Summer 2015

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Recommended Citation
Dendinger, Dana, "Raising Awareness: Polycystic Kidney Disease" (2015). Master of Science in Nursing (MSN) Student Scholarship. 70.
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Raising Awareness: Polycystic Kidney Disease
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Introduction
A young male patient was recently admitted to the intensive care unit at a local hospital with hypertension, kidney failure, and acute renal failure of unknown etiology. A renal ultrasound showed polycystic kidneys, and while performing the investigations, it was discovered that his mother passed away one year ago from stage renal failure related to polycystic kidney disease (PKD). The aforementioned information is recent, yet inherited disorders are what prompted further investigation into the patient’s medical history. Chang and Ong (2013) explain that “inherited cystic kidney diseases, autosomal dominant polycystic kidney disease (ADPKD) and autosomal recessive polycystic kidney disease (ARPKD), are the most common monogenic causes of end-stage renal disease (ESRD) in children and adults” (p. 1771). It was estimated that one in a thousand individuals will be diagnosed with PKD in adulthood and one in twenty will have ESRD (Liu et al., 2012, p. 1). Considering this statistic, patients with PKD may be at risk to develop complications related to renal failure. As a health care provider, primary or otherwise, it is important to educate and appropriately treat PKD patients in an effort to prevent unnecessary damage to their bodies related to complications of the disease. The utilization of genetic testing is not standard treatment; however, study into this may provide some benefits to certain at-risk patients in identifying the disease early on. As a result, early healthy lifestyle choices are necessary in order to slow disease progression.

Pathophysiology
Eisenberger, et al. (2015) state that “renal cysts are clinically and genetically heterogeneous conditions” (p. 1). ADPKD is the most commonly diagnosed form of PKD and is typically diagnosed during adulthood; it results from a mutation in either of two genes, PKD1, which encodes polycystin 1 (PC1), or PKD2, which encodes polycystin 2 (PC2), both of which are membrane proteins (Reed, Gitomer, 2014, p. 17). Srivastava & Patel (2014) explain that “mutations of PKD 1 (eight-five percent of cases) or PKD 2 (fifteen percent of cases) can lead to signal dysregulation and increased levels of intracellular calcium, monophosphorylating, culminating in cytogenesis. Given the dominant nature of inheritance, there is a fifty-three percent probability that a child of an affected parent will inherit the disease. A spontaneous mutation causes ADPKD in five percent of cases” (p. 303). It is difficult to determine genetic makeup for ADPKD early on due to the size and structures of PKD1 and PKD2 (Chang and Ong, 2013). Since the detection of the aforementioned genetic mutations can be difficult, genetic testing can provide definitive diagnosis of PKD is based on two factors, a positive family history and the eventual lead to progressive cystic growth, renal fibrosis, increased systolic blood pressure, and the retention of electrolytes” (Halvorson, Bremmer, & Jacobs, 2010, p. 74). The detection of the renal parenchyma and the ratio of function nephrons that result from this process is irreversible. The normal function of polycystin 1 and 2 also plays a role in the manifestations, due to the fact that these gene products may be expressed in the vascular endothelium and smooth muscle. Polycystin 1 and 2 function as a receptor-ion channel complex on the membranes of cells, resulting in abnormalities. The polycystins are membrane proteins (Reed, Gitomer, 2014, p. 170). When the polycystins are present with defects, intracellular sodium, calcium, and magnesium homeostasis is affected which produces alterations of endothelium-dependent relaxation and increased systolic blood pressures” (Halvorson, Bremmer, & Jacobs, 2010, p. 74).

Current research suggests that the primary reason behind renal cyst formation in both ADPKD and ARPKD is related to defects in cilia-mediated signals (Halvorson, Bremmer, & Jacobs, 2010). The gene products, polycystin 1 and 2, that are affected in ADPKD, regulate the growth of the epithelium in the renal tubules. The genetic disorders associated with PKD affect the formation of the epithelial cells and adenosis, resulting in the formation of cysts. The presence of cilium is found on almost all surfaces of nephron cells, which means that the cysts have the potential to form on these areas as well. In PKD, the “function of the primary cilium is impaired, resulting in the disruption of a number of intracellular signaling cascades that produce dedifferentiation of cyst epithelium, increased cell division, increased apoptosis, and loss of regenerative capacity” (Halvorson, Bremmer, & Jacobs, 2010, p. 73). Additionally, polycystic kidneys and increased intracellular calcium and extracellular matrix defects result in kidney enlargement and interstitial fibrosis” (p. 524).

Clinical Manifestations
The enlarging kidneys of PKD progress over decades, which leads to renal failure in the majority of patients by age sixty (Ma, Tian, Igarashi, Pazour, & Somlo, 2013, p. 1054). Many patients are asymptomatic until later in life; most are diagnosed between the ages of thirty and forty. The gradual loss of nephrons related to the formation and increasing size of cysts is not usually detectable during the first few decades of life (Srivastava & Patel, 2014, p. 303). The presentation of the disease varies based on the presence of other comorbidities. Early signs or symptoms include:

- Flank or abdominal pain
- Macroscopic hematuria
- Frequent urinary tract infections (UTIs)
- Early-onset hypertension

If patients are at a higher risk for:
- Renal stones
- Intracranial aneurysms
- Gross hematuria
- Abdominal manifestations, such as cysts that may appear on surrounding organs
- Renal dysfunction
- Cardiovascular complications

associated with hypertension

Implications for Nursing Practice
As a primary health care provider it is important to obtain a detailed family history. Patients may have to go through a full evaluation of the patient, including signs, symptoms, social behaviors, diet, and exercise, in order to determine the care provided to PKD patients is education; key topics include exercise, diet, and medication compliance. Adequate exercise and an appropriate diet are important components in almost any disease process. However, due to the fact that PKD patients may have a diet that is low in cholesterol to reduce cardiovascular risks, as well as high in potassium, it is important that patients be educated that they should not be consuming too many potassium rich foods. Patients who have PKD are at a higher risk of hypertension. However, research has shown that PKD patients to monitor their blood pressure frequently and maintain medication compliance in regards to hypertension, if applicable. If hypertension has progressed to ESRD, patients will require hemodialysis and/or a renal transplant. However, over the last few decades, due to advancements in clinical trials with regards to slowing the progression of PKD have made advancements in the field (Sitomer, 2014, p. 177).

Conclusion
In the grand scheme of diseases, PKD is considered rare. However, once it is diagnosed the complications that can/will result from it are life threatening. PKD is frequently misdiagnosed because many of the signs and symptoms that go along with it are similar to other ailments/diseases, such as primary hypertension or a typical UTI. Once a patient is diagnosed with PKD, he or she must be educated to take preventive measures of complications that may arise, and diet recommendations. Efforts must be made to maintain the cardiovascular system as well, including smoking cessation and exercise routines. If it is determined that a patient has PKD, it may be beneficial for the patient to map/computer tomography his on his siblings as well as his or her children if applicable. Renal pain is often a symptom of PKD; however, a quick way to get a patient that is up to him or her and their primary care provider.

References


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