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Gastrointestinal Tract Complications in Autosomal Dominant Polycystic Kidney Disease: A Brief Review

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1. Abstract

Autosomal Dominant Polycystic Kidney Disease (ADPKD) represents the most common hereditary nephropathy and is increasingly recognized as a multisystem disease with substantial gastrointestinal (GI) involvement. Extrarenal GI complications include polycystic liver disease (PLD), diverticular disease, abdominal wall hernias, pancreatic cysts, biliary tract abnormalities, and mechanical intestinal compression syndromes. Notably, PLD stands as the most common extrarenal manifestation, affecting up to 90% of the adult ADPKD population. The pathogenesis involves abnormalities in epithelial signaling, extracellular matrix integrity, and connective tissue architecture associated with PKD1 and PKD2 mutations. Recognition of GI manifestations is crucial because a delayed diagnosis may be catastrophic, particularly in kidney transplant recipients and patients with advanced disease. This review comprehensively summarizes the epidemiology, pathogenesis, clinical manifestations, and management strategies of gastrointestinal complications in ADPKD.

2. Introduction

Autosomal Dominant Polycystic Kidney Disease (ADPKD) affects approximately 1 in 400–1000 individuals worldwide and accounts for a significant portion of the end-stage renal disease population requiring renal replacement therapy [1-5, 8]. Although progressive renal cyst formation is well known as its defining hallmark, ADPKD is fundamentally a systemic ciliopathy that involves multiple organs including the liver, pancreas, cardiovascular system, and gastrointestinal tract [1, 7, 9, 10]. The current clinical framework for understanding ADPKD as a multiorgan systemic disorder is rooted in landmark studies from the 1990s, which redefined the disease far beyond its localized renal manifestations [11]. Mutations in PKD1 and PKD2 genes lead to the dysfunction of polycystin proteins, resulting in deregulated epithelial proliferation, disrupted intracellular calcium homeostasis, and subsequent cyst formation [3, 5]. Gastrointestinal manifestations significantly contribute to morbidity, hospitalization, impaired quality of life, and mortality [1, 2]. These complications typically arise from direct cystic involvement of abdominal organs, connective tissue abnormalities, or mass effects from enlarged kidneys and liver cysts [2, 12].

3. Polycystic Liver Disease

Polycystic liver disease (PLD) constitutes the most common extrarenal feature of ADPKD identified in approximately 80–90% of older adults with the disease [2, 4, 13]. Risk factors such as female sex, multiparity, and exogenous estrogen exposure are associated with exacerbated liver involvement [4, 14]. Although most hepatic cysts remain clinically asymptomatic, progressive hepatomegaly can cause abdominal fullness, early satiety, gastroesophageal reflux, dyspnea, and chronic abdominal pain [2, 4, 15]. Severe hepatomegaly may compress adjacent vascular structures, leading to portal hypertension or inferior vena cava obstruction [2]. Complications include cyst hemorrhage, rupture, and infection [2, 14]. Hepatic cyst infection commonly presents with fever, abdominal pain, leukocytosis, and elevated inflammatory markers [2]. 18F-Fluorodeoxyglucose Positron Emission Tomography (FDG-PET) imaging may improve diagnostic sensitivity for infected cysts [2]. Therapeutic interventions range from conservative treatment, percutaneous aspiration with sclerotherapy, laparoscopic fenestration, somatostatin analogues, to hepatic resection and, in severe cases, liver transplantation [4, 14, 15, 16]. The selection of therapeutic interventions depends largely on the morphology and distribution of the cysts, often classified using systems such as the Gigot or

Schnelldorfer criteria [15].

4. Diverticular Disease

Colonic diverticulosis is observed at a higher frequency in the ADPKD population compared to the general public [1, 6, 17]. Broad clinical evidence has confirmed that this increased prevalence is a direct consequence of the underlying genetic mutations that compromise connective tissue integrity throughout the gastrointestinal tract [17, 18]. Connective tissue abnormalities and altered extracellular matrix integrity are believed to weaken the colonic wall and predispose patients to diverticula formation [2, 9, 18]. While uncomplicated diverticulosis is often asymptomatic, diverticulitis may become severe, especially in kidney transplant recipients receiving immunosuppressive therapy [6]. Complications include colonic perforation, peritonitis, abscess formation, and sepsis [6]. Clinicians should maintain a high index of suspicion and a low threshold for abdominal imaging and aggressive intervention in ADPKD patients presenting with unexplained abdominal pain or fever [1, 6]. Management includes antibiotics, bowel rest, drainage procedures, and surgical intervention when necessary [6].

5. Abdominal Wall Hernias

Abdominal wall hernias-specifically umbilical, inguinal, and incisional hernias-are highly prevalent in ADPKD [2, 9]. The pathogenesis is multifactorial, involving connective tissue weakness coupled with chronically elevated intra-abdominal pressure caused by enlarged kidneys and liver cysts [2, 7]. These hernias may impair functional status and may jeopardize peritoneal dialysis access [7]. Surgical repair with mesh is generally preferred, considering the higher recurrence rates in the ADPKD population [2].

6. Pancreatic and Biliary Manifestations

Pancreatic cysts are identified in approximately 10–20% of ADPKD patients, and are usually asymptomatic [2, 3]. Rare complications include pancreatitis, infection, and pancreatic duct obstruction [2]. Biliary tract abnormalities include bile duct dilatation, cholangitis, and biliary obstruction caused by large hepatic cysts [2, 12]. Differentiating cholangitis from hepatic cyst infection may be clinically challenging [2].

7. Mechanical Gastrointestinal Complications

Massive enlargement of the kidneys and liver cysts may directly compress gastrointestinal structures [2, 4]. Patients commonly experience early satiety, gastroesophageal reflux, nausea, dyspepsia, and reduced oral intake [2]. Severe hepatomegaly may contribute to malnutrition and sarcopenia [4]. Rare cases of bowel obstruction secondary to a giant cyst burden or distorted abdominal anatomy have been reported [2, 12]. Nutritional assessment and multidisciplinary management are, therefore, essential in advanced disease [4].

Pain and Quality of Life

Chronic abdominal pain is common in ADPKD and may result from liver cyst expansion, renal cyst hemorrhage, abdominal wall hernias, gastroesophageal reflux, or diverticular disease [19]. Such pain significantly impairs quality of life and frequently requires multidisciplinary management [19]. Treatment strategies include analgesics, cyst decompression procedures, physical therapy, and psychological support [19].

8. Summary

Figure 1 showed common gastrointestinal tract complications in autosomal dominant polycystic kidney disease. ADPKD is a systemic disease with substantial gastrointestinal involvement [1, 9]. Polycystic liver disease persists as the most prevalent GI manifestation, while diverticular disease, abdominal wall hernias, pancreatic cysts, biliary abnormalities, and intestinal compression syndromes contribute significantly to clinical morbidity and impaired quality of life [1, 2, 4]. Proactive recognition and multidisciplinary management are essential to optimize patient outcomes and mitigate GI-related complications, particularly in transplant recipients and patients with advanced disease [6, 7]. Current KDIGO guidelines emphasize that managing these extrarenal manifestations requires a transition to a multidisciplinary care paradigm [20]. Moreover, the integration of targeted molecular therapies and specialized imaging biomarkers is now central to monitoring and mitigating systemic disease progression [21, 22]. Future research should focus on advancing biomarkers, targeted therapies, and quality-of-life interventions for the gastrointestinal manifestations of ADPKD [3, 21, 22].

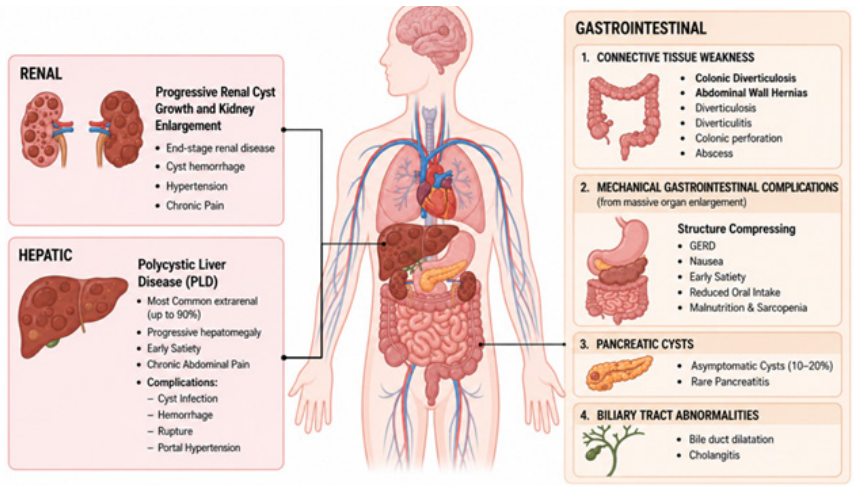

Figure 1: Common gastrointestinal tract complications in autosomal dominant polycystic kidney disease.

9. Conclusion

In conclusion, gastrointestinal tract complications are increasingly recognized as important extrarenal manifestations of ADPKD and can significantly affect patient quality of life and clinical outcomes. Common complications include diverticular disease, abdominal hernias, hepatic cyst-related symptoms, constipation, gastroesophageal reflux, and, in severe cases, bowel obstruction or cyst infection. These manifestations often arise from increased intra-abdominal pressure, connective tissue abnormalities, and progressive enlargement of renal and hepatic cysts. Early recognition and multidisciplinary management are essential to reduce morbidity and improve patient care. Clinicians should maintain a high index of suspicion for gastrointestinal symptoms in patients with ADPKD, particularly in advanced disease stages or after kidney transplantation. Appropriate diagnostic evaluation, lifestyle modification, medical therapy, and timely surgical intervention when necessary can help minimize complications. Further research is needed to better understand the pathophysiological mechanisms linking ADPKD with gastrointestinal disorders and to develop targeted therapeutic strategies that enhance long-term outcomes and patient well-being.

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