SUPPORTING EXAMINATION OF CT-SCAN ON POLYCYSTIC KIDNEY DISEASE PATIENT AT DUSTIRA HOSPITAL CIMAHI INDONESIA

(PEMERIKSAAN PENUNJANG CT-SCAN PADA PASIEN PENYAKIT GINJAL POLISITIK DI RUMAH SAKIT DUSTIRA CIMAHI INDONESIA)

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ABSTRACT

Polycystic Kidney Disease (PKD) is the growth of many kidney cysts. PKD is divided into Autosomal Dominant Polycystic Kidney Disease (ADPKD) and Autosomal Recessive Polycystic Kidney Disease (ARPKD). ADPKD prevalence 1:500 and ARPKD 1:6000-40000. The patient, a 34-year-old male, came to the hospital with the main complaint of bilateral low back pain. Patients are given symptomatic treatment by diclofenac sodium 50mg 2 dd 1—supporting Examination for PKD diagnosis with CT-Scan. The author is interested in studying more about PKD because it is asymptomatic and only becomes symptomatic at the age of 30-40 years in ADPKD and has high mortality in the ARPKD type.

Keywords: CT-scan; polycystic kidney disease

ABSTRAK

Penyakit Ginjal Polikistik (PKD) adalah tumbuhnya banyak kista pada

*ginjal. PKD terbagi menjadi Penyakit Ginjal Polikistik Dominan Autosom (ADPKD) dan Penyakit Ginjal Polikistik Resesif Autosom (ARPKD). Prevalensi ADPKD 1: 500 dan ARPKD 1: 6000-40000. Pasien laki-laki berusia 34 tahun, datang ke rumah sakit dengan keluhan utama nyeri punggung bawah bilateral. Pasien diberikan pengobatan simtomatik dengan natrium diklofenak 50mg 2 dd 1. Pemeriksaan Penunjang diagnosis PKD dengan CT-Scan. Penulis tertarik mempelajari lebih lanjut tentang PKD karena asimtomatik dan baru menjadi simtomatik pada usia 30-40 tahun pada ADPKD dan angka kematian yang tinggi pada t*ipe *ARPKD.*

Kata kunci: *CT-scan; penyakit ginjal polikistik*

INTRODUCTION

PKD is divided into Autosomal Dominant Polycystic Kidney Disease (ADPKD) and Autosomal Recessive Polycystic Kidney Disease (ARPKD). ADPKD prevalence 1:500 and ARPKD 1:6000-40000. ADPKD was mostly found at the age of 30-40 years, while ARPKD founded in neonates or during the womb. ARPKD caused by mutations in the PKD-1 (polycystic kidney disease 1) gene in 85% of cases and PKD-2 (polycystic kidney disease 2) in 15% of cases. The two genes produce polycystin-1 and polycystin-2, which regulate growth and cell proliferation in primary cilia. Meanwhile, ARPKD is caused by mutations in the PKHD-1 (polycystic kidney and hepatic disease 1) gene, which produces fibrocystin with a function like polycystin.[1-9]

PKD was one of the causes of hypertension, haematuria, and low back pain. On physical examination, bilateral kidney enlargement can be found and confirmed by supporting examinations in the form of radiological examination. The radiological examination consists of ultrasonography which can show an enlarged kidney with the appearance of multiple cysts varying marked by a round hypoechoic area bounded by hyperechoic areas. The quality of intravenous urography is usually poor due to the large number of cysts and is rarely performed. CT scan was the better radiological modality. It can show more clearly, especially in small cysts (<0.5 cm) not visible on ultrasound examination, to monitor enlargement cysts and kidneys.[2,8,10-15] Treatment for PKD includes supportive treatment, such as pain

management and operative treatment to reduce pain.

The disease can develop progressively if kidney failure occurs, which allows dialysis and kidney transplantation. This management aims to prevent complications and maintain kidney function. Early detection of this disease is necessary to avoid future complications. Therapeutic options are given according to the patient's clinical condition. If there is hypertension, then giving ACE inhibitors can be an option. If there are complaints of decreased urinary frequency leading to kidney failure, haemodialysis is considered. Other therapies include administering mTOR inhibitors, including agents in the form of rapamycin and everolimus can slow cyst expansion and maintain kidney function, which can be given to the ADPKD type. Besides that, administration of vasopressin receptor antagonists can reduce fluid secretion in the cyst, reducing cyst enlargement.[1-3] Complications associated with polycystic kidney diseases (ADPKD and ARPKD) such as terminal kidney disease, hypertension, urinary tract infection (UTIs), formation of extrarenal cysts (liver, heart (mitral valve prolapse), blood vessels (cerebral aneurysm), intestine (diverticulosis), nephrolithiasis, and metabolic disorders (decreased urine citrate levels, triggering the formation of uric acid stones), and anaemia.[14,16-20] Based on the doctor's professional education at Universitas Jenderal Achmad Yani Faculty of Medicine at Dustira Cimahi Hospital, the incidence of Polycystic Kidney Disease is rare, but the disease diagnosed at the productive age of Indonesia for the ADPKD type and high mortality in neonates with ARPKD type. Therefore, the authors are interested in studying about Polycystic Kidney Disease patients at Dustira Cimahi Hospital. This case report aims to describe the radiological picture of a CT-Scan in a patient with Polycystic Kidney Disease.

CASE REPORT

A man, Mr B, 34 years old, came with primary complaints of right and left low back pain two months ago. The pain has worsened lately to the point where it interferes with activities. It was no other complaints, such as the reddish colour of urine, manifestation on the liver such as enlargement of the abdomen, blood emesis, and blood on the face.

On physical examination, the patient was in compos mentis awareness with GCS 15 and looked moderately ill. The patient's blood pressure was 120/80 mmHg, pulse was 82x/minute, respiration was 20x/minute, and body temperature was 36.5^{o}C. In the head examination, no

abnormalities on the eye or anaemic conjunctiva were not founded, and no exceptions were found in the nose and mouth. There were no abnormalities on the neck. The chest examinations were in the normal range with vesicular breath sound dextra, same on sinistra. On the abdominal examination, there was no palpation of the kidneys on both sides on physical inspection of the abdomen, and there was no enlargement palpation of both lobes of the liver. In extremity superior and inferior examination, there was no abnormality found.

On neurological examination, there were no abnormalities in the cranial nerves, sensory examination was within normal limits, and motor examination was within normal limits, no pathology reflex was found.

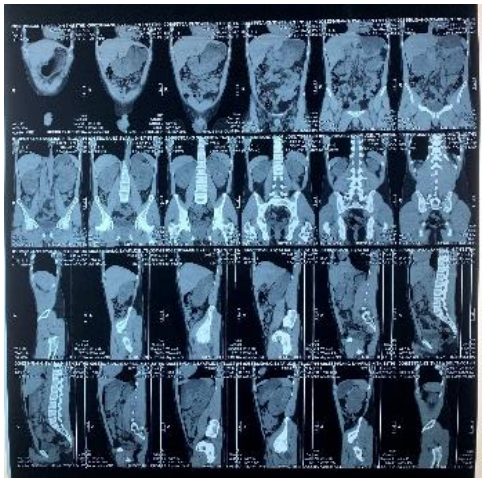


Figure 1. The CT scan shows polycystic kidney disease (PKD) longitudinal view.

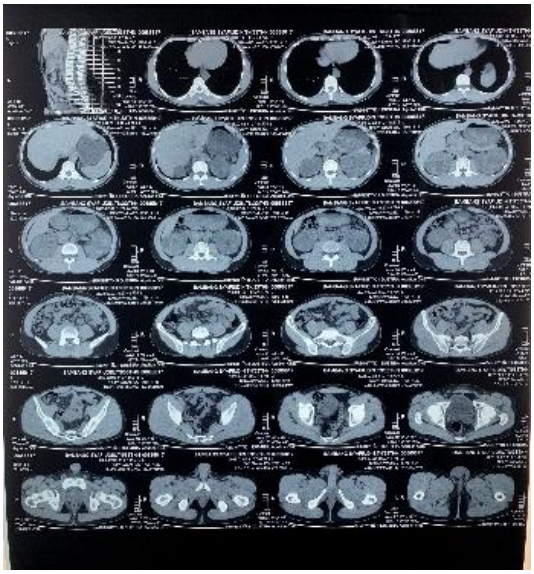


Figure 2. CT-Scan shows polycystic kidney disease (PKD) transverse view

A CT scan of the abdomen with an axial section has been carried out (Figure 1 and Figure 2). Scanning was performed without IV contrast, and a coronal and sagittal section was reconstructed. The liver is not enlarged with a flat surface. The parenchyma texture is homogeneous with average density—no visible s.o.l. The portal vein and hepatic vein are not dilated. There is no dilation of the intra and extra-hepatic bile ducts. Normal gallbladder, the wall not thickened, no stones are seen—average size and shape of the pancreas. No dilation of the pancreatic duct was seen. No masses and no calcifications were seen. Normal size and shape of the spleen with a flat surface and homogeneous parenchymal density. No visible mass/s.o.l, no splenic vein dilation.

The right kidney has normal size and shape, no visible s.o.l., and no dilation

of the pelvicalyceal system. No stone is visible. Hypoechoic shadows, homogeneous ovoid, multiple sharp boundaries with the most significant size being 3.78 cm. In the left kidney, normal size and shape, no visible s.o.l., no dilation of the pelvicocalyces system. No stones were seen, hyperechoic, homogeneous, ovoid, multiple clear boundaries, with the largest size being 4.74 cm. The ureters are not dilated, no masses are seen, no stones are seen, the urinary bladder is large and normal in shape, no stones are seen, no masses are seen, and the walls are not thickened. It was the normal prostate size and shaped with a flat surface, homogeneous parenchymal density, and no visible soles or calcifications.

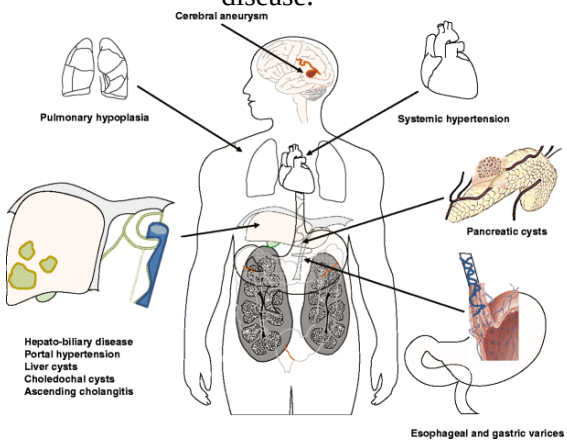
The conclusion showed a bilateral polycystic kidney—liver, bile, spleen, bladder, and prostate within normal limits. The patient has undergone medical therapy in the form of diclofenac sodium 50 mg 2 dd 1.

DISCUSSION

Polycystic Kidney Disease (PKD) is a genetic disorder characterized by the growth of multiple cysts in the cortex and medulla of the kidney. PKD is divided into Autosomal Dominant Polycystic Kidney Disease (ADPKD) and Autosomal Recessive Polycystic Kidney Disease (ARPKD).[2,9]

Polycystic kidney disease (PKD) greatly impacts morbidity, hospitalization, mortality, and cost to society. PKD is one of the life-threatening diseases of the kidney. ADPKD prevalence 1:500 and ARPKD 1:6000-40000. ADPKD is commonly found at 30-40 years; only 10% of cases can be found in <10 years, while ARPKD is found in neonates or can be found during the gestation period.[1-4] In the ADPD type, symptoms arise when the age of 30-40 years is the productive age of the Indonesian people which can be a problem.[2-3,16-17]

Figure 3. Manifestation of polycystic renal disease.[20]



Clinical manifestations of polycystic kidney disease (PKD) include hypertension, especially diastolic blood pressure (50-75% case), haematuria, and low back or flank pain. The pain characterized as severe and debilitating in the back, abdomen, or flank remains parallel to hypertension, with an incidence of about 60%. ARPKD usually shows

clinical manifestations in 2 organs, the kidneys and the liver—symptoms of portal hypertension as a liver manifestation. The manifestation was found, such as hematemesis, abdominal enlargement due to an enlarged spleen, melena, and other symptoms. ARPKD type is classified by perinatal ARPKD, neonatal ARPKD, infantile ARPKD, and Juvenile ARPKD (Table 1).[2,9,20]

Table 1. Classification ARPKD[20]

Form of ARPKD	Presentation
Perinatal	Pulmonary Hypoplasia
Neonatal	Mild hepatic fibrosis, cystic distortion of 90% of nephrons, renal malfunctioning.
Infantile	Cystic involvement of only a few nephrons, portal hypertension, hypersplenism
Juvenile	<10% nephrons involved in cystic distortion, little or no renal malfunctioning, portal hypertension, and hepatic fibrosis.

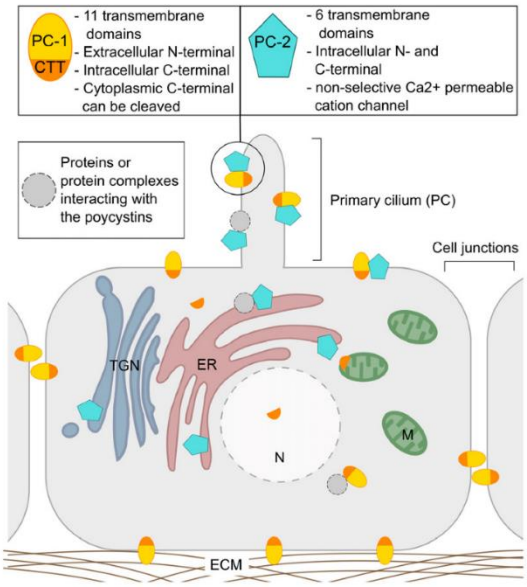
On physical examination, severe oligohydramnios can be detected with prenatal ultrasonography, as bilateral kidney enlargement during pregnancy as ARPKD manifestations usually in the third trimester, with placentomegaly , normal renal architecture, and bilateral, enlarged, smooth and hyperechogenic kidneys, small or absent urinary bladder are suggestive of ARPKD. The ARPKD antenatal ultrasound is diagnostic and only detected in severe cases. The findings are confirmed by supporting examinations in the form of radiological examinations. On radiology examination, based on plain abdominal radiographs, showed bilateral enlargement of the kidneys accompanied by multiple cystic calcifications and can be accompanied by displacement of nearby organs such as the intestine. Based on abdominal ultrasound examination found, enlarged kidneys with multiple cysts. Based on radiological examination, it is easy to find multiple cysts in ADPKD, while in ARPKD, it is difficult to find cysts. There are diagnostic criteria for ADPKD based on ultrasound examination, namely, at least two cysts in 1 kidney or one cyst in each kidney in patients aged <30 years or children, at least two cysts in each kidney in patients aged 30-49 years, and at least four cysts in each kidney. Kidney in patients >60 years of age. Based on the CT-Scan examination, bilateral kidney enlargement can be found accompanied by a picture of hypodense lesions in the kidney parenchyma, especially in the ADPKD type. Anatomical, pathological examination of lung tissue can reveal pulmonary hypoplasia, specifically in ARPKD. [2,9,13,16-18,20]

Denaturing high-performance liquid chromatography (DHPLC) was used for the mutation scanning with detection rates of 65-70% for the mutation of PKD1

and PKD2. Blood tests are also performed to check the variations in plasma concentrations of specific agents like serum creatinine, and a reduced concentration of Na (hypernatremia) is diagnostic for ARPKD. Other blood tests like haemoglobin level and leucocyte profile can be used as a diagnostics for PKDs Complication.[20-21]

In polycystic kidney disease, the cysts can develop anywhere on the kidney's nephrons due to abnormal cell division. It is due to gene mutations of two types of genes. Was PKD-1 located on primary cilium, tight junctions, adherent junctions, desmosomes, focal adhesions, and was PKD-2 located in the primary cilium, centrosome, endoplasmic reticulum gene in ADPKD. A third mutated (monoallelic) gene, DNAJB11, was recently identified to trigger ADPKD. There was an autosomal dominant disease so that it can be found both in males and females equally, and each offspring has a chance of 50% of inheriting the disease. *PKD1* codes for polycystin 1, a 4304 amino acid protein. Then, Polycystin 1 interacts with polycystin two and is involved in cell cycle regulation and intracellularcalcium transport. *PKD2* codes for polycystin two, which is structurally similar to polycystin 1. It is a member of the family of voltage-activated calcium channels. Polycystin 1 and polycystin two are located in the epithelial cells of the renal tubules and other areas of the renal cell epithelium (figure 4). Both form heteromeric complexes found in the primary cilium of kidney epithelial cells. The primary cilium was considered a mechanical receptor that can sense changes in tubular fluid flow, and that can transduce them into intracellular calcium signalling (Figure 4).[14,16-23]

Figure 4. Subcellular localization of the polycystins in ADPKD.[23]



ADPKD1 was more severe than ADPKD2. According to the normal physiological conditions, the interaction of polycystin 1 and 2 design was a "receptor-ion channel complex" in which polycystin 1 is responsible for the sensation of luminal sheer stress and opens polycystin 2, a non-selective channel Ca2+. Polycystin 2 interacts with store-operated Ca2+ transient

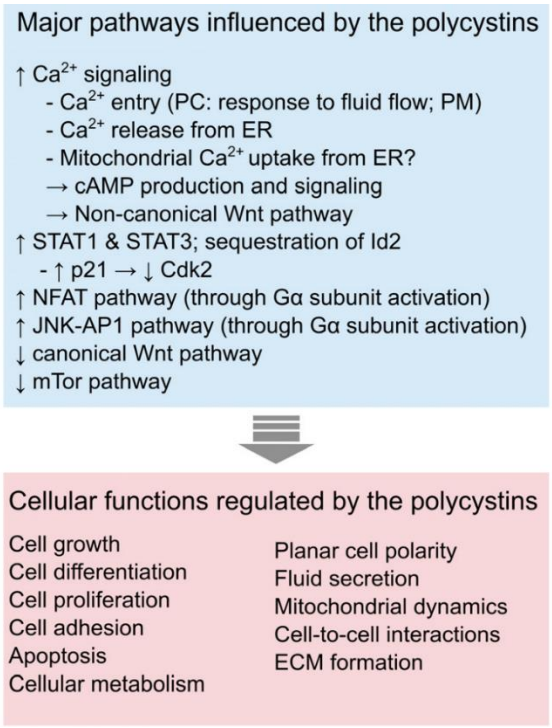
receptor potential channel 1 (TRPC 1) it is proposed that the primary cilia become curved due to flow of the fluid through renal epithelium and induce intracellular Ca2+ influx. The changes in intraluminal pressure induce the arterial response, which affected by polycystin 1 and polycystin 2 ratio in the arterial myocytes by regulating "*stress activated cation channels*" thus plays a key role in maintenance of intracellular Ca2+ homeostasis in vascular smooth muscles. Planar Cell Polarity (PCP) controls polarized cell division, direction of cell migration and cellular differentiation to assist in organogenesis of organ systems. The disruption of these signalling cascades produce differentiation of cystic epithelium, abnormalities of basement membrane, uncontrolled division of cells, impaired secretory characteristics of epithelia, and altered polarized state of cell. Ultimately the cystic expansion compresses the normal renal tissues causing apoptosis and intrarenal ischemia due to compression of renal vessels leading to stimulation of renin angiotensin aldosteron system that in turn is responsible to increased vascular resistance, sodium retention, progressive cystic enlargement and finally renal fibrosis. In vitro studies suggest the proliferation of epithelial cells of ADPKD patient supporting the idea that formation of normal renal tubular structures are controlled by apoptosis after birth, but it becomes persistent and proliferation continued. The polycystin proteins are also found in other subcellular locations and seem to be implicated in numerous cell signaling pathways regulating gene expression, cellular differentiation and proliferation, as well as apoptosis (figure 5). [14,16-23]

And then, for the ARPKD type, there was one type of gene, there were PKHD-1 gene mutations located in chromosome 6p21 characterized by the development of cysts in the kidneys of fetuses or young children and responsible for high infant mortality. The gene encodes a protein called Fibrocystin or polydactyls. Fibrocystin is found excessively in infants' collecting tubules of the kidneys and serves as a membrane receptor to interact extra-cellular and intracellularly with proteins and phosphorylation sites, respectively. The recessive form means that 25% of children of two carrier parents will be affected by cystic kidneys. In recessive conditions, the dilatation usually remains limited to collecting tubules of the nephrons because of the typical development of cysts here. The disease is rare compared to ADPKD and is always bilateral, with severe outcomes due to manifestation in early infancy and early or late childhood. Still, the disease also manifests in early adulthood in rare cases. Based on the age of occurrence,

number of collecting ducts dilatation and extent of hepatic fibrosis. The disturbance in the PCP pathway is responsible for ARPKD because mutation on the PKHD1 gene causes loss of cell-oriented division which, unlike PKD1 and PKD 2, appears as an aetiology of cyst formation.[14,16-23]

Figure 5. Polycystine function.[23]

Even though the precise pathophysiological mechanisms of PKD remain elusive, extensive research has offered valuable insights regarding the function of the polycystin proteins. The pathogenesis and pathophysiology of ADPKD were similar to ARPKD. The



anatomical changes in renal architecture include bilaterally distended kidneys with sonographically identified multiple small cysts and medullary to cortical parenchyma containing cylindrical spaces arranged along the radius. The gene mutation can make the cyst grow larger. The cysts that grow not only damage the glomerulus and tubular membranes then fill the nephrons. The fluid enters the cyst instead of leaving it because of baso-laterally oriented Na+/K+ ATPase, which is abnormally located at the apical position (figure 5). Shifting of another channel, Na/K/2Cl symporters to epithelial cells basal surface. The cyst enlargement causes decreased kidney function but can press the other organ. The kidney tissue is eventually replaced with a cyst. The kidneys become very large and larger. Each kidney will enlarge two to three times from its normal size, which can reach a total volume exceeding 3500 mL for both kidneys combined. This abnormal size causes other abdominal pressure, shifts to the side of their normal position, and pulls it on the renal capsule cause activation of the nociceptor, resulting in flank pain and eventually low back pain. Hypertension occurs by activating the renin-angiotensin-aldosterone system in response to decreased glomerular function. Cysts filled with fluid can rupture or compress the surrounding vasculature causing urine to mix with blood and resulting in hematuria and pain. [14,16-19,20-23]

The complications of these ADPKD and ARPKD are similar, but there are differences in the onset and severity of the disease. The ARPKD type is commonly encountered during gestation, so severe manifestations are often found at birth, so the baby can die quickly due to severe kidney and liver failure. Patients with ARPKD who can pass this period generally experience complications in the form of cirrhosis of the liver (congenital liver fibrosis). The most common risk factors of PKD is positive family history. Family history is a cheap and easy way to approach the diagnosis of PKD. Other risk factors like age, drug and hormone consumption. Multiple cysts of the kidney and long-term dialysis considered risk factors for progression to ESRD[1,14,16-23]

Complications associated with polycystic kidney disease (ADPKD and ARPKD) include terminal kidney disease, hypertension, urinary tract infection (UTI) in 30-50%, especially in women, formation of extrarenal cysts (liver, heart (mitral valve prolapse), blood vessels (cerebral aneurysm) occurring in 4-10%, and intestine (diverticulosis), nephrolithiasis, and metabolic disorders (decreased urine citrate levels, triggering the formation of uric acid or calcium stones). Kidney stones are found in 20% to 30% of patients. Stone formation caused by impaired medullary trapping of ammonia and its conversion to urine leads to reduced ammonia excretion. Low citrate concentration and low urine pH are contributory factors for stone formation. In patients with PKD (ADPKD and ARPKD), anaemia can occur due to genetic mutations causing macrophages in the blood to infiltrate tissues causing erythrocyte fragmentation and haemolysis of red blood cells.[1,14,16-20]

Cardiovascular is The most common extrarenal complication that contributes to morbidity and mortality in ADPKD patients. It is believed that distortion of the renal parenchyma leads to structural damage, tubular dysfunction, and renal vascular ischemia and activates the Renin Angiotensin Aldosterone System (RAAS). Furthermore, GFR decline occurs after extensive vascular remodelling, the altered intima-media thickness of carotid arteries, impaired endothelial-dependent vascular relaxation, and vascular ultrasound thickness. As Cilia are a local regulator of blood vessels, ciliopathy plays a crucial role in CVD development.[16-23]

PKD treatment is supportive and can develop progressively if kidney failure occurs, which allows dialysis and kidney transplantation. ADPKD patients account for about 6% of all patients under renal replacement therapy. Patient education, including a low salt diet, is recommended if

hypertension or renal failure symptoms are present. No special diet founded for kidney failure has been reported to be useful in PKD therapy. Routine blood pressure monitoring is necessary. Achievement of blood pressure is recommended at <130/80mmHg. If the investigation found that there is >1gr/day protein in the urine (proteinuria), the blood pressure target is suggested to be <125/75mmHg. It was found that blood pressure monitoring can reduce mortality in patients with ADPKD. Haematuria may result from a ruptured cyst. Haematuria is self-limiting. In patients with haematuria, physicians can educate adequate drinking and rest to the patients. Hospitalization is necessary if haematuria persists for several days and if large amounts of blood are found. Segmental artery embolization is indicated in case of excessive bleeding or hematoma formation. In the case of ARPKD, intensive care and prolonged ventilation are required to cope with respiratory complications. In patients with PKD, moderate physical activity is recommended for 30 minutes 5 times weekly. [2,16-18,22-24]

Treatment for PKD patients is conservative. The treatment is given in the form of diclofenac sodium for pain management. Diclofenac sodium therapy was a symptomatic therapy. Diclofenac Sodium is one of the NSAIDs (nonsteroidal anti-inflammatory) that works by inhibiting the COX enzyme (cyclooxygenase), thereby reducing the production of prostaglandins so that the effect is as an analgesic (pain reliever). However, this therapy is debated because of the potential to cause hyperkalaemia. For acute pain management, narcotic analgesics (methadone, fentanyl, and codeine) can be recommended, and local anaesthesia's blockage of the splanchnic nerve might be helpful. The best therapeutic management is surgical therapy in the form of cyst decompression, which relieves pain in 60-80% of patients. Pain management is given together with periodic monitoring of kidney function and liver function. This patient was given pain management therapy, namely administering diclofenac sodium. In this patient, cyst decompression was not performed by cyst aspiration because there were multiple cysts. According to the literature, cyst aspiration has a high recurrence rate. In patients with hypertension, the recommended antihypertensive therapy class is angiotensin-converting enzyme (ACE) inhibitors; examples of drugs in the ACE inhibitor class are captopril, enalapril, lisinopril or angiotensin II receptor blockers (ARBs). Examples of ARB class drugs are telmisartan, losartan, irbesartan, and candesartan. It is not recommended to give

calcium channel blockers such as amlodipine. Antihypertensive drugs were recommended for ACE inhibitor group work by inhibiting ACE (angiotensin-converting enzyme), causing vasodilation of blood vessels resulting in decreased blood pressure and inhibiting aldosterone. The ARB group is also recommended as one of the antihypertensive drugs that work by inhibiting angiotensin II receptors, causing vasodilation of blood vessels which results in a decrease in blood pressure and inhibits aldosterone so that they can have side effects in the form of hyperkalaemia similar to the ACE inhibitor group but because they do not inhibit ACE so they do not have side effects in the form of coughing and ARB group was safer used for patient PKD with hypertensive. ACE inhibitors and ARBs can improve renal functions by increasing renal blood flow, reducing proteinuria, and decreasing left ventricular hypertrophy. Monitoring blood electrolyte levels is necessary for patients with renal failure in administering antihypertensive drugs. If complications are found in the form of Urinary Tract Infection (UTI), antibiotics in the ciprofloxacin class, trimethoprim-sulfamethoxazole,
clindamycin, and chloramphenicol can be given. The most common bacteria that cause UTIs are gram-negative bacteria.[2,16-18,20-25]

When Kidneys are not excessively enlarged, peritoneal dialysis is suitable, while renal transplantation remains the treatment of choice in end-stage renal disease. Certain complications, such as diabetes mellitus and cardiovascular diseases, follow renal transplantation. For hepatic cysts, no treatment is recommended until it becomes symptomatic. The interventions such as percutaneous cyst aspiration, laparoscopic hepatic cyst fenestration, combined liver resection and cyst fenestration, and liver transplantation are available options to reduce the size of the cyst as well as the liver. Conservative treatment is recommended for intracranial aneurysms less than 7 mm in length and detected on presymptomatic screening. Patient education, for example, avoiding smoking, controlling blood pressure, and controlling lipid profile, show beneficial results in patients with intracranial aneurysms. If cysts are found in the liver or kidneys that are infected, which are not effective with conventional antibiotic therapy, surgical treatment in the form of cyst drainage can be planned. The procedure is performed with a puncture assisted by ultrasonography (USG).

Acute abdominal pain related to cystic bleeding or obstruction by a clot, stone, or infection. On cysts. 25% of patients with severe pain not relieved by

operative treatment can be given narcotics. The cause is an inaccessible cyst, such as a part of the renal medulla. Nephrectomy can be suggested as a last resort for pain control in patients. In cases where bilateral nephrectomy is necessary, be aware of refractory ascites as a side effect. Partial hepatectomy may be recommended in patients with extensive liver enlargement, causing difficulty getting adequate nutrition or severe abdominal pain due to liver enlargement. Liver transplantation is suggested in patients with complicated portal hypertension with large inaccessible areas.

Based on recent studies, tolvaptan can be used as a vasopressin V2 receptor agonist; tolvaptan diminishes cAMP production and, thus, cAMP-associated cyst cell proliferation and cyst fluid secretion. However, this treatment is only approved in at-risk adult patients or shows evidence of rapidly progressing disease. Furthermore, it is unsuitable for all patients due to its risk of hepatotoxicity and other adverse effects, like polyuria and increased thirst, that can significantly lower the quality of life of people under treatment, but further research is needed. Other drugs, like mTOR inhibitors in the form of rapamycin, can slow cyst expansion and maintain kidney function. Rapamycin, combined with ARB, has been shown to be useful in treating polycystic kidney disease. Specific therapy for ARPKD is lacking and is focused on managing complications such as respiratory insufficiency, hypertension, growth retardation, renal insufficiency, and hepatic manifestations. Mechanical ventilation with neonatal intensive care support is required. Growth delays are related to increased metabolic demands, treated with feeding through a nasogastric tube, correction of existing electrolyte abnormalities, and periodic endoscopy to check whether oesophagal varices appear. And liver transplantation or dialysis is required in treating end-stage renal failure. [1-3,18,20,23-26]

Polycystic kidney disease of both the ADPKD and ARPKD types is a major cause of end-stage renal failure and a common indication for dialysis or kidney transplantation. The patient becomes suggestive to transplantation or most commonly, dialysis at 70 in 50% of cases. In ADPKD, mutations in both PKD-1 and PKD-2 genes show a worse severity than mutations in just one gene. Life expectancy for ADPKD patients is estimated to be 53 to 70 years. Cardiac diseases are the most common cause of death (34%) in ADPKD patients with end-stage renal disease (ESRD). Next to cardiac diseases are infections that cause approximately 20.4% of deaths in such patients. There is a high risk of ruptured intracranial aneurysms,

where 8% to 11% of ADPKD patients die due to ruptured intracranial aneurysms. Properly detecting risk factors for intracranial aneurysm formation in ADPKD is crucial and can allow for targeted patient screening and risk factor reduction. These data are critical given the significant morbidity and mortality (between 40% and 60%) associated with intracranial aneurysm rupture in ADPKD. In ARPKD, 30% of babies can die quickly due to severe kidney and liver failure after birth. ARPKD sufferers who can pass this period generally experience complications in the form of cirrhosis of the liver (congenital liver fibrosis) but have a good chance of living into adulthood but need lifelong medical therapy.[2,18,20-26]

CONCLUSION

The patient has right and left low back pain. No neurological abnormalities were found. The development of cysts in the bilateral kidney causes pressure on surrounding organs and pulls on the renal capsule, causing nociceptor activation and flank pain. The results of the CT scan showed bilateral polycystic kidneys. Therapeutic management of polycystic kidney disease includes symptomatic therapy, haemodialysis, kidney transplantation and management of complications. Diclofenac sodium therapy is a symptomatic therapy to reduce pain in patients. Further management aims to prevent complications and maintain kidney function. Early detection of this disease is necessary to avoid future complications.

CONFLICT OF INTEREST

The authors state that in this case report, there is no conflict of interest

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