Introduction:

Choledochal cysts are congenital anomalies of the bile ducts. They consist of cystic dilatations of the extrahepatic biliary tree, intrahepatic biliary radicles, or both. The first anatomic study of a choledochal cyst in the Western literature was published by Vater and Ezler in 1723. ***Alonso-Lej et al 1959*** provided the first systematic description of choledochal cysts based on the clinical and anatomic findings in 96 cases. The resultant system classified choledochal cysts into 3 types and outlined therapeutic strategies for each. The classification system for choledochal cysts was further refined by ***Todani et al 1977*** and currently includes 5 major types. ***(Kumar,et al;2005).***

Choledochal cysts are exceedingly rare in the general population, and reports of familial occurrence are scarce. Choledochal cysts occur more commonly in female than male individuals (with a ratio of 3–4:1), and the incidence in Western nations (Europe and North and South America) is 1 in 100,000 to 150,000 live births. Although classically considered a disease of infancy, an increased number of patients are being discovered in adulthood. **(Shi,et al;2001).**

Familial case reports of congenital choledochal cysts are extremely rare. Choledochal cysts occur most commonly in patients of Asian descent, with more than two thirds of the reported cases occurring in Japanese patients. **(Iwata,et al;1998).**

Aim of the Essay :

Is to review the incidence of choledochal cysts and associated anomalies and to discuss the different methods in the diagnosis and surgical modalities of its treatment.