

by the microinsertion c.33–34insC, Phe152Ile, Gly156Arg then Ile244Thr. Twenty-eight novel variants were detected. None of the 12 new AGXT missense mutations was detected in a population of 50 normal controls.

The whole-gene sequencing method yielded 98% sensitivity when applied to the 48 probands with liver-biopsy-proven PH1. Diagnostic sensitivity was 77% when only the three exons with the highest mutation frequencies were sequenced, still higher than the 62% sensitivity of a previously published partial-gene screen specific for the Gly170Arg, c.33–34insC and Ile244Thr mutations.

These data indicate even greater variety in the AGXT coding region of PH1 patients than was previously thought. As AGXT is a relatively small gene, the authors believe that whole-gene sequencing is a financially viable technique for molecular diagnosis of PH1 that could obviate the need for biopsy in many patients.

Original article Monico CG *et al.* (2007) Comprehensive mutation screening in 55 probands with type 1 primary hyperoxaluria shows feasibility of a gene-based diagnosis. *J Am Soc Nephrol* **18**: 1905–1914

ACE inhibitors do not delay progression of hypodysplastic nephropathy in children

Angiotensin-converting-enzyme (ACE) inhibitors have been shown to have a renoprotective effect in adults with chronic renal insufficiency (CRI). There is a lack of evidence for similar effects in pediatric patients; nevertheless, ACE inhibitors are commonly used as antiprogession agents in these individuals, despite the very different etiologies, clinical courses and outcomes of CRI in adults and children. Most notably, children with chronic kidney diseases often do not have hypertension or proteinuria.

After analyzing data from the Italian Pediatric Registry of Chronic Renal Failure (ItaKid Project), Ardissino and co-workers have reported no apparent benefits of ACE inhibitors for delaying progression of renal disease in children with hypodysplastic nephropathy, the most common cause of pediatric CRI. In comparison with 123 controls matched by age, gender, diagnosis and baseline creatinine clearance, the rate of progression of CRI (measured as the slope of creatinine clearance over time) was not significantly reduced in 41

children treated with ACE inhibitors. There was, however, a trend towards slower progression in the treatment group (slope -1.08 ± 2.08 vs -1.80 ± 4.42 ml/min/1.73 m²/year; $P=0.31$). No significant differences in creatinine clearance slopes were observed when participants were grouped by initial severity of renal impairment, suggesting that selection bias did not influence the results of the analysis.

The authors conclude that use of ACE inhibitors as antiprogession agents should continue to be regarded as experimental in children with hypodysplastic chronic kidney diseases.

Original article Ardissino G *et al.* (2007) No clear evidence of ACEi efficacy on the progression of chronic kidney disease in children with hypodysplastic nephropathy—report from the ItaKid Project database. *Nephrol Dial Transplant* [doi: 10.1093/ndt/gfm237]

Polyflux® 6H dialyzer: a new option for small children requiring dialysis

Hemodialysis is the only option in children who require renal replacement therapy but are not candidates for renal transplantation or peritoneal dialysis. During hemodialysis, the extracorporeal circuit should not contain more than 10% of the patient's blood volume. Until recently, dialyzers meeting this specification for children weighing <15 kg were only available from one company in the US (Fresenius Medical Care), but FDA approval has now been obtained for another dialyzer suitable for small children, the Polyflux® 6H dialyzer (Gambro AB, Lund, Sweden).

Goldstein *et al.* evaluated use of this new high-flux dialyzer in six children (mean \pm SD age 10.3 ± 3.8 years, mean weight 24.4 ± 6.5 kg) who had been receiving maintenance hemodialysis with the Fresenius F3 ($n=2$), F4 ($n=2$), or F5 ($n=2$) dialyzer for at least 2 months. All patients switched to maintenance hemodialysis with the Polyflux® 6H dialyzer for 2 consecutive months.

No adverse events occurred during Polyflux® 6H dialyzer use. Outcome variables (serum sodium, potassium and CO₂ levels, post-dialysis weight, total ultrafiltration, single-pool Kt/V and estimated equilibrated Kt/V) did not differ significantly between the pre-evaluation period (with Fresenius dialyzers) and the period of treatment with the Polyflux® 6H dialyzer. There