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# A RARE ASSOCIATION OF CHOLEDOCHAL CYST WITH ADULT POLYCYSTIC KIDNEY DISEASE

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| **Dr. M. N. Karthikeyan** | Postgraduate, Department of Medicine, Kurnool Medical College, Kurnool-518002. A.P. |
| **Dr. A. Gopi Krishna*** | Postgraduate, Department of Medicine, Kurnool Medical College, Kurnool-518002. A.P. *Corresponding Author |
| **Dr. P. Vidyadhar** | Postgraduate, Department of Medicine, Kurnool Medical College, Kurnool-518002. A.P. |

**ABSTRACT** Combination of different types of choledochal cysts are very rare, seldom reported in literature. Their association with polycystic kidney disease has been never reported before. Caroli's disease is the most commonly associated fibrocystic disease with polycystic kidney disease. Here, we present an a middle aged woman, in whom, evaluation for led to diagnosis of Type IV associated with polycystic kidney disease.

**KEYWORDS :** choledochal cysts, Biliary tract, Polycystic Kidney Disases, Choledochocele

## INTRODUCTION

Choledochal cyst is thought to be a type of hepatobiliary fibropolystic disease and it have been recognised to be caused by pancereati cobiliary maljunction. Polycystic kidney disease is often associated with polycystic liver disease and carol is intrahepatic biliary dilation. It is rarely reported to be associated with choledochal cyst. We describe a case of 56 yrs old female with rare association of polycystic kidney disease with choledochal cyst without pancreaticobiliary maljunction.

## CASE REPORT

A 56yrs old female presented to OPD with complaint of vomiting, high grade fever, decreased urine output, hematuria, right hypochondrial pain since one week. On examination patient is conscious and coherent, icterus present, asterixis present. On per abdomen examination tenderness in right hypochondrial region noted. On USG abdomen reveals multiple polycystic kidney in both kidneys. On computed tomography cystic lesion of size 5x3.8 in right lobe of liver with communication between common bile duct and cyst, dilation of common bile duct measuring 24mm suggestive of choledochal cyst multiple hypo dense cystic lesion of varying sizes and few of them showing hyper density in bilateral kidney. On MRCP both intrahepatic and extra hepatic dilatation are seen without pancreaticobiliary maljunction. Laboratory date were serum total bilirubin 12.8mg/dl, direct:7.1mg/dl, indirect:5.7mg/dl, AST:110µ/I, ALT: 105µ/I, Alkaline phosphate:2.5µ/I, Serum Creatinine :7.4mg/dl, Total WBC count 19000 cells/cumm.

## DISCUSSION

Polycystic kidney disease and polycystic liver disease is a tpical combination of fibropolycystic disease. However association of choledochal cyst and polycystic kidney disease is extremely rare. Babbit's theorem suggest that malformation of pancreaticobiliary system is an important cause of choledochal cyst mostly, the presence of a long common channel with anomalous insertion. He proposed that reflux of pancreatic enzymes into bile ductresulted in continued inflammation, weakening of duct wall and cystic dilatation of biliary tree.

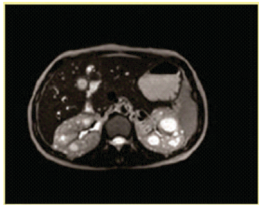

**Fig-1**

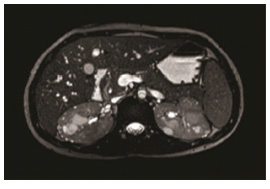

**Fig-2**

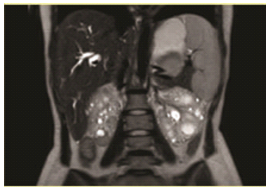

**Fig-3**

This theory fails to explain isolated ductal dilatation seen with Type III and Type V CC due to sphincter of oddi dysfunction. Embryoogically the hepatobiliary abnormalities are thought to stem from ductal plate mal development in different parts of biliary tree. So far only 6 cases of type IV choledochal cyst associated with adult polycystic kidney disease is reported.

## CONCLUSION

In conclusion, association of choedochal cyst and adult polycystic kidney disease is very rare. In the case of choledochal cyst combined with real fibropolystic disease, pancreaticobiliary maljunction may not contribute to etiology of choledochal cyst.

## REFERENCES

1. Wechsler, R.L. ad Thiel V.D (1976), Fibropolycystic disease of the hepatobiliary system and kidney's Digestive Disease, 21, 1058-1069.
2. Summerfield, J.A., Nagafuchi, y.Sherlock S. Cadafalch.J.and Scheuer, P.J.(1986), Hepatobiliary fibropolycystic diseases: A clinical and histological review of 51 patients, Journal of Hepatology,2,141-156.
3. Komi N, Takehara H, Kunitomo K, Miyoshi Y, Yagi T.Does the type of anomalus arrangement of pancreaticobliliary ducts influence the surgery and prognosis of choledochal cyst? Pediatr Surg.1992;27(6): 728-31
4. Alonso-Lej F, Rever WB Jr, Pessagno DJ, Congenial choledochal cyst, with a report of 2, and an analysis of 94 cases. Int absr Surg. 1959;108(1): 1-30.