British Association for Paediatric Nephrology

Congenital single functioning kidney (CSFK)

Draft Scope

1 Guidance title

Congenital single functioning kidney (CSFK)

2 The remit

Management strategies for Congenital single functioning kidney vary across the UK. The purpose of national guideline development is to reduce unwarranted variation in practice and facilitate an equitable approach to management.

3 Clinical need for the guideline

Congenital single functioning kidney (CSFK) refers to the anatomical or functional absence of one kidney, present at birth.

CSFK can result from:

- Agenesis: Complete failure of one kidney to develop during embryogenesis.
- Aplasia: Presence of a rudimentary, non-functioning kidney.
- Multicystic Dysplastic Kidney (MCDK): A severely dysplastic kidney replaced by cysts that lose function and can involute before birth.
- Dysplastic non-functioning kidney
- Non-functioning kidney secondary to obstruction; e.g., at the pelvi-ureteric junction

CSFK occurs in about 1 in 2,000 births; unilateral renal agenesis (one kidney absent) is more common than bilateral (both kidneys absent, which is more likely than not to be incompatible with life). Most children, young people and adults with CSFK require only monitoring after birth, due to an increased risk of developing high blood pressure or progression through later stages of chronic kidney disease (CKD). A minority of babies have an abnormality of the solitary kidney. Rarely, this is severe

and babies do not survive the pregnancy or early neonatal period, or may require dialysis in the first few days or weeks of life.

Guideline Content

The guideline will be developed according to UKKA standards for guideline development and in line with NICE and RCPCH standards for guideline development. Where evidence is lacking, a Delphi consensus process may be employed to increase the rigour of development of recommendations.

- This document is the scope. It defines exactly what this guidance will (and will not) examine, and what the guidance developers will consider.
- The areas to be addressed by the guideline are in the following sections.

4.1 Population

3.1.1 Groups that will be covered

Babies, children and young people with CSFK

3.1.2 Groups that will not be covered

- Babies, children and young people with acquired single kidney e.g., due to removal, trauma, infection
- Adults with CSFK

4.2 Healthcare settings and services

Primary and secondary care, in-patient (including neonatal units) and outpatient settings

4.3 Key areas

4.3.1 Clinical issues that will be covered

1 Referral pathways following antenatal detection of CSK

2 Postnatal management of babies, children and young people born with CSFK, including:

2.1.1 Indications for, timing and frequency of postnatal imaging, including: Ultrasound, DMSA, MCUG

2.1.2 Investigation and counselling for extrarenal manifestations, e.g., mullerian anomalies

2.1.3 Indications for nephrectomy
2.1.4 Indications for genetic testing
3 Location and frequency of follow-up
4 Surveillance of:
4.1.1 Blood pressure
4.1.2 Urine albumin: creatinine
4.1.3 Tests of kidney function, including blood tests and urine biomarkers
5 Physical activity restrictions
6 Medication restrictions
7 Dietary recommendations

4.3.2 Clinical issues that will not be covered

Management of complications of CSFK, e.g., hypertension, proteinuria, chronic kidney disease, urinary tract infection

4 Clinical questions

5.1 What is the optimal referral pathway following antenatal detection of CSFK?
5.2 What imaging investigations should be undertaken in babies, children and young people born with CSFK?
5.3 Where, when and for how long should babies, children and young people with CSFK be followed up?
5.4 What is the recommended frequency of surveillance for:
5.4.1 Blood pressure
5.4.2 Urine albumin: creatinine
5.4.3 Tests of kidney function
5.4.3.1 Including blood tests and urine biomarkers
5.5 What are the indications, if any, for genetic testing in CYP with CSFK
5.6 What are the indications, if any, for nephrectomy?
5.7 In children and young people born with CSFK, do dietary modifications (e.g, salt restriction, improve outcomes compared with no dietary modifications?
5.8 In children and young people born with CSFK, does restriction of certain physical activities (e.g., contact sports, cycling) improve outcomes compared with no restriction?
5.9 In children and young people born with CSFK, does restriction of certain medications (e.g., ibuprofen) improve outcomes compared with no restriction?

6 Related guidance

NICE guidance on chronic kidney disease: Overview | Chronic kidney disease: assessment and management | Guidance | NICE

7 Further information

8 References

8.1 La Scola C, Ammenti A, Bertulli C, Bodria M, Brugnara M, Camilla R, Capone V, Casadio L, Chimenz R, Conte ML, Conversano E, Corrado C, Guarino S, Luongo I, Marsciani M, Marzuillo P, Meneghesso D, Pennesi M, Pugliese F, Pusceddu S, Ravaioli E, Taroni F, Vergine G, Peruzzi L, Montini G. Management of the congenital solitary kidney: consensus recommendations of the Italian Society of Pediatric Nephrology. Pediatr Nephrol. 2022 Sep;37(9):2185-2207. doi: 10.1007/s00467-022-05528-y. Epub 2022 Jun 17.

8.2 Solitary or Single-functioning Kidney - NIDDK

8.3 Renal agenesis | infoKID