Multiple Fractures from Metabolic Bone Disease,
being Falsely Attributed to Inflicted Child Abuse:

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One of the defining differences between traumatic fractures and those of metabolic origin is a prevalence of shearing**,** severance**,** and/or major dislocations in fractures of traumatic origin as compared with their absence or rarity on those of metabolic origin.(1)

A second defining difference is the absence of pain and discomfort in metabolic fractures. Traumatic fractures, in contrast, are almost always extremely painful. This is easily explained. It is well known that bone has no pain fibers, while surrounding connective tissues are abundantly supplied with pain fibers. In metabolic fractures, which can take place with ordinary infant handling, surrounding connective tissue is not involved. In traumatic fractures, surrounding connective tissues are torn resulting in significant pain, as anyone who has experienced a sprained ankle or traumatic rib fractures can testify.

Current Standards of Medical Practice Concerning Multiple Fractures**,** and the Requirement for Differential Diagnosis

As reported by Jenny C, Committee on Child Abuse and Neglect, in "Evaluating infants and young children with multiple fractures," *Pediatrics,*(2)(2006), *a differential diagnosis of child abuse should include the following list, with appropriate evaluation for each, according to currently recognized medical standards. Anything less should be considered as substandard medical practice.*

- Osteogenesis imperfecta
- Preterm birth (osteopenia of prematurity)
- Rickets
- Osteomyelitis
- Copper deficiency
- Disuse demineralization from paralysis
- Other rare conditions (e.g. Menkes Syndrome)

Two additions might legitimately be added to this; **First,** classical scurvy from vitamin C was characterized by multiple fractures. Vitamin C deficiency is still quite prevalent and may be a contributory factor in many cases of brittle bone disease. This is due to the essential role of vitamin C as an enzymatic cofactor for the conversion of the amino acid, proline, into procollagen and collagen tissue, the latter making up 90 % of bone mass. (3) **Second,** based largely on the pioneering work of Marvin Miller (reviewed below), disuse demineralization of bone will take place in any situation involving prolonged immobilization of bone. This is well documented in children with club feet subjected to prolonged immobilization in corrective casts, as well as in animals sent into the weightlessness of space in early experiments by the Soviet Union. As a matter of common observation, it may be a complication of prolonged bed rest. Finally,

the immobilization of the fetus from any cause during the last trimester of pregnancy may result in "**temporary brittle bone disease**," according to the work of Marvin Miller. (Emphasis added)

Flawed Medical Consensus that Multiple Fractures in Infants, in the Absence of Known Accidental Trauma, are Diagnostic of Child Abuse

In 1990 Garcia reported in *Journal of Trauma* on a series of 33 children brought into a trauma center with rib fractures, all brought about by blunt trauma. Nearly 70 percent were from auto accidents, 21.2 percent from child abuse, and 9.1 percent from falls. Mortality was 42 percent. 72 percent of the children with three rib fractures had internal chest injuries, such as lung punctures or tears and/or injuries to other internal chest organs. With four or more rib fractures there were 100 percent internal chest injuries. (1)

By way of explanation, when rib fractures are brought about by severe blunt force, such as in auto accidents or falls, a significant portion of ribs will be severed and sheared, the severed pieces acting like spears that are plunged into the deeper chest organs. In the present series of 30 infants, there was only one report of internal chest injury (pleural effusions). From a statistical standpoint, this small incidence would have been a virtual impossibility, had these fractures resulted from violent, inflicted force.

Atrophy of Disuse as Applied to Fetal Bone Development: Spontaneous Fractures Taking Place during Childbirth or the Neonatal Period, Commonly Attributed to Child Abuse

Atrophy-of-Disuse is a universal principle as applied to human organs, tissues, and physiology. Bone is no exception. Common examples include bone weakening and rapid decalcification known to take place during prolonged bed confinement, thereby predisposing to spontaneous fractures or fractures with minimal trauma. As reported by Grayev *et al*, eight children with clubfeet experienced metaphyseal fractures during physical therapy when their legs were passively manipulated, their legs having been immobilized for prolonged periods in corrective casts. (4) In studies by Rodriguez *et al* of the long bones in newborn infants with congenital muscular dystrophy (with marked muscle weakening/paralysis), the bones were found to be thin, hypomineralized, and elongated. In most of the bones there were multiple diaphyseal (shaft area of bone) or metaphyseal (at ends of bones) fractures or both. (5, 6) A study of rat fetuses that were curarized (paralyzed) during the later phases of pregnancy revealed marked thinning and delay in ossification of bone.(7) Conversely, as a matter of common observation, exercise such as weight lifting strengthens bone as well as muscle, or else stress fractures would be near universal in more advanced weight training.(8,9)

The following paragraph from the text, *Skeletal Tissue Mechanics*, by R Martin et al,(1998) vividly describes the "atrophy of disuse" process as applied to bone:

> "It is commonly observed clinically that the intact portions of the fractured bone become osteoporotic as healing occurs. This generalized osteopenia of the intact regions, called *posttraumatic osteoporosis* or *posttraumatic bone atrophy*, is

> caused by two factors. First, in addition to the healing response, the fracture causes a remodeling….so that osteonal BMUs riddle the entire cortex with resorption cavities. The second factor is the removal of mechanical loading from the fractured bone…If the fracture is well fixed and sufficient loadbearing is resumed, the resorption spaces will refill and the osteopenia will be transient."(10)

In other words, without movement or weight bearing, such as takes place when a fractured limb is immobilized in a cast, there may be significant decalcification of the bone; but there will be rapid recalcification once movement, weight bearing, and other mechanical stresses of daily living are resumed.

With the above material as background, one of the frequent causes of osteopenia with spontaneous fractures during the perinatal period (shortly before and shortly after birth) is prematurity. (11-13) As reviewed by Marvin Miller, (12)(2003), premature infants are at increased risk to develop temporary brittle bone state. It has traditionally been thought that the primary cause was insufficient calcium and phosphate in the diet of the premature infant. However, there is emerging evidence that the bone disease of prematurity may be more of a mechanical issue than one of nutritional mineral deficiency. Miller suggested that this increased bone resorption in the premature infant compared to the term infant is secondary to inadequate "bone loading" in the form of fetal muscular movement. During the last trimester of a full-term pregnancy the fetus is actively kicking and bouncing against the mother's uterus. This fetal activity with associated muscle development is the primary determinant of fetal bone formation, **without which the bone remains poorly ossified**, **weak**, **and brittle. (Emphasis added)**

It has been shown that preterm infants who receive 5-10 minutes of daily physical activity, with passive movements of extremities by nurse attendants, realize a 76% greater gain in bone density by one month of life compared to control premature infants who receive no physical activity. (8)

M. Miller and T Hangartner have observed a comparable clinical situation referred to as "temporary brittle bone disease" associated with lack of fetal movement during the last trimester of pregnancy, **in which the baby remains susceptible to spontaneous fractures with minimal trauma for 6 or more months following birth**.(**14-15**)(**Emphasis added**) Risk factors that may lead to reduced fetal movement from limited uterine confinement include twin or triplet pregnancies, cephalopelvic disproportion, oligohydramnios (reduced amniotic fluid), large maternal uterine fibroids, or other maternal structural uterine abnormalities. Fetal structural defects such as clubfoot and dislocated hips may also result in decreased fetal movement; also short umbilical cords (16) and depressive-type drugs taken by the mother during pregnancy. (17)

Radiology Interpretations:

It is likely that many cases of metabolic bone disease are being missed in hospitals for the following reason: *In the earlier phases of metabolic bone diseases,*

x-ray studies are of limited value since there must be at least a 30 to 40 percent loss of bone density (*calcification*) *before there is any detectable reduction of whiteness on the films,*(*18-21*) *a level at which fractures may take place with minimal trauma or ordinary infant handling*, (a physiology unknown to many prosecutors.)

Congenital Rickets from Vitamin D Deficiency

It is well established that there is a re-emergence of vitamin D-deficient rickets with "an alarming prevalence of low circulating levels of vitamin D in the United States population, leading to an increased incidence of infant fractures, especially when premature." These conclusions were announced by the National Institute of Health (NIH) following a conference on vitamin D, October 9-10, 2003.(22)

In a study conducted at the Pittsburgh Graduate School of Public Health, (23)(2007) serum 25-hydroxy vitamin D was measured at 4-21 week gestation and predelivery in 200 white and 200 black pregnant women and in cord blood of their neonates. Over 90 percent of women used prenatal vitamins. Women and neonates were classified as vitamin D deficient at less than 37.5 nmol/LI, insufficient between 37.5 and 80, and sufficient at over 80. At delivery, Vitamin D deficiency and insufficiency occurred in 29.2% and 54.1% of black women and 45.6% and 46.8 of black neonates, respectively; 5 % and 42.1% of white women and 9.7% and 56.4% of white neonates were vitamin D deficient and insufficient respectively. In other words, over 92% of black neonates and 66% of white neonates were born with grossly deficient or suboptimal vitamin D levels. It was concluded that black and white pregnant women and neonates residing in the northern USA are at high risk of vitamin D insufficiency, even when mothers are compliant with prenatal vitamins. Causes of the reemergence include reduction in milk intake (milk allergies, lactose intolerance, reduction in vitamin D-containing fats, and increased use of sun screens (sun acts on skin oils to generate vitamin D precursors).

An article entitled, "Rickets vs. Abuse: A National and International Epidemic," by Kathy Keller and Patrick Barnes,(24)(2008) provided a review of the literature and four case reports of infants with multiple fractures demonstrating classical x-ray findings of rickets. Also, in each case vitamin D deficiencies were documented in the mothers.

*Classical **x**-ray findings of congenital rickets include the following:*

- Washed-out appearance of skull from side view.
- Skull sutures widened and irregular.
- Pseudo fractures occur in weakened bone with normal infant handling.
- Rachitic rosaries: (Bulging irregularities in the growth centers at the anterior ends of the ribs.)
- Irregularities of the paired forearm bones at their endings in the wrists.
- Curved diaphyses of leg bones ("bowed legs") even before walking.
- Absence of dense white lines in growth centers of epiphyses, such as at the wrists.
- Maternal and infant diet histories are of highest importance in these cases.

Nutritional rickets has also been described with normal circulating 25-hydroxy vitamin D attributed to calcium deficiency in infants.(20) Elevated parathormone (PTH) levels are generally found in these cases.

Infantile Scurvy **(**Barlow's Disease**)**

Infantile scurvy is another possible cause of spontaneous fractures, which may be more common than generally appreciated. The probability of Barlow's Disease can be increased by maternal malnutrition, by hyperemesis gravidarum (excessive vomiting in pregnancy), by viral or bacterial infections in the mother or the infant. (26) The bones of infants may be vulnerable to fracture and defective formation before radiological signs appear. (18-21) Scurvy or subclinical scurvy would contribute to the deficiencies of proline and lysine hydroxylase (amino acid enzymes) that affect connective tissue components of bone formation. The hydroxylation (oxidation) of proline and lysine into procollagen is carried out by the enzyme prolyl hydroxylase, ***which requires vitamin C as a cofactor.*** (27,28) Collagen provides the bone's tensile strength, comprising 90 percent of bone matrix. Deficiencies in vitamin C would compromise the prolyl hydroxylase enzyme system, resulting in imperfect bone formation.

Far from being uncommon, vitamin C deficiency does still commonly occur in the Western World. When people attending a Health Maintenance Organization (HMO) in Tempe Arizona were tested for plasma vitamin C, it was found to be depleted (between 0.2 and 0.5 mgs/100 ml) in 30 percent and deficient (below 0.2 mgs/100 ml) in 6 percent. (29) As reviewed by Clemetson, when the human plasma ascorbic acid level falls below 0.2 mg/100 ml, whole blood histamine level is doubled or quadrupled. (30) It has been shown that bleeding from scurvy results from increased blood histamine, or histaminemia, which causes separation of endothelial cells from one another in capillaries and small venules, leading to spontaneous bruising. (31) When these are seen by a physician, almost always inflicted abuse is erroneously suspected. It follows then that the diagnosis of non-accidental trauma based on multiple bruises cannot be ethically or professionally justified without first ruling out scurvy by a plasma vitamin C blood test.

Metabolic Bone Disease and Vitamin K Deficiency

Maternal vitamin K deficiency during pregnancy is a risk factor for hemorrhagic disease of the newborn (HDN), usually a self-limited disorder taking place within 24 to 72 hours following birth. The primary dietary source of vitamin K is from green, leafy vegetables. It is for this reason that a maternal dietary history is of highest importance in adequate evaluation of infant fractures.

Although calcium absorption from the gut into the blood stream is dependent on vitamin D, it is less well known that vitamin K delivers calcium from the blood into the bone.(32) Consequently deficiencies of vitamin K in early infancy may be a contributory cause of metabolic bone disease. Also, bone matrix proteins necessary for normal bone metabolism are vitamin K-dependent,(33-35) so that vitamin K deficiency in infants can lead to fractures as well as hemorrhagic disease..

Osteopenia of Prematurity

As reviewed above under "Atrophy of disuse," prematurity poses one of the most common sources of infant fractures. (11-13) This is due to the fact that the fetal kicking and other vigorous body movements, which are essential for promotion of bone strength and integrity, are only in their beginning phases when preterm births occur. These movements are intrinsically necessary for bony maturation, just as they are necessary to maintain bone strength at all ages following birth.

In *Nelson's Textbook of Pediatrics,* 16th Edition, the following quotation is found:

> "**Osteopenia of prematurity**. Very small premature infants with chronic illnesses often develop a rickets-like syndrome with pathologic fractures and demineralized bone. There may be associated cholestasis and vitamin D or calcium malabsorption; urine calcium loss due to diuretics; and poor calcium, phosphorus, or vitamin D intake, or aluminum toxicity. The treatment of fractures requires immobilization and administration of calcium and, if needed, phosphorus (for hypophosphatemia) and vitamin D (not more than 1,000 IU/day unless severe cholestasis or vitamin D resistance). Appropriate formulas for premature infants should provide a more optimal intake of calcium, phosphorus, and Vit. D."(36)

As reviewed by FR Greer:

> "Osteopenia of prematurity refers to the hypomineralized skeleton of the premature infant compared with that of the normal fetal skeleton…..In growing, low birth-weight infants with birth weight less than 1500 grams (3.3 lbs) and less than 32 weeks gestational age, it occurs almost without exception. This high incidence (of hypomineralization) is not surprising considering that 80% if fetal skeletal mineralization takes place during the last trimester of pregnancy. Thus one would expect an increasing degree of osteopenia in premature infants with decreasing gestational age. …Even term infants may have decreased stores owing to maternal complications such as severe preeclampsia."(37)

Three reviews of fractures occurring during the first year of life in premature infants (38-40) found that rib fractures often remain undetected and are only discovered on x-rays taken for other reasons. In one series, clinical suspicion of fractures was documented prior to ordering the radiographs in only 3 of the 19 (16%) infants.(41) Hence the true incidence of fractures in infants born prematurely remains unknown, but it probably is much more common tan reported in the literature.

Timing of Fractures

In court cases involving multiple fractures in which parent and/or caretaker have been accused of child abuse, the dating or timing of the fracture often plays a critical role.

As reviewed by Amir *et al*,(11) from 1977 to 1984, 973 premature infants were admitted to the neonatal intensive care unit of Beilinson Medical Center, Petah Tiquva (Isreal). Among those who survived over 6 months, 12 suffered from fractures that appeared during their hospitalization between ages 24 and 60 days. All of these were without clinical signs. All were diagnosed on routine chest x-rays. Callus was always present when first diagnosed. In six instances angulation was present, but there were no instances of separation or dislocation in the fractures. According to the authors, fractures usually occur a few weeks after birth, and are almost always pathologic, the most common cause being metabolic bone disease.

According to N. Bishop, who observed a somewhat different timing, fractures due to osteopenia of prematurity and preterm rickets occur typically from 10 weeks age and usually stop before 6 months.(39)

In one of the earliest prospective studies of the clinical course of fractures and rickets in very low birth weight infants (less than 1,500 grams) by WH Koo *et al*, (13) 78 infants were enrolled solely on the criteria of birth weight <1,500 grams. There was a distribution of 82 fractures in 19 infants in the study. 73 fractures (89%) originated during the course of hospitalizations ranging from 32 to 131 days. Clinical suspicion was documented prior to ordering the radiographs in only three of the 19 infants (16%). It was further determined that physical therapy was the source of some of the fractures. Fractures from congenital rickets, when followed serially on radiographs, showed complete resolution beyond six months after birth.

Conclusions and Recommendations:

The Jenny report in 2006, which established a standard for the differential diagnosis of multiple fractures in infants and children, might justifiably be considered a major landmark in medical history. Since the publication of this report, it has been incumbent on physicians to rule out various forms of metabolic bone disease before diagnosing inflicted child abuse directed against parent or caretaker. In my opinion, failure to do this must be considered substandard medical practice, bringing disrepute on the medical profession.

According to the time-honored principle of “considered innocent until proven guilty,” appropriate medical evaluation of multiple fractures would require a formal listing of a differential diagnosis and appropriate laboratory tests. In addition to routine chemistries and blood counts, blood tests should include 25-hydroxy vitamin D, alkaline phosphatase, parathormone, calcium, phosphorus, serum histamine, plasma vitamin C, and tissue exams for osteogenesis imperfecta. These tests should be performed immediately on finding of fractures, as later tests might be irrelevant. A careful medical history of the mother’s diet and vitamin supplements during pregnancy and of the infant’s diet and supplements following birth are of paramount importance. Lacking these criteria, diagnosis of inflicted child abuse cannot be justified.

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