



Case Report

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## Perioperative management of giant symptomatic autosomal dominant polycystic kidney disease: A case report

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### ARTICLE INFO

### ABSTRACT

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Autosomal dominant polycystic kidney disease (ADPKD) is an inherited systemic disorder characterized by renal cyst enlargement which results in abnormal kidney structure and renal insufficiency. The anaesthesia management of patients with ADPKD is often complex and carries considerable risks of perioperative complications. Safe and successful anaesthesia management requires an increased understanding of ADPKD pathophysiology. We report successful perioperative management of a bilateral open surgical radical nephrectomy on a male patient with giant symptomatic ADPKD. The patient had chronic renal failure and extremely large kidneys. We believe that the description and observations seen in this case will aid in the recognition and anaesthetic management of this syndrome.

#### Keywords:

Anaesthetic management  
Autosomal dominant  
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### 1. Introduction

Polycystic kidney disease (PKD) is an inherited genetic disease characterized by progressive cystic formations (Chapman, 2008; Wuthrich et al., 2009). Although the involvement of complex mechanisms in the development of cysts is suggested, ciliary dysfunction is thought to play a role in the main pathology. Cystic expansion progressively enlarges the kidney, leads to intrarenal ischemia and activates the renin-angiotensin-aldosterone system (RAAS). An activated RAAS causes hypertension, related proteinuria, cardiovascular complications and progression to end-stage renal

disease. Apart from these, extra renal findings are seen, but they rarely emerge during the early stages of the disease (Chapman, 2008; Wuthrich et al., 2009). Autosomal dominant or recessive genetic inheritance is often seen. Autosomal dominant polycystic kidney disease (ADPKD) is more frequently seen during the fifth and sixth decades of life; it slowly progresses to renal enlargement with a resultant development of renal failure (Chapman, 2008). Its prevalence is nearly 1:800–1:1,000, and constitutes 2.5% of all end-stage renal diseases (Chapman, 2008; Wuthrich et al., 2009). ADPKD is a heterogeneous disease which

develops as a mutation of at least two genes (mutations on chromosomes 16 and 4, chromosomes at PKD1 and PKD2 genes, respectively). As a consequence of these mutations, epithelial de-differentiation, increased cell division and apoptosis and decreased reabsorptive capacity of the kidney are seen. Thus, cyst growth and expansion and, finally, enlarged kidneys and progressive renal injury occur (Chapman, 2008). Clinically, the patient may present with hypertension, flank pain, haematuria and cyst infection. Cysts may develop slowly or rapidly. Generally, the glomerular filtration rate is maintained until 30–40 years of age, and then it quickly worsens (Franz and Reubi, 1983; Wuthrich et al., 2009). During the seventh decade of life, nearly 50% of ADPKD patients become dependent on dialysis (Halvorson et al., 2010).

However, unlike the other forms of the disease, the autosomal recessive form evidences itself more frequently during the neonatal period and during infancy (Capisonda et al., 2003; Guay-Woodford and Desmond, 2003; Kaimori and Germino, 2008). It is seen 20 times less frequently than ADPKD, and its clinical manifestations generally lead to a more severe course (Kaimori and Germino, 2008). It is the most frequently seen renal cystic disease during childhood. Most of the cases are discerned during intrauterine life or at birth. In a seriously affected foetus, oligohydramnios and facial, vertebral and extremity anomalies are frequently seen. Clinically, it is characterized by cystic dilation of the collecting ducts of the kidneys, malformation of the biliary ductal system and hepatic fibrosis and portal hypertension; frequently, during the prenatal period, the foetus is lost secondary to pulmonary hypoplasia and respiratory failure (Zerres et al., 1998; Capisonda et al., 2003; Guay-Woodford and Desmond, 2003).

In our case, we report a successful perioperative management of a bilateral open surgical radical nephrectomy in a male patient. The patient had giant symptomatic autosomal dominant polycystic kidney disease, chronic renal failure (CRF) and extremely large kidneys. The anaesthesia management of patients with ADPKD is complex and carries with it considerable risks of perioperative complications. We believe that the description of this case will help in the recognition and anaesthetic management of this syndrome.

## 2. Case report

A 52-year-old male patient who had been followed for 20 years with diagnoses of ADPKD and CRF was admitted to the renal transplantation unit. The patient, when he was being prepared for transplantation in the unit, complained of shortness of breath which was found to be related to coronary artery disease. In the cardiology department, a drug-eluting stent was implanted, and in order to prevent stent thrombosis, a dual antiplatelet treatment, employing a combination

of acetylsalicylic acid (100 mg/d) and clopidogrel (75 mg/d), was initiated. One week later, he complained of abdominal pain and persistent fever, and a computed tomographic examination revealed intracystic bleeding. Based on the antibiotic susceptibility test results of the cyst contents, meropenem (2x1 g/d) and teikoplanin (1x400 mg/d) were initiated without any effect on his febrile state. Meanwhile, abdominal tenderness developed, and ultrasonography examination demonstrated a cyst rupture. Dual antiplatelet treatment was then discontinued, and a bilateral nephrectomy was planned. His anamnesis revealed the presence of ankylosing spondylitis for 25 years and hypertension for 20 years. He was using a tumour necrosis factor (etanercept 50mg/d) for the treatment of the ankylosing spondylitis and valsartan (160 mg/d), doxazosin (4mg/d) and nifedipin (60mg/d) for the management of his hypertension. It was also learnt that his brother had been followed-up with diagnoses of ADPKD and CRF. Systemic examination was unremarkable (temperature 37.5 °C). His airway was graded as Mallampatti class I with normal neck movements. Preoperatively, excluding his haemoglobin (Hb, 9 g/dL), his blood urea (63 mg/dL), serum creatinine (5mg/dL) levels and other parameters (serum sodium and potassium, blood cell count, liver function tests and coagulation profile) were within normal limits. On an abdominal ultrasoundogram, in addition to renal cysts, two small simple hepatic cysts were detected. An electrocardiogram showed sinus rhythm, and a chest X-ray showed normal lung fields. The patient was categorized as ASA III (American Society of Anesthesia). He was informed about all potential risks, and then he provided his consent for all procedures. He was advised to continue antihypertensive medication on the morning of the surgery. The Department of Rheumatology recommended cessation of etanerceptin 15 days prior to the operation because of potential infectious complications. In addition, the cardiology department was again consulted about the patient who discontinued his dual antiplatelet therapy. Accordingly, infusion of thyrofiban at a rate of 12.5 mg/24 hr was initiated up to eight hours before the operation, and re-initiation as soon as possible of dual antiplatelet therapy during the postoperative period was indicated. During the two weeks prior to the surgery, he underwent three sessions of dialysis each week; his blood urea nitrogen, serum creatinine and potassium values were maintained within acceptable ranges. His last haemodialysis was performed 24 hours prior to surgery.

In addition to standard monitorization (pulse oximeter, non-invasive blood pressure, urine output measurement, electrocardiogram and core body temperature), central venous pressure (CVP, measured from the right subclavian vein) and invasive blood pressure (intra-arterial, radial artery) measurements

were performed. Pleth variability index (PVI) was calculated, and total haemoglobin (SpHb) monitorization was carried out. For the estimation of PVI and the measurement of SpHb, a pulse co-oximetry (Rainbow R1 25 Adhesive Sensors, Adult; Masimo Corp., Irvine, CA, USA) probe was used. This probe was placed on the index finger of one hand and connected to a Masimo Radical-7 monitor with PVI software (Masimo Rainbow SET Radical-7 Pulse CO-Oximeter; Masimo Corp., Irvine, CA, USA). Before induction of anaesthesia, his heart rate (HR, 140/min), systemic blood pressure (120/80 mmHg), blood oxygen saturation (SpO<sub>2</sub>) (98%), SpHb (9 g/dl) and PVI (12%) were estimated. For the induction of anaesthesia propofol (1.5 mg/kg IV), rocuronium (0.6 mg/kg IV) and remifentanyl (0.5 mcg/kg/min), infusions were used, and the patient was intubated. Central venous pressure (CVP, 14 cm H<sub>2</sub>O), urine output (50 ml) and core body temperature (37 °C) were detected as indicated. The surgery was performed with the patient in the supine position and maintenance of the anaesthesia was achieved using desflurane (1 minimum alveolar concentration)/air (1L/min)/oxygen (1L/min), and remifentanyl infusion (0.1-0.25 mcg/kg/min). Fluid replacement therapy was adjusted according to pulse oximeter-derived PVI (goal-directed fluid management). Accordingly, PVI values were maintained at 13-14% (Cannesson et al., 2008). After the first hour of the operation, development of hypotension (70/40 mmHg) necessitated initiation of inotropic support (norepinephrine 0.04-0.1 µg/kg/min, and dopamine 4-10 µg/kg/min). During the operation, CVP (14-16 mmHg), SpHb (9-10 g/dL), core body temperature (36 °C), systolic and diastolic blood pressures (100-110 mmHg and 60-80 mmHg) and heart rate (110-120/min) were maintained within indicated limits. Arterial pH did not drop below 7.25, and decreased ionised calcium level (< 3.5 mg/dl) was treated with infusion of four ampoules of calcium (calcium gluconate levulinate 10%, 10 ml). During the surgery, which lasted five and one-half hours, infusions of various solutions and blood components, including a total of 5000 cc 0.9% sodium chloride, 2000 cc lactated Ringer's solution, 1500 gelofusine (0.04 g/500 ml; B. Braun-Irengun, Istanbul, Turkey), nine units of erythrocyte suspension and 11 units of fresh frozen plasma were made. The polycystic kidneys, which were removed bilaterally, weighed nearly eight kg (Fig. 1 and Fig. 2). At the termination of the operation, the patient was intubated under dual inotropic support and transferred to the intensive care unit (ICU). After nine days in the ICU, the patient was extubated and transferred to the Organ Transplantation Unit. The patient was admitted to a three times per week haemodialysis program and kidney transplantation was planned.

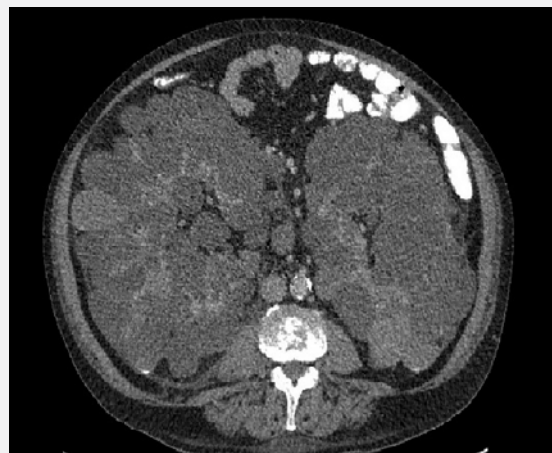


Fig. 1. Computed tomographic scan shows multiple bilateral kidney cysts, resulting in enlargement of the organ.



Fig. 2. Explanting giant polycystic kidneys.

### 3. Discussion

Although ADPKD is the most prevalently seen inherited disease, it is essentially a systemic disease (Gabow, 1993; Chapman, 2008; Kaimori and Germino, 2008). In addition to the kidneys, it is also seen in the liver (primarily as an extra renal lesion), pancreas, thyroid, subarachnoid space, seminal vesicles and inguinal hernias (Chapman, 2008; Kaimori and Germino, 2008). In addition, its vascular phenotype can manifest itself as intracranial aneurysms, mitral valve prolapses and biventricular dysfunctions. However, its clinically characteristic features are gradual and include massive cystic enlargement and, finally, the development of renal failure (Chapman, 2008). Because of increased RAAS activity, its earliest symptom is hypertension, which is seen in nearly 60% of the patients (Suvarna and Fernandes, 2011). During the advanced stages of the disease, a decrease in the concentration ability of the kidneys is seen (Arulkumaran et al., 2012). ADPKD is responsible for 8-10% of all cases with end-stage renal

disease (Gabow, 1993). Especially in patients who are diagnosed during its early stage, the coexistence of the male gender and hypertension indicate the progressive course of the disease (Wilson, 2004).

Our patient had hypertension and CRF which had been kept under control for 20 years with medication and with a low-salt and low-protein diet. In patients with CRF, coronary artery disease is frequently (40%) seen because of the presence of hypertension, systolic and diastolic dysfunction, diabetes, dyslipidaemia, peripheral vascular disease and anaemia (Gupta et al., 2004). Our patient had experienced a coronary ischemic attack before transplantation, and as a result of antiplatelet therapy, intracystic bleeding occurred. This, together with infection, worsened the clinical condition of the patient, further leading to the development of end-stage renal disease and azotaemia.

During the preoperative evaluation of such patients, a detailed medical history should be taken, and a thorough physical examination should be performed. Signs and symptoms of uraemia, fluid overload and inadequate dialysis should be emphasized, especially in the presence of comorbidities. Laboratory parameters, such as electrolyte concentrations, acid-base status, urea and creatinine levels, haematocrit, platelet count and coagulation, should be analysed in case dialysis needs to be performed before the operation. Chest radiography is usually helpful to rule out fluid overload, and an electrocardiogram is necessary to assess for changes caused by myocardial ischemia and by electrolyte abnormalities (Maurizio et al., 2012). Blood pressure (below 130/80 mm Hg) and haemoglobin values ( $\geq 10$  g/dL) should be brought to indicated optimal levels (Gupta et al., 2004; Locatelli et al., 2008).

In this patient group, the pharmacokinetic effects of several anaesthetic drugs were significantly altered due to CRF. Since free fractions of hypnotic agents increase, doses used in the induction of anaesthesia should be significantly reduced (SarinKapoor et al., 2007; Wagener and Brentjens, 2010). With this type of patient, after the induction of anaesthesia, hypotension is frequently seen because of autonomic dysfunction, hypovolemia and the preoperative use of angiotensin-converting-enzyme inhibitors. Therefore, in addition to dose reduction, slow titration of anaesthetic and sedative agents is required. In this patient group, because of the high incidence of gastroparesis rapid-sequence induction was frequently used. To this end, rocuronium can be used safely (Maurizio et al., 2012). Since plasma clearance of rocuronium is not affected by renal dysfunction, it can be also used in the maintenance of anaesthesia (Cooper et al., 1995; SarinKapoor et al., 2007). In our case, we used lower doses of propofol and remifentanyl for the maintenance of anaesthesia. Rocuronium was the preferred muscle relaxant.

Although the safe use of total intravenous anaesthesia (propofol, remifentanyl, and cisatracurium) in the maintenance of anaesthesia in this patient group had been indicated, we preferred to use the combination of an inhalational aesthetic agent (desflurane) and remifentanyl (Dahaba et al., 1999). Remifentanyl does not have active metabolites and is well tolerated in patients with CRF (SarinKapoor et al., 2007). It has been demonstrated that desflurane can be used in CRF patients without increasing renal dysfunction (Litz et al., 2002; SarinKapoor et al., 2007). Since it is minimally metabolized in the body, even with chronic use, the liberation of inorganic fluoride does not appear to cause renal injury (Maurizio et al., 2012).

Successful perioperative haemodynamic management of these patients is challenging. Intravenous fluid administration should be planned so as to avoid fluid overload or hypovolemia. However, the frequent coexistence of systolic and diastolic left ventricular dysfunction and poorly controlled hypertension causes low cardiac output and large swings in blood pressure. Therefore, these patients often need more advanced haemodynamic monitoring (arterial line, central venous and pulmonary artery catheters or the oesophageal Doppler, stroke volume variation and pulse pressure variation) (Maurizio et al., 2012). In meta-analyses that have been performed, it has been reported that goal-directed fluid therapy for the management of critically ill patients, and the major surgeries of these patients, will decrease organ-specific complications and improve post-operative outcomes (Corcoran et al., 2012; Davies et al., 2013). Herein, haemodynamic targets to be considered are achieved by measuring either cardiac output (CO) or fluid responsiveness. In these measurements, invasive methods (pulmonary artery catheterization) have been replaced by non-invasive methods (stroke volume variation [SVV], pulse pressure variation [PPV] or PVI) (Davies et al., 2013). The Radical-7 PVI that we used in our clinic allows measurement of these parameters. PVI is an algorithm that automatically and continuously measures the variability of the pulse oximeter plethysmographic waveform amplitude during the respiratory cycle in a mechanically ventilated patient (Hood and Wilson, 2011; Siswojo et al., 2014). Thus, we may be able to predict fluid responsiveness in major surgery. The response to the fluid administered to our patient was evaluated by continuous PVI measurements to optimize the patient's fluid status.

Anaesthesia management of nephrectomy for giant polycystic kidneys is challenging. Safe anaesthetic management requires an understanding of APDK pathophysiology and careful patient assessment. The changes in pharmacokinetics and pharmacodynamics of drugs used in the perioperative period must be taken into consideration. However, optimisation of volume status and blood pressure are fundamental to reducing morbidity and mortality.

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