bone at widely spaced intervals, often causes a productive reaction and new bone formation. Years ago Hugh Owen Thomas treated dislocating patellae by means of staccato blows of a small hammer delivered to the anterior surface of the lateral femoral condylar ridge forming the outer border of the patellar groove. Time between treatments was sufficient for the subperiosteal hematoma to organize and calcify. This treatment, in certain instances, resulted in the formation of an exostosis at the hematoma site.

Roentgenograms of bunions often show on the medial side of the metatarsal heads areas of bone repair and bone absorption occurring simultaneously. These irregularities reflect the reaction of bone to various pressures which the prominent portion of the metatarsal heads receive (fig. 1).

Bone tends to absorb or disappear in the face of constant abnormal pressure placed upon it. This pressure may be of the slightest degree. Bone, nevertheless, will be absorbed at the point of abnormal pressure. Absorption continues until the bone frees itself sufficiently from this abnormal pressure to maintain a physiological balance of pressure between its structure and the force acting upon it.

Parham bands have fallen into disfavor mainly because of the normal physiological response of bone to constant and unusual pressure. An unyielding constrictor surrounding bone causes rapid absorption of the bone under the band. The cortex is thus greatly weakened in this area by the disappearance of much of its substance. This process ceases only when a physiological balance is achieved between the bone and the band, or when the constrictor is removed. Because of these facts and failure to appreciate their significance, insufficiency fracture at the band site may follow treatment of this sort.

Contiguous soft tissue tumors may press upon bone. This pressure, though constant, may be slight, for in certain cases the pressure effect is distributed in all directions and to all tissues. Bone, however, compensates for this pressure and strikes its physiological balance, not by stretching or moving, but by absorption. If the tumor continues to grow, the bone retreats before the soft tissue advance. If the tumor ceases to grow, a physiological balance is quickly set up between the tumor and the bone. Condensation and evidence of bone repair are then visible in the bone at the juxtaposition of its surface and the soft tissue mass. Bone reacts in like fashion to soft tissue tumors within its substance (fig. 2).

A growing long bone in a young child may be subjected to marked angulation due to fracture. This deformity with subsequent healing may occur in the midshaft far away from the area of influence of the epiphyseal plates. Yet, over the years this bone may partially or completely straighten out. Bone that has ceased to grow or has completed most of its longitudinal growth will not exhibit this property.

Following fracture with uncorrected angulation, young long bone forms new bone substance on the concave side of the angulation. This area is subjected to compression forces in line of bone length. At the same time, lateral pressure is diminished by the relaxation of surrounding tissues. This affords a space for the confinement of a hematoma.

On the convex side at the apex of this

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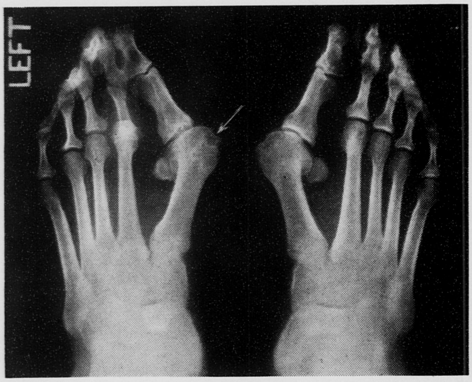

Fig. 1. The effect of extraneous pressures on bone is here illustrated.

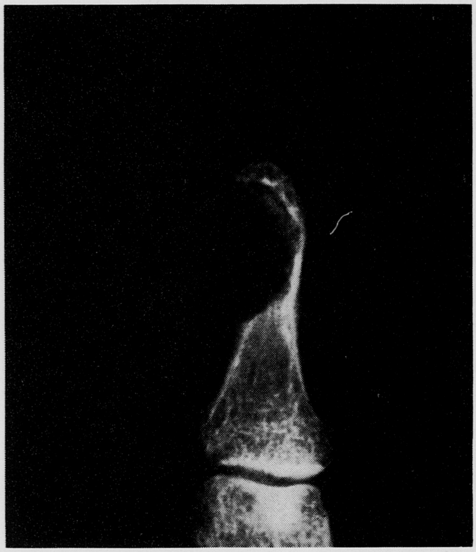

Fig. 2. The effects of continuous pressure on bone of a contiguous, non-malignant, soft tissue tumor is illustrated in this x-ray of a terminal phalanx.

fracture the situation is different. The force is more or less one of distraction in line of bone length. The lateral pressure of the tissue against the bone is increased over normal, due to the stretching of the surrounding tissue over the apex of the angle.

As the bone grows and its length is increased by pushing out of new bone from both ends towards the middle, the tissue tensions tend to remain constant, with the greatest pressure exerted on the bone at the apex of angulation. This pressure favors absorption of bone at the apex on the convex side. Gradual removal of this bone, with development of new bone on the concave side, diminishes the actual degree of angulation. This process continues until a physiological balance is struck between bone and tissue. If growth persists long enough, the physiological balance demand forced upon the bone by the soft tissue runs concurrently. If longitudinal growth ceases, the abnormal pressure increments also cease. The bone quickly strikes its balance with the soft tissue and a residual angulation, if present, will remain.

Other factors are also present, but it would seem that the forces mentioned above play some part in the moulding or straightening of rapidly growing long bone in the young.

The inhibitory effects on bone production due to the pressure of a dense scar or due to the pressure of a split thickness skin graft, when these pressures are directly applied to bone, are only mentioned to be recalled.

As stated previously, intermittent pressure widely spaced may lead to bone production. Conversely, intermittent pressures applied to bone at shortly spaced intervals have the physiological effect of constant pressure. The continued patency of a foramen for a nutrient artery, the ability of the pulsating dura mater to free itself from normal bone, and the scalloping out of vertebral bodies that may be produced by an aortic aneurysm are all examples of this type of intermittent pressure.

Certain constriction rings starting in the skin can eventually go deeper and, through pressure, amputate the part, including the bone. Ainhum is a disease to point.

The well known facts cited above indicate that normal bone responds in a predictable way to the influence of abnormal pressure. The type and nature of the pressure determine the response in bone.

The purpose of this paper is threefold. First, we wish to suggest that congenital pseudo-arthrosis of the tibia may well be related to and possibly grouped with congenital constriction ring, congenital absence of a portion of long bone, and congenital amputation of limbs.

Second, we wish to entertain the thought that congenital pseudo-arthrosis

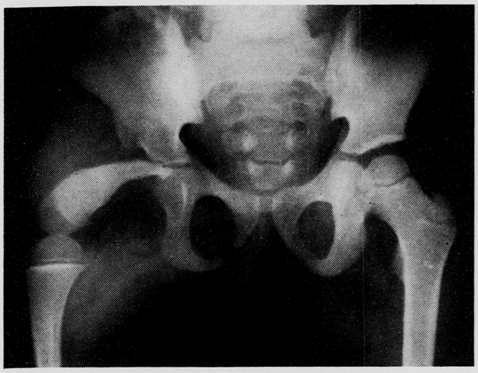

Fig. 3. Congenital absence of femur with tapering of bone well illustrated.

of the tibia may not be due to any inherent fault in the bone, but that it may result from a normal physiological response of bone to soft tissue pressures of a peculiar and extraordinary nature.

Third, pseudo-arthrosis of the type mentioned is not a common condition. Cases of the disease tend to gravitate into hospitals for children.

The statements put forth here and the treatment suggested in this one case can be proved wrong or right if a number of men pool their experiences. This writer has had no real clinical adventure with congenital constriction ring or congenital amputation of limbs.

In congenital absence of the upper four-fifths of the femur, the condyles are often present and articulate at the knee with the tibia. Superior to the condyles the shaft rapidly tapers off in carrot-like fashion to form a point. Above the termination of the shaft no bone exists (fig. 3).

In a specimen examined, this writer distinctly remembers that at the distal end of the shaft and at the upper portion of the condyles, the periosteum appeared normal in texture and thickness. As the shaft was approached proximally, the so-called periosteum became thicker and thicker as the bone tapered to become smaller and smaller.

Many tibiae presenting congenital pseudo-arthrosis taper to needle-like points at the fracture site. Other tibiae contain multiple cysts within the bone. The site of fracture may or may not be through a cyst. Still other tibiae that are continuous at birth show marked anterior bowing. These at times develop pseudo-arthrosis. In each instance some form of fibrous dysplasia in the region of the pseudo-arthrosis seems to be present. Does the bone become insufficient in structure and give away because of abnormal soft tissue pressure? Is healing delayed or prevented by a soft tissue mass of tumor-like consistency?

Case Report

This female child was born of English and Swedish parents on July 2, 1939. The family history is immaterial.

An anterior bowing of the right tibia was present at birth. A fracture of this tibia was first noticed when the girl was about 30 months of age. X-rays at this time showed two cysts in the tibial shaft superior to the fracture. No cyst was visible at or below the fracture line. The fracture was directly transverse with no feathering of the bone ends. No displacement was present. No marked sclerosis or evidence of bone healing was revealed by the x-ray plates. There was a generalized decalcification of the tibia and fibula. Both bones were smaller than their fellows. The fibula was relatively smaller than the tibia. Marked anterior bowing was present.

An operation was performed using bone chips taken locally. No attempt was made to correct the angulation. A long leg plaster was applied, and later this was changed to a long leg brace. The anterior bowing and nonunion persisted, so the brace was worn continuously.

When the child was 6 years of age, an operation was performed using an inlay graft from her ilium. This graft was placed in the tibia from the front, and it went through about one-half the depth of the tibia. Union occurred at the graft site. Some weeks later the plaster was removed and a well fitted long leg brace with complete leg cuff was substituted. The anterior bowing was not corrected. Refracture occurred about six months after surgery.

Study of x-rays taken during this time showed no evidence of any attempt by bone to develop healing in the posterior half of the tibia. The fracture site in this area remained distinct. The graft appeared continuous. There was no evidence of any new subperiosteal bone production anywhere about the fracture site. Sclerosis was not marked, but each end of the fracture site appeared to be closed. The medullary cavity stopped about one-half inch short of each bone end. The bone cysts were still present.

The patient was first seen by us during June, 1947. Examination at that time showed a well formed youngster. The right leg exhibited scarring and was smaller than its mate. At this time the patient presented frank nonunion and marked anterior angulation of the right tibia at the junction of its middle and lower thirds. X-rays showed the thin fibula to be intact. One and one-half inches of actual shortening was present. The right foot was in equinus and was much smaller than the left. Attempts to dorsiflex the right foot were halted by a tough, thick band of palpable tissue lying between the calcaneal tendon and the tibia. A small dimple in the skin was present on the medial side of the tibia in the region of the fracture site. This dimple was not related to any scar and was firmly attached to the thick underlying tissue. The dimple was not movable, although the surrounding skin was freely so.

About the fracture site, the leg appeared much fuller and thicker than normal. It appeared as though a collar of heavy tissue were in the area. This tissue appeared to taper from its center to either end. Palpation anteriorly revealed the tibia. Palpation laterally, medially, and posteriorly revealed the tibia to be much more deeply seated than the same portion of tibia on the left (fig. 4).

During the two years this child has been under observation other findings are of interest:

1. No scoliosis has developed to date.
2. Following the loss of deciduous teeth, the permanent teeth appeared months later than is expected in the average child.
3. The patient has silky, very fine, blonde hair and blue eyes.
4. The skin is light and thin. There are no areas of abnormal pigmentation.
5. Pain sense is less in the right leg and foot than in the corresponding areas on the left. The fracture site could be manipulated freely and the patient would complain of little, if any, pain.

The Course and the Findings:

An operation was performed July 1, 1947. Under tourniquet an anterolateral incision, beginning at the level of the tibial tubercle and extending to one inch above the ankle, exposed the whole tibia between the epiphyseal plates. A longitudinal incision of the complete extent of the periosteum was then made. This was striking. The upper portion of the periosteum was of normal thickness and color. As the fracture site was approached, the periosteum became much thicker and reached its maximum thickness within one inch of either side of the fracture site. The color changed from the normal pearl gray to a yellowish white, tough-looking fibrous tissue. Passing by the fracture site, the periosteum gradually regained its normal appearance as the lower end of the tibia was reached. The point of greatest thickness of this fusiform mass was estimated to be one-quarter of an inch on the lateral and medial sides of the bone and somewhat thicker posteriorly. The tissue stripped easily from the bone except at the defect. Here it occupied spaces within the bone as well as being continuous throughout the extent of pseudo-arthrosis. Because of its mass and toughness, retraction of it from the bone was very difficult.

The fracture site was thoroughly curetted. A subcutaneous osteotomy of the fibula was done three or four inches above its distal epiphyseal plate. This allowed better contact of the tibial fragments and also allowed correction of the anterior bowing. The medullary cavities were then opened by means of curettes.

Following this, a window was made on the medial side of the tibia about an inch below the tibial tubercle. A curved metal rod was inserted through the window and run repeatedly up and down the medullary portion of bone, entering and breaking up the continuity of the two bone cysts.

A tibial graft taken from the other leg was then split longitudinally. Notches were made medially and laterally in the tibia well above and well below the fracture site. The ends of the grafts were forced into these notches. This gave a slight outward bow to the lateral graft. The ends of the grafts were fixed with

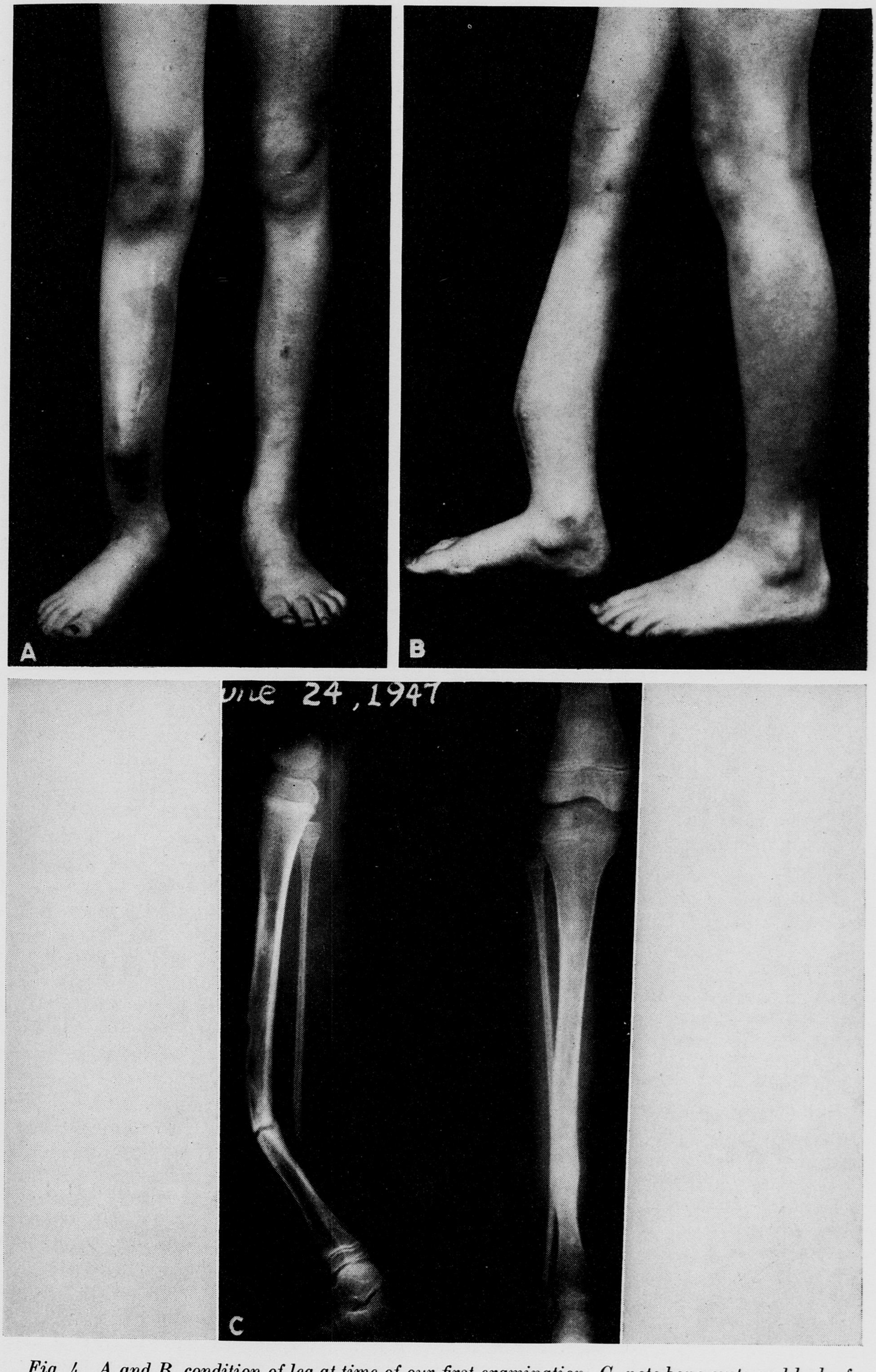

Fig. 4. A and B, condition of leg at time of our first examination. C, note bone cysts and lack of productive reaction about fracture site.

small Kirschner wires cut flush to the grafts. An inlay tibial graft was put across the fracture line in the anterior portion of the tibia. Contact between the ends of tibia at the fracture site was good. After fish-scaling of all exposed ends of the tibia not covered by grafts, many bone chips were placed about the fracture site.

Because of the correction of the angulation, it was possible to close the thick cuff of tissue easily and with excellent apposition. Plain gut was used here and silk was used for the skin. A thin dressing was then applied, and one thickness of elastic bandage was run from foot to groin. Over this, a single plaster spica was applied which included the foot and extended to the nipple line. The knee was in fifteen degrees of flexion. Recovery was uneventful.

The spica remained on for ten weeks. At ten weeks, gentle manipulation showed rigidity at the fracture. X-rays at this time showed continuity of the grafts and some new bone production. In a comparison of these x-rays with postoperative ones taken before the spica was applied, some decrease in the outward bowing of the lateral graft was seen, although the pins did not appear to have shifted. A skin tight plaster was applied from toes to groin and the patient was allowed to use crutches without bearing any weight on this leg. Serial x-rays were taken.

At the end of five months, x-rays showed fragmentation and complete flattening out of the grafts. The pins had shifted and migrated, and there was springy motion at the fracture site. The patient was placed in a skin-tight, long plaster with a walking sole. Full weight bearing without any other support was then started.

Nine months after surgery, after four months of weight bearing in plaster, nonunion still was present. Bone production was not progressing but rather was retrogressing. Decrease in callus and absorption of the bone grafts were evident. Motion had increased at the fracture site (fig 5).

At this time it was decided to build the patient a bone to act in the nature of a second tibia. To accomplish this, the fibula was to be moved at each end into the tibia, well above and well below the fracture site, following which, a tibia or a femur from the bone bank was to be fashioned to slip over this transposed fibula. The ends of the transplant would be fitted into the tibia at the new tibiofibular junctions and then further attached to the tibia by means of transverse grafts run through both bones at set intervals. In line with this plan, the upper portion of the fibula was moved into the tibia.

In preparation for the other stages to come, the x-rays were frequently viewed and studied. We became impressed by the flattening of the tibial grafts and their fracture, which seemed strange, since neither graft had been firmly fixed. Furthermore, bone production began normally, went well for a few weeks, and then seemed to be choked off. Then a retrogression of the normal bone healing seemed to have taken place (fig. 5).

It seemed worth gambling that constriction and pressure might have been the cause of failure of the bone to take hold and maintain itself in accordance with the well known laws regarding bone's response to function. We then reasoned that the abnormally thick periosteal cuff which we had so carefully replaced might be a factor. For this reason, the fracture site was opened on June 17, 1948. The loose pins were removed and a complete and radical resection of the thick periosteal cuff was done throughout its extent. Careful dissection was necessary to free the skin from the cuff at the dimple site. The cuff was about six inches long; it was not as thick as when first viewed, but was still many times thicker than normal periosteum. It fitted the bone tightly and continued through the pseudo-arthrosis. Nothing more was done. The pseudo-arthrosis was left untouched except for the removal of a small piece of bone for section, and no bone was added. The wound was then closed. A snugly-fitting, long-leg plaster with sole piece was applied. The patient was kept in bed for ten days and then allowed full weight-bearing without support other than the plaster (fig. 6).

Plasters were changed and x-rays taken every six weeks. At twelve weeks, the fracture site was much firmer than it

Fig. 5. A, operation with tibial grafts and copious amounts of bone chips. B, two months later. One pin has extruded and has been removed as it was subcutaneous. There is evidence of bone production. C, later bone production continues. Grafts have not fragmented. Invagination of fragments remain evident. D, grafts fractured. Bone production has ceased. E, absorption of bone is now evident, with marked recession of bone production. F, definite nonunion. Compare with C. Absorption and lack of further bone production is evident.

had been six weeks before. Only a suggestion of motion could be elicited. New bone was evident on the lateral, medial, anterior, and posterior portions of the fracture site. At the end of eighteen weeks, the fracture site was clinically solid. X-rays at this time showed maturing of the new bone, which extended completely around the fracture site. The site began resembling the typical "wiped joint" appearance of normal healing bone. This had never occurred previously in this case. Since that time, the bone has continued to mature, and the leg has remained solid (fig. 7). The patient is still in full weight-bearing supports, for which Castex has been substituted for plaster of paris (fig. 8).

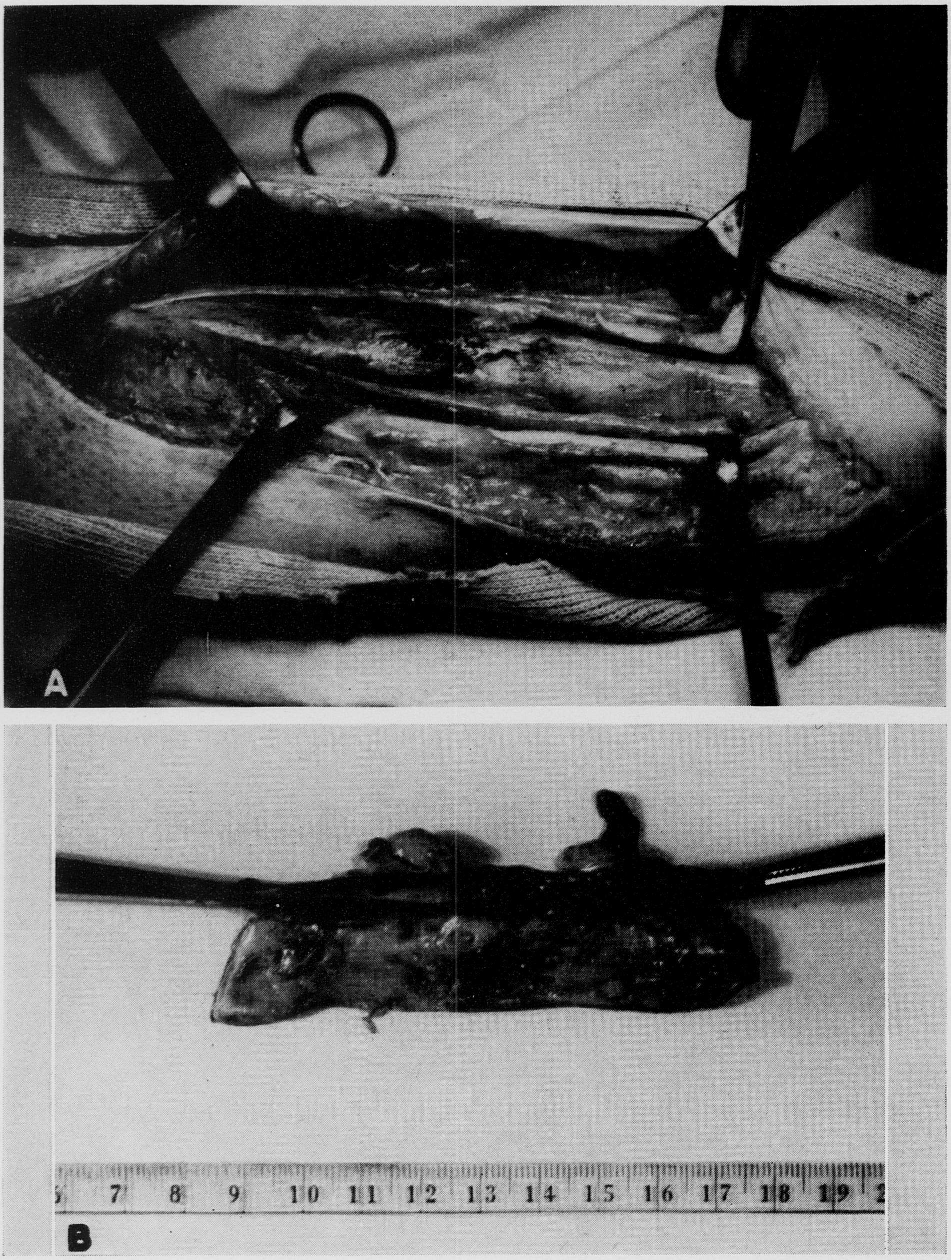

Fig. 6. A and B, operative findings. A, note thickness of periosteal cuff and tag of periosteum that exactly fits the defect on the anterior surface of the tibia shown in fig. 5, F. B, the resected cuff that was completely removed. (PATHOLOGICAL REPORT: *Bone consists of irregularly shaped piece of tissue 0.4 and 0.5 cm. in maximum dimensions. The external surfaces are irregular, white and tan.*

Sections prepared after decalcification show a large amount of dense collagenous connective tissue in which there are scattered bony trabeculae. The connective tissue is for the most part sparsely cellular and at a few sites it contains lymphocytes and large mononuclear cells. The trabeculae vary in size and have irregular margins. DIAGNOSIS: *Marked fibrosis of bone.*

(Legend concluded at bottom of facing page.)

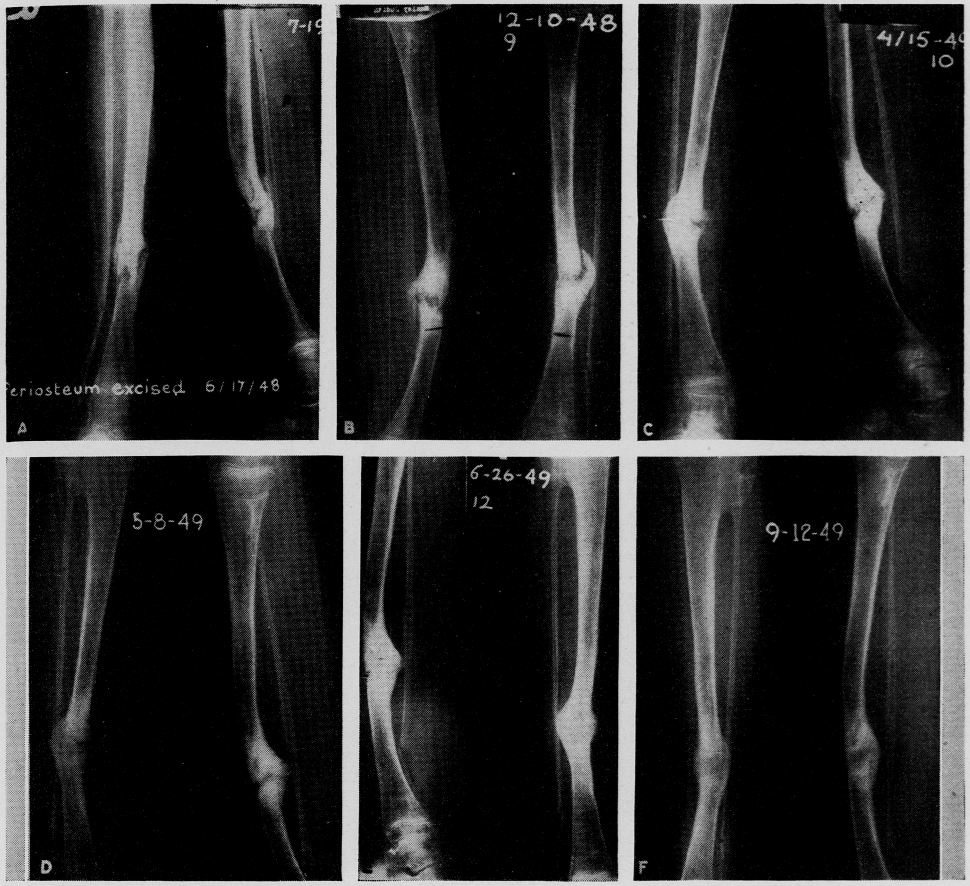

Fig. 7. A, note anterior defect with small piece of bone raised while taking biopsy. B, bone production continues. Light union at this time. Note how anterior portion of tibia has proliferated. Note building up of bone all about fracture site. C, while playing badminton, patient broke cast just below fracture site, angulating fragments. No real free motion demonstrable. Without anesthesia, tibia was straightened manually through gradual, continued pressure over a period of few minutes while plaster was setting. D, one month later. Fracture again solid with evidence of bone healing. E, complete bridging practically established. F, bone growth continues.

Comment

At present there is no evidence that union will be maintained in this leg. The child is now 10 years of age, which might be a deciding factor in this case. There is no proof that really substantiates the contention that the removal of this very thick, constricting fibrous mass had anything to do with the union that occurred and that has been maintained with steady improvement in the bone's appearance.

The removal of this tissue, however, seemed to:

1. Allow rapid, spontaneous union, in spite of the fact that there was no disturbance of the fracture site and no addition of bone. Spontaneous union in a case of congenital pseudo-

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Periosteal cuff. *Irregular shaped tissue measuring 2.0 cm. in maximum dimension. The external surfaces are irregular, pink, green and white. The tissue is firm in consistency and sections with marked resistance. The cut surfaces are white, pink and green.*

Sections prepared after decalcification show a small amount of bone and a large amount of connective tissue with attached fat and skeletal muscle. The bone shows no noteworthy abnormality. Dense collagenous connective tissue shows focal infiltration with lymphocytes and large mononuclear cells. In some parts there is an occasional multinucleated giant cell. There is no evidence of malignant tumor in sections examined. Diagnosis: *Focal chronic inflammation of collagenous connective tissue.)*

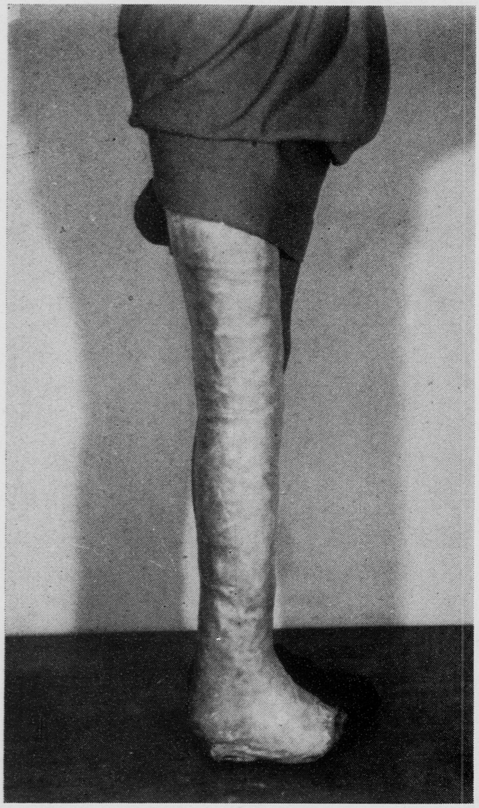

Fig. 8. Patient at present. Shoe to fit over Castex cast not shown.

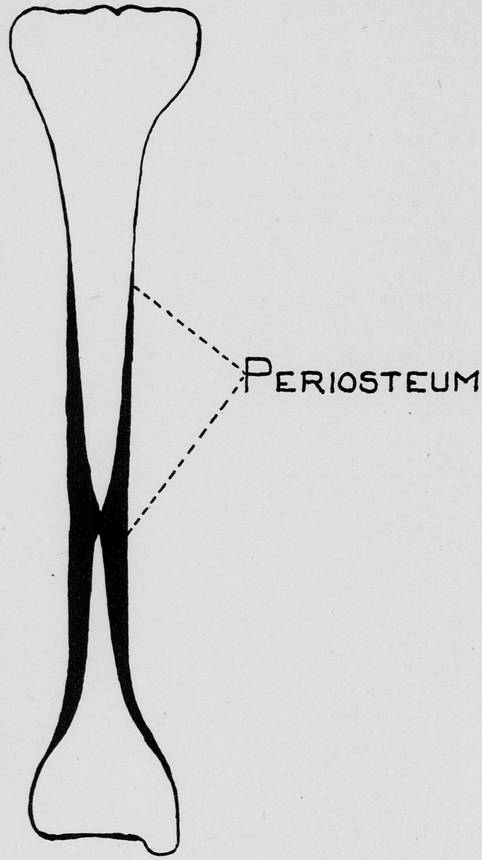

Fig. 9. Diagram showing a conception of how cuff might appear in cases where bone tapers. If cuff is replaced over graft or if bone is placed beneath it, or if cuff is left open but contiguous to bone, the abnormal pressure of this cuff tending to establish its normal pattern may be one reason the bone fails to remain and flourish. Resection of this cuff may be another aid in combating congenital pseudo-arthrosis of the tibia.

arthrosis is, in itself, remarkable.

2. Once union arrived, it progressed satisfactorily. The pattern of bone healing closely resembles that in normal fracture healing. This had not been the case prior to the removal of the tissue. Previously, lack of subperiosteal bone formation, failure of bone production, some sclerosis, and retrogression of healing after a few weeks seemed to be the pattern in this case.

The heavy cuff of fibrous tissue that enveloped the fracture site may have been secondary to the nonunion. It may be nature's way of attempting structural support at the weakened bone site. This cuff was thinner at time of removal following good support than when it was first viewed months before. The cuff may have been primary. This can be determined by study of other cases. Primary or secondary, a cuff as found in this case might diminish the productive reaction of bone as in other known conditions that cause abnormal confinement, constriction, and pressure about bone.

If the periosteal cuff is thick and abnormal in the condition of congenital pseudo-arthrosis, it would seem that complete, meticulous, and radical removal of this cuff is indicated prior to repair of the fracture site. Certainly, bone for transplant, that has been either stored for weeks or used immediately, acts well and with purpose when its surrounding bed is favorable. It is almost inconceivable that one small portion of a tibia fails to unite because of congenital fault of bone, or failure of bone to heal itself if given a normal bed in which it can flourish. Perhaps tibiae with congenital pseudo-arthrosis, that have been or will be amputated, if devoid of their surrounding tissue, might make normal bone transplants. It seems to this writer that the bone itself is good, but that some extrinsic factor is working on it unfavorably (fig. 9).

Healing and refracture following repair of congenital pseudo-arthrosis is not uncommon. It may be that temporary relaxation of the fibrous tissue through surgery allows bone healing to begin. As the fibrous tissue reorganizes, a vicious circle is, however, again set up, leading to disappearance of bone retrogression and fracture.

If, after removal of the fibrous tissue bone progresses normally for awhile, only to show signs of retrogression later, exploration would be indicated. A new mass of fibrous tissue may have formed, which should be removed. This may have to be done again and again, until the structures surrounding the tibia offer circumstances similar to those in which we find bone beneath normal tissue.

We believe that in a case of this type a brace is no substitute for a well fitting plaster. If bone healing continues to progress satisfactorily, the patient will be kept in long-leg casts until union is complete, until there is no sclerosis, until the bone is sufficient in size, and until the medullary cavity is continuous throughout the affected area.