# Fibrous Dysplasia/McCune Albright Syndrome module

Meeting 11-09-2023





#### Evolving of the Registries in time













E-reporting -RarE Conditions tool (e-REC)



Calcium & Phosphate

& Bone



Condition
- Specific
Modules



Integration of the Registries in WP 5 of the European Reference Networks

2023

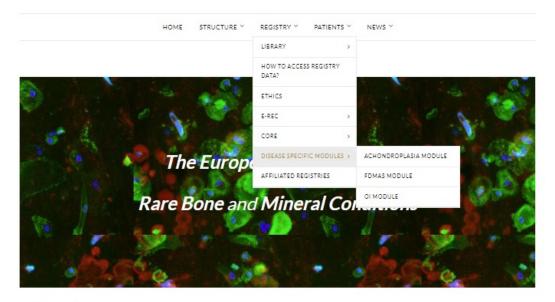
2017 2018 2020 2022

Core

Registry







#### What is EuRR-Bone?

Expert centers for rare bone and mineral conditions and patient representatives have joined their forces and started the European Registries for Rare Bone and Mineral Conditions (EuRR-Bone) in April 2020. This project works closely with the European Registries for Rare Endocrine Conditions project (EURRECa) and the European Reference Networks for Rare Bone and Rare Endocrine conditions and includes:

#### FIBROUS DYSPLASIA/MCCUNE ALBRIGHT SYNDROME MODULE

The disease specific module was launched April 2022 and is the work of the vertical theme **Fibrous Dysplasia/McCune Albright Syndrome** (FDMAS)

Want to start using the module? Get access to the Core registry **here**. After entering the patient in the Core Registry, the FDMAS module appears automatically and is ready to use.

If you have questions or interest in joining the working group, please contact us on EuRR\_Bone@lumc.nl

The full data dictionary can be downloaded here when available.

The modules should be updated every 12 months in adults and every 6 months in children when patients are under active control. If not updates should be provided every 3 years. Of course additional data can be entered on demand.

You only have to fill out the relevant modules to your patient eg when no Cranial Facial Fibrous Dysplasia or Endocrinopathies are present, the modules can be left empty.

Do not forget for all bone conditions EQ5D and mobility should be assessed (either by clinician or patient with access) yearly. You will need to set those within the general bone dysplasia module

Tools used in the module:

Brief Pain Inventory (coming soon in 10 languages) in adults and children

EQ5D

Calculation of the TmP/GFR for the assessment of hypophosphatemia



#### Brief Pain Inventory (Short Form)

Ziter i am inventory (cherri cim,	
. Throughout our lives, most of us have had pain from time to time (such as minor headaches, sprains, and toothaches). Have you had pain other than these everyday kinds of pain today?	7. What treatments or medications are you receiving for your pain?
☐ Yes ☐ No	
2. On the diagram, shade in the areas where you feel pain. Put an X on the area that hurts the most.	
Prigni Front Back	8. In the last 24 hours, how much relief have pain treatments or medications provided? Please mark the box below the percentage that most shows how much relief you have received.
	0% 10% 20% 30% 40% 50% 60% 70% 80% 90% 100%  No Relief  Complete Relief
	<ol><li>Mark the box beside the number that describes how, during the past 24 hours, pain has interfered with your:</li></ol>
	A. General Activity         □ 0       □ 1       □ 2       □ 3       □ 4       □ 5       □ 6       □ 7       □ 8       □ 9       □ 11         Does Not Interfere       Interfere       Interfere
3. Please rate your pain by marking the box beside the number that best describes your pain at its worst	B. Mood  □ 0 □ 1 □ 2 □ 3 □ 4 □ 5 □ 6 □ 7 □ 8 □ 9 □ 10  Does Not
in the last 24 hours.    0	C. Walking ability           □ 0 □ 1 □ 2 □ 3 □ 4 □ 5 □ 6 □ 7 □ 8 □ 9 □ 11           Does Not Interfere         Comple Interfere
4. Please rate your pain by marking the box beside the number that best describes your pain at its least in the last 24 hours.	D. Normal Work (includes both work outside the home and housework)  0 0 1 2 3 4 5 6 7 8 9 10  Comple
□ 0 □ 1 □ 2 □ 3 □ 4 □ 5 □ 6 □ 7 □ 8 □ 9 □ 10  No Pain As Bad As You Can Imagine	Interfere Interfere  E. Relations with other people
5. Please rate your pain by marking the box beside the number that best describes your pain on the average.	01234567891[  Does Not
0 1 2 3 4 5 6 7 8 9 10  No Pain As Bad As Pain	F. Sleep         □ 0       □ 1       □ 2       □ 3       □ 4       □ 5       □ 6       □ 7       □ 8       □ 9       □ 10         Does Not Interfere       Comple Interfere
6. Please rate your pain by marking the box beside the number that tells how much pain you have right now.  0 1 2 3 4 5 6 7 8 9 10  No Pain As Bad As You Can Imagine	Therefore





Anthropometry	Basic disease module	Disease characteristics	McCune-Albright syndrome	Craniofacial FD	Therapy
Assessment Da	nte		2	mm/dd/yy	уу
Height (cm)			•		
Height SDS			<b>2</b>		
Weight (kg)			•		
Weight SDS			•		
ВМІ			<b>2</b>		
BMI SDS			•		



Anthropometry	Basic disease module	Diseas	e characteristics	McCune-Albright syndrome	Craniofacial FD	Therapy		
Assessment	t Date	0	mm/dd/yyyy				Musculoskeletal	
Age at Asse	ssment Date	0					Fractures due to fibrous dysplasia	- Select -
							Limb deformity due to fibrous dysplasia	- Select - Progressive scoliosis: scoliosis that worsens
							Scoliosis due to fibrous dysplasia	by more than 5 degrees or more between x-rays
							Pain at fibrous dysplasia site	- Select -
Endocrino	pathies						Bone and mineral metabolism	1
Endocrinopa	athy	0	- Select -		Ÿ		Alkaline phosphatase	- Select -
							CtX-1	- Select -
							Hypophosphatemia, serum confirmation	- Select -



Anthropometry	Basic disease module	Disease characteristics	McCune-Albright syn	drome	Craniofacial FD	Therap	ру	
Assessment Da	nte		0		mm/dd/yy	ууу		
Age at Assessn	ment Date		? Age at	Assessr	ment Date for this S	ection		
Endocrinopathy	,		•		None Neonatal I Precociou Thyroid in Growth ho	s pubert volveme ormone e	ty ent excess	
Age at presenta	ation of neonatal hyper	cortisolism if applicable	2					
Age at presenta	ation of precocious pub	erty if applicable	2					
Age at presenta	ation of hyperthyroidisr	n if applicable	2					
Age at presenta	ation of GH excess if ap	pplicable	•					
Age at presenta	ation of hyperprolactine	emia if applicable	•					





ge at Assessment Date	ropometry	Basic disease mod	ule I	Disease characteristics	McCune-Albright syndrome	Craniofacial FD	Therapy
Age at Assessment Date    None	Past Thera	ру					
D related medication  None Active vitamine D Bisphosphonates oral Bisphosphonates intravenous Cholecalciferol Denosumab Phosphate Not known	ssessment l	Date	0	mm/dd/yyyy	/		
Active vitamine D  Bisphosphonates oral  Bisphosphonates intravenous  Cholecalciferol  Denosumab  Phosphate  Not known	ge at Asses:	sment Date	•				
	FD related me	edication		Active vitam Bisphosphol Bisphosphol Cholecalcife Denosumab Phosphate Not known	nates oral nates intravenous erol		
	dication for ledication	FD related		Growth Low phosph Pain Prophylaxis To improve o			



Questionnaire: Fibrous dysplasia/McCune Albright Condition specific patient reported outcomes

Professionals I/the patient see			
Assessment Date	0	mm/dd/yyyy	
ge at Assessment Date	0		
How many health professionals do you/the patient see?		<ul><li>○ 1</li><li>○ 2</li><li>○ 3</li><li>○ 4</li><li>○ 5</li></ul>	
Which health professionals do you/the patient see regularly?	•	Professionals that take part in your follow up, therefore, you have appointments at regular intervals.    Opntnaimologist	
Other (freetext)	0		