GLOSSARY

OSTEOBLAST
Cell that arises from fibroblasts and which, as it matures, is associated with the production of bone

OSTEOCLAST
Large multinuclear cell associated with the absorption and removal of bone

METAPHYSES
Transitional zones at which a shaft of a long bone meets the part of bone where growth occurs

JOUBERT SYNDROME
A syndrome in which the cerebellar vermis fails to develop; characterized by tachypnea, irregular muscular action and mental retardation

experienced a significant decline in renal function. Thirteen patients experienced infusion-related reactions, but these did not compromise study compliance.

In another study, Linthorst *et al*.—who followed 30 patients during self-infusion of a median of 44 doses (0.2–1.0 mg/kg) of recombinant human α-Gal A over more than 3 years—also found a high rate of compliance. Only two patients (both male) experienced minor infusion-related adverse events during home-based treatment.

Together, the results from these new studies support the feasibility and safety of home-based enzyme replacement therapy for Fabry disease, which has the potential to reduce the burden on healthcare providers.

Kate Matthews

Original articles Schiffmann R *et al.* (2006) Long-term therapy with agalsidase alfa for Fabry disease: safety and effects on renal function in a home infusion setting. *Nephrol Dial Transplant* **21:** 345–354
Linthorst GE *et al.* (2006) Home treatment for Fabry disease: practice guidelines based on 3 years experience in The Netherlands. *Nephrol Dial Transplant* **21:** 355–360

Pamidronate for steroid-induced osteoporosis in pediatric nephropathy patients

Corticosteroids, the first-line therapy for nephrotic syndrome in children, can cause adverse osteoporotic effects. Pamidronate, a bisphosphonate, can be used to treat such complications. A single-center prospective study in Korea has investigated whether this drug is safe and effective in children.

Following a short, intensive course of the steroid methylprednisolone, 44 children with nephropathy (including Henoch–Schönlein purpura nephritis, membranous glomerulonephritis and IgA nephropathy) were randomized 1:1 to receive oral calcium and 125 mg pamidronate or oral calcium alone. After 3 months, mean concentrations of urine pyridinoline, serum parathyroid hormone and serum osteocalcin, which are all well-known markers of OSTEOBLAST and OSTEOCLAST function, were similar in both groups. In control patients, bone mineral density—measured in the vertebrae of the lumbar spine—reduced significantly over 3 months, from 0.654 ± 0.069 g/cm^2 to 0.631 ± 0.070 g/cm^2 (P <0.05). In the pamidronate group, there was no significant change in bone mineral density, but five patients who had received pamidronate for longer than 5 months developed sclerotic lines at METAPHYSES. Pamidronate therapy was also associated with mild abdominal discomfort.

Preventing and treating bone loss in children with nephropathy would have a positive effect on bone strength and quality of life in adulthood. Bisphosphonates inhibit osteoclast activity and bone resorption, but the sclerotic patterns observed in the present study indicate that these drugs might also perturb skeletal growth. Pamidronate is a promising treatment for steroid-induced osteoporosis but further trials are necessary to assess its safety in the pediatric setting.

Rachael Williams

Original article Kim S-D and Cho B-S (2006) Pamidronate therapy for preventing steroid-induced osteoporosis in children with nephropathy. *Nephron Clin Pract* **102:** c81–c87

Excellent kidney transplant outcomes for children with mental retardation

It has been suggested that pediatric kidney transplant recipients with mental retardation might be at greater risk of post-transplant infectious and malignant complications than children without mental retardation. In addition, many patients with mental retardation have a decreased life expectancy, which has caused concern about the validity of kidney transplantation in this population. Authors of a prospective, multicenter investigation in Japan, however, conclude that mental retardation should not preclude kidney transplantation in children.

The study included 25 pediatric renal transplant recipients with mental retardation, five of whom had severe mental retardation and immobility. Causes of mental retardation included acquired diseases and genetic syndromes such as JOUBERT SYNDROME. All patients had end-stage renal disease, secondary to congenital urinary tract abnormality, focal segmental glomerulosclerosis or other etiologies. Comorbidities included central nervous system and cardiovascular diseases, such as epilepsy and hypertrophic cardiomyopathy.

After a median follow-up of 20 months, all patients survived, had good graft function (mean serum creatinine 0.60 ± 0.29 mg/dl [53.0 ± 25.6 μmol/l]), were compliant with immunosuppressive therapy and showed improved quality of life. Incidences of acute rejection and

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