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REVIEW ARTICLE

Renal transplantation and polycystic: Surgical considerations☆

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KEYWORDS
Autosomal dominant polycystic kidney disease;
Nephrectomy;
Transplant

Abstract

Background: The indication and timing of nephrectomy in patients with autosomal dominant polycystic kidney disease (ADPKD) remain controversial, especially in patients who are candidates to renal transplantation (RT). The main surgical options such as unilateral vs. bilateral nephrectomy, nephrectomy before vs. after RT, or simultaneous nephrectomy and transplantation, are herein discussed.

Objective: Evidence acquisition of the best surgical management available for ADPKD in the context of kidney transplantation.

Acquisition of evidence: Systematic literature review in PubMed from 1978 to 2013 was conducted. Articles selected included: randomized controlled trials and cohort studies. Furthermore, well designed ADPKD reviews were considered for this study.

Synthesis of evidence: Laparoscopic nephrectomy in ADPKD is a safe procedure with an acceptable complication rate. Unilateral nephrectomy has advantages over the bilateral one regarding the perioperative complication rate. Although the timing of nephrectomy is controversial, it seems that simultaneous nephrectomy and renal transplantation neither increase surgical morbidity nor affect graft survival.

Conclusions: Simultaneous nephrectomy and RT appear to be an acceptable alternative to conventional two-stage procedure without any increased morbidity, in the context of ADPKD. Furthermore, laparoscopic nephrectomy performed in experienced centers is a safe alternative to conventional approach.

PALABRAS CLAVE
Poliquistosis renal autosómica dominante;
Nefrectomía;
Trasplante

Trasplante renal y poliquistosis: consideraciones quirúrgicas

Resumen

Contexto: La indicación y el momento de realizar una nefrectomía en pacientes con poliquistosis renal autosómica dominante (PQRAD) continúa siendo en la actualidad un tema controvertido, especialmente en aquellos pacientes candidatos a trasplante renal (TR). La nefrectomía uni o bilateral, el realizar la nefrectomía antes o después del TR, y la realización de la nefrectomía y el TR en el mismo acto son las principales opciones en la actualidad.

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Objetivo: Realizar una revisión de las diferentes opciones de manejo quirúrgico de la PQRAD en el contexto del TR, adaptada a la evidencia científica actual, y de otros aspectos que rodean la indicación.
Adquisición de la evidencia: Se realizó una revisión sistemática en PubMed (1978-2013), que incluye revisiones previas, estudios clínicos aleatorizados controlados y estudios de cohortes de los aspectos quirúrgicos de la PQRAD.
Síntesis de la evidencia: La nefrectomía laparoscópica en la PQRAD constituye una técnica segura y con un aceptable porcentaje de complicaciones. La realización de una nefrectomía unilateral presenta ventajas respecto a la bilateral en cuanto a un menor porcentaje de complicaciones perioperatorias. Aunque existe controversia en cuanto al momento en el que realizar la nefrectomía, parece que la realización en un mismo acto del TR y la nefrectomía no incrementa la morbilidad quirúrgica ni la supervivencia del injerto.
Conclusiones: En el contexto de la PQRAD, la realización de nefrectomía y TR simultáneo puede ser llevado a cabo sin que exista una morbilidad añadida con respecto al TR convencional. En caso de ser necesario la nefrectomía laparoscópica realizada en centros con experiencia es una alternativa segura al abordaje convencional.

Background

Autosomal dominant polycystic kidney disease (ADPKD) is a multisystem, monogenic disease characterized by the development of cysts in both kidneys and other extrarenal manifestations such as cysts in other organs and vascular, cardiac, digestive and musculoskeletal abnormalities, which develop at variable degrees.[1,2] ADPKD has a prevalence of 0.1–0.25% and is responsible for 10% of all cases of end-stage renal failure (ESRF).[3] ADPKD is a genetically heterogeneous disease in which 2 genes are involved: PKD1 (chromosome 16p13.3), responsible for 85% of cases, and PKD2 (chromosome 4q21-23), responsible for 15% of cases. Patients with PKD1 mutations have a more serious clinical presentation and have a higher tendency of developing ESRF at an earlier age (mean, 54 years). In contrast, individuals with PKD2 mutations develop ESRF later (mean, 74 years).[4] Cysts are present in 17% of cases at age 29–39 years and in 75% of those aged 70 years or more.[5] Overall, 50% of affected individuals progress to ESRF.[6]

Pathogenesis

The polycystin-1 (PC1) and polycystin-2 (PC2) proteins, which are associated with the membrane, are encoded by *PKD1* and *PKD2*, and their main function is the regulation of intracellular calcium. In the primary cilia, the polycystin complex receives and translates the mechanical stimulation into a calcium influx, which triggers an increased release of calcium from the endoplasmic reticulum. The reduction of one of the 2 polycystins below a critical level results in a phenotypic change that is characterized by the inability to maintain cell polarity, an increase in the rate of proliferation and apoptosis, the expression of a secretory phenotype and the remodeling of the extracellular matrix. The molecular mechanisms involved in these phenotypic changes include an alteration in intracellular calcium homeostasis, the activation of cyclic adenosine monophosphate (cAMP), tyrosine-kinase receptors, mammalian target of rapamycin (mTOR), the canonical Wnt pathway and other intracellular signaling mechanisms.[7] The formation of cysts occurs as a result of dilation of renal tubules. In the initial phases, the renal parenchyma presents a normal appearance, but in the final phases the kidneys are very large and have a distorted collecting system and numerous fluid-filled cysts. AHT is the most common clinical manifestation[8] and the main contributing factor to the disease progression. In addition, there is associated abdominal pain in 60% of cases.[9] Other forms of presentation include recurrent urinary infections, early satiety and hematuria. Overall, 50% of cases will present ESRF at 60 years of age, with a loss of 4.4–5.9 mL/min in the GFR.[10] Patients with PKD1 will experience ESRF at a lower mean age than patients with PKD2 (54.3 vs. 74.0).[11]

The purpose of current treatments is to limit morbidity and mortality due to complications of the disease. Kidney transplantation (KT) is the treatment of choice for ESRF in patients with ADPKD. The majority of patients with ADPKD do not require nephrectomy (unilateral or bilateral)[12,13]; however, in cases where there are space restrictions or symptoms, nephrectomy is required to facilitate KT. The timing and indication for nephrectomy in patients with ADPKD continues to be a controversial subject. Traditionally, nephrectomy before transplantation has been reserved for patients with a history of infected cysts, frequent haemorrhaging or a massive increase in size.[6,14]

Acquisition of evidence

A systematic review was performed in PubMed (1978–2013), which included prior reviews; randomized, controlled clinical studies; and cohort studies of the surgical aspects of ADPKD. The controversial issues that were assessed were the justification for nephrectomy, unilateral or bilateral nephrectomy and the timing of the nephrectomy (before, during or after the KT).

Justification for nephrectomy

Only 20% of the patients with ADPKD will require a nephrectomy.[12] The justification for indicating this procedure is based on the potential risk of presenting renal