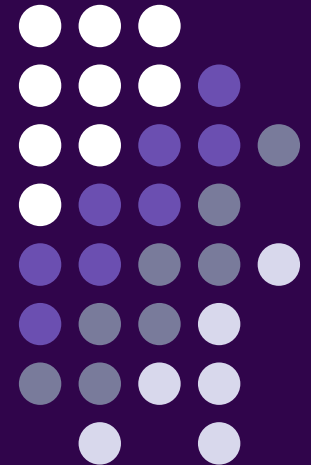


Osteogenesis Imperfecta

Michelle Abercrombie-Peterson

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What is Osteogenesis Imperfecta?



- Genetic defect
- Characterized by fragile bones
- Poor quality of collagen or smaller quantity of collagen
- “Brittle Bone Disease”

Four types or levels

- Type I
- Type II
- Type III
- Type IV

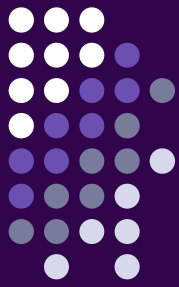


Type I



- One per 30,000 live US births
- Onset of symptoms in infancy
- Mildest form of OI
- Near-normal or normal stature
- Loose joints
- Low muscle tone
- Blue sclera
- Triangular face
- Possible scoliosis/kyphosis
- Possible brittle teeth
- Absent or minimal bone deformities
- Possible hearing loss (onset in 20's or 30's)
- Less collagen than normal

Type II



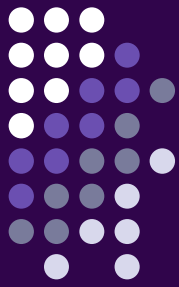
- One per 60,000 live US births
- Onset of symptoms in utero
- Most severe form of OI
- Often lethal at or shortly after birth
- Numerous fractures
- Severe bone deformities
- Small stature
- Underdeveloped lungs
- Improperly formed collagen
- Possible blue sclera
- Small nose

Type III



- One per 70,000 live US births
- Half the cases show symptoms in utero, the other half in the neonatal period
- Bones fracture easily
- Short stature
- Blue sclera
- Loose joints
- Poor muscle development in arms and legs
- Barrel-shaped rib cage
- Triangular face
- Possible respiratory problems
- Possible scoliosis/kyphosis
- Possible hearing loss
- Often severe bone deformity
- Possible brittle teeth
- Improperly formed collagen

Type IV



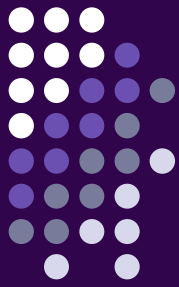
- Rare
- Onset of symptoms usually in infancy
- Between Type I and Type III in severity
- Sclera are normal or near-normal in color
- Mild to moderate bone deformity
- Short stature
- Barrel-shaped rib cage
- Triangular face
- Possible brittle teeth
- Possible scoliosis/kyphosis
- Possible hearing loss
- Collagen is improperly formed



Treatments

- No known cure
- Prevention and control of symptoms
- Rodding surgery
- Exercise
- Maintain healthy weight
- Nutritious diet
- Avoid smoking, excessive alcohol & caffeine consumption, and steroid medications

Caring for a child with OI



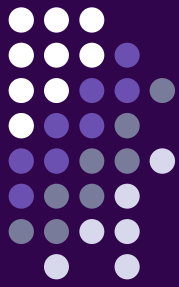
- Reclining car seat and stroller with additional padding
- Lift baby from behind head and buttocks with fingers spread
- Lift baby from under buttocks when changing diaper
- Keep baby's fingers and toes free from catching onto your clothing
- Dress child in lightweight, cotton clothing
- Use molded sponge bathing aid with infants and a safety bath ring with sitting children during bath time
- Bathe baby with sling on and change when finished with bath; sponge bathe baby with a cast



Child Abuse

- False accusations of child abuse may occur when a child has not been diagnosed with a mild form of OI or when working with medical staff that is new to your child's medical history
- Fractures in multiple stages of healing – rib fractures, spiral fractures, fractures with no adequate explanation of trauma
- Explanation of injury doesn't appear to match the injury found by the medical staff

Practical Advise for Families

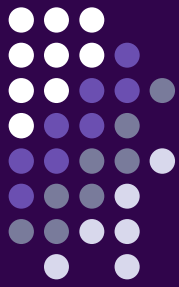


- Carry on your person a letter from your doctor describing your child's condition
- Consult with a Family Law Attorney when you've been accused of child abuse
- Remember: doctors, teachers, and social service workers are mandated to report suspected child abuse – they are concerned for your child, too
- Do not change the hospital or physician treating your child without consulting with your case worker – your actions may be interpreted as an attempt to hide abuse

Practical Advice - continued



- Work with your case worker – keep them up to date on new information, make sure the case worker understands what you have told them
- If your child is taken from your home, you may request that s/he stay with a relative
- When the problem is resolved, insist that the charges be taken off your record or it is possible that you may be registered as an abusive parent until your youngest child is 18



Internet Resources

- Children's Brittle Bone Foundation
<http://www.cbbf.org>
- March of Dimes Birth Defects Foundation
<http://www.marchofdimes.com>
- National Institute of Arthritis and Musculoskeletal and Skin Diseases <http://www.nih.gov/niams/>
- National Institute of Child Health and Human Development <http://www.hih.gov/hichd>
- Osteogenesis Imperfecta Foundation, Inc.
<http://www.oif.org>