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International Journal of Clinical Nephrology

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Review Article

The Rare Association of Adult Polycystic Kidney with Hearing Impairment and Further Experience with Intestinal Dialysis: An Educational Article

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Received Date: November 07, 2023; **Accepted Date:** November 17, 2023; **Published Date:** December 04, 2023

Citation: Aamir Jalal Al-Mosawi (2023), The Rare Association of Adult Polycystic Kidney with Hearing Impairment and Further Experience with Intestinal Dialysis: An Educational Article, *International Journal of Clinical Nephrology*. 5(5); **DOI:10.31579/2834-5142/071**

Abstract

Background: Renal failure remains a significant healthcare problem in Iraq and other countries in the developing world. Renal failure accounted for 4.1 % of the deaths registered by the Iraqi Ministry of health, and it accounted for 4.9 % of deaths in males during the year 2020.The availability of renal replacements therapy and its quality are much less in Iraq and many other developing countries than in the advanced countries. Many patients with chronic renal failure in Iraq has been reported to be reluctant to accept dialysis therapies because of the wide spread notion of its association with high mortality. Therefore, the need for a more robust and more convenient therapy for chronic renal failure has been increasingly emphasized during the previous two decades.

The association of adult polycystic kidneys with sensorineural deafness has been very rarely been reported.

Patients and methods: The case of a 52-year male patient who had progressive hearing impairment over the few years and developed progressive symptomatic uremia despite low protein diet is described. He was advised to perform dialysis by more than one physician, but he and his family continued to refuse treatment by dialysis even when he was admitted to the emergency room on the 21st of October because of vomiting and dehydration.

Results: On the 14th of October, 2023, the patient had symptomatic uremia (Serum creatinine: 9.2 mg, blood urea: 289 mg/dL) and was dehydrated. anemic (Hb: 7g/dL), and hypocalcemic (Serum calcium: 5.5 mg/dL). Abdominal ultrasound showed polycystic kidney disease. The patient was admitted to the emergency room and received intravenous fluids, and started treatment with intestinal dialysis. On the 28st of October, 2023, the patient experienced significant symptomatic improvement which occurred in association with lowering of blood urea, and elevation of hemoglobin and calcium levels. Hemoglobin was 9.3 g/dL (PCV: 22%), serum calcium was 8.2 mg/dL (Normal: 9-11 mg/dL), and blood urea was 104 mg/dL.

Conclusion: The association of autosomal dominant polycystic kidneys with sensorineural deafness has been very rarely reported. Mora et al from Spain were most probably the first to report the association of autosomal dominant polycystic kidneys with bilateral sensorineural deafness. The paper of Mayssa Abdelwahed and her research group was most probably the second paper reporting the association of autosomal dominant polycystic kidney disease with deafness, and this paper is most probably the third paper reporting this association.
This paper also further emphasized the clinical therapeutic values and effectiveness of intestinal dialysis in lowering blood urea level and improving symptoms of symptomatic uremia associate with chronic renal failure.

Keywords: adult polycystic kidney disease; hearing impairment; intestinal dialysis

Introduction

Renal failure remains a significant healthcare problem in Iraq and other countries in the developing world. Renal failure accounted for 4.1 % of the deaths registered by the Iraqi Ministry of health, and it accounted for 4.9 % of deaths in males during the year 2020 [1,2].

The availability of renal replacement therapy and its quality are much less in Iraq and many other developing countries than in the advanced countries. Many patients with chronic renal failure in Iraq has been reported to be reluctant to accept dialysis therapies because of the wide spread notion of its association with high mortality [3-8].

Therefore, the need for a more robust and more convenient therapy for chronic renal failure has been increasingly emphasized during the previous two decades. In a series of eighty patients with chronic renal failure. Fourteen (16.5%) patients were treated with intestinal dialysis, and treatment was associated with amelioration of the uremic symptoms and lowering of blood urea levels and delaying the need for dialysis. In these 80 patients, the longest survival of 5 years was achieved in 2 patients, both treated initially with intermittent peritoneal dialysis. One of them was transplanted and the other patient was treated with intestinal dialysis [9, 10, 11, 12].

A previous study suggested that polycystic disease of the kidneys is a relatively uncommon cause of chronic renal failure in Iraq as in 80 patients with chronic renal failure, there was only one patient had polycystic disease of the kidneys [13].

The association of autosomal dominant polycystic kidneys with sensorineural deafness has been very rarely been reported [14, 15].

Patients and methods

The case of a 52-year male patient who had progressive hearing impairment over the few years and developed progressive symptomatic uremia despite low protein diet is described. He was advised to perform dialysis by more than one physician, but he and his family continued to refuse treatment by dialysis even when he was admitted to the emergency room on the 21st of October because of vomiting and dehydration.

Results

On the 14th of October, 2023, the patient had symptomatic uremia with weakness, pruritus, nausea, and vomiting. His urine output was good, but he was dehydrated and his blood pressure was 105/70 mm Hg. He also has muscle spasm. He was anemic and his hemoglobin level was 7g/dL Serum creatinine was 9.2 mg, blood urea was 289mg/dL, and serum sodium was 104 mmol/L (Normal: 136-149 mmol/L), serum calcium was 5,5 mg/dL (Normal:9-11 mg/dL) .and serum potassium was 4.2 mmol/L (Normal: 3.8-5 mmol/L).

Abdominal ultrasound **(Figure-1A)** showed bilaterally enlarged kidneys (15x8 cm), and both kidneys were heavily occupied by different size cysts. The largest cyst was 3.5 cm in diameter. There was no mass or stone, and the pelvi-calyceal system, and ureters were not dilated. The urinary bladder, liver, and spleen were normal, but the gall bladder showed several stones, each 7mm in diameter. The gall bladder was thin and there was no sign of cholecystitis.

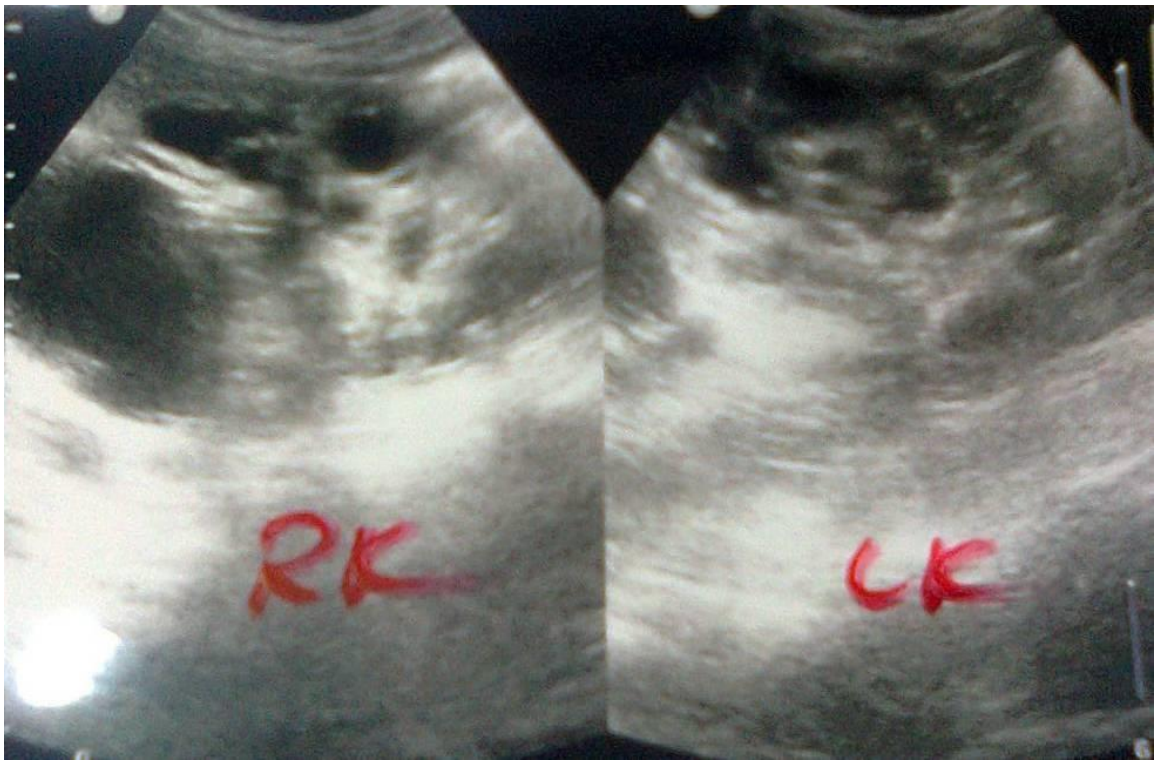

Figure-1A: Abdominal ultrasound showed polycystic kidney disease

The patient has a sister who died before the onset of chronic renal failure and also had hearing impairment.

The patient was admitted to the emergency room and received intravenous fluids.

The patient was treated based on the published guidelines of the conservative dietary and pharmacological management of chronic renal failure and intestinal dialysis with acacia gum [16-24].

He received:

Intramuscular pyridoxine 100 mg daily for 7 days.
Intramuscular Vitamine B complex daily for 7 days.

Intramuscular Iron dextran 100 mg daily for 7 days.

Subcutaneous erythropoietin 4000 units three doses over one week.

Oral alphacalcidol 2 mcg daily, to be continued with dose adjustment according to the serum calcium level.

Calcium carbonate 1500 mg daily, to be continued indefinitely.

He also received acacia gum powder 25g dissolved in 250ml Diet 7Up and given three times daily.

It was necessary to give him oral domperidone (Motilium) 10 mg three times daily to prevent and reduce the occurrence of abdominal distention and discomfort of the intake.

However, it was necessary during the first week of treatment to eliminate almost all dietary protein, and his diet was consisting mainly of high calorie juices, grapes, and water melon. Therefore, nutritional support with oral royal jelly capsules was added to prevent any unexpected nutritional deficiency.

On the 21st of October, 2023, the patient experienced significant symptomatic improvement in weakness, appetite, pruritus and general well-being.

Treatment was also associated in lowering of serum creatinine and blood urea, and elevation of hemoglobin level.

Serum creatinine was 6.2 mg, and blood urea was 116 mg/dL. Hemoglobin was 9g/dL (PCV: 27%).

Screening of other family members with renal ultrasound was advised. A daughter of one of the patient's sister who was not known to have polycystic kidney disease was also found to have polycystic kidney disease. She was in her thirties and her ultrasound showed bilaterally enlarged kidneys (15x8 cm), and both kidneys showed multiple cortical cysts of different sizes. The largest cyst in the right kidney was 3.1x3.3 cm, and largest cyst in the right kidney was 4.3x3.4 cm. Some cysts contained septa with no vascularity on color Doppler **(Figure-1B, C).**

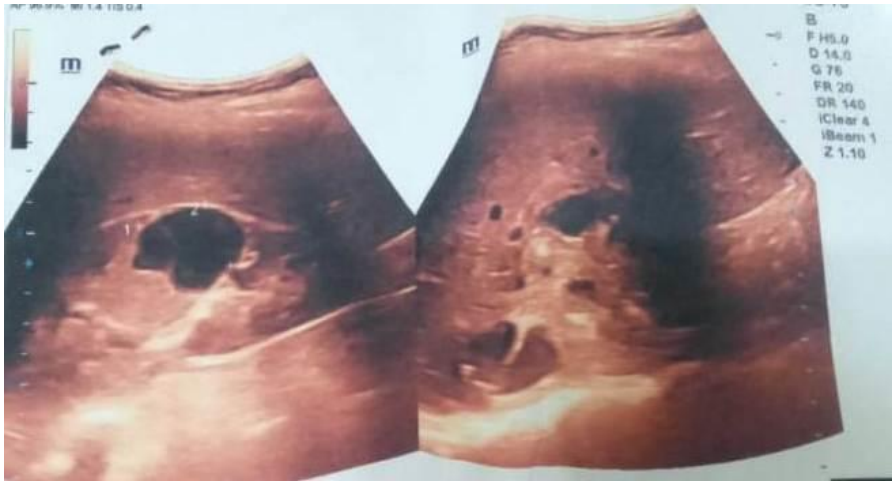

Figure-1B: The patient's nephew's ultrasound

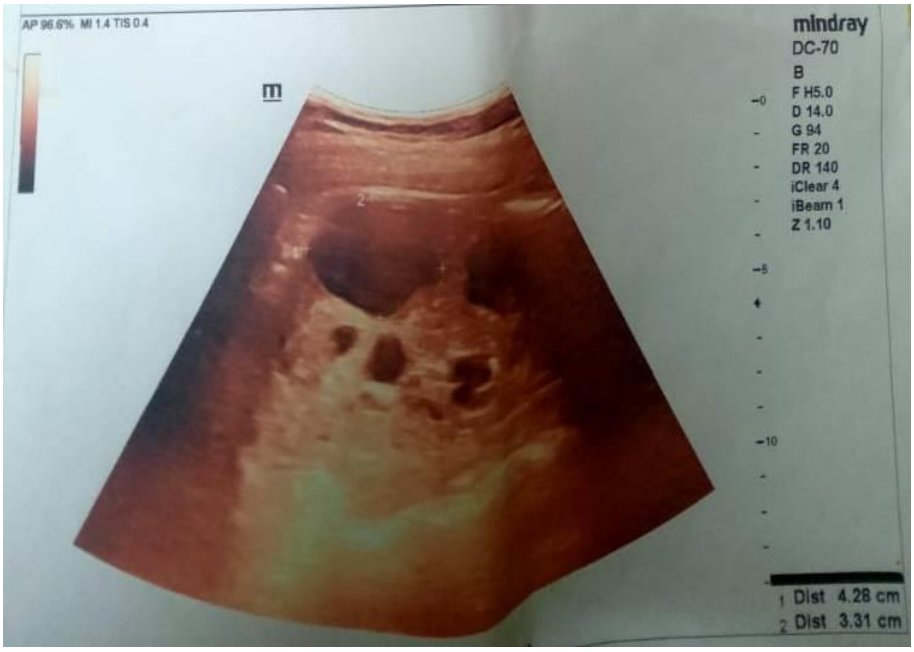

Figure-1C: The patient's nephew's ultrasound

Thereafter, the dietary therapy was given according to the latest published guideline of intestinal dialysis [21, 24]. Oral ferrous sulfate 200 mg three times daily was given instead of intramuscular iron.

On the 28st of October, 2023, the patient was much better (Figure-1D), and other than pruritus, the patient didn't have any other symptoms. Serum creatinine was 6.2 mg, and blood urea was 116 mg/dL. Hemoglobin was 9g/dL (PCV: 27%). Hemoglobin was 9.3 g/dL (PCV: 22%), serum calcium was 8.2 mg/dL (Normal: 9-11 mg/dL), and blood urea was 104 mg/dL.

The family asked if it is possible to return to the very low protein diet to ensure further and rapid lowering of blood urea, and their suggestion was welcomed and the patient was encouraged to return to the very low protein

diet (A diet consisting mainly of high calorie juices, grapes, and water melon).

Oral ferrous sulfate was stopped temporarily and intramuscular iron dextran 100 mg every other day (10 doses was prescribed}.

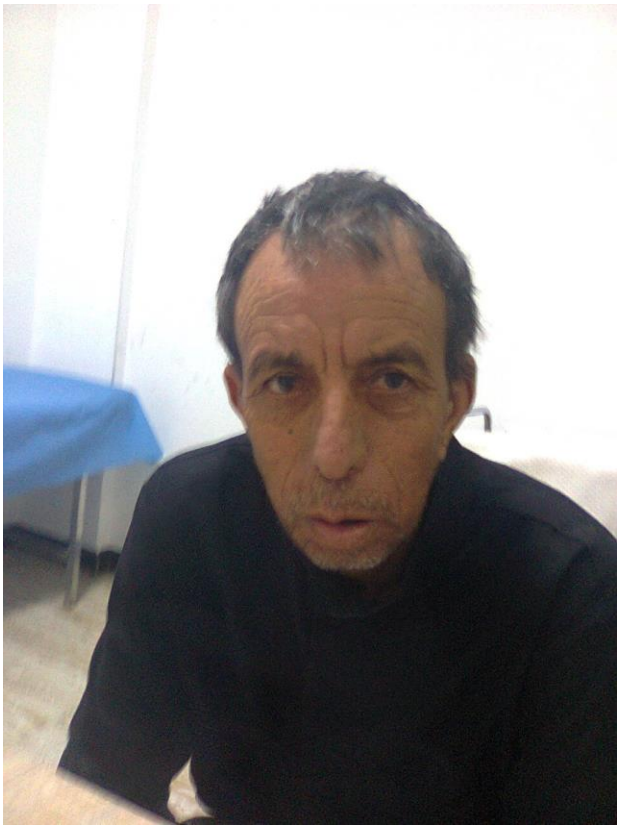

Figure-1B: On the 28st of October, 2023, the patient was much better

Discussion

Stephen Báthory (Figure-2A) one of the kings of Poland was most probably the first person known to have adult polycystic kidney disease.

The king died at about the age of 53 years. During the process of extracting internal organs for preserving the king's dead body as a mummy, surgeon Jan Zigulitz who was assisted by Dr.Buccella described the cysts in king's kidneys as large like the kidneys of a bull, and had an uneven and bumpy surface.

Figure-2A: Stephen Báthory (September 27, 1533-12 December 12, 1586), King of Poland and Grand Duke of Lithuania (1576-1586)

They also reported that there was a stone in gall bladder. The heart, lungs, liver, stomach, and spleen were considered to be normal.
The renal abnormalities were not thought to be the cause of the death during that that time. However, in 1933, Professor Franciszek Walter (Figure-2B) from Krakow Medical School, invited doctors and historians to a meeting to discuss the autopsy findings of King Stephen Bathory, they concluded that the cause of death was uremia caused by polycystic kidneys disease [25,26].

Figure-2B: Franciszek Ksawery Walter (1885-1950)

In 1793, Matthew Baillie (Figure-3C) emphasized that the vesicular cysts in kidneys in this disorder were different from hydatid cysts, and named the condition "False hydatids of kidney" [27].

Figure-3C: Matthew Baillie (October 27, 1761-September 23, 1823), a British physician and pathologist

In 1888, Félix Lejars (Figure-3D) from Paris emphasized that the cysts in this disorder were present on both sides and named the condition "Polycystic kidney" [28].

Figure-3D: Félix Lejars from Paris

In 1983, Szabó and colleagues emphasized that diagnostic ultrasound has been used since early 1970s for the early diagnosis of polycystic kidney disease and has been increasingly considered as a reliable diagnostic tool [30]. Figure-3 shows the diagnostic ultrasound of a female patient with polycystic kidney disease, and the diagnosis was made before the age of 50 years, and before the development of advanced chronic renal failure. Renal ultrasound showed bilaterally enlarged kidneys (Right kidney: 18x7 cm, left kidney: 16x6 cm), and both kidneys were affected by different size cysts. The largest cyst was 3.5 cm in diameter. There was

no mass or stone, and the pelvi-calyceal system, and ureters were not dilated.

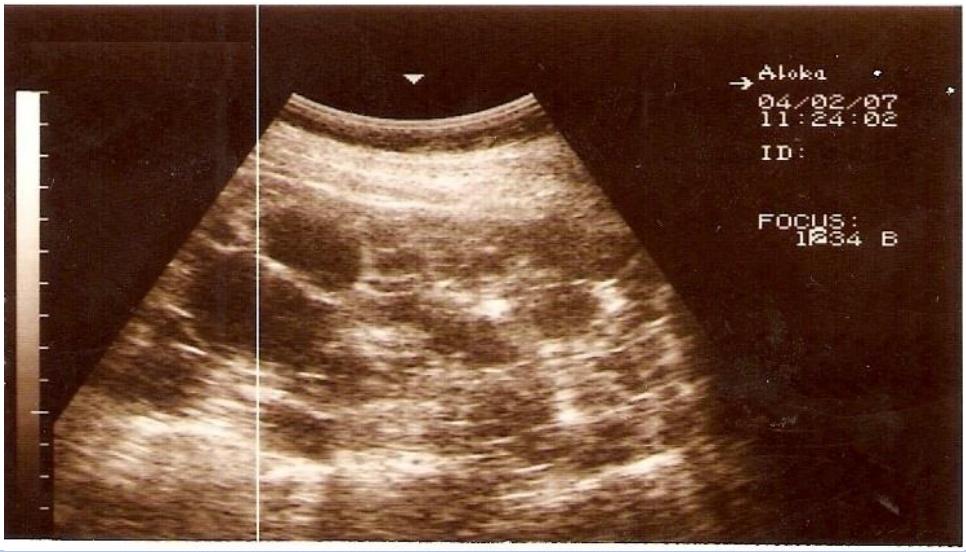

Figure-3: The diagnostic ultrasound of a female patient with polycystic kidney disease

The association of autosomal dominant polycystic kidneys with sensorineural deafness has been very rarely reported [14, 15].

Mora et al from Spain were most probably the first to report the association of autosomal dominant polycystic kidneys with bilateral sensorineural deafness. They reported four patients from four generations of one family who had autosomal dominant polycystic kidney disease associated with bilateral sensorineural deafness [14].

In 2022, Mayssa Abdelwahed (Figure-4) and her research group reported the association of autosomal dominant polycystic kidney disease in Tunisian patients with deafness [15]. The paper of Mayssa Abdelwahed and her research group was most probably the second paper reporting the association of autosomal dominant polycystic kidney disease with deafness, and this paper is most probably the third paper reporting this association.

Figure-4: Mayssa Abdelwahed from Tunisia

It is interesting to find that our patient had gall bladder stones, just like Stephen Báthory the first known patient to have polycystic kidneys. However, in 1996, Ishikawa et al from Japan studied 55 patients with autosomal dominant polycystic kidneys and 55 patients with chronic renal failure (Control group) who didn't have autosomal dominant polycystic kidneys with CT-scans. Eight patients with autosomal dominant polycystic kidneys and nine patients with chronic renal failure (Control group) who didn't have autosomal dominant polycystic kidneys had gall stones on CT-scans [31].

We have also recently reported an adult patient with chronic renal failure who didn't have polycystic kidneys, but had a gall stone disease [32].

The symptomatic uremia associated with chronic renal failure is caused largely by the retention of nitrogenous waste products which results from the metabolism of ingested proteins. Urea accounts for 80% or more of nitrogenous materials excreted into the urine in patients with chronic renal failure on diet containing 40 g or more of proteins. 25 to 40 % of the produced urea are recycled through the gastrointestinal tract and excreted through the intestine into the feces. This urea extra-renal excretion dose not reach 3 g daily even in symptomatic uremia.

Peritoneal dialysis lowers elevated blood urea through shifting urea excretion from the kidneys and urine to the peritoneal excretion by using intraperitoneal dialysis fluids **(Figure-5).**

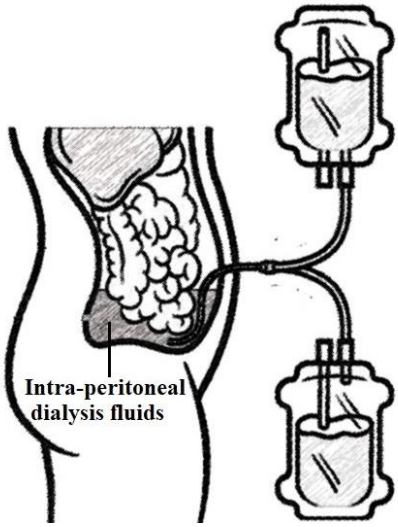

Figure-5: Peritoneal dialysis lowers elevated blood urea through shifting urea excretion from the kidneys and urine to the peritoneal excretion by using intraperitoneal dialysis fluids

Intestinal dialysis lowers urea by restricting protein intake and shifting the excretion of urea from the kidneys to intestinal excretion by increasing the fecal excretion of nitrogenous through modifying the enterhepatic urea cycle by consuming a relatively large amount of acacia gum which is a soluble fiber digested by colonic flora [32-35].

Intestinal dialysis improves uremic symptoms and other abnormalities of chronic renal failure through the use of a urea lowering agent, powdered acacia gum in association with use of the dietary and pharmacologic therapies of chronic renal failure [32].

Powdered acacia gum (Figure-6A) is a complex polysaccharide produced from the dried gummy substance (Figure-6B) of acacia trees' stem and branches [Senegal family leguminosae] (Figure-6C).It is considered safe by the FDA. It's widely used in the production of foods such as candies, puddings, and beverages. Powdered acacia gum has demulcent properties and therefore, it is often added to medicines [21, 24, 32].

Figure-6A: Powdered acacia gum

Figure-6B: The dried gummy material of the stem and branches of acacia trees

Figure-6C: Acacia trees, Senegal family leguminosae

Conclusion

The association of autosomal dominant polycystic kidneys with sensorineural deafness has been very rarely reported. Mora et al from Spain were most probably the first to report the association of autosomal dominant polycystic kidneys with bilateral sensorineural deafness. The paper of Maysa Abdelwahed and her research group was most probably the second paper reporting the association of autosomal dominant polycystic kidney disease with deafness, and this paper is most probably the third paper reporting this association.

This paper also further emphasized the clinical therapeutic values and effectiveness of intestinal dialysis in lowering blood urea level and improving symptoms of symptomatic uremia associate with chronic renal failure.

Acknowledgment

The patient kindly accepted publishing his photo to enhance the documentation of his case.

Some of the figures in this paper were included in previous author's publications, but the author has the copyright of all the figures and sketches included in this paper.

Conflict of interest: None.

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