

**Abstract Submission No.: A-1322****Polycystic Kidney Disease of infants: too large to grow together****Seokyoung Chung**, Hee Gyung Kang, Yo Han Ahn

Department of Pediatrics-Nephrology, Seoul National University Hospital, Korea, Republic of

**Case Study :** Autosomal recessive polycystic kidney disease (ARPKD) is often fatal in newborns due to pulmonary hypoplasia caused by oligohydramnios. If an infant with PKD survived the neonatal period, sometimes the PKD is so large that it impedes the patient's growth or proper management. Here, we present two such cases. A 9-month-old girl presented to our center for PKD management. An ultrasound scan revealed markedly enlarged kidneys with numerous cysts and hepatic fibrosis. At 13 months, she lost her kidney function. The intra-abdominal space was filled with PKD (kidney volumes 315cm<sup>3</sup> & 344cm<sup>3</sup>, right kidney length 12.3cm [height 74.6cm]). To ensure sufficient space for peritoneal dialysis (PD), a left unilateral nephrectomy (345gm [weight 10.6kg]) was performed at the time of PD catheter insertion. Ten months later, she received deceased donor kidney transplantation. At 41 months, her right kidney was still 11.5 cm long, and she was growing well, with a height of 97cm (25-50p) and a weight of 17.7kg (95-97p). The second case was hospitalized for respiratory support at 7 months age. The baby was prenatally diagnosed with PKD and had recurrent respiratory infection, the last of which was COVID19. He was 69cm (25p) tall on admission and weighed 11.1kg with ascites (8.3kg (25p) on discharge). Imaging revealed enlarged, polycystic kidneys (approximate length 18cm (left) & 15cm (right)) with mass effect on the intra-abdominal organs. Oral intake was insufficient due to respiratory difficulty and abdominal distention with poor growth (1 cm and 0.3 kg over three months). He is actively managed with medication, and early nephrectomy and PD are being considered for better growth. Although the size of infantile PKD is generally smaller than that of ADPKD, it can still have a deleterious effect. We need to balance between the risk of nephrectomy and the benefit of volume reduction.