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Syndromic hearing loss with extrarenal symptom is common in childhood-onset chronic kidney disease.

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Objectives: The study evaluated the prevalence and underlying diseases of hearing loss (HL) in patients with childhood-onset chronic kidney disease (CKD).

Methods: In this retrospective study in single center, pediatric patients with CKD stage 2-5 were recruited. Audiometric examinations were performed using pure tone audiometry (PTA) or automated brainstem response test (ABRT). Genetic or syndromic disease in addition to HL and CKD was defined as syndromic HL.

Results: A total of 421 patients (279 males, 142 females) were recruited. Audiometric examinations were performed in 178. The mean onset age of renal symptom was 4.1 ± 4.8 years and the mean follow-up period was 9.4 ± 6.2 years. The patients were grouped according to the causes of CKD: congenital anomalies of the kidney and urinary tract (CAKUT, n=184), glomerulopathy (GP, n=105), cystic kidney diseases (CYST, n=39), perinatal problem (PP, n=29), and others (n=64). HL was detected in 82, including 51 with sensorineural hearing loss (SNHL), 30 with conductive hearing loss (CHL), and 1 with mixed HL.

The prevalence rates of HL were as follows: 16.8% (SNHL:CHL:mixed HL 7.6:8.7: 0.5) in the CAKUT, 28.6% (22.9:5.7:0) in the GP, 12.8% (5.1:7.7:0) in the CYST, 24.1% (20.7:3.4:0) in the PP, and 14.1% (7.8:6.3:0) in the others group. The prevalence of HL increased as the CKD stage increased ($P=0.002$), and the prevalence of HL and SNHL were higher in GP ($P=0.007$ and 0.021 , respectively). Of a total 82 with HL, 41 had syndromic HL (SNHL:CHL:mixed HL 30:10:1). Among 51 with SNHL, 30 patients were syndromic HL. Otitis media with effusion was present in 27 of 30 patients with CHL.

Conclusions: The overall prevalence of HL were 19.5%, including 12.1% of SNHL and 7.1% of CHL. SNHL was a manifestation of genetic or syndromic disease in more than half, while CHL was associated with otitis media with effusion mostly.

Table 1

Table 1. Baseline characteristics of the patients (n = 421)

Items	
Gender (male:female)	279:142
Mean age	15.0 ± 6.4 years
Median onset age of renal symptom	1.7 years (0-16.7 years)
Mean follow-up duration	9.4 ± 6.2 years
Stage of CKD at the last follow-up	
Stage 2	68 (16.2%)
Stage 3	82 (19.5%)
Stage 4	30 (7.1%)
ESRD	241 (57.2%)
Mean age at ESRD	9.2 ± 5.9 years
Causes of CKD	
CAKUT	184 (43.7%)
Glomerulopathy	105 (24.9%)
Hereditary glomerular diseases	57 (13.5%)
Cystic kidney disease	39 (9.3%)
Perinatal problem	29 (6.9%)
Others	64 (15.2%)
Hearing loss	82 (19.5%)
SNHL	51 (12.1%)
CHL	30 (7.1%)
Mixed	1 (0.2%)
Otitis media with effusion	43 (10.2%)
VTI	21 (5.0%)

CKD, chronic kidney disease; ESRD, end-stage renal disease, CAKUT, congenital anomalies of the kidney and urinary tract; SNHL, sensorineural hearing loss; CHL, conductive hearing loss; VTI, ventilation tube insertion.

Table 2

Table 2. Prevalence of hearing loss

Items	Patients with SNHL	Patients with CHL	Patients with Mixed HL	Total patients with HL	Total patients
No. of patients (M:F)	51 (32:19)	30 (19:11)	1 (0:1)	82 (51:31)	421 (279:142)
CKD stage					
Stage 2	5/68 (7.4%)	3/68 (4.4%)	-	8/68 (11.8%)	68
Stage 3	9/82 (11.0%)	1/82 (1.2%)	-	10/82 (12.2%)	82
Stage 4	3/30 (10.0%)	1/30 (3.3%)	-	4/30 (13.3%)	30
ESRD	34/241 (14.1%)	25/241 (10.4%)	1/241 (0.4%)	60/241 (24.9%)	241
Disease group					
CAKUT	14/184 (7.6%)	16/184 (8.7%)	1/184 (0.5%)	31/184 (16.8%)	184
Glomerulopathy	24/105 (22.9%)	6/105 (5.7%)	-	30/105 (28.6%)	105
Cystic kidney disease	2/39 (5.1%)	3/39 (7.7%)	-	5/39 (12.8%)	39
Perinatal problem	6/29 (20.7%)	1/29 (3.4%)	-	7/29 (24.1%)	29
Others	5/64 (7.8%)	4/64 (6.3%)	-	9/64 (14.1%)	64

CKD, chronic kidney disease; ESRD, end-stage renal disease, CAKUT, congenital anomalies of the kidney and urinary tract; SNHL, sensorineural hearing loss; CHL, conductive hearing loss; HL, hearing loss.