The device was used within an individualised care plan to assess acute or routine review with a member of the team from home.

3. Feedback on the usability, workflow and key outcomes was gathered at various stages of the project:

a. A feedback survey completed by the healthcare professional after each consult.

b. Data was collected via the TytoCare system for each consultation.

c. End of pilot surveys were completed by staff and families.

Results 48 consultations were undertaken using TytoCare during the pilot. We had healthcare professional feedback for 46 of them reporting the following impact: 100% of consultations felt to provide reassurance to families, 98% had a positive impact on the CYP. Two hospital assessments, 3 inpatient admissions, 13 face to face clinic appointments, 4 home visits, 23 face to face physiotherapy reviews and approximately 329 miles travelled were saved.

Conclusions In this pilot the TytoCare device was found to be easy to use by professionals and carers and to be reliable and effective in providing safe and quality care for a select group of CYP at home. The pilot highlighted the impact technology can have in reducing the burden of chronic illness for families. It also demonstrated that technology could be used successfully to improve access to care for some of our most vulnerable families.

British Association for Paediatric Nephrology

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ALPORT SYNDROME IN CHILDREN: A 15-YEAR SINGLE CENTRE EXPERIENCE

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Background Alport syndrome (AS) accounts for 0.2%-0.6% adult and 2-3% paediatric patients with end stage renal disease (ESRD), with increased risk of chronic kidney disease in those with heterozygous mutations. Angiotensin-converting enzyme inhibition (ACEi), the current first line therapy, can delay the onset of ESRD. Clinical trials of newer therapies eg, bardoxolone methyl and anti-microRNA-21 are underway. Recent classifications adopt an inclusive genetic-based approach to help early diagnosis, monitoring and treatment.

Objectives The objectives of this study are to review the characteristics in our cohort of patients with AS, and to compare patient characteristics between the different genetic-based classifications.

Methods A retrospective review of all patients with pathogenic COL4A3, COL4A4 and COL4A5 variants followed up at Great Ormond Street Hospital between 2005 and 2020 was undertaken. Patients were classified as: (i) males with X-linked AS (XLAS); (ii) autosomal recessive AS (ARAS); (iii) autosomal dominant AS (ADAS); and (iv) females with XLAS.

Demographics, genetics, presenting symptoms, length of follow up, clinical parameters at the last follow up were collected.

Results Twenty-eight children with AS were included. The median age of presentation was 5.4 years (IQR 4.5-11.1) with a median follow up of 3.0 years (IQR 1.3-7.3). Median age at latest follow-up was 11.8 years (IQR 8.1-14.6). The median urine albumin-creatinine ratio at latest follow-up was 38.6mg/mmol (IQR 3.3-127.4), with median serum albumin of 41.5g/L (IQR 38.0-44.0) and median eGFR of 115.4ml/min/1.73m² (IQR 99.2-138.3). The most common presenting symptom was isolated microscopic haematuria (39%).

Eight patients transitioned to adults: 3 had male XLAS, 4 had female XLAS and 1 had ADAS. The median age of these patients at last follow-up was 15.9 years (IQR 14.6-16.4), with a median follow up time of 4.8 years (IQR 3.0-9.1). The median eGFR at last follow-up was 100.8 ml/min/1.73 m (IQR 95.6-107.5), median serum albumin was 46.0g/L (IQR 39.0-42.0), median urine albumin creatinine ratio was 45 mg/mmol (IQR 5.3-201.0). One patient with male XLAS progressed to ESRD at the age of 14.

Conclusions Microalbuminuria was the earliest event observed in the progression of AS in this cohort of patients, most significant in male X-linked and autosomal-recessive AS patients. Majority of children had normal serum albumin and eGFR during the study period but we have identified 2 patients for consideration of enrolment in clinical trials.

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Abstract 1397 Table 1 Comparison between the different groups of patients. Data presented as median (interquartile)

Males with XLAS	n=8 ARAS	n=7 ADAS	n=5 Females with XLAS	n=8	p-value
Age of presentation (years)	5.8 (4.6-10.5)	4.7 (3.4-9.2)	7.5 (4.9-12.8)	5.4 (4.1-11.8)	0.69
Age of genetic confirmation (years)	12.7 (6.2-13.6)	5.1 (3.9-8.8)	8.5 (5.8-13.2)	11.8 (5.0-13.8)	0.22
Length of follow up (years)	5.4 (2.0-9.1)	1.4 (0.5-4.9)	2.6 (1.4-3.0)	4.8 (1.6-8.5)	0.23
Age at latest follow up (years)	14.1 (9.3-15.0)	10.2 (5.3-11.9)	10.5 (6.7-15.1)	14.2 (7.7-16.1)	0.33
Urine albumin-creatinine ratio at latest follow up (mg/mmol)	40.0 (1.6-172.5)	135.8 (65.5-421.0)	1.2 (0.8-21.7)	9.5 (6.0-301.0)	0.03
Serum albumin at last follow up	39.0 (38.0-44.0)	38.0 (31.0-43.0)	45.0 (43.3-46.8)	41.5 (37.8-46.0)	0.07
eGFR at last follow up (ml/min/1.73 m ²)	131.6 (98.8-152.5)	104.8 (97.4-135.1)	142.6 (97.6-168.4)	106.0 (100.5-132.7)	0.58