Pediatrics Masterclass 2017 REPORT

Full success for the advanced MedEd Orthofix initiative addressed to expert surgeons, which took place on the 30th and 31st October in Verona

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Fractures are quite common in childhood, as children play games and sports and do risky activities. If trauma is one the most common causes of a bone fracture and/or an eventual consequent deformity, we know that some children have an inherited condition called *osteogenesis imperfecta* that makes their bones more fragile and susceptible to breaking. Children may also suffer of congenital bone deformities such as the *Pseudoarthrosis of the Tibia* or the *Fibrous Dysplasia*, where it is evident an atypical, structural deviation or distortion of the bone's shape from its normal alignment, length and size.

On paediatrics bone diseases management – congenital and/or as a consequence of an acute trauma or even adverse life conditions – the Orthofix Masterclass was focused, in a two days advanced training program with the contribution of a top level scientific faculty, a team of accredited international experts mainly from Australia, USA, Germany and France. The Masterclass objective was mainly educational, and centred on free discussion on a significant number of clinical cases presented by the speakers. A really worldly event: more than 50 participants – all very active and ready to discuss, deepen and share their practical experiences – from 14 countries: Australia, Brazil, China, Colombia, Denmark, France, Germany, Italy, Netherlands, Spain, South Africa, Switzerland, UK and US.

Particularly appreciated by all participants the quality of the seminar organization and the innovative, interactive training method. "Different pathologies require different treatment strategies. Besides, as paediatric orthopaedic surgeons, we must always respect our patients' quality of life, knowing their natural history and considering the familiar and community contexts where children live. Each individual child needs an individual approach. Differential diagnosis is more important than in adults, and children's compliance is fundamental. Our common mission is to bring each child healthy back to his/her feet", is the final faculty take home message.

First session

The first session was on **Blount's disease** (*tibia vara*), from the name of Walter Putman Blount, who brought the attention to this pathology in 1937 describing 13 children with *tibia vara* or *osteochondrosis deformans*. This session was moderated by Prof. **Christopher Iobst** from Nationwide Children's Hospital, Columbus Ohio, US.

Blount disease is a developmental disorder characterized by the abnormal growth of the shinbone resulting in progressive lower limb deformity, more frequent in US than in Europe – may be also due to juvenile obesity problems. Common risk is to underestimate the complexity of this disease.

The infantile form, occurring in children from 2 to 4 years of age, is generally bilateral, progressive, associated with internal tibial torsion; it appears as a pathologic development of the physiologic *genu varum*. The adolescent form is less common, occurring in children between 9 and 14 years of age, typically monolateral and less often associated with internal tibial torsion. In bowlegs (*genu varum*) the knees appear rotated away from each other; if physiological, this condition is due to the position of the hips and inward curve of the shinbone (tibial torsion), created by the position of the legs in the uterus before birth; quite frequent in newborn and infants up to 12 months, it improves with growth.

When the child starts standing and walking the lower limbs gradually straighten (with normal growth) up to a zero tibiofemoral angle (18-24 months). At this stage, knees generally turn to valgus (knock-knee). Finally the *genu valgum* spontaneously corrects at the age of 7, until the adult alignment of the lower limbs of 8 degrees of valgus in the female and 7 degrees in the male. Therefore, angular bowlegs are to be considered pathologic after 2 years (while Knock-knees quite often correct themselves by the age of 9 without a treatment. If they persist after the age of 10, they may require surgical treatment).

Femoral torsion (twisting), either internal or ex-

ternal, can gradually decrease without treatment, but sometimes (after the age of 8) it may require surgery.

Aetiology

It seems to be a multifactorial disease, a combination of hereditary and developmental factors. Presence of strong genetic and biomechanical components (more blacks, more obese patients, with the overload of the physis) which produce the disturbance in growth and ossification of the medial part of the proximal tibial epiphysis and metaphysis.

Symptoms and Diagnosis

Children with physiologic varus angulation at the knee generally do not exhibit a thrust. On the contrary, in pathologic conditions a lateral thrust is often present, suggesting incompetence of the knee ligaments, which increases the potential for progression of the deformity. The child can also walk or run in an unusual way on the toes, or rotating the leg.

Diagnosis should include family history, any description of onset and information about the progression of the deformity, plus a selective use of X-rays, CT and MR imaging (see **Fig. 1**). The child will be observed walking, with attention to her/his knees during the stance phase, to determine if lateral thrust (genu varum) occur. The physician should know the diet and amount of vitamins taken by the child, inquire about milk allergy, or intoxication to metal – specifically to lead and fluoride.

Imaging

X-rays (preferable not earlier than 2 years) are particularly useful to distinguish physiological bowing and *tibia vara* from Blount's disease; children who present an MDA more than 10°, an EMA (Epiphyseal Metaphyseal Angle) more than 20° (at 2 years old) or 10° (at 5 years old), and with a BMI greater than 22, are at risk and must be followed over time (even if, despite these numbers, certain cases can evolve spontaneously towards resolution). Doctors should follow the spectrum of the disease, and decide if and when to intervene along that spectrum.

Management

Avoid unnecessary surgery. Treatment may vary depending on the condition and age of the child. Some children may outgrow some disorders, but some others may require bracing and/or surgical intervention. For children younger than 3 years old the suggested treatment is leg braces or splints, but in some cases experts are not sure if it is the bracing or the natural history of the kid which solves the pathology spontaneously. For older children the suggested treatment is surgery, always accompanied by physiotherapy, particularly in presence of significant deformity (stage III-V). When surgery is necessary, if performed at an early stage the surgeon will usually obtain complete and permanent correction of the child's limb deformity, whereas there is a greater chance for recurrence if it is done in later years.

Surgical procedures

1. *Guided growth*, a benign procedure with the correction only at the deformity apex

- minimally invasive
- no immobilization necessary

Fig. 1. Radiological and MRI imaging of Blount's disease from authorised source: Dr. Christopher lobst





Fig. 2. Elevation osteotomy from authorised source: Dr. Christopher lobst

- stapling 61% of success; more frequent failures with obese kids
- screws work better with obese kids (BMI more than 35)
- guided growth plates 78% of success at younger age, i.e. 2¹/₂ years old

With screws and plates the risk is screw's (more frequent with titanium and definitely less with stainless steel) or plate's breakage: to avoid it, a possible solution is using double screws and plates. If one of the two breaks, the other resists.

With younger patients, a new thought is the "sleeper plate": only remove the metaphyseal screw, and let plate ride along with tibial screw, then just reinsert distal screw.

2. *Osteotomy*, preferable in older patients (over 3 years old), but difficult to determine the best osteotomy technique for Blount disease:

- "M" osteotomy with K wires, frequent in East countries where there is a lack of instruments
- Serrated "W/M" osteotomy
- Inverted arcuate osteotomy with external fixation for adolescent *tibia vara*
- Rab osteotomy, minimally invasive
- Elevation osteotomy with K wire and barrel vault, for example in case of depression of the medial tibial plateau in early Blount's disease, a deformity which can be treated with medial plateau elevation (see **Fig. 2**). It can be combined with metaphyseal osteotomy. Recurrence prevented by performing a concomitant lateral epiphysiodesis.

3. Acute correction with internal fixation (plate, rod), for mild deformity, easier on the patient, avoid pin site issues, consider soft tissues limitations.

4. *Gradual correction with an external fixator* – with Ilizarov method and hexapod system (see **Fig. 3**), in case of severe/advanced *tibia vara*, to find the right position and slowly improve the correction.

Consensus vs. controversial issues

There are no defined rules, but the faculty tip is "start with the simplest and then pass to more complex procedures". The problem is to find the right time when to intervene, and choose the right procedure. Doctors should follow the spectrum of the disease, and decide if and when intervene along that spectrum. Osteotomy can be considered a controversial matter referred to the child's age as, for example, Dr. O. Birke doesn't apply it on small kids. Some other experts, i.e. Dr. M. Samchukov and Dr. A. Cherkashin, can decide for a correction osteotomy now, to prevent further deformity and other operations in time. In obese young patients, the choice to proceed with osteotomy should be accompanied by a whole change of life-style for the kid, with a proper diet, more movement and sport, and sometimes some psychological help. Other experts, i.e. Dr. JD. Metaizeau, prefer to use precise flexible nails to other procedures, a choice which decreases the operation time and requires less amount of X-rays.

Future questions: Strategies for avoiding recurrence?

There is still an unresolved problem: how much to correct as patients don't bear valgus after being in varus for long. Future direction: three-dimensional analises.

Fig. 3. TL-HEX in Blount's disease from authorised source



Second session

Second session on **Limb equalization**, moderated by Dr. **Mikhail L. Samchukov** from the Scottish Rite Hospital for Children in Dallas, Texas, US.

Suggested surgical procedure

Distraction and gradual correction with external fixation (i.e. TL-HEX hexapod system) for the deformity correction and sequential lengthening.

Indications for external fixation (contraindications for a lengthening nail):

- 1. Open growth plates
- 2. Bad conditions of soft tissues

3. Patients with no or limited non-weightbearing capability

- **4.** Anatomical restrictions for using nail: bone shape, length, diameter
- 5. Infections, with post traumatic chronic osteomyelitis
- **6.** Pathological bone structure (enchondromatosis, fibrous dysplasia, etc)
- **7.** Associated limb deformities which benefit from gradual corrections
- **8.** Complex limb reconstruction with simultaneous lengthening
- **9.** Adjacent joint control (stiffness, contractures, subluxation/dislocation)

Consensus

Prior to any surgical correction of lower limb deformity, it is very important to proceed with a precise preoperative planning (which includes an accurate, exact geometrical analysis of the malalignment), and then to share the final treatment plan with the kid's family. The objective must be to correct as much as the patient can tolerate (try to avoid obsessing correction), and it cannot be more than 20% of the length the kid had before surgery (max. 5cm-8cm). Vascular, soft tissues and muscles evaluation is fundamental. Orthopedic surgeons must be aware of the patient's overall status. A supplement of vitamin D may be necessary.

Third session

Third session was focused on the **Metabolic Bone Disease in Skeletal Dysplasia**, moderated by Prof. **Franck Launay** from Timone Children's Hospital in Marseille, France.

• Fibrous Dysplasia, coexistence of healthy and diseased tissues: plates and screws are contraindicated so is curettage. Elastic and telescopic nails are the gold standard. • **Rickets** (presented by Dr. M. **Samchukov**) is a bone disorder caused – if nutritional – by a deficiency of vitamin D, calcium or phosphate. It leads to softening and weakening of the bones and it is seen most commonly in children of 6-24 months of age.

Three common types include nutritional rickets, hypophosphatemic rickets and renal rickets.

Signs and symptoms

Sign and symptoms may include bone pain, decreased muscle tone and strength, short stature and a number of skeletal bone deformities such as bowlegs, rib-cage abnormalities (rachitic rosary) and breastbone, craniotabes, pelvic and spinal deformity, multiple fractures at different healing stages.

Goal of treatment

Goal of treatment is to correct the clinical, biochemical and radiological findings, and restore the deformed extremities to functional alignment. Treatment outcome depends mainly on early recognition and correct management of the underlying disease.

Management

Vitamin D and supplement of calcium and phosphorus are used for the treatment of nutritional rickets. For severe extremity deformities, due to the impaired bones growth, and multiple fractures due to the bone fragility, the golden treatment is surgical. Corrective osteotomy with epiphysiodesis and external fixation with circular hexapod, intramedullary flexible nailing, Kirshner's wires, plates and casting, are the recommended procedures.

The author, Prof. Launay, prefers acute correction because his patients come from underdeveloped countries.

• Osteogenesis Imperfecta (illustrated by Dr. C. Iobst) is a genetic, connective tissue bone disease commonly known as "brittle bone disease" which can show in a wide spectrum from mild to lethal forms.

Cause

Usually due to a deficiency of normal type 1 collagen, an organic component necessary for the proper bone formation. It is a rare pathology (0.005%).

Common clinical manifestation

Multiple/atypical fractures, short stature, basilar skull deformities, spine curves, hearing loss, increased laxity of ligaments and skin, easy bruising, accelerated osteoporosis - among others.

Laboratory findings

Elevated level of serum alkaline phosphatise, hyper-

calciuria (the magnitude reflects the severity of the disease). Both C- terminal peptide (marker of bone formation) and C-telopeptide (marker of bone resorption) can be higher.

Imaging findings

Along a spectrum from type 1 (mild) OI: thin cortex and few fractures with normal skull development; 2)to type 4 (more severe) OI: hyperplastic callus formation (from thickened periosteum), shortened long bones with multiple fractures, osteoporosis, bowing deformities, past healed multiple fractures.

Goal of treatment

Goal of treatment is to minimize fractures, enhance independent function, reduce pain and promote health.

Management

Bisphosphonate therapy to increase bone mass and supplement of vitamin D, with physical and occupational therapy since the early infancy; in presence of no deformity the treatment may be conservative – casting, splinting, bracing fractured bones can help them heal properly; in presence of increasing bowing, the indication is to proceed with the deformity analysis: if acute deformity (which was the illustrated case of a 14 years old boy), the pillar of treatments is surgical: gradual correction with external fixation and controlled dynamization (pre-assembled prior to surgery for a perfect bone segments fixation), osteotomy and distraction for lengthening, followed by one month of non weight bearing. Telescopic nails (J. Lauen).

Another indicated surgical procedure is intramedullary rod placement to support long bones, and soft tissues surgery in case of lower limb or Achilles tendon contractures. Avoid the use of plates and screws for fracture stabilisation.

Close follow-ups are vital to ensure fracture healing and restoration of correct function.

- Fibrous Dysplasia (*Jaffe-Lichtenstein Disease*, presented by Dr. Joachim. Lauen, Trauma Center Murnau, Murnau, Germany) is a developmental pathology, not malignant, that results in abnormal growth, pain and deformity of the affected bones. Some children experience severe symptoms such as difficulty walking, deformity in arms and legs, limb length discrepancy, rickets, light to dark brown pigmentation but some others appear perfectly healthy, and show no evidence of bone or endocrine involvement. There are three types of this pathology:
- Monomelic: it affects only one bone, most often the femur (thigh), the tibia (shin), the rib cage or one of the facial bones

- *Polyostotic:* it affects numerous bones at a younger age, sometimes half of all skeleton's bones.
- *Mc Cune-Albright Syndrome:* it is the most complex, associated with abnormalities of the endocrine system, can lead to fractures, skin lesions and legs at different lengths.

Goal of treatment

Goal of treatment is usually to maintain bone strength and integrity, and correct the deformities, as it is very difficult to totally eradicate the disease.

Management

- **1.** In presence of no fracture, the suggested treatment is conservative, with splint and cast;
- **2.** Curettage-bone grafting, an operation which involves a surgical removal of the bony lesions by surgically scraping it out. The cavity is packed with donor bone tissue (allograft), bone chips taken from an other bone (autograft) or other material on the choice of the surgeon;
- **3.** In presence of increasing bowing and deformity, the indication is to proceed with the deformity analysis: if acute deformity, gradual correction with external fixation (pre-assembled prior to surgery for a perfect bone segments fixation); osteotomy and distraction for lengthening; one month of non-weight bearing.
- **4.** Drugs that alter bone resorption may help the pain generated by the fibrous dysplasia and strengthen the bone, but at the moment there is not much evidence of their effective value in the therapy, in any case it is a promising development for the future.

Fourth session

Fourth session on **Congenital Pseudoarthrosis of the Tibia (CPT)**, a rare and not completely understood disease, which remains one of the most complex and challenging problem in orthopaedics. It is usually associated with neurofibromatosis type 1 that features deficient bone anabolism and excessive catabolism.

The disease becomes evident within a child's first year of life. Its natural history is extremely unfavourable: once a fracture occurs, there is little or no tendency for the lesion to heal spontaneously. This session was moderated by Dr. Jean Damien Metaizeau from CHU, Centre Hospitalier Universitaire, Dijon Bourgogne, France.

Goal of treatment

- **1.** To obtain long-term bone union without recurrence of pathological refracture;
- **2.** to prevent leg length discrepancy and angular deformities;

- **3.** to restore soft tissue alterations and prevent adjacent joint stiffness;
- **4.** to avoid amputation.

Management

The treatment is mainly surgical, with a wide variety of different reconstructive strategies after CPT excision. It is often difficult to predict outcomes, but there is a **consensus** around the importance of *fixation and stability*.

Suggested standard treatment for CPT

See the results from a cohort of 39 patients directly illustrated by Dr. **Björn Vogt**, from the University Hospital Muenster, Children Orthopaedics, Deformity Reconstruction and Foot Surgery, Muenster, Germany (**Broeking JN** et al 2017).

• CPT excision followed by:

either

A. bone transport using stable circular external fixation and secondary autologous bone grafting at the docking site keeping original tibial length (Group A),

or

- **B.** primary autologous with acute compression of the bone ends either using external fixation or intramedullary rods resulting in tibial shortening. Extrafocal tibial lengthening by distraction osteogenesis using external fixation to reconstitute length (Group B)
- intramedullary fixation and orthoses (day and night) till skeletal maturity (only in 4 cases rods had to be removed for infection), with constant follow up of the child to avoid malalignment and residual deformities.

High rates of long-term bone healing and bone union obtained in 33 cases, in both Groups A and B. Only three amputations: 2 in Group A, 1 in Group B. Because of the duration of the treatment, much longer in Group A, the *recommended method is excision and extrafocal lengthening* as applied in Group B.

As an adjunct to surgery, some experts suggest a combined use of bone morphogenetic protein (BMP) and Bisphosphonate (BP) – "*a promising biological concept*" (**Birke** O 2017) (**Schindeler** A. et al. 2001).

From literature: "Circular external fixation as the treatment of choice of CPT allows total resection of pathological tissue, ensures stability regardless of the amount of resected tissue and allows extension of the member, correction of axial deformities and full support immediately after the intervention" (Shah 2012).

No surgery under the age of 3 years, preferably to be postponed until 5 (EPOS recommendation). External fixation as a primary treatment for children older than 8 years of age, with adding intramedullary nailing and bracing until skeletal maturity.

Amputation must be reserved for failure to achieve union after two or more (*controversial matter*) good attempts, and in case of worse deformity than that produced by prosthesis.

Fifth session

Fifth session centred on **Perthes Disease** (*Legg-Calvé-Perthes Disease*, from the names of the three doctors who first described this pathology) and moderated by Dr. **Oliver Birke** from The Sidney Children's Hospitals Network at Westmead and Randwick, Sidney, Australia. Perthes is a rare childhood condition that affects the hip. It occurs when the blood supply to the rounded head of the femur (thighbone) is temporarily disrupted. More than a simple disease, Perthes is a complex process of four stages that can last for years (2-5 years); as the condition progresses, the weakened bone of the femur's head (the ball of the "ball-and-socket" joint of the hip) gradually begins to break apart. Over time the blood supply of the femur returns, and the bone starts growing back.

WALDENSTROM CLASSIFICATION (Consensus):	
1 st stage	Initial. The blood supply to the femoral head is disrupted and bone cells die. Inflammation, some pain. Limp and different way of walking.
2 nd stage	Fragmentation. In 2 years, the body removes the dead bone and replace it with an initial softer bone (<i>woven bone</i>). The head of the femur in this stage is weaker and can break apart and collapse.
3 rd stage	Reossification. New stronger bone develops and begins to take shape in the head of the femur. This stage can last a few years.
4 th stage	Healing. The bone re-growth is complete, the fem- oral shape has reached its final shape. How close is to round depends – among other factors – on the extent of damage during the fragmentation phase, and the child's age at the onset of the condition.

Aetiology

Aetiology is still unknown. Recent studies reveal a genetic link, but more research must be conducted.

Symptoms

One of the earliest sign is a change in the way the child walks and runs, developing a very peculiar style. He/she may limp or have limited motion.

Other symptoms are pain in the hip or groin, or in other parts of the leg, such as the thigh or the knee, that

increases with activities and releases with rest, and painful muscle spasms.

Goal of treatment

- **1.** to relieve painful symptoms;
- to help and protect the bone grow back into a more rounded shape that fits into the socket of the hip joint, in order to allow the hip joint move normally;
- **3.** to prevent hip problems in adulthood. "*A stiff hip is a hard hip, a moving hip is a good one*" (**Birke** O 2017)

Imaging

X-rays, after the physical examination with the assessment of the range of the child's hip motion. A child with Perthes can expect several X-rays taken during the treatment.

Duration of the treatment

From 18 months to 2 years. This condition manifests itself in children between 4 and 10 years old. Five times more common in boys than girls, even if the bone damage is generally more extensive in girls.

Long-term prognosis is good in most cases. After treatment children generally return to their daily activities without major limitations, and grow into adulthood without further hip problems.

Management

The doctor's treatment plan depends on the child's age, the degree of the damage to the femoral head, and the stage of the condition when the child is diagnosed.

Non-surgical Management

Recommended:

- For very young children (2-6 years old) therapy should be simple observation ("*watch and wait*"), with regular monitoring through X-rays to make sure that the re-growth of the femoral head is on track as the disease runs its course.
- Pain, generally caused by inflammation of the hip joint, can be treated with steroids and anti-inflammatory drugs such as ibuprofen.
- Limiting high impact sport activities (rugby, running, jumping). Sometimes also limiting the weight bearing (through crutches or walker) to reduce the force across the softened femoral head. Use of wheelchair if the hip remains irritable after activity modification.

- Regular physical therapy to increase the hip joint's range of motion, with hip abduction and internal ro-tation.
- Casting and bracing (such as "Petrie casts" to keep the legs spread apart in a shape which remind of an "A" to maintain the hips in the best position for healing), if the range of motion becomes limited.
- Arthrogram, a sequential series of special X-rays images obtained with the injection of a dye into the hip joint to mark the degree of deformity and show better the anatomy of the hip. Also useful if planning surgery.
- Tenotomy, if the adductor longus muscle in the groin is very tight and obstacles the hip from rotating into the right position. Doctor performs tenotomy, a minor procedure to release this tightness, before applying the Petrie casts. After the removal of the cast, physical therapy is recommended. Casting can continue at intermittent intervals until the hip enters the final stage of the healing process.

Surgical Management

Recommended:

- if the child is older than 8 years at the time of diagnosis, very active and sportive, and the potential of deformity during the re-ossification stage is greater because of older age
- more than 50% of the femoral head is damaged
- in presence of instability
- failure of previous conservative treatment (controversial in small children under 6, to check with consensus).

Goal of surgery

Re-establish the proper alignment of the hips bones, contain the femoral head (femoral head alive and covered) and produce minimal secondary deformity.

Surgical procedures

1. *Osteotomy:* pelvic osteotomy, femoral head reduction osteotomy, varus osteotomy with Trochanter Flip distalisation* if lateral pillar is healing outside the joint (uncontained but moving).

"Statistically significant benefit from surgery" (Herring JA et al 2004).

2. Release cut to incorporate traditional plate-offset

^{* &}quot;Trochanter flip approach with retinacular soft tissue flap to preserve the blood supply to the femoral head" (from authorised source: Dr. Oliver Birke et al. 2017 unpublished technique). The new release cut technique allows varisation without creating abductor leverarm dysfunction. The greater trochanter can be optimally positioned and the hip offset can be maintained and improved.





Fig. 4.

Fig. 5



Fig. 6



Fig. 7

All pictures from authorised source: Dr. Oliver Birke

into osteotomy (see **Fig. 4**). See also the case of a 4 years old child treated with a varus derotational osteotomy and internal fixation (**Fig. 5**).

- **3.** Decompression treatment followed by hip distraction technique with external fixation (see **Fig. 6**).
- **4.** Reconstruction of residual deformity, muscular relief (see **Fig.** 7).
- 5. Casting for 6-8 weeks to protect alignment.
- **6.** Physiotherapy after casting removal to restore muscles' strength. Temporary crutches and walker to reduce weight bearing.

As an adjunct to surgery, some experts suggest a combined use of bone morphogenetic protein (BMP)

and Bisphosphonate (BP) "*a promising biological concept*" (Schindeler A 2011) currently under study in a prospective multi-centre trial in Sidney: preserves architecture, allows revascularization and may favour new bone formation.

Controversial issues

Mobile or fixed distraction? Age of the child? For some experts surgery is suggested over 6 years of age (other sources: POSNA 2015).

J. Lauen, 118 cases of Perthes disease: what is evident is that a long delay both in diagnosis and in decision for surgery can compromise the surgical outcome.

Sixth session

Sixth session dedicated to **Clubfoot** (*Talipes equinovarus*) and moderated by Dr. **Ludwig Schwering** from Mariannen Hospital – Paediatrics Orthopaedics, Foot Surgery and Rehabilitation, Werl, Germany. Clubfoot is a fairly common birth defect for an (often) otherwise healthy newborn, in which the top of the foot is twisted out of normal shape, downward or inward, increasing the arch and turning the heel inward. In clubfoot tendons connecting the muscles to the bone are shorter than usual, and in some cases the foot may be turned so severely that is actually looks if it was upside down.

Aetiology

It can be an inherited or an idiopathic condition, with unknown causes. Some risks factors may include congenital skeleton abnormalities such as *spina bifida*, or smoking during pregnancy, or too little amniotic fluid during pregnancy. Males are twice as likely as females to be born with clubfoot. Clubfoot can be mild or severe; 50% of the affected children have clubfoot in both feet.

Goal of treatment

To improve the aesthetics and the functionality of the child's foot before he or she learns to walk, in order to prevent long-term deformities and disabilities.

In older children, often from neglected areas in the world, to restore the proper functionality and prevent arthritis, further deformities and more pain.

Management

Small children have soft growing joints, tendons and bones – partly or totally made of cartilage. This is the reason why the treatment for clubfoot should usually begin in the first weeks after birth.

Non-surgical Management

Dr. Schwering prefers a pain free, conservative ap-

proach as golden treatment of the clubfoot, using the Ponseti method, with gentle manipulation (ligaments, joint capsules and tendons are stretched) and casting.

Procedures: the doctor moves the baby's foot into the correct position and place it in a cast to hold it there. One/twice a week the foot is repositioned and recasted for several months on (up to 10 new casts). Towards the end of the therapy time, the doctor will eventually perform a percutaneous Achilles tenotomy to lengthen the Achilles tendon.

After the foot's proper shape is obtained, the child will go on with stretching exercises and will wear special shoes and braces constantly and properly for about three months.

Recommendation: Ponseti technique is painless, non invasive, cost-effective and successful in almost 100% newborn cases of congenital clubfoot; it is supported (among others) by WHO (2012), NHS (2011), AAOS (*American Academy of Orthopedics Surgeons* 2011) and EPOS (2011).

Surgical Management

Recommended:

If the clubfoot condition is severe, if the patient is older and does not respond to non surgical-conservative approaches, or if there is a residual clubfoot, or untreated clubfoot, surgery may be needed.

Options/Procedures: taken from the clinical case of a 10 years old female with a story of bilateral clubfoot and multiple surgeries before (see **Fig. 8**), illustrated by Dr. A. **Cherkashin:**

1. Achilles tendon lengthening (ATL), to ease the foot into a better and proper position. After surgery, cast-

Fig. 8. From authorised source: Dr. Alexander Cherkashin

10 years old female

- History of bilateral clubfeet
- Multiple surgeries...
- Triple arthrodesis on the left
- Talectomy on the right foot
- Severe equino-varus deformity







ing up to 2 months and bracing for one year to prevent clubfoot from coming back.

- 2. Calcaneal and midfoot osteotomies (V-osteotomy) (see Fig. 9).
- **3.** Gradual correction of the deformities with 1) circular external fixation frame (TrueLok circular fixator); the set frame is **gradually adjusted** for calcaneal, equinus, varus forefoot abduction correction (see **Fig. 9**).
- **4.** Joshi fixator, which is light, thin, flexible, when not much strength is needed. On circular external fixation: *"Ilizarov-type frame is great in foot applications but requires frequent adjust*ments..." (Cherkashin A 2017).
- 5. Frame removal after 3 months (see Fig. 10);
- **4.** Cast and physiotherapy. *Outcome* in 4 years with plantigrade feet. The patient must be informed that he will recover functionality, aesthetics and pain, but with limited motion.

Birkholtz described the four steps of Ponseti technique with the Hexapod Fixator:

- **1.** correction of varus-valgus
- 2. correction of internal-external rotation
- 3. distraction
- 4. correction of equinus deformity

Consensus

First start with Ponseti method, surgery when needed always "a la carte", rarely in long term the deformity can relapse (a problem associated with older children, non compliant with the brace treatment). **Seventh session**

Seventh session was dedicated to Acute trauma, moderated by Dr. Jean-Damien Metaizeau, a paediatric orthopaedic surgeon from CHU, Centre Hospitalier Universitaire, Dijon Bourgogne, France. Most common causes of trauma fractures are falls from a height, injuries or accidents, but also (considering the recent dramatic worldly events) mass casualty terror attacks.

Paediatric fractures heal at different rates, depending on the age of the child and type of fracture, but in general a child's bone has a high remodelling potential and heals faster than an adult's one. This is due to some differences in the bone structure: in children the periosteum is thicker, stronger and more active to better supply oxygen and nutrients to the growing bones, and this helps in case of fracture. Its inner part contains very vital cells able to produce new bone; if an injury or an accident unfortunately occur to the child's bone, these cells are able to renew and remodel the broken bone in a shorter time.

Management

Non-surgical vs surgical management

- **1.** In case of traumatic *stable*, closed fractures *non articular*, non displaced the suggested treatment is casting.
- 2. With traumatic *stable*, *displaced* or *angulated*, closed articular fractures, (even physeal/growth plate fractures type 1 and 2) the suggested treatment is closed reduction (90% traction, 10% manipulation) with cast or flexion immobilization. In case of residual displacement the indication is surgery with Kirschner

Fig. 9. From authorised source: Dr. Alexander Cherkashin



Fig. 10. Three months after removal. From authorised source: Dr. Alexander **Cherkashin**



wires (K-wire) fixation and immobilization, with casting to avoid redisplacement (see Fig. 11).

- **3.** In presence of *unstable*, placed or displaced, articular or non articular fractures, surgery is the suggested treatment: closed reduction (in some cases also open reduction) and reposition-stable osteosynthesis with elastic nailing (ESIN), pins or elastic pins/screws; if needed additional casting and/or slings. In unstable fractures, ESIN can be used to hold the anatomical reduction (See **Fig. 12**). To be noticed: ESIN in children are not a smaller version of intramedullary nails in adults.
- **4.** In presence of *unstable*, displaced, *very open fracture* (*type II or III*) external fixation frame is indicated (see **Fig. 13**), also in association with ESIN.
- In **Humerus supra-condylar** fractures, prior check if no vascular compromise, eventual neurological in-

Fig. 11. From authorised source: Dr. Joachim Lauen



jury and suspicion of compartment syndrome. Initial management includes splinting the limb in a comfortable position (20-40° flexion) and pain control. Historically a majority of these fractures were treated with closed reduction and long arm casting/splinting with the elbow in position greater then 100° (with often consequent vascular problems and Volkman's contracture). Recently, closed reduction and percutanous pinning without the need for immobilizing the elbow in significant flexion is commonly recommended, and considered the safest form of treatment. Open reduction is **controversial**, indicated when the fracture is open and irreducible by closed methods (Brubacher JW et al 2008). Metaizeau JD (2017): "Do not pin a fracture that does not need it. If it is stable, flexion immobilization works very well").

Femoral neck: rare injuries in children, generally consequence of a high energy trauma. About 80% of these fractures are displaced with distal fragment. In small children the fracture may be perfectly anatomically reduced by manipulation, and leg immobilized (i.e. with full plaster spica in abduction) for 8-10 weeks. In older children whenever feasible open reduction with internal stable/rigid fixation is the golden standard treatment (as it is an unstable fracture) with nails or cannulated screws, or multiple thin Austin Moore's pinning, or osteotomy plate with screws, and capsular decompression*.

* **Controversial issue:** conservative vs. operative treatment; ORIF vs. CRIF. In **Bali** K (2011): "Femoral neck fractures in children need aggressive operative treatment aiming at anatomical reposition of femoral neck rather than conservative treatment. There should not be any hesitation to perform open reduction to achieve a stable anatomical reduction".

Fig. 13. Case of a 15 years old male, tibia and femur multiple displaced open fractures, caused by a motorcycle accident: traction femur, tibia/







Fig. 12. From authorised source: Dr. Jean Damien Metaizeau

Mass-casualty trauma/polytrauma: Goal of treatment is the quickest and safest stabilization, not the perfect one. JD Metaizeau: "Cast and traction are not good in mass trauma; plating and rigid nailing are too long procedures. Best method is flexible nailing - smaller diameter than usual, stainless steel, not much bending. With this procedure a well-trained surgeon should use an open approach, no traction for femur; not care for balance. Nails must not be cut too short. One can use flexible nailing also in association with external fixation".

Take home message

"With a child always choose the least invasive technique and best stabilization solution, and when possible keep it simple. In emergency mass trauma there is no time for perfect solutions, look for the simplest, quickest and safest solution".

Controversial issues

Material of ESIN (better stainless steel than titanium); superiority of flexible nailing to rigid intramedullary nailing, or to external fixation in most paediatric femoral fractures treatment. When to use the plate: age? Weight?

Eigth session

Eight session was on **Trauma sequelae**, moderated by Dr. **Oliver Birke**.

• Non-union, introduced by J. Lauen.

Non union in children is a very rare event (1/500 fractures), and it could be considered a complication of paediatric fracture management - post surgical complications, relating to the quality of surgical technical procedures, even if in some children non-union occur despite appropriate treatment (Arslan H 2003). When it happens, biological, mechanic and metabolic aspects must be explored. It can also be due to infection, increased age and/or gender of the children, and anatomical region of the fractures. In a study collecting regional fracture data by NHS Scotland, regarding Scottish children population from 2005 to 2010, it is emerged that the risk of non-union per fracture under the age of 15 is 1 in 500 for both boys and girls; over 15 (15-19) is approximately 1 in 500 for girls and 1 in 200 for boys (Mills LA et al. 2013).

Management

Non-surgical management

Cast immobilization until clinical and radiological union.

Surgical management

External fixation.

Reconstituted bone xenograft (RBX) can be safely used and has a good biocompatibility, does not cause immunological rejections and promotes bone healing.

Consensus vs. controversial issues

Age limitations?

• Bone Defect (Shortening/Lengthening), presented by dr. A. Cherkashin.

Limb length discrepancies can be due to congenital anomalies, or to acute trauma and/or infection, (or even to post-surgical complications) and it is more typical in lower extremity, although upper extremity bones can be affected as well.

Management

Treatment decision for children with length discrepancies are based not only on the extent of the disparity at the time of diagnosis, but also on predicted increases in discrepancies over time. In order to make this prediction, the surgeon takes into account the growth place location, the age of the child (the amount of growth remaining), if the discrepancy is due to trauma or infection, and the extent of injury to the growth plate.

Non-surgical management

If the discrepancy is less than 2 cm: a shoe lift is commonly used.

Surgical management

- For children with a discrepancy of about 2-5 cm, whose bones are still growing, epiphysiodesis may be appropriate. This procedure slows down the growth of the opposite leg or arm by altering the activity of the growth plate, and allowing the affected limb to catch up by the end of the growth. Proper time of the treatment is fundamental for best outcome.
- For discrepancies of more than (3)-5 cm. limb lengthening may be the effective treatment choice.
- Lengthening a shorter leg/shortening a longer leg may be recommended for children whose bones are no longer growing. In the treatment of bone defects, external fixation has the advantage of small trauma, simple operation, elastic fixation without stress shielding and no-limitation from local soft tissue conditions; besides there is a satisfactory functional recovery of the affected extremities. Average healing time 4-6 months.

It is important to check vascular and neurological aspects. Soft tissue coverage: is soft tissue movement predictable?

Procedures:

Ilizarov method: Osteotomy (Metaphysis); place circular external fixation with pins and wires for distraction osteogenesis. Tissues modulation under distraction. Flaps, skin graft. Continual frame adjustment. Great care to protect blood supply. External fixation is good for soft tissues healing and weight bearing. No more than +/- 3-10 cm (an amount associated with a lower number of complications). After frame removal, casting (*for how long? To check with consensus*). A physical therapy must be followed during the frame period and after, in order to keep flexible the soft tissues and maintain the muscle strength.

• Bone Defect: transport, presented by Dr. M. Samchukov.

Bone transport is a simple, reliable, biological procedure, with low complications but a high rate of success for managing segmental bone defects in children. Untreated or badly managed osteomyelitis can lead to bone loss.

Open fractures, in children who often have not completed their bone development, can provoke true or in situ bone loss with resulting limb length discrepancies. It can be useful to consider bone loss as an "anticipated non-union", and decide the strategies in order to prevent this possibility, and shorten the healing period.

Goal of treatment

Filling the bone gaps and promote bone union. Obtain limb equalization and functionality (*to check with consensus*).

Management and procedures

Surgical management

Osteosynthesis/osteotomy/sequestrectomy and bone transport (average – seven days after osteotomy, 1 mm per day). External fixation frame, circular or monolateral, with gradual adjustment. Commonly a successful bone healing, very rare refracture after frame removal.

Outcome parameters

Bone parameters – union, deformity, infection, limb length discrepancy; functional parameters; external fixation parameters (time in external fixation and external fixation index – number of days the external fixator is attached to the bone per centimetre of length gained). Ilizarov method has the advantage of early weightbearing, and treatment infection and bone defect in onestage surgery.

• Growth disturbance for partial/complete physeal arrest, presented by Dr. Franz Birkholtz, a consultant

orthopaedic surgeon from Unitas Hospital, Mediclinic Midstream and Zuid-Afrikaans Hospital, Pretoria, South Africa.

The most common growth plate injury is that of the distal radius, followed by the distal tibia. Permanent damage to the growth plate can be produced by illnesses, trauma or inadequate treatment.

Impairment of growth after fractures which involve the distal tibial physis may cause a length discrepancy, an angular deformity or both (Ogden JA 1982).

Imaging

"X-rays must be interpreted in order to understand the trauma mechanism and evaluate the grade of damage to the physeal plate correctly. The young patient must be carefully assessed and the fracture displacement must be assessed by X-Rays or if necessary by CT scan" (Schurz M et al. 2010).

Growth disturbance lines can be seen on standard radiographs and (for the distal tibial physis) are reliable indicators of growth plate activity after fracture: "Future function of the physis may be predicted from the appearance of the growth disturbance line as early as three months after injury, and used to assess the need for surgery before any gross deformity" (Hynes D et al. 1988).

Controversial

Using algorithm to predict the pathway?

Consensus

Tailored made therapy designed on patient's needs and family's capacity of sacrifice. Flexible and combined approach strongly recommended.

Management

It can be treated conservatively (i.e. shoe lift) or surgically.

Undisplaced fractures can be treated with cast and immobilization.

Displaced fractures can be treated with open or closed reduction, through internal and or external fixation, to reach an absolutely anatomical reduction in all planes, followed by cast immobilization unless external fixation has been used. Weight bearing or non-weight bearing depends on patient's age and compliance, fracture location, fragment size, stable or unstable fracture, comminution, stability of osteosynthesis. Follow-up period (*duration?* average of two-years).

Final take home message from the faculty

"We warmly recommend reasonable, tailored made and flexible/combined approaches, decided on patient's situation and personal needs, always with the family (or adult caring context) involvement and compliance".

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