

Ruolo della fissazione esterna nella correzione di deformità nei pazienti pediatrici in Spagna

Role of external fixation in the correction of deformities in pediatric patients in Spain

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- **Annual activity:**
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 - **16,323** surgical interventions
 - **25,747** hospital discharges



- Lower Limb Deformities
- Length Discrepancies
- Congenital Lower Limb Pathology
- Skeletal dysplasia
 - Short Stature
 - Deformity
- Collaboration with the Trauma and Upper Limb Unit in Cases with External Fixation



Dra. Méndez

Dr. G. Fontecha



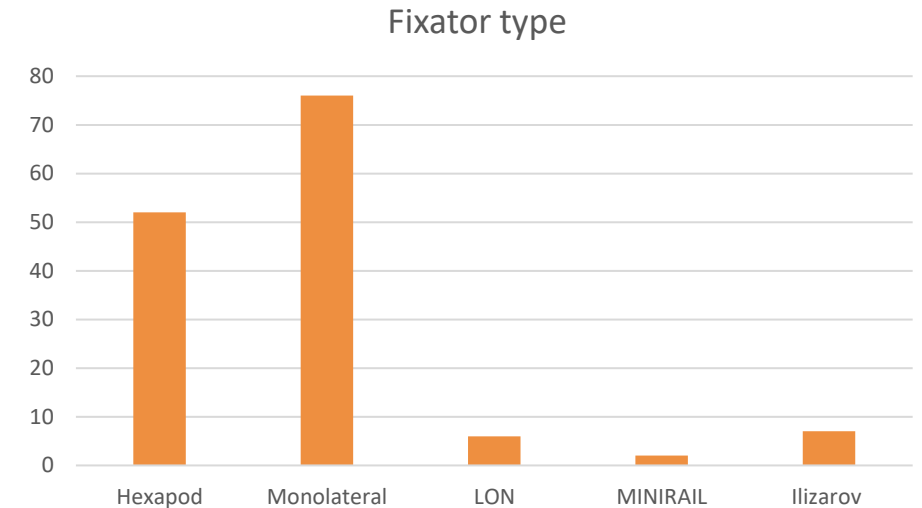
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EUROPEAN REFERENCE NETWORK
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- **Patient Data Overview (2020-2024)**
- **Total Patients:** 109
- **Gender Distribution:**
 - Men: 52
 - Women: 57
- **Bilateral Cases:** 34 (affecting 143 extremities)



Skeletal Dysplasias (41, 37.6%)

- Achondroplasia (n=25)
- Other dysplasias (n=7)
- Oler disease (n=6)
- Hypochondroplasia (n=3)

Upper Limb Pathology (3, 2.8%)

- Hand with short metacarpal (n=1)
- Radial spoon hand (n=1)
- Brachymetacarpia (n=1)

Short Stature (2, 1.8%)

(n=2)

Length Discrepancies (20, 18.3%)

(n=20)

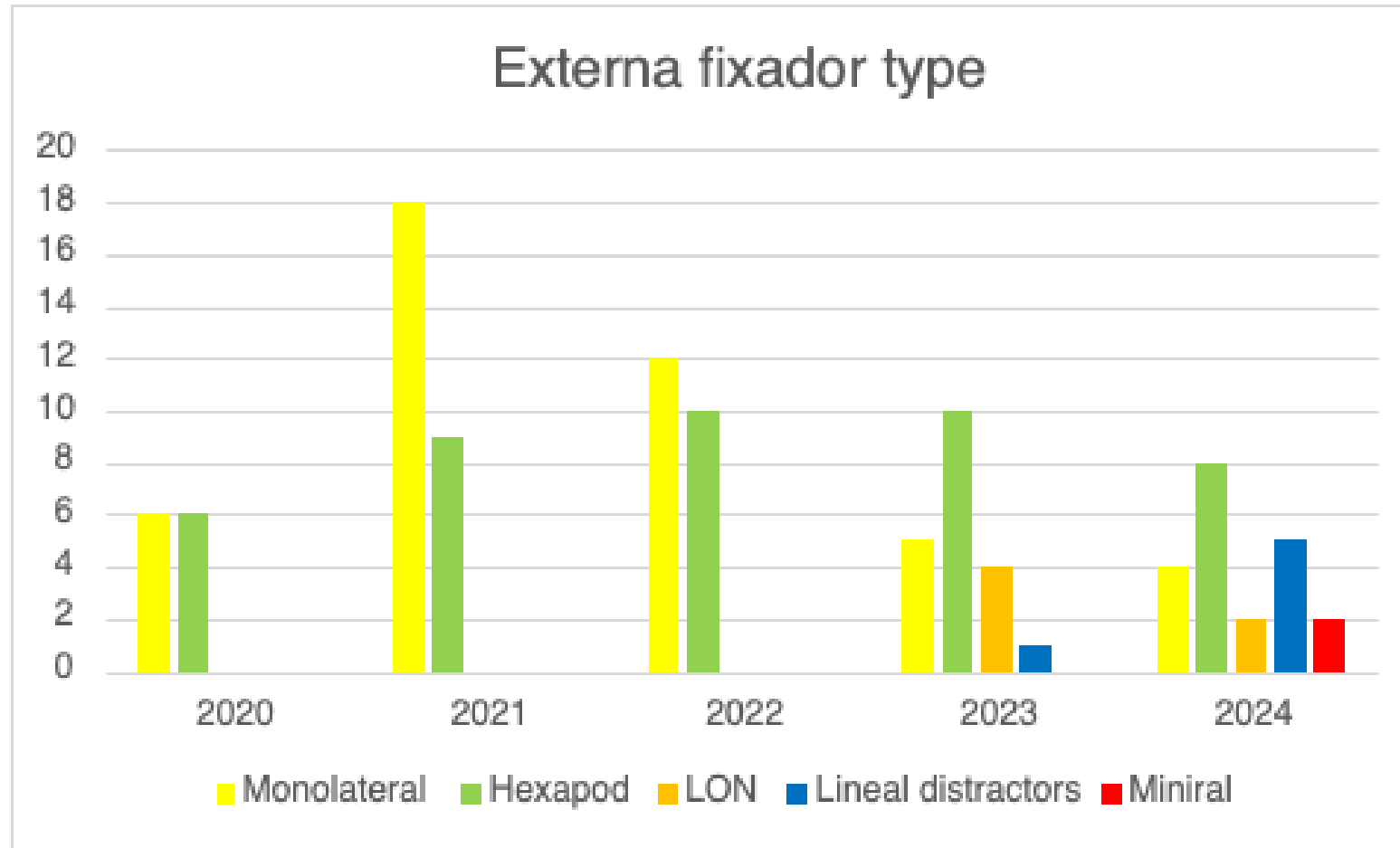
Congenital Pathology (36, 33%)

- Postaxial deficiency (n=15)
- CFD (n=5)
- Clubfoot (n=5)
- Blount's disease (n=5)
- Popliteal pterygium syndrome (n=3)
- Tibial hypoplasia (n=3)

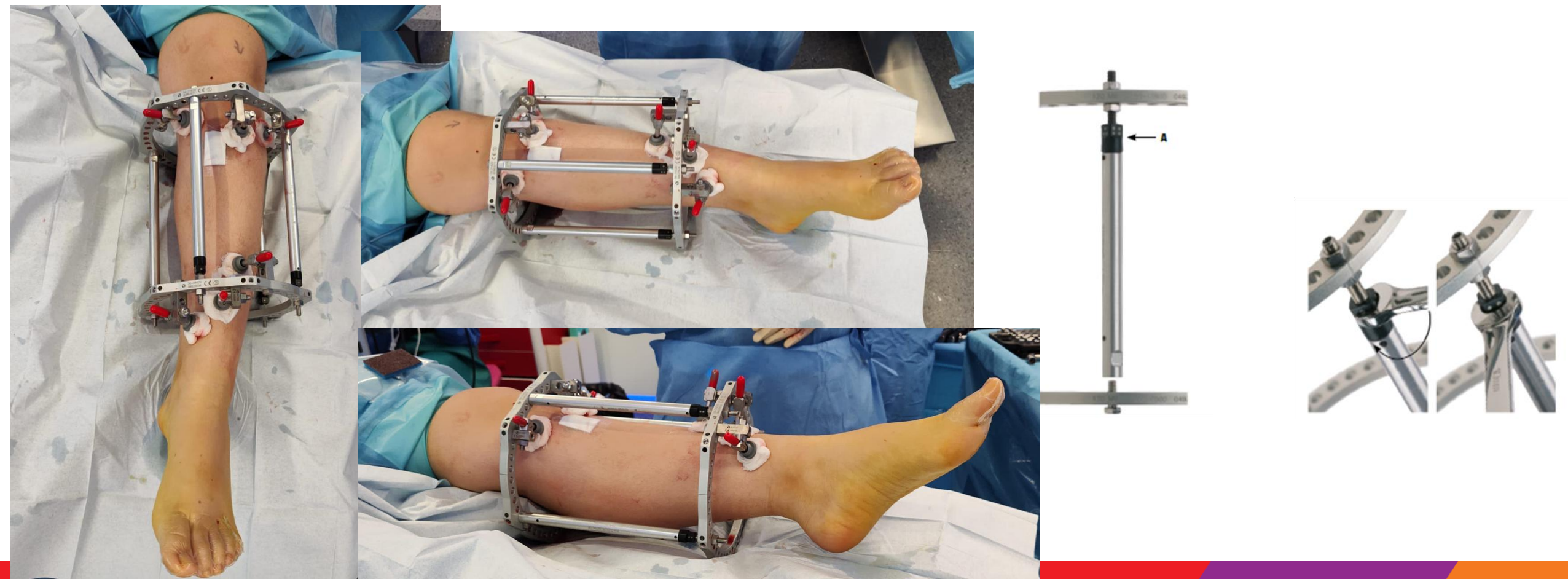
Physeal Arrest (7, 6.4%)

(n=7)



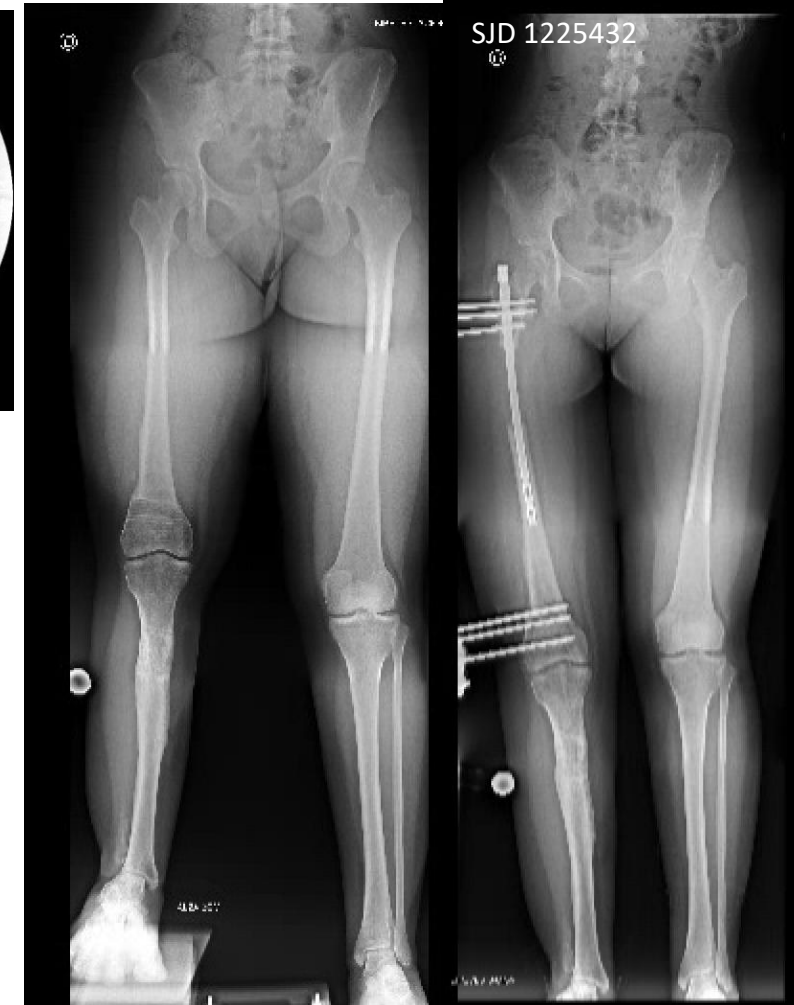
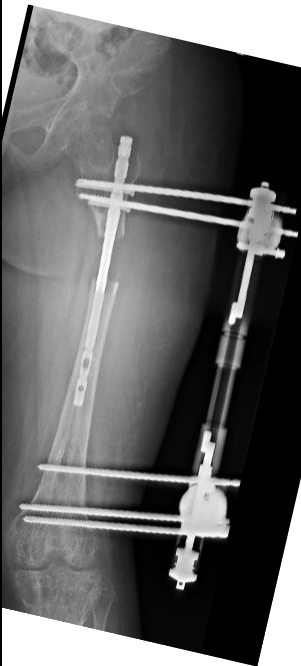


- From monolateral fixators to circular frames with distractors
- Similar cost and permits conversion to hexapod in case of angular deviations.



LON (Lengthening over nail)

- Reduce the time patients spend in external fixation
- Reduce complications (deviations and fractures)
- Cost-conscious alternative to lengthening nails
- Technically complex



- Lengthening and after nail

Prophylactic Intramedullary Rodding Following Femoral Lengthening in Congenital Deficiency of the Femur

Amr A. Abdelgawad, MD, Julio J. Jauregui, MD,† Shawn C. Standard, MD,†
Dror Paley, MD,‡ and John E. Herzenberg, MD, FRCSC†*

(J Pediatr Orthop 2017;37:416–423)

- Reduction in number of fractures from 34% to 18%



Prophylactic Intramedullary Rodding After Femoral Lengthening in Patients With Achondroplasia and Hypochondroplasia

Pilar Rovira Martí, MD, Ignacio Ginebreda Martí, MD,† and César García Fontecha, MD, PhD**
J Pediatr Orthop • Volume 44, Number 3, March 2024



	N without /with		% post removal fractures without / with		Lengthening withput / Con (cm)		% lengthening Sin / Con		EFI (days/cm)	
<i>Rovira et al. (2023)</i>	44	42	15.9%	2.4%	15.1	14.5	61.5	52.6	24.5	25.8
<i>Park et al. (2015)</i>	56		26.8		8.4		37		28.1	
<i>Venkatesh et al. (2009)</i>	40		15		9.2		39.3		39	
<i>Chibule et al. (2016)</i>	10		10%		9.9		52.8		25.6	

- Limb lengthening protocol**

Short	Age	Lengthening (cm)	Total
Tibial lengthening	14 years of age	15 cm	30cm
Femoral lengthening	16 years of age	15 cm	
Humeral lengthening	17 years of age	9 cm	

Long	Age	Lengthening (cm)	Total
Tibial lengthening	10 years of age	10 cm	40cm
Femoral lengthening	12 years of age	10 cm	
Tibial lengthening	14 years of age	10 cm	
Femoral lengthening	16 years of age	10 cm	
Humeral lengthening	17 years of age	9 cm	



Acondroplasia	
Código: A-COT-PC-0012-01	Ámbito: Hospital
Autores: Pilar Rovira Martí, César Galo García Fontecha, Rosa Bou Torrent, Meritxell Vigo Moráncho, Miriam Pérez Cruz, Martín Tronzo Sanz, Isabel Gascón Casareil, Ignacio Barber Martínez de la Torre, Oliver Heinz Haag, José Hinojosa Mena Bernál, Yndira Carolina González Chopite	
Fecha aprobación: Abril de 2022	Fecha revisión:
Palabra clave: Acondroplasia	
Fecha revisión Comisión de Farmacia:	

1. Descripción

La acondroplasia es una displasia esquelética que afecta al crecimiento normal del cartilago (condrodisplasia), siendo la causa más frecuente de talla baja desproporcionada. Se caracteriza por una talla baja rizomélica con tronco normal, hiperlordosis lumbar e hipercifosis dorsal, braquidactilia con mano en tridente, dificultad para la extensión completa de los codos, macrocefalia con abombamiento frontal y nariz deprimida en forma de "silla de montar", discreta hipotonía, inteligencia normal, aunque puede haber retraso del desarrollo al inicio.

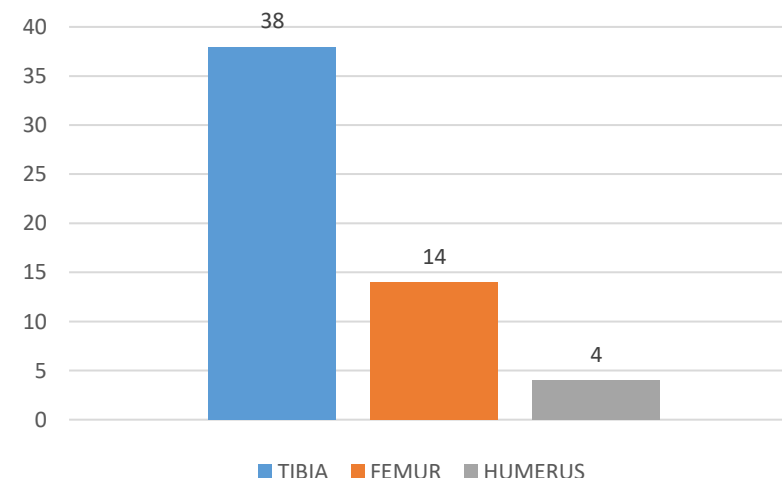
La acondroplasia puede presentar complicaciones como apneas, obesidad, obitis de repetición, hiperlordosis lumbar con estenosis del canal lumbar, estenosis del foramen magno e hidrocefalia.

La prevalencia estimada es de 1 por cada 25000-30000 recién nacidos.

2. Etiopatogenia

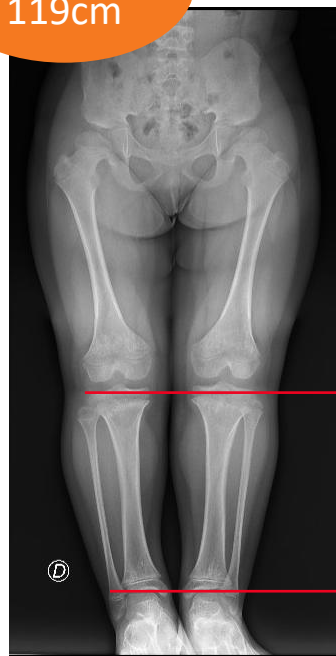
La acondroplasia se produce por mutaciones en el gen del receptor 3 del factor de crecimiento del fibroblasto (FGFR3) que codifica a un receptor transmembrana importante para la regulación del crecimiento lineal de los huesos. La mayoría de las mutaciones aparecen en el mismo par de nucleótidos, que resultan en una sustitución de una glicina por una arginina c.1138G >A (G380R). Tiene una herencia autosómica dominante, aunque el 80% de las mutaciones se producen de novo. El riesgo de tener un hijo con acondroplasia si hay un progenitor afecto, es del 50%. Si los dos progenitores están afectados, existe una probabilidad del 50% de tener un hijo/a con acondroplasia (mutación en heterocigosis), del 25% de tener un hijo/a sano (sin mutación) y del 25% de tener un hijo/a con mutación en homocigosis (que generalmente será letal).

Achondroplasia /Hypochondroplasia

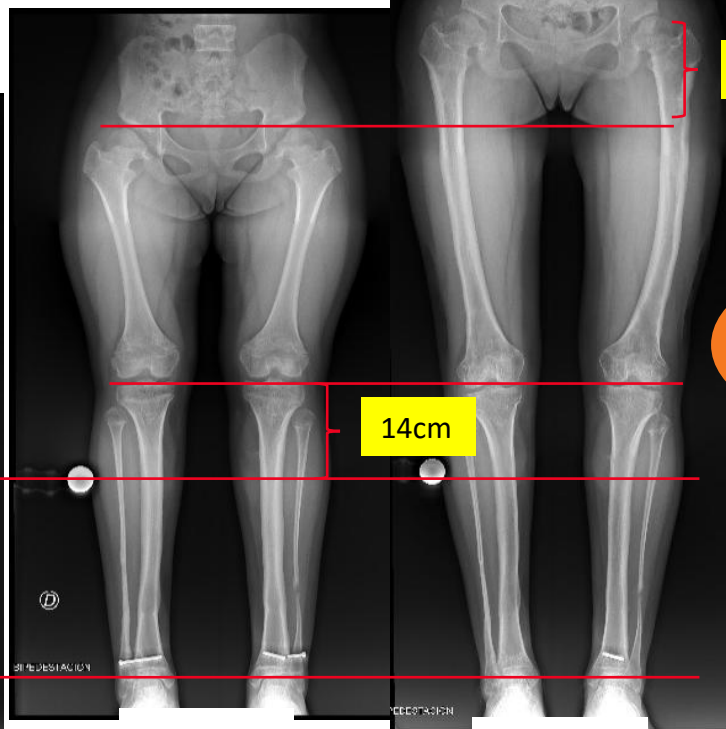




Height
119cm



12 years



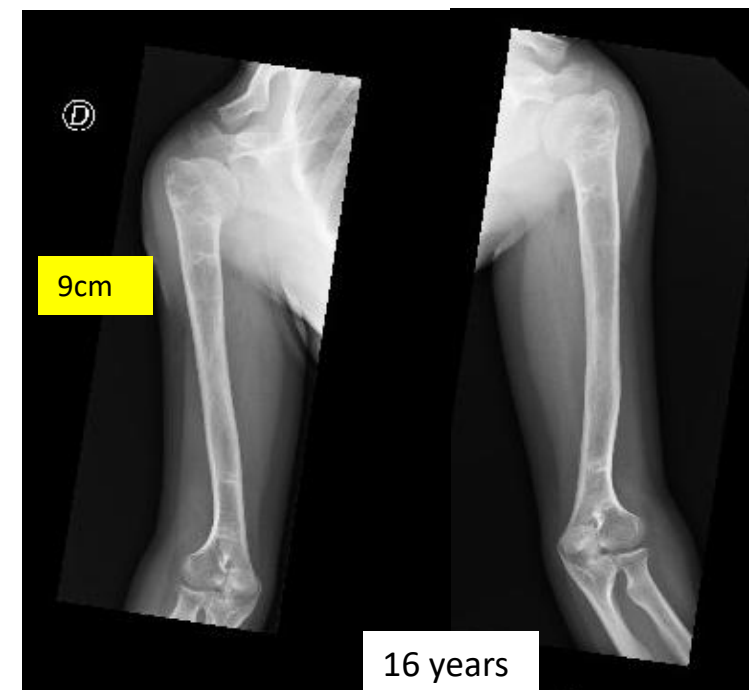
14 years

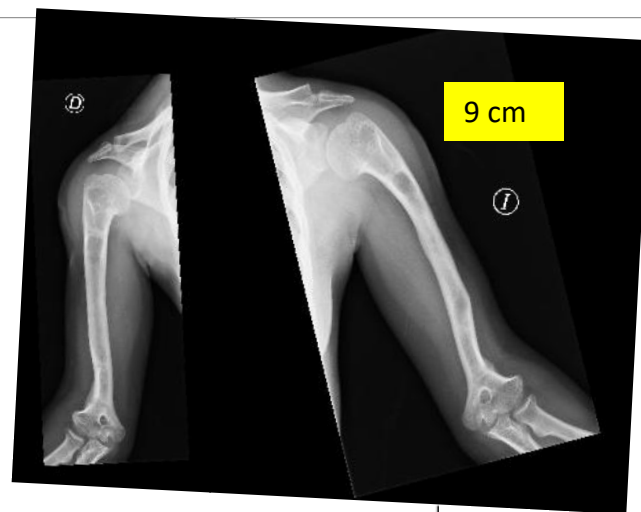
14cm

Height
149cm

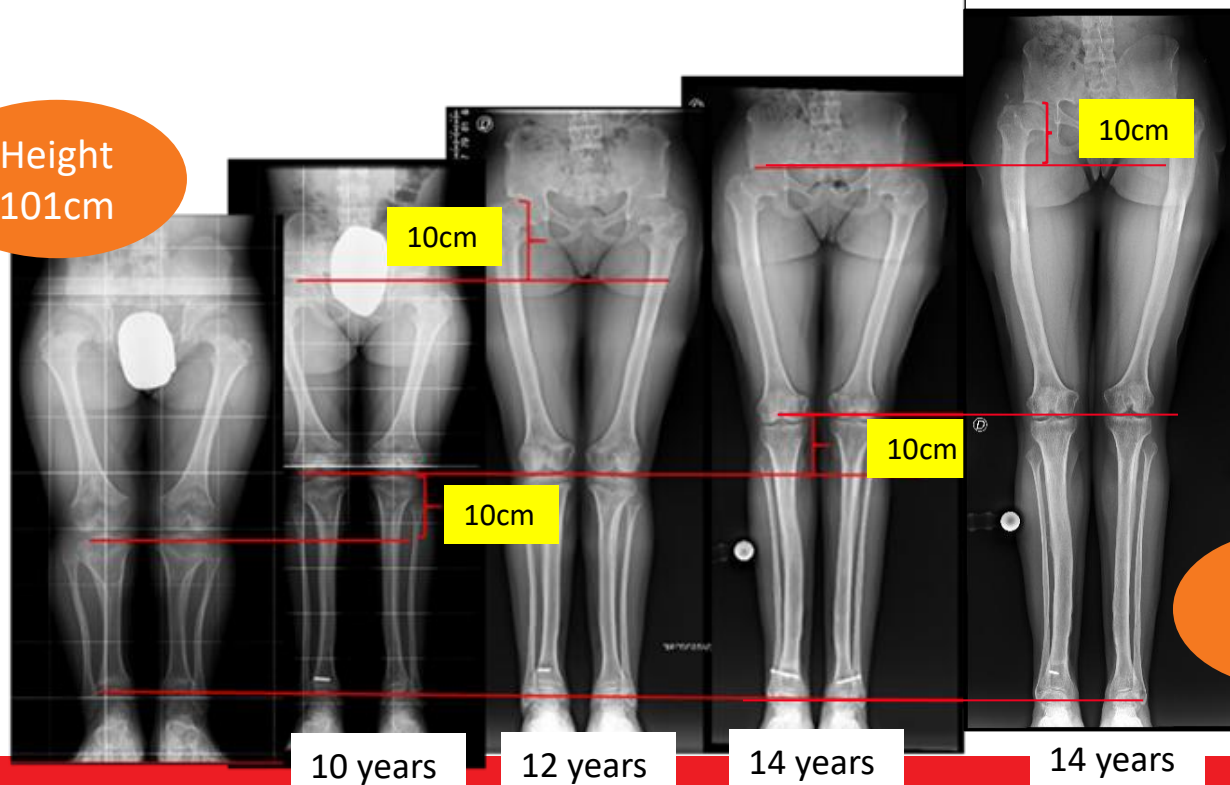
14cm

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Humeral lengthening	17 years of age	9 cm	





Height
101cm



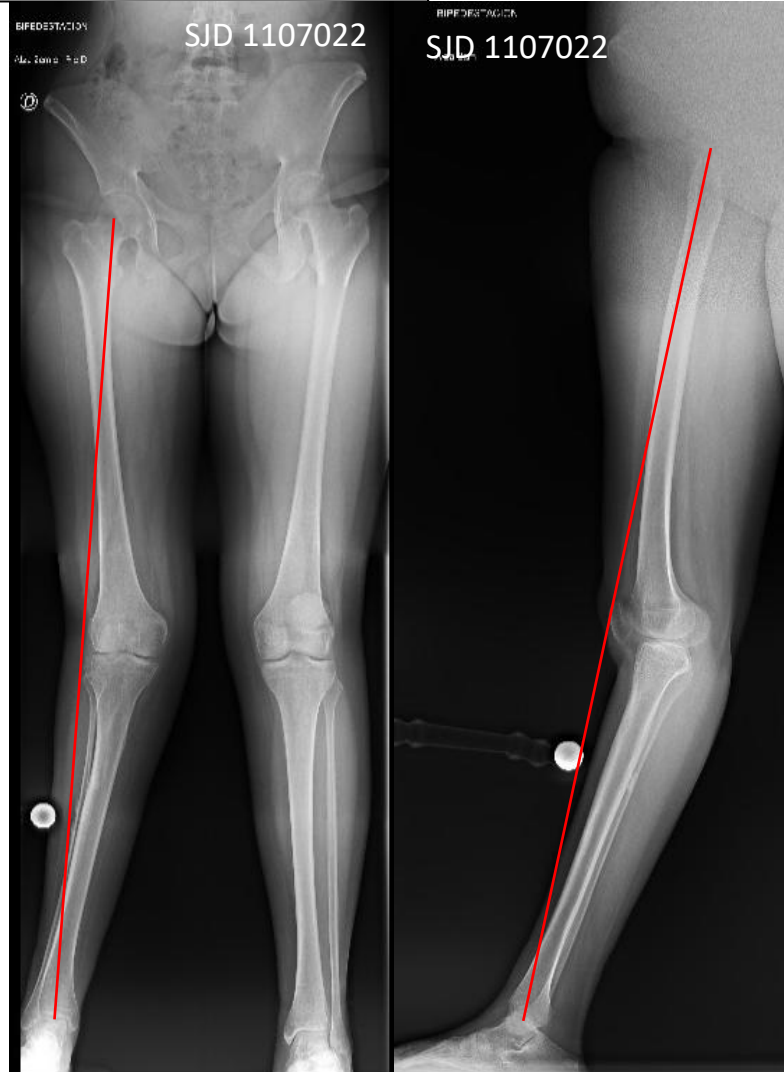
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Femoral lengthening	16 years of age	10 cm	
Humeral lengthening	17 years of age	9 cm	

Height
156cm

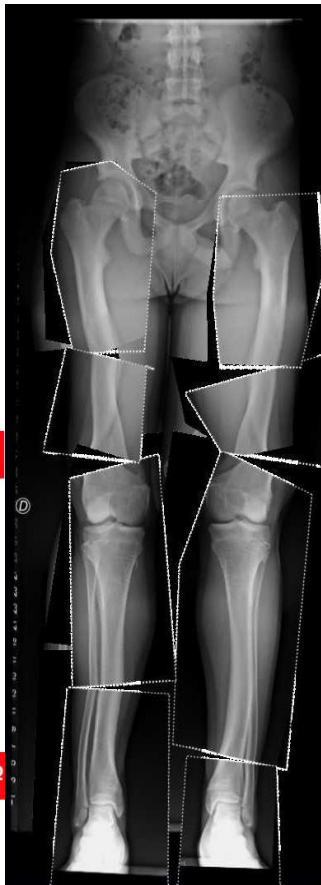
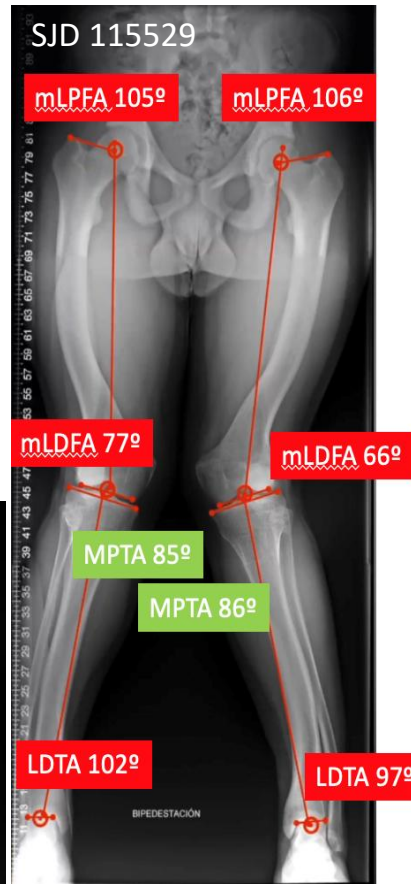
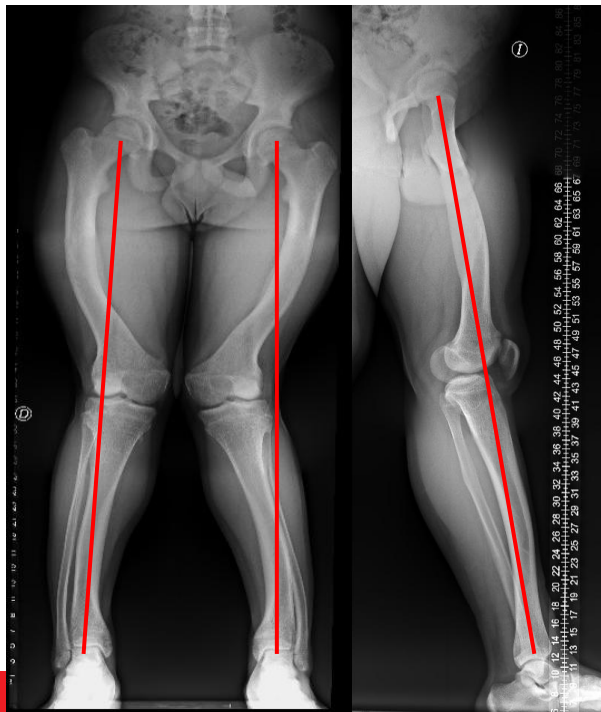
Sepsis at age 7

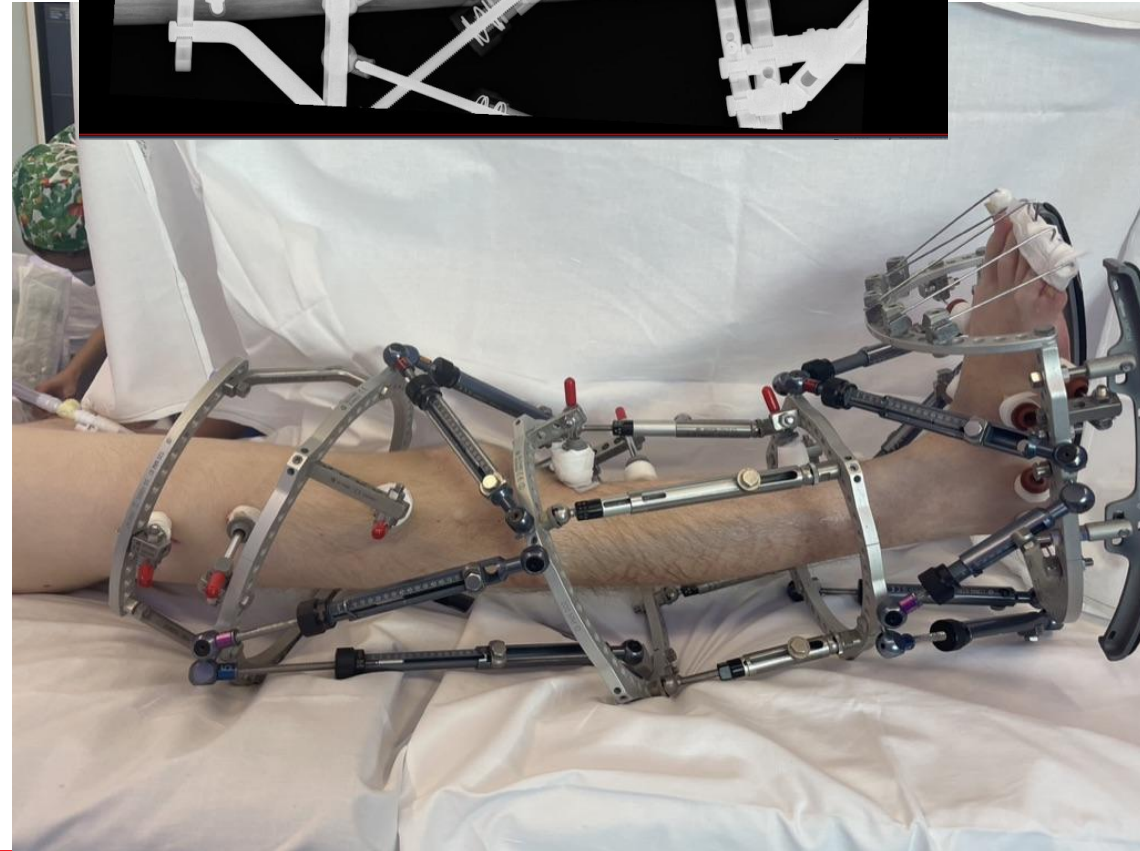
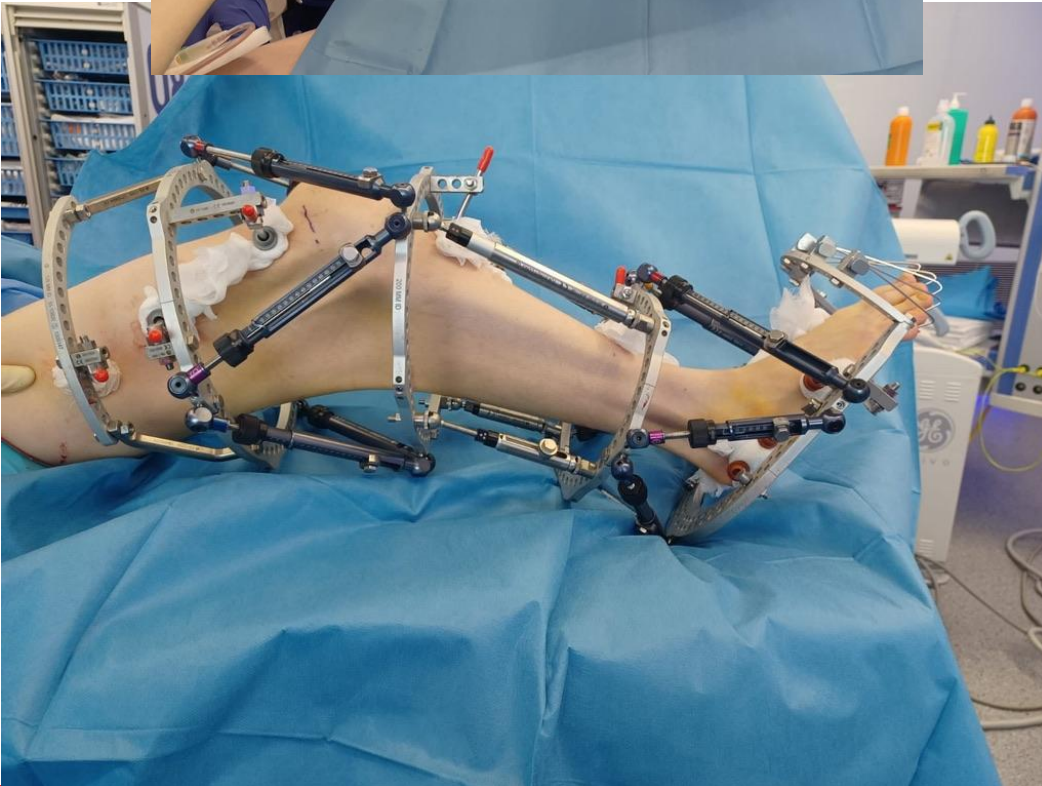
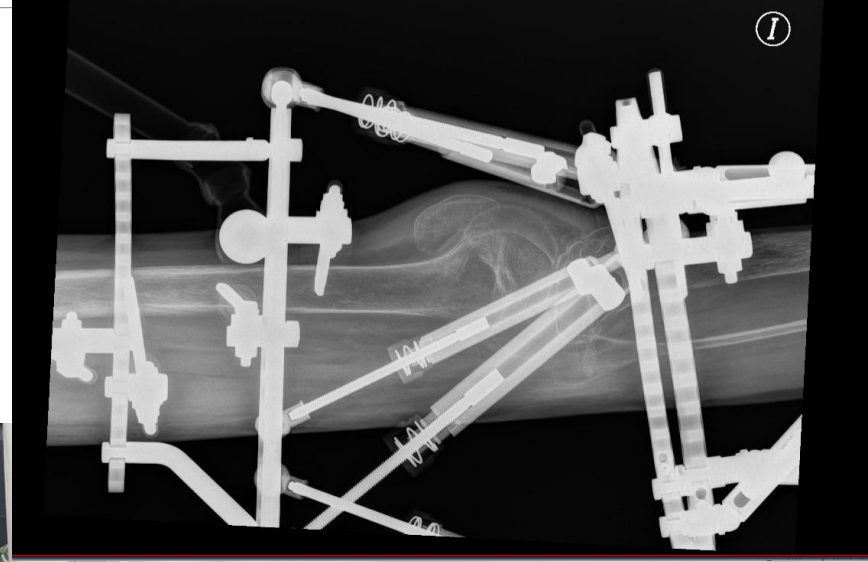
Neurological impairment with dorsal flexion deficit of the ankle

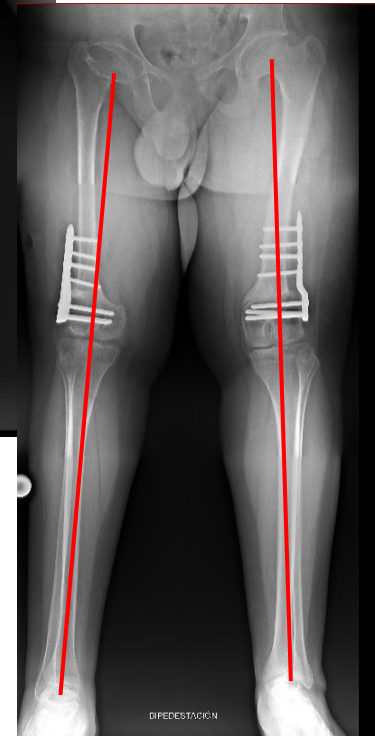
Recurvatum 30-40° and limb length discrepancy 2cm



- X-linked hypophosphatemic rickets

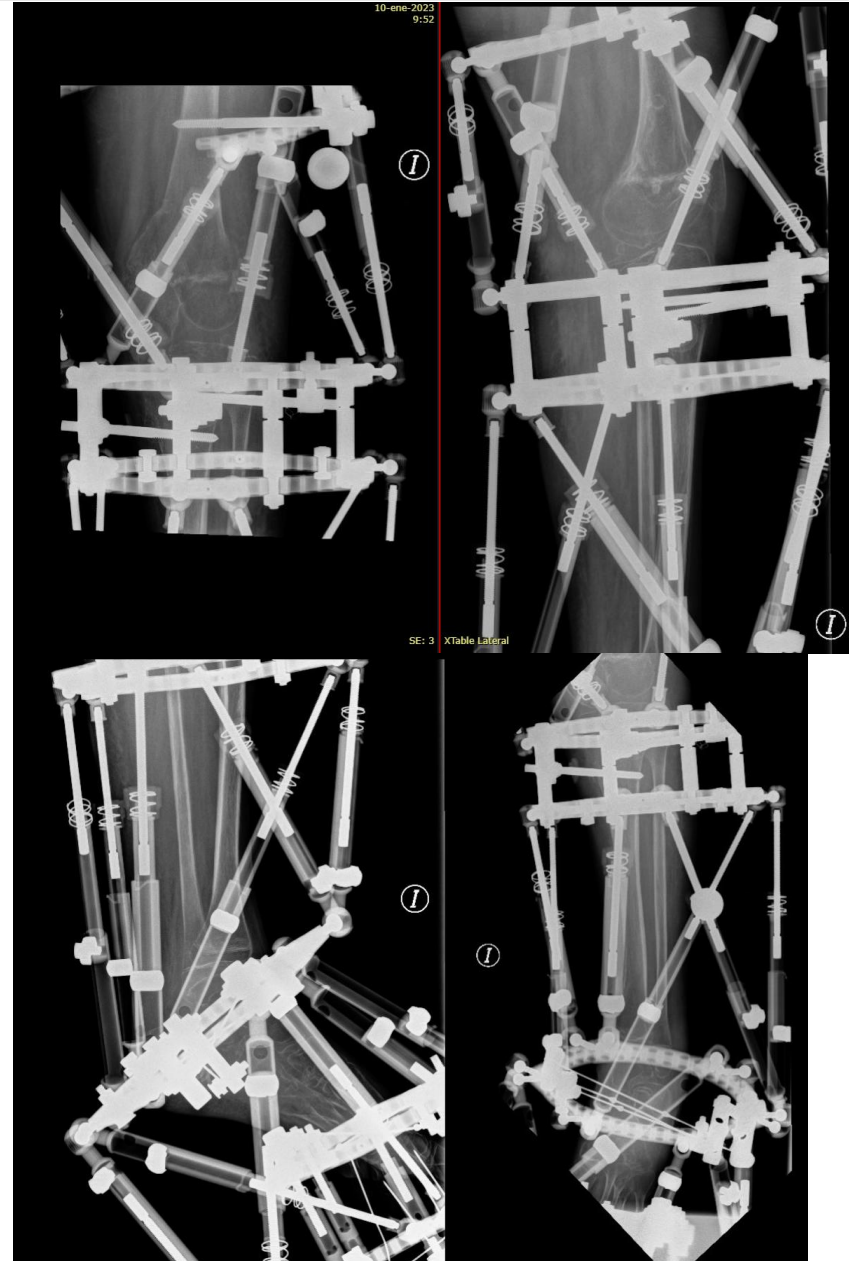
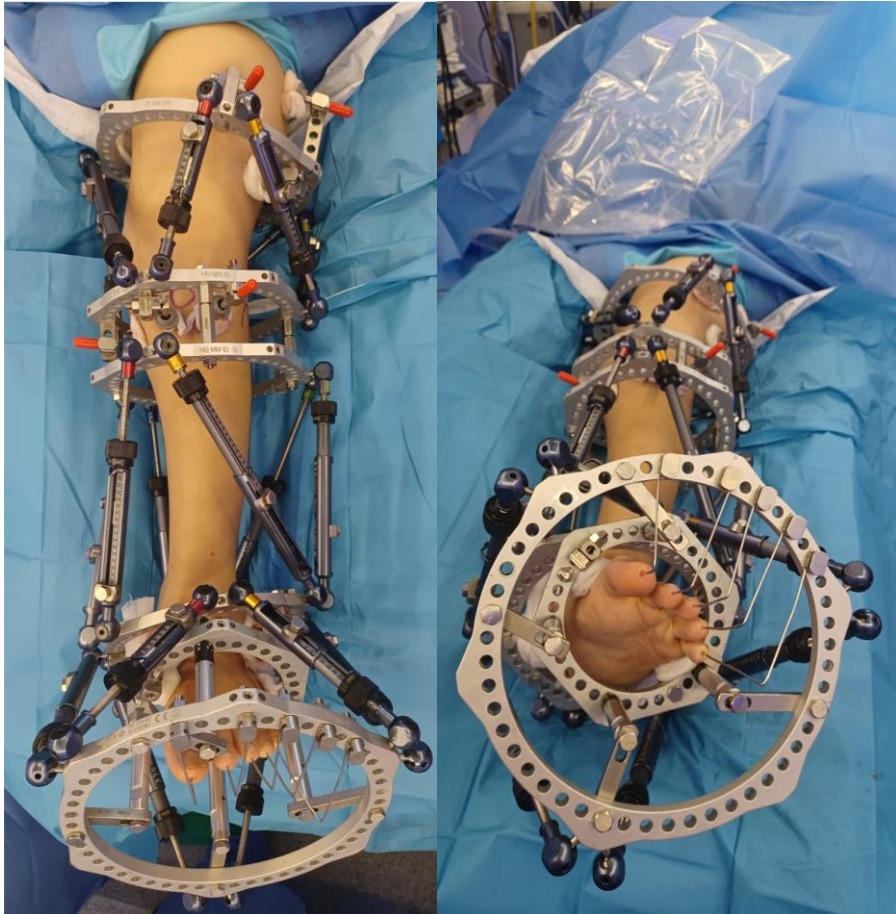






- Spondyloepiphyseal dysplasia with congenital joint dislocation







8 y 9 m
BMI 28 Kg/m²
50° Medial physeal slop

Gradual Deformity Correction with a Computer-assisted Hexapod External Fixator in Blount's Disease

Pieter H Mare¹, Leonard C Marais²

2022 STLR

>30° medial physeal slop → Medial elevation osteotomy

The Medial Elevation Osteotomy for Late-presenting and Recurrent Infantile Blount Disease

*Pieter Herman Maré, MBChB, FC Orth (SA),**

*David Mungo Thompson, MBChB, FRCS (Glasgow),**

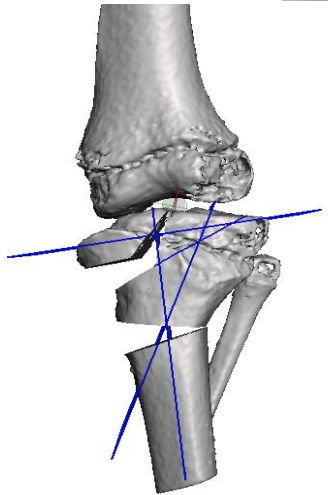
and Leonard Charles Marais, MBChB, FC Orth (SA), MMed (Ortho), PhD†

2021 JPO

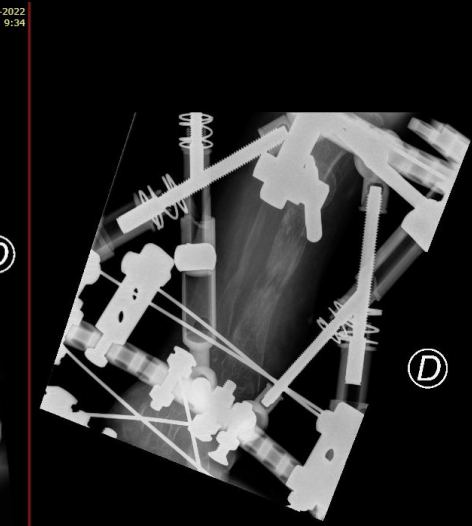
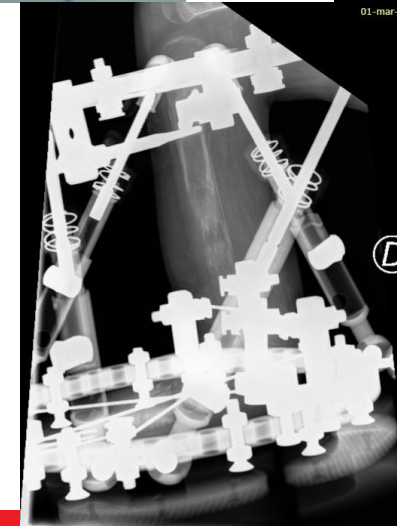
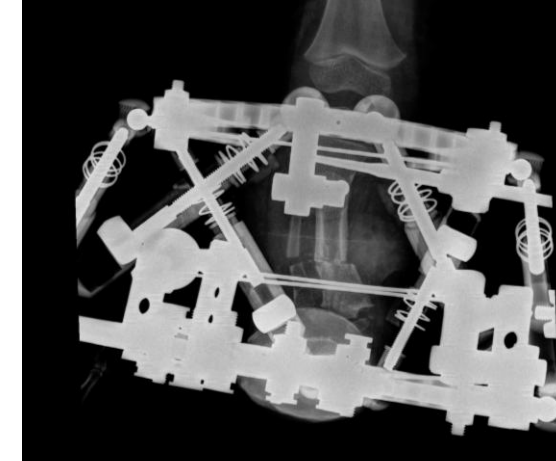
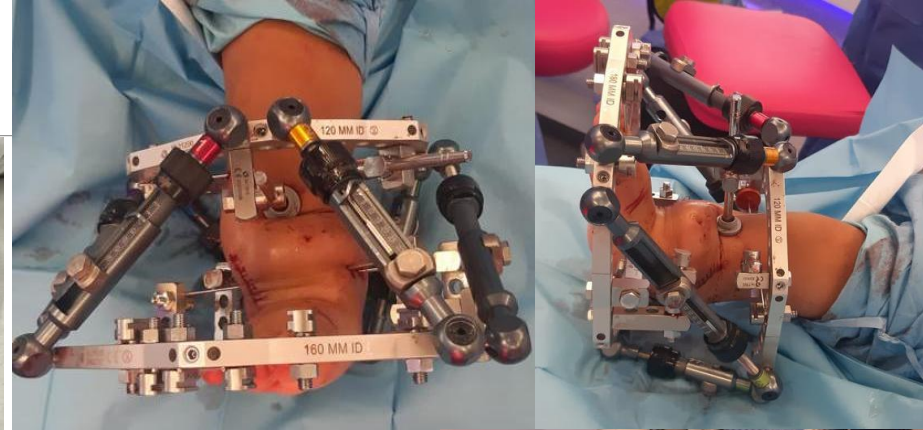
>40° medial physeal slop

Medial elevation osteotomy + lateral epiphysiodesis + metaphyseal osteotomy

75% good results

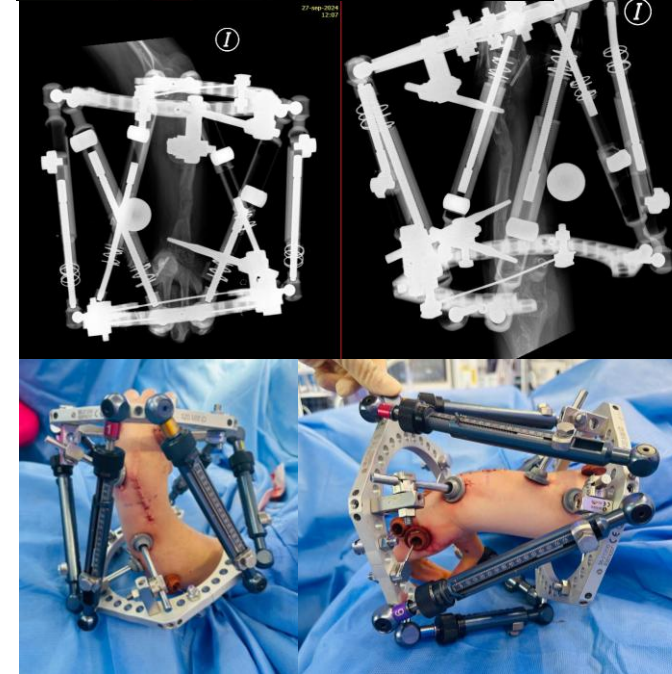
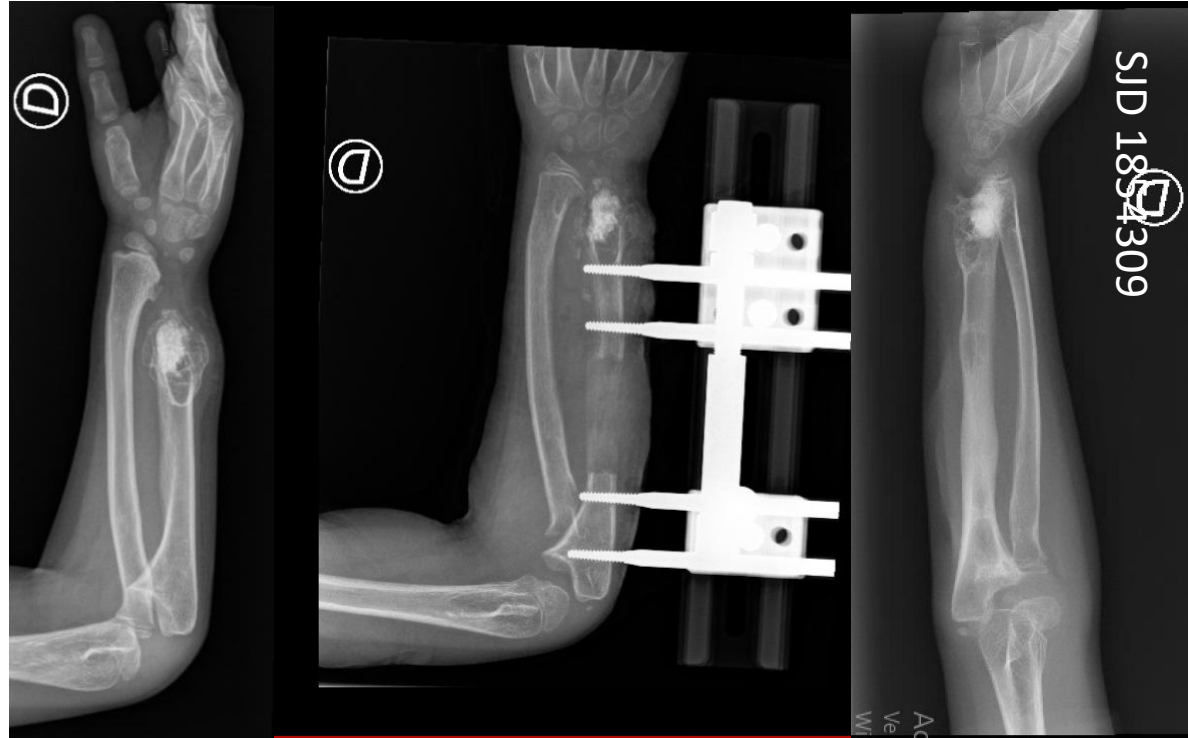


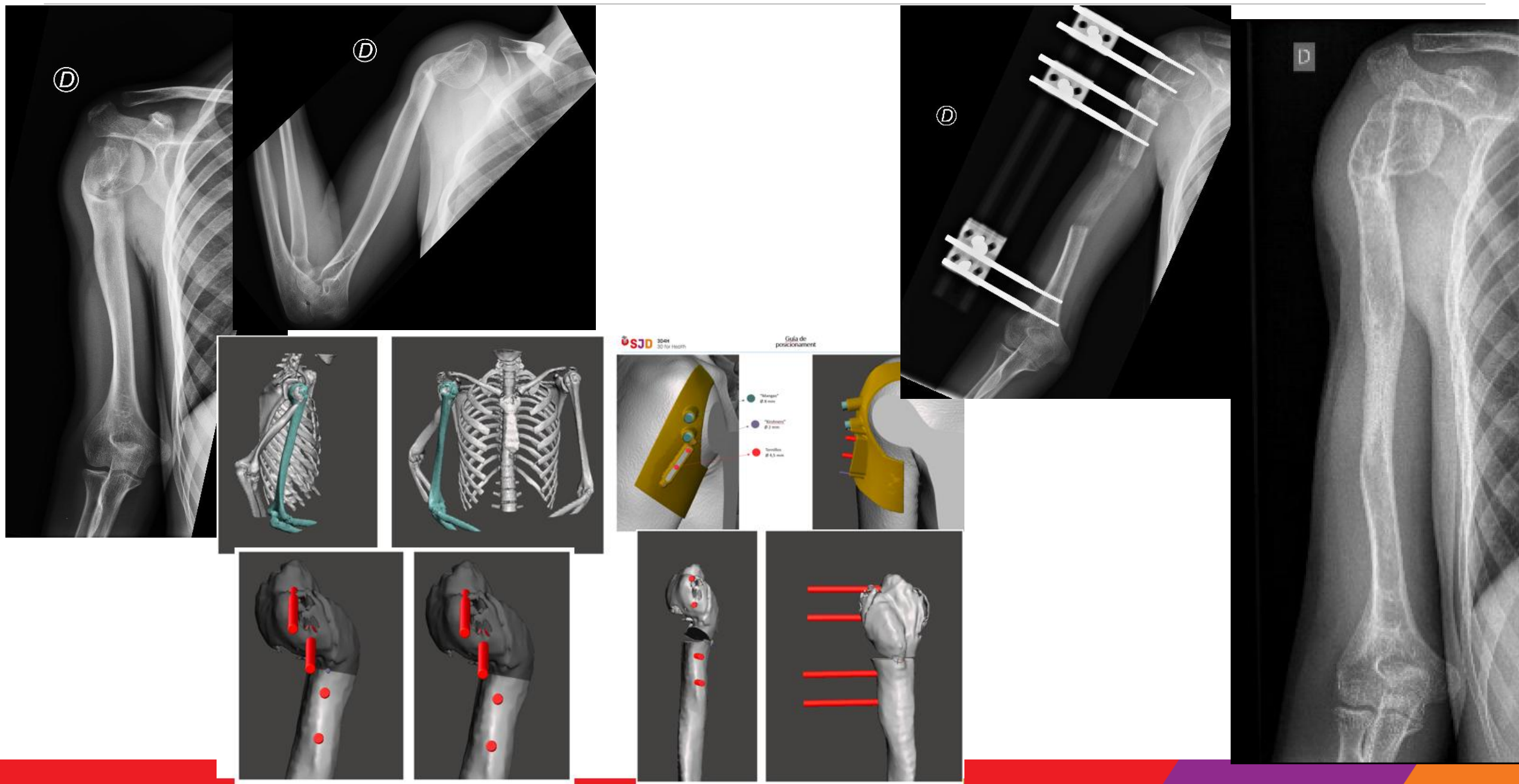










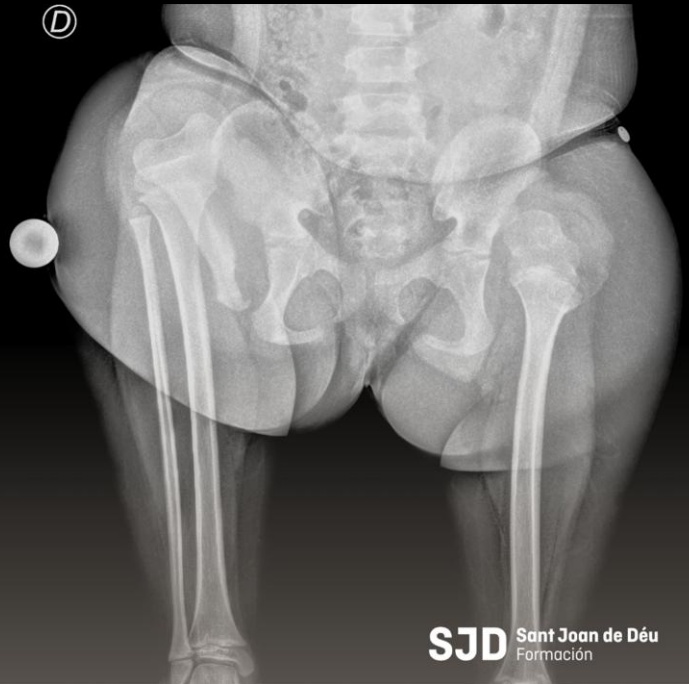


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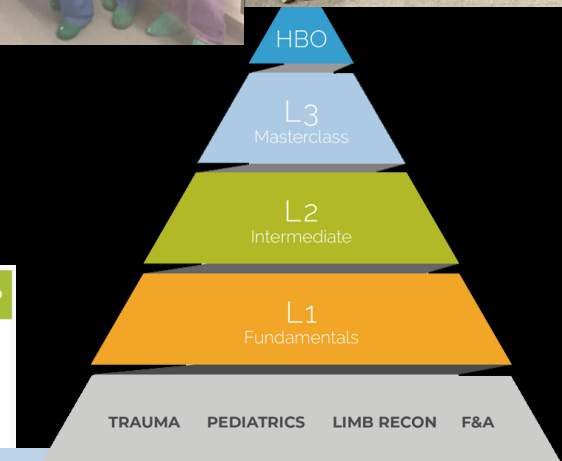


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