

Follow-up study in patients with *HNF1B*- Nephropathy – Basic data

Date of examination:

Medical center:

Patient number:

Year of birth:

Gender: ♀ ♂

Body height: cm Bodyweight: kg

Clinical data

Sonographic abnormality of the kidneys prenatal:

None

Unilateral

Bilateral

→ Hyperechogenicity

→ Hyperechogenicity

→ Renal cysts

→ Renal cysts

→ Urinary tract malformation

→ Urinary tract malformation

Others: _____

Oligohydramnios: Yes No Unknown

Birth: week of gestation

Abnormality of genital tract:

None

Yes, _____

Initially documented renal function (Date _____):

Body height cm Body weight kg

Creatinine $\mu\text{mol/l}$ or mg/dl

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Genetics:

HNF1B:

please provide detailed result of mutational analysis (change on nucleotide and protein level), if possible

Family genetics:

- de novo
- maternal
- paternal
- unknown

Family anamnesis

Family member	Renal dysplasia	MODY type 5	Malformation of genital tract	Elevated liver enzymes
Mother	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Father	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Sibling 1	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Sibling 2	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Others	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Family tree

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Current sonography:

	right kidney	left kidney
Size (Length x Width x Depth in cm)	_____	_____
Echogenicity	<input type="checkbox"/> normal <input type="checkbox"/> increased	<input type="checkbox"/> normal <input type="checkbox"/> increased
Corticomedullary differentiation	<input type="checkbox"/> normal <input type="checkbox"/> reduced <input type="checkbox"/> lost	<input type="checkbox"/> normal <input type="checkbox"/> reduced <input type="checkbox"/> lost
Cysts	<input type="checkbox"/> none <input type="checkbox"/> n ≤ 5 <input type="checkbox"/> n > 5 <input type="checkbox"/> d ≤ 10mm <input type="checkbox"/> d > 10mm	<input type="checkbox"/> none <input type="checkbox"/> n ≤ 5 <input type="checkbox"/> n > 5 <input type="checkbox"/> d ≤ 10mm <input type="checkbox"/> d > 10mm

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Laboratory investigations (serum)

Creatinine	<input type="text"/>	$\mu\text{mol/l}$ or	<input type="text"/>	mg/dl
Uric acid	<input type="text"/>	$\mu\text{mol/l}$ or	<input type="text"/>	mg/dl
GOT	<input type="text"/>	U/l		
GPT	<input type="text"/>	U/l		
γ -GT	<input type="text"/>	U/l		
AP	<input type="text"/>	U/l		
Bilirubin (total)	<input type="text"/>	mg/dl or	<input type="text"/>	$\mu\text{mol/l}$
Magnesium	<input type="text"/>	mmol/l		
Calcium	<input type="text"/>	mmol/l		
Total protein	<input type="text"/>	g/dl		
Albumin	<input type="text"/>	g/dl		
Blood glucose	<input type="text"/>	mg/dl (fasting)		
HbA1c	<input type="text"/>	%		

Laboratory investigation (urine)

Creatinine	<input type="text"/>	$\mu\text{mol/l}$ or	<input type="text"/>	mg/dl
Uric acid	<input type="text"/>	$\mu\text{mol/l}$		
Magnesium	<input type="text"/>	mmol/l		
Calcium	<input type="text"/>	mmol/l		
Total protein	<input type="text"/>	mg/dl and	<input type="text"/>	g/day
Albumin	<input type="text"/>	mg/dl		

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Extrarenal symptoms

MODY type 5	no <input type="checkbox"/>	yes <input type="checkbox"/>	manifestation _____
Hyperuricaemia	no <input type="checkbox"/>	yes <input type="checkbox"/>	manifestation _____
Elevated liver enzymes	no <input type="checkbox"/>	yes <input type="checkbox"/>	manifestation _____
Exocrine pancreas insufficiency	no <input type="checkbox"/>	yes <input type="checkbox"/>	manifestation _____
Others	_____		