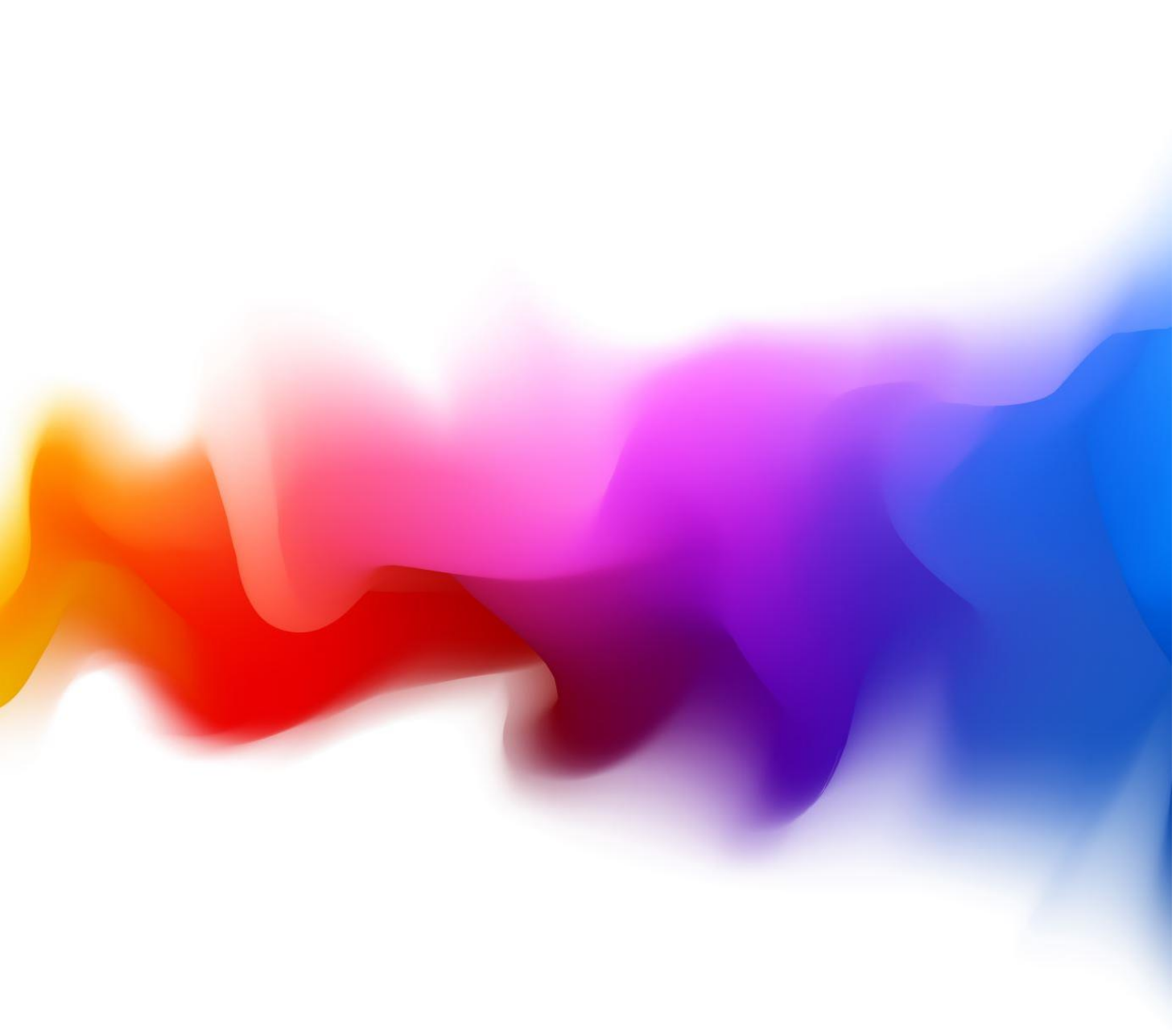

Dietary Management of Nephrotic Syndrome

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Nephrotic Syndrome (NS)

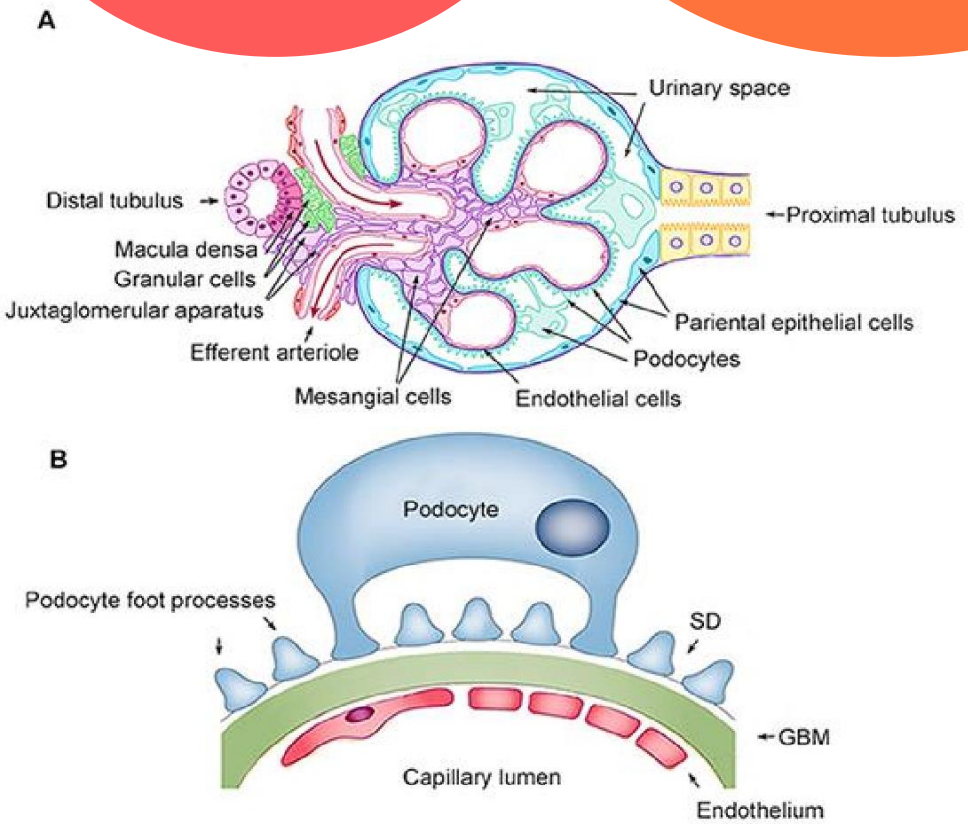
Proteinuria:
urine protein–creatinine ratio ≥ 200 mg/mmol

Hypoalbuminaemia:
<30g/L

Oedema

+/- Hyperlipidaemia

(KDIGO, 2025)



Regulate the amount of water in blood vessels

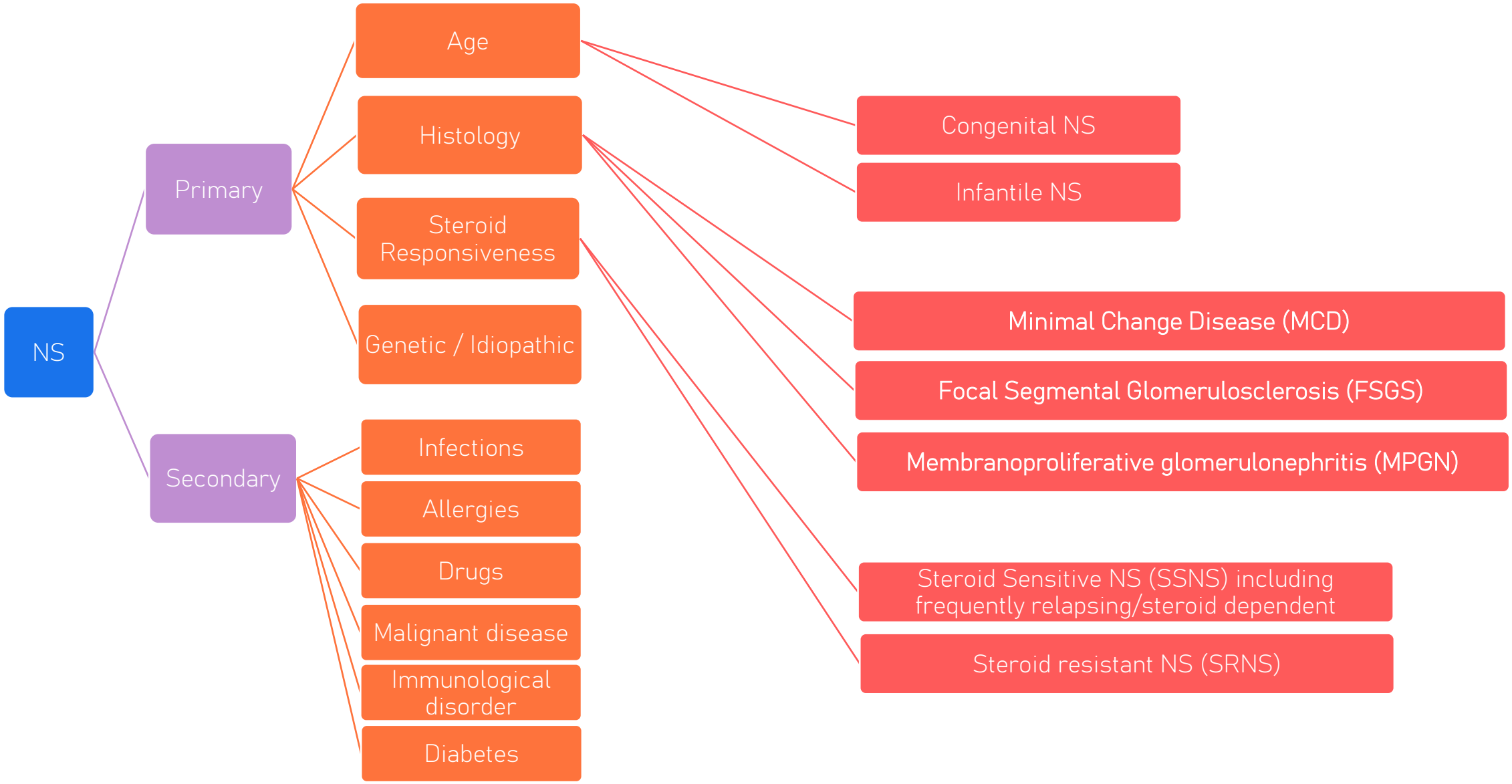
Act as carriers for many substances in the blood, such as hormones and lipids

Prevent blood from clotting

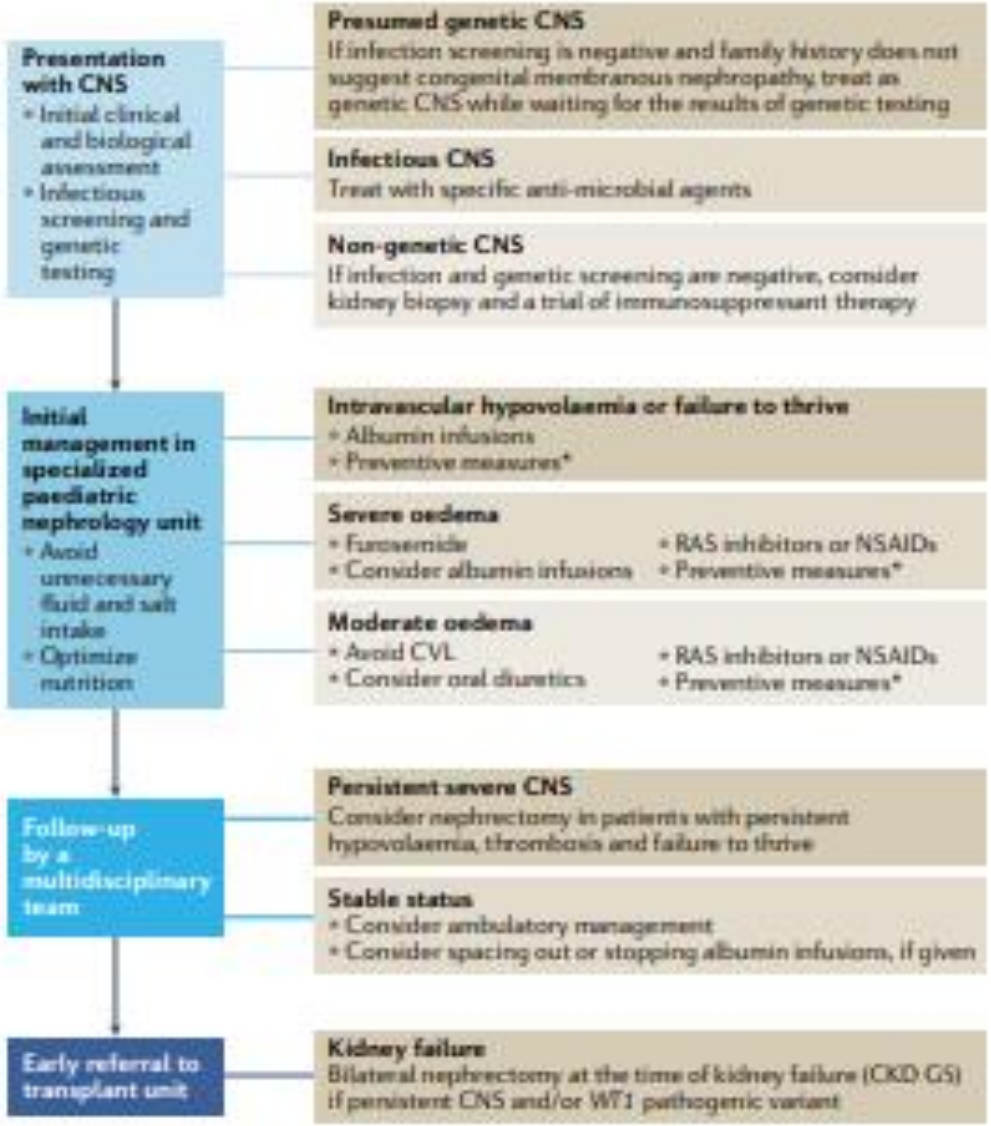
Are important for the growth and development of children

Proteins, in the form of antibiotics, are an important part of the immune system

Classification overview of NS



CNS Medical Management Overview



CONSENSUS STATEMENT

OPEN

Management of congenital nephrotic syndrome: consensus recommendations of the ERKNet-ESPN Working Group

Olivia Boyer[1,2✉], Franz Schaefer[3], Dieter Haffner[4,5], Detlef Bockenhauer[6], Tuula Holtta[7], Sandra Bérody[1], Hazel Webb[6], Marie Heseldenl[8], Beata S. Lipska-Ziętkiewicz[9,10], Fatih Ozaltin[11], Elena Levtchenko[12] and Marina Vivarelli[13]

Fig. 1 | **Opinion-based management algorithm for CNS.** At presentation with congenital nephrotic syndrome (CNS), a clinical and biological assessment including screening for congenital infections and genetic analysis is recommended. Initial treatment should be based on the results of these assessments. Patients should be managed at diagnosis by a specialized paediatric nephrology team. Blood volume should be assessed and symptomatic treatments instituted to maintain blood volume and prevent complications. Follow-up must be managed by a multidisciplinary team. Nephrectomy can be considered for children with persistent, severe CNS despite optimal management. Stable children can be managed on an outpatient basis with spacing or even stopping of albumin infusions. All children should be referred promptly to a kidney transplant team. Bilateral nephrectomy is recommended at the time of kidney failure (chronic kidney disease (CKD) G5) if nephrotic syndrome persists and/or if the patient has a *WT1* pathogenic variant. *Preventive measures: prophylaxis for thrombosis, infection and anaemia, adequate nutrition and growth hormone substitution. RAS, renin–angiotensin system; CVL, central venous line.

NS Medical Management Overview

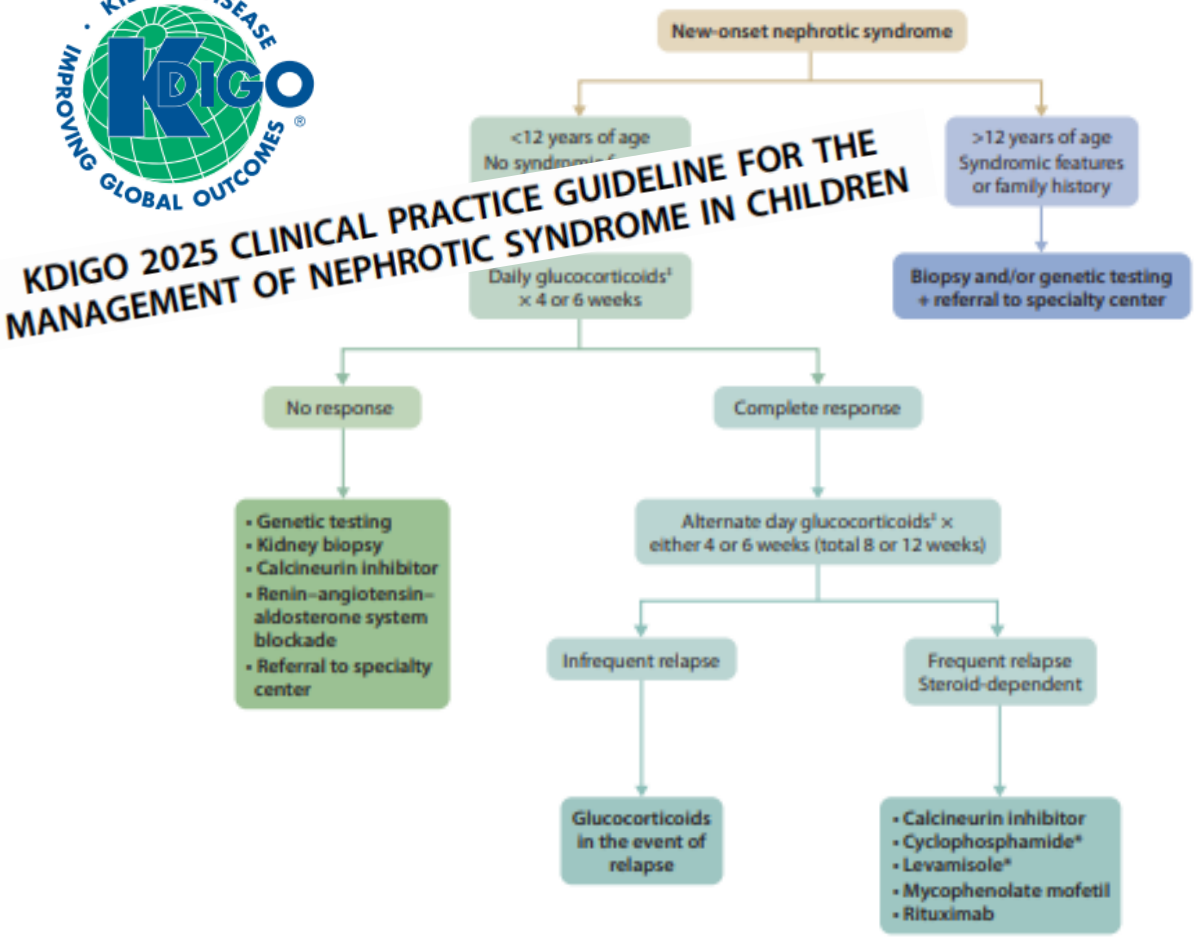
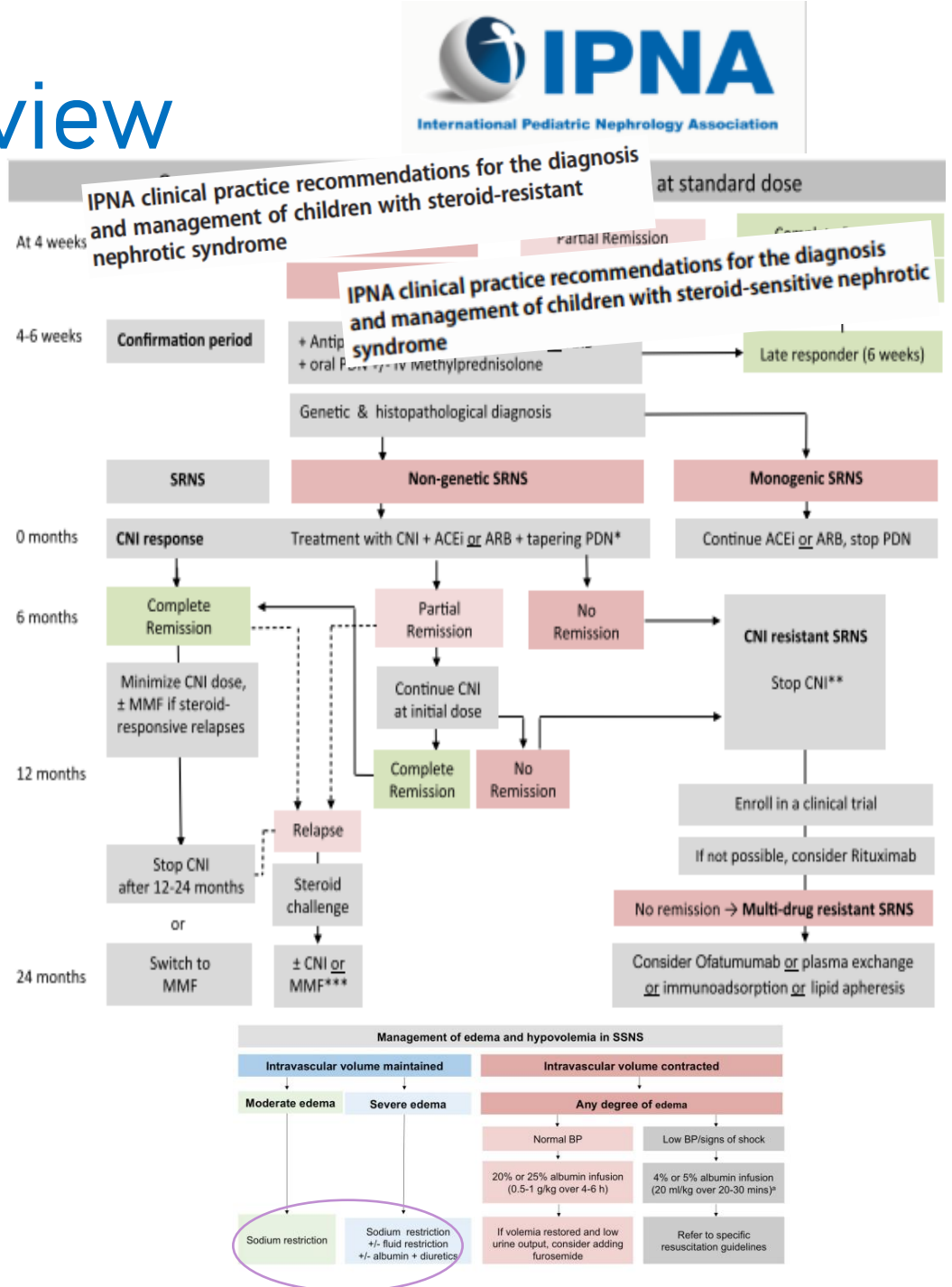


Figure 2 | Treatment algorithm for nephrotic syndrome (NS) from onset. Therapeutic approach to NS in children from onset. Refer to the clinical trial where appropriate. Syndromic features are defined as impaired statural growth; skeletal, neurodevelopmental, and ocular abnormalities; deafness; genital ambiguity; facial dysmorphisms; and so on. Glucocorticoid-sparing agents for children with frequent relapses or steroid-dependent nephrotic syndrome are listed here in an unbiased order. For the management of partial remission, please refer to the 2020 International Pediatric Nephrology Association guideline.[2,3] *May be more indicated for frequent relapses. In patients with frequent relapses without glucocorticoid complications, low-dose, alternate-day oral prednisone/prednisolone may also be considered before introducing a glucocorticoid-sparing agent (see Practice Point 1.3.3.4). [‡]Glucocorticoids: oral prednisone/prednisolone.



Dietary Principles

	IPNA, 2020 SRNS & 2023 SSNS guidelines	ERKNet-ESPN working group, 2021 CNS consensus statement
Sodium	During relapse (moderate-severe oedema): low salt diet - max dose of 2-3 mmol/kg/d *(similar to SACN on Salt and Health 2003)* During remission: normal salt intake	Avoid unnecessary salt intake, advise low salt: <6mo: <0.5g/d 7-12mo: <1g/d >1yrs: <3g/d
Fluid	Routine restriction: not recommended with SSNS or SRNS as at risk for symptomatic hypovolaemia (if intravascular volume depletion) Restriction: hyponatraemia, severe oedema considering volume status and urine output	Fluid restrict in hyponatraemia & severe oedema Avoid unnecessary fluid intake - Fluid prescriptions should primarily be used to provide nutrition - Use concentrated formulas
Energy	adequate energy - child's age or height age	130kcal/kg/d but do not specify age - Concentration of feeds/high energy & protein feeds/ addition of glucose polymers/fat emulsions/protein powders
Protein	normal protein intake	3-4g/kg/d but do not specify age - Addition of protein modules

Dietary Principles

	IPNA, 2020 SRNS & 2023 SSNS guidelines	ERKNet-ESPN working group, 2021 CNS consensus statement
Fat	SSNS: usually resolves with remission of NS SRNS: persistently high LDL >3.4mmol/L suggests lipid lowering treatment starting with lifestyle changes, including dietary modifications, physical activity and weight control	Consider use of statins when fasting LDL cholesterol is persistently >4.1mmol/l or >3.4mmol/l in patients with additional cardiovascular risk factors
Vitamin D & Ca	SSNS: adequate dietary Ca +/- supplementation SDNS/FRNS: assessing 25(OH)D levels annually during remission phase (after 3mo) aiming for >50nmol/L SRNS: hypocalcaemia (iCa/ Albumin corrected Ca)+/- low 25(OH)D levels then supplement	Measure Vit D every 6months + yearly > 12months of age Measure ionized calcium and parathyroid hormone (PTH) levels Supplement with cholecalciferol/calcifediol + Ca (250–500mg/day if low and/or elevated PTH
Micronutrients		Persistent anaemia after 4 weeks of iron and erythropoietin therapy: other factors Cu/Caeruloplasmin/Vit B12

Other factors to consider

- Anthropometry:
  - Dry weight
  - Steroid treatment: impairs growth and increases BMI
- Reduced appetite:
  - Sip feeds may be required
  - Tube feeding
- Malabsorption +/- diarrhoea
  - Hydrolysed protein feeds
- Food allergy:
  - Relapses have been described after exposure to allergens and patients with idiopathic NS can show increased IgE levels
  - Small clinical trials and case reports

Case Study – CNS

6 week

Birth Weight		Current Est. Dry Weight	Length	HC
3 kg	→	3.5 kg	54 cm	37 cm
25th P		2nd P	25th P	50th P

- Daily infusions of albumin start
- Fluid restricted to 120ml/kg, aiming for energy 130kcal/kg/d and protein 3g/kg/d
- Mum is not breastfeeding or expressing and wishes to use SMA Pro 1
- Feeding orally

Feed Recipe	Energy (kcal)	Protein (g)	CHO (g)	Fat (g)
20g SMA Pro 1	102	1.9	11	6
1g Vitajoule	3.8	0	1	0
1g Protifar	3.6	0.9	0	0
Total per 100ml	109	2.8	12	6
Total per 420ml (120ml/kg)	458	11.8	50	25
Total per kg	**131**	**3.4**	**14**	**7**

Case Study – CNS

Birth Weight
3 kg
25th P

→

Current Est. Dry Weight
3.6 kg

Requirements:
130kcal/kg/d
3g prt/kg/d

- More oedematous + frequent vomiting
- Fluid restricted further to 110ml/kg
- Oral feeding reduces -> NG inserted, and feeds given continuously
- Vomiting reduces but continues -> feed type changed

Feed Recipe	Energy (kcal)	Protein (g)	CHO (g)	Fat (g)
100ml Infatrini	100	2.6	10.3	5.4
5g Vitajoule	19	0	4.8	0
Total per 100ml	119	2.6	15.1	5.4
Total per 400ml (110ml/kg)	476	10.4	60.4	21.6
Total per kg	**132**	**2.9**	**16.8**	**6**

Learning points

Advise a reduced salt ('no added salt') diet when the child is in an oedematous state

CNS leads to protein losses, increased infections, diarrhoea & vomiting

CNS babies will need high energy and protein feeds, fluid restriction, likely tube feeds

If resistant to steroids – reduced appetite, diarrhoea & malabsorption. Sip/tube feeds may be needed

Monitoring of trace elements and 25(OH)D in steroid resistant NS & CNS