



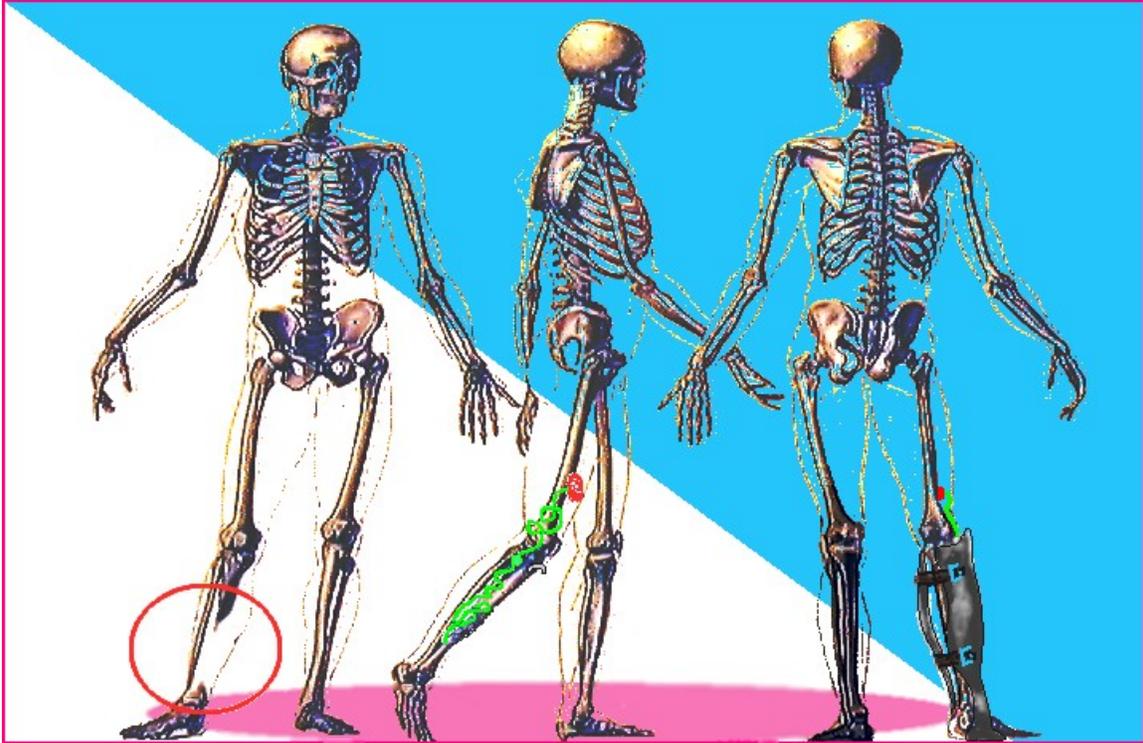
CONGENITAL PSEUDARTHROSIS OF THE TIBIA

A whole new way of thinking about it.

Abstract

Just dropping the words 'failure to make bone' and considering a backward running formation-lysis cascade, changed everything.

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Disappearing Bone variant of Congenital Pseudarthrosis of the Tibia

a possible insight to disorders of osteoclast metabolism.

Preamble to the original older report:

This is a public offering at over 32 years follow up from the beginnings or [?] 19 years post 14 year report (presented at several orthopaedic meetings – see cv for details). It is hard to get comprehensive publication for single cases (paragraph to page if lucky). But this case defies such format without gutting all that is important about it.

A second case, NV, is separately included. Though not as extensive a defect it was a multiple level defect with the severe one more central and the lesser ones more at the periphery (less structural problem). The treatment was targeted at the ‘problem’ leaving the lesser areas as controls provided by nature. Later the lesser areas were included in a partial way then more completely.

What was learned from this was applied to other conditions with dissimilar appearances and names but with theoretical possible connection. We will address all of it, the solidly determined and why not then the less defined and less proven maybe-could-be stuff as fuel for thought (though not for conviction).

It really begins not with CPT but with abject failure of every known form of treatment to even show poor response and the meeting of the ethics committee the night before planned amputation with parents to review the reports from just about every well known expert in the world – all of whom had a single agreed plan – amputate. Only one had reservations, John Hall, MD. He warned that though amputation was a definite immediate requirement, be sure that the parents are NOT told that it would make life so much easier (as if a cure). The reason was that this form of CPT – known informally as the ‘disappearing bone’ variant – is very difficult to get to a long term a stable stump for prosthetics. He warned that revisions would be many.

The mother then drops her head with breath release. She lifts it and asks “Where does it go?” Where did all that graft and transplant go?”

The non doctors offered quick dissolving based light weight offerings. The medical doctors were layering on about the actual published concepts of why bone does not form and even with the implanted bone stimulants and external stimulators it *failed to form*.

I choked on her question. As close a quote as I can remember, “This cannot be a failure to FORM bone. We put live donor bone in there with every trick to keep it alive known and there was no trace of it ever having existed within three weeks. Bone does NOT dissolve. Neither alligators nor snakes digest bone. Prehistoric bones are scattered all over the earth. Dogs rely on bones not dissolving when they bury them. This bone is being digested!”

By what?, from the group.

Has to be osteoclasts, I said without hesitation. They are the only cells that digest bone. It’s a complicated process.

But all the pathology reports from everywhere. Nobody mentioned...

It is only an osteoclast, *by name*, if under the microscope it looks like an osteoclast. But maybe a tumor of osteoclasts – or maybe the very cells which were seen by everybody in great excess – fibroblasts – had somehow come into possession of osteoclast active digestive metabolism – a DNA spillover? They act like osteoclasts as they have the recipe!! Bone IS being made. It is being digested faster.

How do you...

CALCITONIN! It stops the pathway to digest bone and it also stops cell division of cells with that chemistry – I hope... over time reducing the nasty cell ratio . . . maybe?

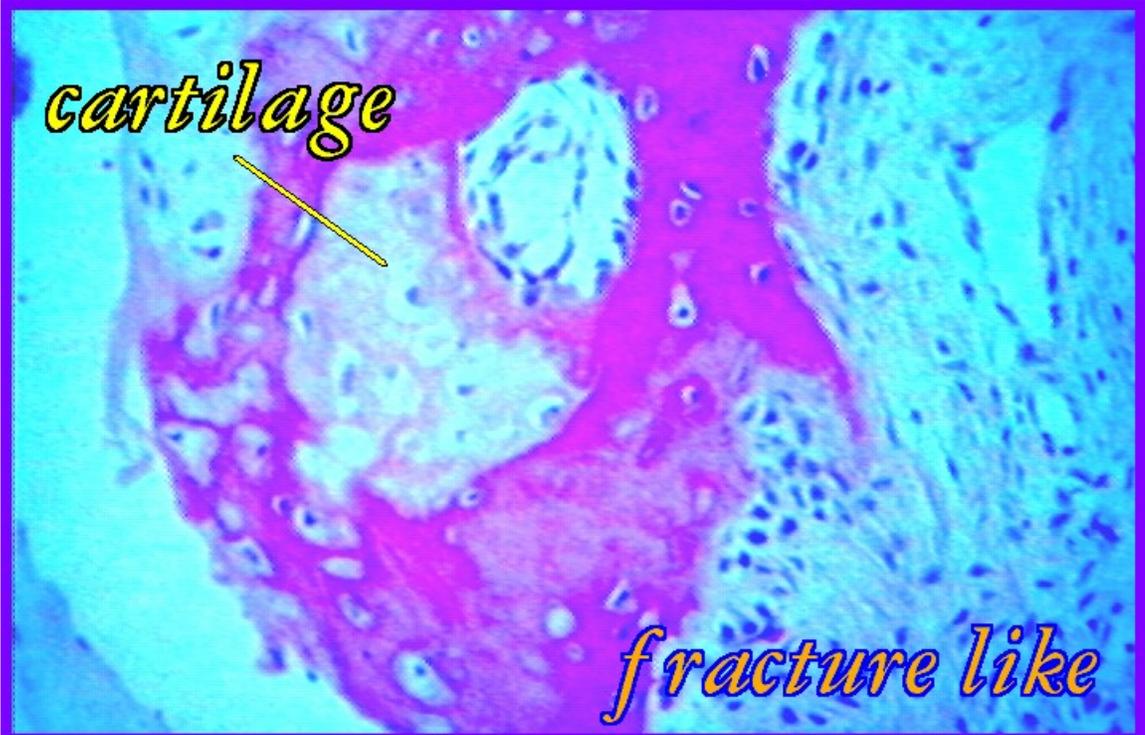
Meaning?

Well, tomorrow we were going to lop a leg off. It can wait, right? Why don't we bathe a new graft in calcitonin and see if THAT changes the game?

The totally quiet room of big eyes looking left and right as if at a tennis match heard only the father and mother in perfect synchrony, "DO IT".

Evening into the late hours, the neurosurgical nurse and I went through all the implants available to the neurosurgeons, looking for small (had to fit in a baby thigh) and slow delivery. Using methylene blue & saline we found which hydrocephalus tubing pump valve when placed backward would leak for sure but slowly. Upstream - a porta cath. That and some loops of tubing at the side of the knee would be the growth loops. Meanwhile, at Ciba Geigy (down the street from our hospital) the veterinarians who responded to our late evening plea for a night time conference were reviewing everything they did with small animals that would give us a safe starting first dose. We would be using their new synthetically made human calcitonin.

I have to share with you that that neurosurgical nurse (LB) stayed with this case and she and several others who had assisted with the prior failed bone grafts were crying as this was done. Where today, just before we cut, we have a 'time out' to say aloud our surgical plan - to reveal any errors - we stopped for prayer and I lost it. That was good. With catharsis we really plowed through this thing. The rest is pretty much how the original presentation less emotionally describes:



Congenital pseudarthrosis of the tibia (CPT) is a focal defect with a spectrum of severity. Categories depict scope at presentation so as to anticipate unresponsiveness to treatment. (2-5,8,12,18,20-21).

A severe form of CPT has substantial segmental narrowing of bone with obliteration of the canal. Cystic appearing loss of bone is seen in the occasional most difficult cases. Though not strictly part of the classification scheme, a subtype of this severe CPT group is called by several names by practitioners in the field, including "disappearing bone variant" (1 A,B) of congenital pseudarthrosis (DB-CPT). Distinguishing characteristics include fine vascular-like honeycomb soft tissue at surgery which is nondescript histologically, and an amazing speed of bone destruction which follows grafting.

CPT interventions have included electrical stimulation, pulsed electromagnetic field stimulation (PEMF), autologous and homologous bone grafting (avascular and vascularized), geometric bone reversal, bone transport, external and internal fixation in assorted configurations, and an array of supportive modalities.

Establishment of a stable vertical column of continuous bone which can be loaded by weight bearing is the goal common to all forms of nonablative treatment. Reports dealing with the most aggressive of surgical methods have had little good to say about the more severe cases. In fact, DB-CPT has maintained so bleak a prognosis that early amputation has been commonly advised. Even so, stump problems are common (1 A).

A singular case is reported for four reasons. This is the only reported case employing a predominantly biochemical attack against CPT. This is the youngest human, known by the drug manufacturer, to be regularly treated with calcitonin (13,23,29). The clinical course of this case, belonging in the worst category and followed fourteen years, is unique

in the abruptness and extent of alteration in the pathology following treatment. Lastly, the severest forms of CPT are rare enough to warrant dissemination of any promising observation.

CASE REPORT:

SB was born of a normal pregnancy at full term but with a floppy deformity of her



distal right leg. Family history was negative for neurofibromatosis or its stigmata (2,9,12) and any other limb anomalies. Evaluation was otherwise negative. X-ray showed a lucent cystic looking area sized about one fourth the

length of the diaphysis replacing most of the distal third of her right tibia. A thin thread of apparent anterior bone, as seen on x-ray was soft to palpation and nonstructural.

At 18 days of age, complete excisional biopsy was performed with autograft placed



in the defect. The thin anterior bone density that was seen on x-ray was gritty soft fibrous-like tissue. A significant margin of bone as seen on x-ray was soft and pliable. Most of the shaft of the entire distal half of the

tibia was soft and mechanically unsound.

Abnormal tissue residing within and around the defect appeared to have many fine lakes similar to those seen in lymphatic or vascular tissue anomalies but without bleeding. They did ooze blood in a fine manner but did not bleed like cut vascular tissue. The fibula was not surgically exposed.

Tissue (including that of the following subsequent surgeries) was reviewed by many

pathologists including those of the United States Armed Forces Institute of Pathology (AFIP). No exact tissue diagnosis was established. It was thought to be some sort of odd fibrous tissue. Although an unusual tumor could not be absolutely excluded, it was deemed benign. By histochemistry, it was not neurologic in origin, although occasional small areas had a passing similarity to neurologic derived tissue. Microscopic characterization was summarized as: Morphology nonspecific, fibrous tissue with focal areas of lamella woven bone, foci of necrotic mineralized debris with palisading histiocytes and giant cells, negative S-100 immunostain (not neurofibromatosis). The bone graft was totally gone by exam and x-ray within two to three weeks.

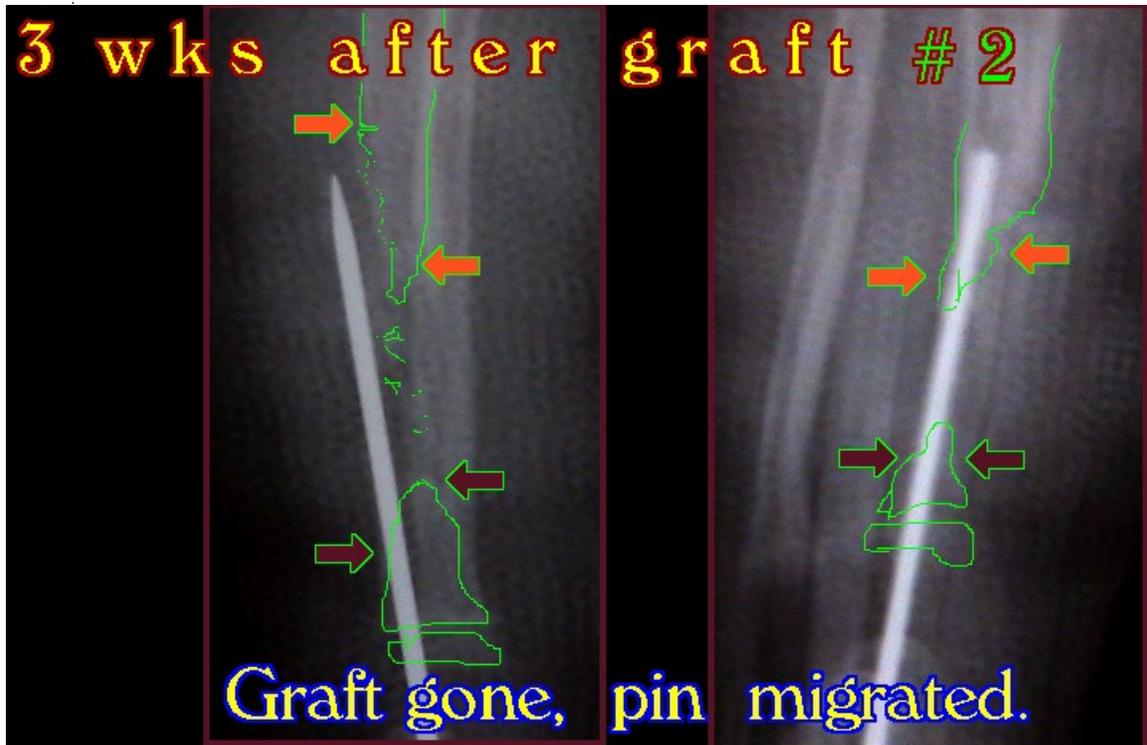
A series of surgeries followed with escalating attempts to span the long gap between



the distal and proximal fragments.

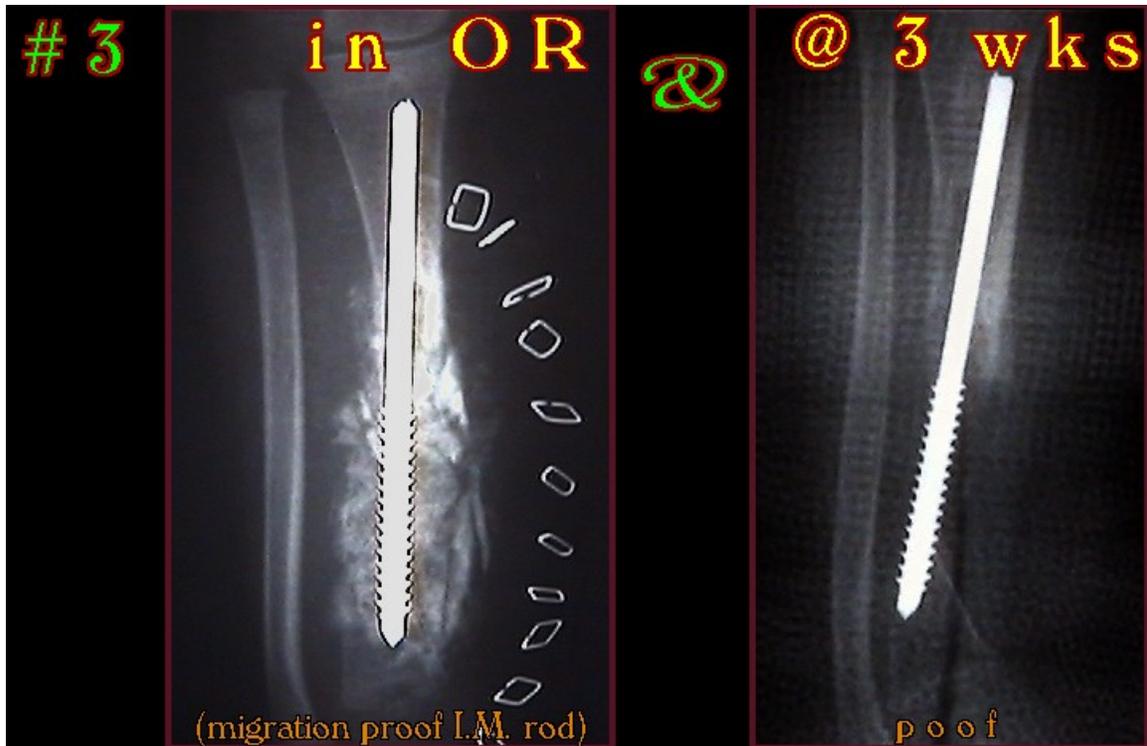
Immobilization with casts was continued throughout early treatment and changed regularly to try to protect as well as maintain the tibial side length and prevent ankle angulation. Casts or

full circumferential braces were used throughout the span of treatments.



At two months of age, repeat resection of all nonessential tissue in the area was performed along with bipolar cautery of margins. Very large iliac crest bone graft plus periosteal (iliac) graft was placed. The periosteal sheet included a strip of iliac cartilagenous apophysis. The chubby leg was defatted to make space. Stability was established by a longitudinal intramedullary rod passing through calcaneus on and into the proximal tibia. Pulsed EMF treatment with a custom made coil was very well adhered to. Surgery & PEMF quickly failed. With no x-ray visible bone graft at three weeks. The floating metal migrated.

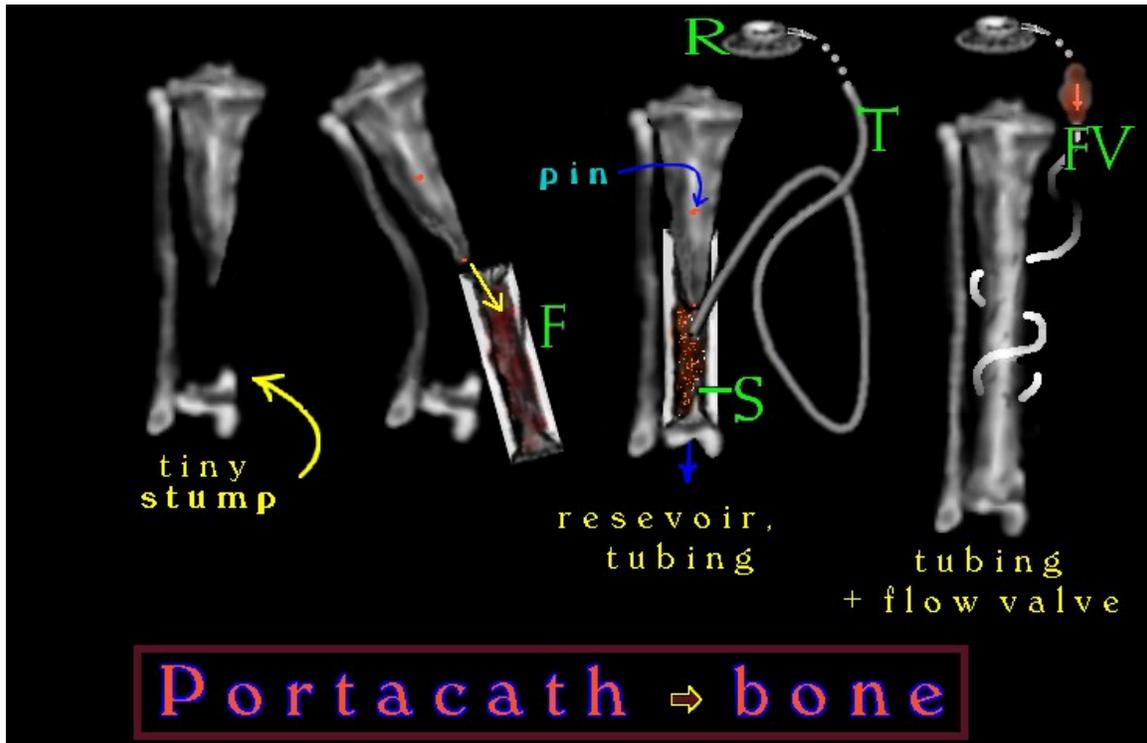
At four months of age, tissue resection of all nonessential tissue was repeated. Large iliac crest graft was used to supplement a dense cadaver bone implant. A different sort of intramedullary rod was implanted (one threaded end to retard metal migration). PEMF was



continued. Surgery & PEMF again quickly failed with minimal residual traces of bone implant on x-ray at three weeks.

Following each surgery very rapid implant loss was apparent within the week of surgery. Bone disappearance was slightly more rapid for her own iliac bone than for the more dense adult cadaver fibula bone, but only slightly so.

At one year of age a different approach was tried. Because the distal fragment was so small with barely any bone proximal to the growth plate, it was decided to envelop it.



An adult cadaver fibula allograft was transplanted. Power burr funneling of both ends formed two vase-like receptacles into which the narrowed tibia ends of the tibia could be nestled (by bending the patient's own fibula and letting it spring back). The allograft had its medullary canal first packed with finely crushed bone (like fine sand) to serve as a biological sponge. A rush pin, passed through the proximal tibial shaft from above, stabilized the structure. As the total distal fragment was less than one cm. in length, the rod was passed through to the calcaneus. The funnel ends of the allograft were finely fluted to allow fluid leakage.

The midshaft of the allograft was drilled very obliquely to just allow entry of a small plastic catheter tubing (PortaCath) which was advanced into the center of the allograft. The

rest of the fibula graft was filled with ground bone similar to sand to serve as a leaching pool. Small drill holes allowed sutures to secure the catheter to the graft. The PortaCath reservoir was placed in a subcutaneous pocket in the anterior thigh. A loop of catheter was also placed in that pocket away from the expected site of thigh injection. The catheter was passed along the medial side of the knee at the neutral axis of flexion and then into the tibia area where a second loop of excess catheter was allowed before threading it into the inside of the allograft. Additional bone 'sand' was placed around the two ends of the allograft to act as a seepage area outside the bone.

Saline was injected through the catheter to assure intact flow and to measure the fluid volume of the system. This volume of saline was selected to suspend Calcitonin to prime the system.

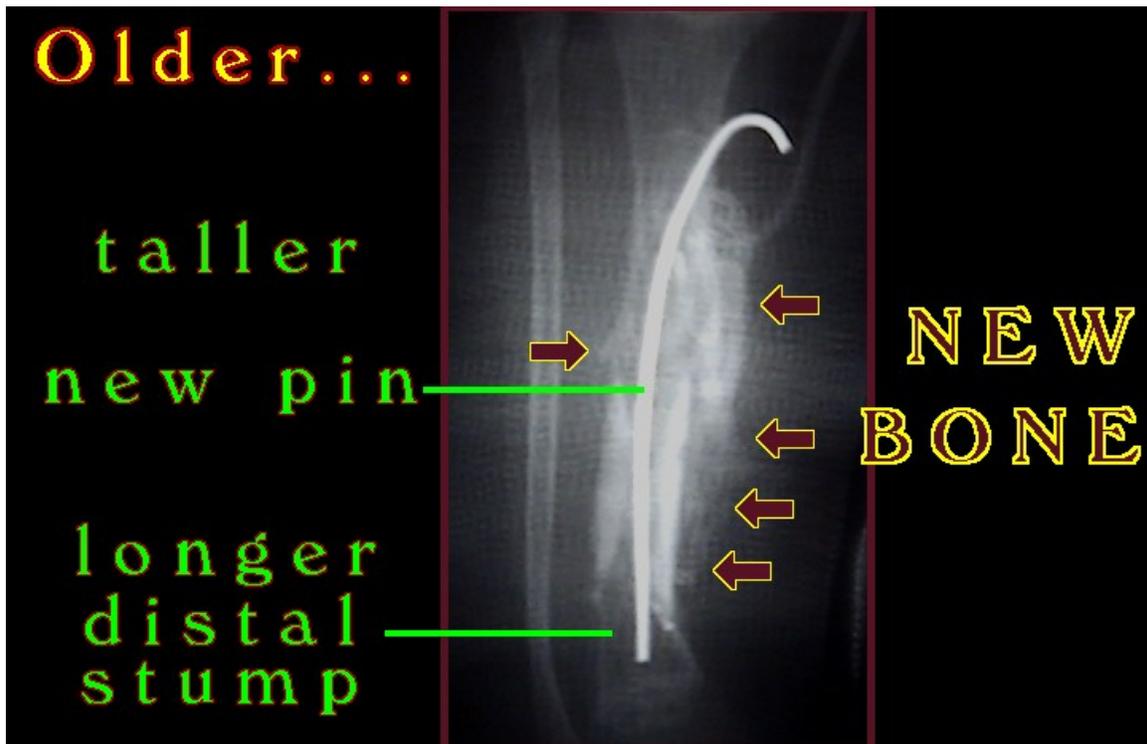
Calcitonin 0.5 mg (synthetic human, Ciba-Geigy) was injected each week into the reservoir. PEMF was discontinued. To minimize fear and allow subtle evaluation (as she was watched for one hour continuously after every injection), a very fine needle injection of 2% Xylocaine anesthetized the skin over the PortaCath reservoir. Later, with diversion, the Calcitonin was injected into the reservoir (noncoring Huber portacath needles).

Initially, there were no outward physical findings noted relative to the weekly administration of calcitonin. Blood tests of electrolytes taken 1 hr. after Calcitonin injection were the similar to pretreatment. One month later, the mother noted that each day following calcitonin injection the baby would saturate each diaper rather than just wet the diaper. She did not think there was any alteration in eating or drinking volume on any one day as compared with any other. On one occasion a brief pink flushing of the palms was noted 30 minutes after injection with no other findings and no distress. The child was playful and

happy through all this.

At about four weeks post op, more resistance to reservoir injection was noticed. Over time each reservoir injection was done more slowly as back resistance further increased. At a point when bone x-ray density increased significantly, the child displayed brief dry retching 45 minutes after injection.

At 18 months of age, new bone engulfed the ends of the allograft which also had additional bone density within. Because of deficient length caused by limb growth, the catheter was replaced. At surgery, the new bone was found to be grossly solid, bonded to and replacing the allograft. The patient's own bone was much more like normal bone than prior to calcitonin treatment in that it was hard and had a durable feel to metallic probing. The canal of the allograft was solid except for the path of the Rush pin.



The plastic tubing configuration was altered. Dual (Pudenze Y-connector, Heyer-Schulte) sump catheters curled about the outside of the bone, one low and one high (lower:

Heyer-Schulte Peritoneal Reflux Control Catheter, higher: Heyer-Schulte ventricular catheter to provide less outflow resistance proximally). The firm incorporation of the lower tibial segment and allograft into one solid unit allowed replacement of the old metal pin with a more proximal one that did not cross the ankle physis.

Calcitonin powder was, from this point on, reconstituted in only one fourth of the supplied diluent, a volume of fluid that was less than one fourth of the combined volume of the well and tubing so as to use diffusion as the mechanism a diminish systemic effects. The same therapeutic dose of 0.5 mg calcitonin was injected weekly via the thigh reservoir.

Eventually, the sump catheter delivery system became plugged. Air study showed blockage within the tubing. Clotted serum inside the tubing had blocked the fine holes of the sump. A new tube with full diameter open end was substituted. A proximal ventricular reservoir with high flow pressure valve was interposed in reverse proximally in the delivery system to create a larger reservoir dead space for slower diffusion distribution. Additional bone graft was placed although the previous graft bone was still present. The old graft appeared to be in continuity proximally. It was in continuity with itself as multiple strips were now joined. The distal part was intricately entwined around and in the distal stump, but not united to it. The new tubing outflow was positioned to favor the distal junction.

Twice weekly PortaCath injections followed. Different dilutions were tried, but always with 0.5 mg calcitonin. Clearly volumes using over 25% of the provided diluent (one cc. provided) caused reddened palms within 20 to 30 minutes and a giddy reaction. Thirst was a definite reaction to larger volumes of injection. Calcitonin injections of under 25% of the provided diluent had no visible effect on the patient.

Continued growth over time required replacement of the tubing system and a new intramedullary pin, confined to the tibia. It was noted that bone growth was most thick and exuberant where tubing was looped. At surgery it was noted that the fluid tracked from the end of the tubing back along the sides of the tubing. Medium pressure one way flow valves were placed to increase the dead space within the tubing PortaCath system. With this configuration calcitonin is injected into the reservoir in a volume that is less than the volume proximal to the flow valve. Diffusion through the closed valve is then the only distribution mechanism after injection.

During periods of illness, injections were suspended. It was noted that remodeling occurred during respites and also that bone loss significantly slowed during these off periods as she got older. Weekly injection was made biweekly then monthly. Injections were then suspended at ten years of age. Rush pins were traded for size during her growth. At 13 years of age, during a routine follow up, a stress fracture of the pin was seen on x-ray, without any significant change in the bone itself. She had been active as a cheerleader. She was unaware of the metallic event. Because of her maturity, leg length discrepancy and moderate midtibial varus it seemed wisest to combine metal retrieval with final limb correction. The patient was referred for limb alteration.

RESULTS:

No broad or sustained systemic reactions to the calcitonin injections were noted. Excellent bone formation continued in the absence of bone destruction. Allograft incorporated into the patient's own bone proximally as well as distally.

Dr. Herzenberg's results here...

Dr. John Herzenberg was consulted and took over the upstream (proximal tibia) lengthening to offset the collapse and undergrowth that had gone on over the years and finalize alignment. With ring devices and a central rod and graft that went quite well. Continuity of bone was not lost and normal length was attained.

Presently this 14 [add 19 more years no losses, but minus braces] year old patient is ambulatory with an external free ankle orthosis as a lateral stress shield allowing full vertical loading. Bone formation and modeling has continued to progress. The incredible destruction of bone witnessed earlier is not seen at this time. Bone age, blood glucose, urine and blood electrolytes, and endocrine studies have been normal to date. Other bones have shown no density or maturation effects of the calcitonin treatment thus far. Behavioral, neurologic and general milestones are all normal.

Periods of no injection occurred during the treatment period. No injections were given when ear aches, sore throat, chicken pox, or the like was ongoing nor if colds appeared imminent. It was noted that remodeling, that tubulation, absent despite bone formation, did occur during these medication lapses. So did some thinning of the previously healed lytic area. A cycle of 5 day, 7 day, and 14 day injections seems to provide both bone formation and retubulation as well as a mechanism for vacations. The robust activity that was allowed by establishment of a longitudinal bone column has also correlated with better bone shape. There remains a mild ankle varus. A leg length difference is noted with continued growth.

DISCUSSION:

Although no therapeutic or mechanistic conclusions can be drawn from a single case, one observation cannot be ignored, a brutally destructive process was altered in a setting where such alteration is not otherwise expected. This concept needs to be considered in several ways.

We are now in an era of bone stimulating biologics. But this case did not respond to electrical bone stimulation, nor was chemical stimulation used. Calcitonin is not a bone stimulant. Calcitonin shuts down osteoclasts. It turns off osteoclast action and it also stops osteoclast reproduction. Tissue growth with osteoclasts not reproducing may shift the relative osteoclast osteoblast ratio. Although abnormal tissue did not look like osteoclast cells, the conclusion must be that it had the chemistry and biologic workings of osteoclast cells. Calcitonin is very rapidly degraded by the body upon injection. Sustained effect would require sustained or pulsed delivery to the disordered site. Because of the initial tiny size, an actual pump was not feasible. Using a reversed neuro VP shunt valve was effective. Calcitonin delivery depended on the fact that such flow valves are not perfect and will leak. That slow leak was the pump.

There were no guidelines as to tolerable or optimum dosage as no patient of this age or size had ever been reported to have been given this hormone. Using weight extrapolations derived from treatments of the smallest individuals with other diseases, an arbitrary dose schedule was guessed at. There is nothing that tells us whether this dose is a good one, or whether the interval selected is appropriate to the characteristics of the drug other than the hard experience of clinical usage.

Calcitonin treatment was conjured from an entirely unproven premise. That premise

was derived, in turn, from the perception that bone was not merely failing to be incorporated, nor merely failing to form nor failing to heal, but rather that it was being rapidly, methodically, and efficiently digested.

That a random focal quirk could come up with the complex biochemical machinery to so thoroughly remove bone in so spectacular a way was deemed unlikely. This was the work of experts - osteoclasts. To limit systemic effects in an otherwise normal infant exposed to a potent hormone with very fast diffusion and absorption properties and potentially far reaching systemic consequences led to implementation of the scheme described above.

Remodeling, a positive function of osteoclasts is suppressed, a negative of calcitonin treatment. With altered cycles of no treatment remodeling has been noted. It is not complete. Over ten years the destructive aspect of the disease was attenuated nearly to zero. And yet, throughout, a boundary between proximal bone and distal bone always seemed at least in part to remain, even when bridged. Stability may have been as much engulfment as linkage. Shortening may be either loss or growth failure. As linear growth is the action of the bone ends and not the diaphysis, we can only assume that there was diaphyseal micro loss through long periods of end growth. That would also account for the varus tendency.

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[This list is as of 19 years ago at time of presentation]

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**Also See Dr. Herzenberg many refs on
Leg Lengthening & Ilazarov Methods**

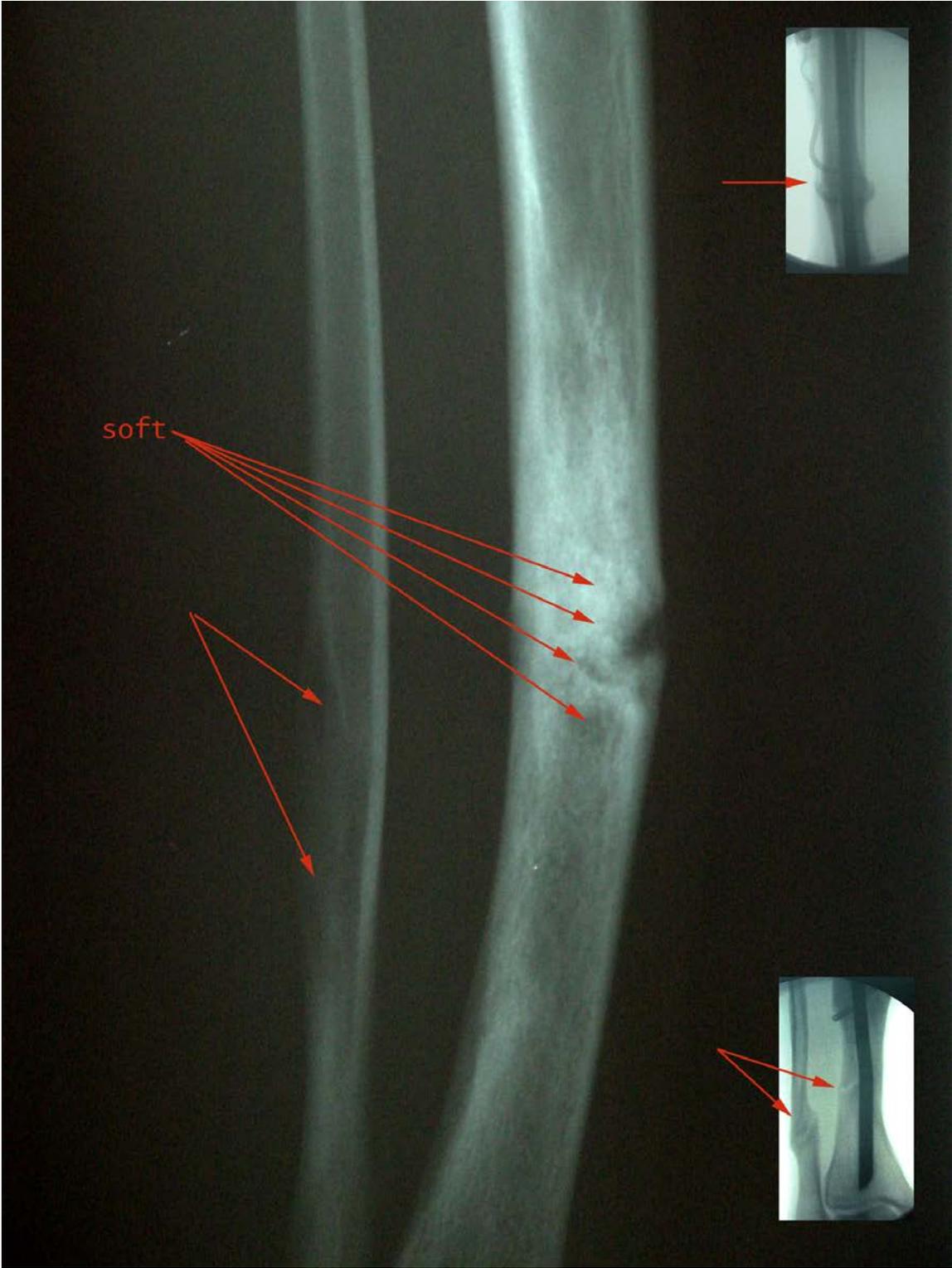
Special thanks to Clinical Reference Systems, Ltd., 5613 DTC Parkway, Suite 350, Englewood, Colorado 80111 for the foreign trace and their translations and to Mr. & Mrs. Bruegel in tracking these sources, and so quickly.

*Thanks to research dept. of Ciba-Geigy for calcitonin data and general assist.
Thank you Lynda Baker, RN for being you.*

*Here is a case of CPT, not of the 'disappearing bone' sort but interesting in being multifocal yet absent any findings outside the involved leg (followed 20 years).
Less words more images?*



Despite electrical bone stimulator and focal bone grafting, the CPT persists. But also note several fibular sites and a very fine (seen only on very inclined views) distal fine line defect.



Bone graft to the main defect just ended in a larger looking defect. However at graft time the scooped away 'bad bone' was not structurally sound but rather soft gritty tissue pretending to be solid looking bone on x-ray. Image insets show what happened after a catheter that leaked calcitonin to the graft area. Bone.

From what was learned from SB (first case presented), calcitonin allows bone formation. But bone piles up and does not remodel. So off periods allow resorption and reshaping. The process has thus altered from a continuous bath to an intermittent one.

As lytic cell population cell volume drops over the years in both cases the severe loss of bone stops at about mid teen (post puberty, not early puberty).

Even so with this patient looking quite womanly, the totally gone initial defect – the one treated had become replaced by the lesser –seemingly trivial defect as the limiting functional item. It caused intermittent pain acting as would a stress fracture.

Falling back on the leaching idea but of lesser scope the half cross section of tibia that seemed to hold this problem was scraped out and Grafton Putty packed with an onQ pain pump filled with saline and 2ml of salmon calcitonin (human not available and salmon having been found to work well also by us). That half of the tibia healed. We did not want to risk a 360 degree discontinuity. But over the next three years have come to do the other half the same way.

The fibula defects persist but are not a clinical problem.



X-rays showing the main defect that was treated with calcitonin as healed. The pretreatment lesser defect persists and will come to two steps of treatment (half at a time fearing 360 degree defect. That worked.

The success of calcitonin in this diagnosis was brought to mind when confronted with a young girl with total femur fibrous dysplasia – presented as neck fracture. The entire length of femur from hip to knee was woefully hard to see – so thin was the calcification. It was possible to simply use a hypodermic needle and make three test injections (at three levels) in that bone of calcitonin. Two weeks later three stripes of more density was obvious (but bone?).

A most difficult threading of the femur with a rod guide wire from greater trochanter distally (staying center with nothing to stop going out of the ‘canal’ region (mush the entire length) took hours. Then stepwise reamers created an actual hollow. That hollow was filled with bone sand that had been soaked in calcitonin. A thoracoscope from lateral mimicking (although percutaneous) the path of a hip nail or screw, hollowed the femoral neck and head - then likewise packed. The inferior head neck junction was very hard to get at this way and so allowed to let be.

Except for the let-be part, the bone became obviously more dense and cortical thickness which was mostly everywhere absent became apparent. Dr. Rosenthal of Harvard’s radiology department, at our request, used his tools (normally used to insert into osteoid osteomas as he had for several of our patients which were part of his series) to inject calcitonin into the portion we could not get at. That worked. Followed to maturity this patient does everything on an intact cosmetically perfect leg.

Several patients with difficult unicameral bone cysts (too close to growth plate but extensive) had healing with one injection which was a mix of the original steroid method to which was added Grafton Gel and 1 cc of calcitonin. A distal humerus case was the impetus as damage to the trochlea was feared by open scraping.

Several other UCBs were quickly healed the same way then one shocked us (real fear) in that bone formation was so fast and so dense that we worried that we may have injected a sarcoma. That case was sent immediately to Sloan Kettering where the fear was not dispelled until open biopsy found ‘dense dense bone’. No tumor. Remodeling over the

following months dispelled the sarcoma theory completely.

So what is the rumination here? Osteoclasts are a cell based pathway of tissue recognition and complex protein digestion all contained in a single cell whose job it is to remove bone. This packet of capability must be passed as a complex of abilities aimed at a single end. Cells so armed can become a focal tumor-like thing and so be seen as a bone defect. Typically thought of as failure to make bone there. Now we think bone manufacture with failure to keep up with local bone digestion there.

Or, the process can be less focal, even regional but focally seen (CPT in two levels with CPF, you did notice that the second case had CPseudarthrosis of the Fibula as well as CPT?). Same idea just less focal. But maybe diffuse to an entire bone (fibrous dysplasia?). Or even spotty and contributory to a lymphatic vessel blown up (UBC)? This is pure speculation. I am too old to defend these thoughts and would not even if younger. This needs a wider more basic science approach. More cases, rare as they are, do not further this where it counts. What are the molecules doing?