

# Case Presentation on Klatskin's Tumour

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# Leena J.

## **Abstract**

This is a case presentation on Klatskin's tumour. He is aged 47 admitted to Gastroenterology ward with the complaints of progressive abdominal distention over a period of one month, with pain in right upper quadrant. His abdominal girth confirmed distention due to ascites and thrill on ballottement. He had hypoalbuminemia in diuretic stage but his Serum Protein levels were within normal limits. He had stent fixation markings and scar due to radiation therapy on his abdomen. His Alkaline Phosphatase indicated an obstruction of biliary system. Paracentesis confirmed infection. He was diagnosed as Hilar Cholangio Carcinoma i.e., Klatskin's Tumor with Stenting and Post Radiation Therapy with Reactive Mesothelial Proliferation. He was found to have Liver Secondaries (Unresectable Tumor) since the growth extended to the under surface of the liver. ERCP was attempted twice and precut was extended. Liver enzymes were given along with symptomatic treatment. His activities were gradually increased as he could tolerate. The case presentation is extended with the application of nursing process as well.

Key words: Klatskin's tumor, Paracentesis; Stent; As cites.

## Introduction

Anatomy and physiology (bile duct)

The bile duct is long tube-like structure that connects the liver to the intestine and transports bile from the liver to the intestine. The top half of the bile duct is associated with the liver while the bottom half of the bile duct is associated the pancreas. The bile duct enters the part of the intestine called the duodenum into a structure called the Ampulla. Blockage of the bile duct causes build up of the bile in the blood since the bile can no longer go into the intestine. This condition is called jaundice and the skin becomes yellow from the accumulated bile in the blood.

# Bile duct cancer

Bile duct cancer or Cholangio carcinoma are tumors that occur in the bile duct. Bile duct cancer usually develops in patients older than 65 years old. Types of bile duct tumors

There are several suggested classifications. Clinically, Intra hepatic and Extra hepatic Cholangio Carcinomas are different entities, but they are linked by cell of origin and etiological factors. Intra hepatic Cholangio Carcinomas arises from small ducts or ductules and presents as an intra hepatic mass. Extra hepatic Cholangio Carcinomas arises from large ducts and usually presents as biliary tract obstruction and further subdivided in to upper duct tumors or Peri Hilar or Klatskin's tumors and lower duct tumors (Feldman, 2002).

# **Courtesy: Cancer Help UK**

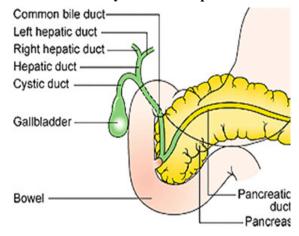


Diagram showing the position of the bile ducts

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# **Classification of Primary Liver Neoplasms**

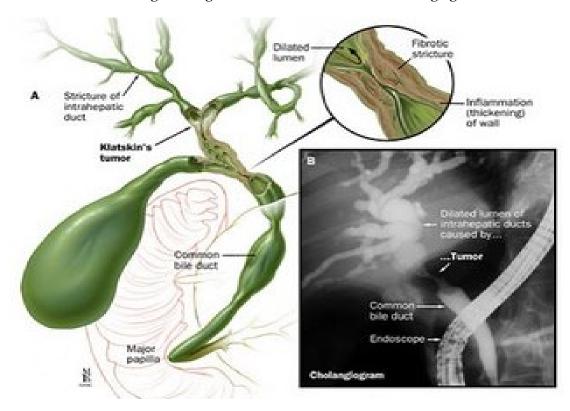
ORIGIN	BENIGN	MALIGNANT
Hepatocytes	Adenoma	Hepato cellular
		Carcinoma
Connective	Fibroma	Sarcoma
tissues		
Blood vessels	Hemangioma	Hemangio
		Endothelioma
Bile ducts	Cholangioma	Cholangio sarcoma

removal of the tumor together with a liver resection (removal) in an attempt to provide a surgical cure.

# Klatskin's tumors

Bile duct cancer of the upper part of the bile duct is also called Klatskin's tumor. Klatskin's tumors involve the upper part of the bile duct as divides to enter the right and the left parts of the liver. The bile

# Radiological Diagnosis of Klatskin"S Tumor- Cholangiogram



Courtesy: Dept of Radiology, Kyungpook National University Hospital, Knuhrad

Two major types of bile duct tumors are found

# Distal bile duct tumors

Tumors affecting the bottom half of the bile duct

# Klatskin's tumors

Tumors affecting the upper part of the bile duct

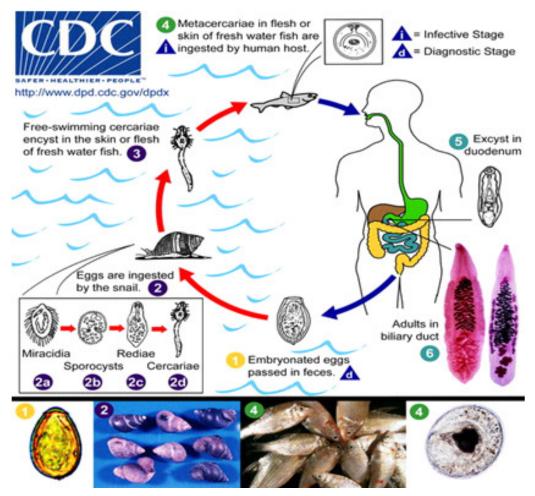
This separation between the two locations is important since the treatment for the tumors in the two locations is different. For tumors affecting the bottom half of the bile duct, the cancer is removed with a Whipple operation. For tumors in the top half of the bile duct, surgical treatment often requires

ducts in the liver are called right and left hepatic ducts. The tumor may involve one or both right and left sides of the hepatic ducts as they enter the liver. The hepatic ducts are closely associated with the blood vessels that supply blood to the liver. Klatskin's tumors are closely associated with liver and as they grow invasion into the blood vessels that supply blood to the liver is often found.

# Cholangio carcinoma

Cholangio Carcinoma is an adeno carcinoma of the intra hepatic bile ducts (Yamada, 2009). Cholangio Carcinoma originates from small intra

# Life Style of Clonorchis Sinensis, a Liver Fluke Associated with Cholangio Carcinoma



Courtesy: www.answers.com, www. Science codex.com

hepatic bile ducts – peripheral Cholangio carcinoma, large intra hepatic bile ducts – hilar Cholangio carcinoma or Klatskin's tumor and extra hepatic ducts- Bile duct carcinoma.

Epidemiology/Etiology (Fledman, M., Friedman, S.L. and M.H. Sleisenger (2002).

# It includes

- Long standing sclerosing cholangitis (7%) (Fledman,M., Friedman, S.L. and M.H. Sleisenger(2002)
- Biliary atresia (Fledman, M., Friedman, S.L. and M.H. Sleisenger (2002)
- Biliary cirrhosis (Sheila Sherlock,2002)
- Biliary dysplasia (10%)
- Intra hepatic cholelithiasis (Fledman, M., Friedman, S.L. and M.H. Sleisenger (2002)

- Post radio graphic contrast and medium Thoridium Dioxide (Fledman, M., Friedman, S.L. and M.H. Sleisenger (2002)
- X anitrypsin deficiency leads to Thorotrast related Cholangio carcinoma (Fledman, M., Friedman, S.L. and M.H. Sleisenger (2002)
- Common in older than young, between 50 & 60 years (Yamada Tadataka, (2009) Common in men than in women (Fledman, M., Friedman, S.L. and M.H. Sleisenger (2002)
- Colorectal neoplasia (Sheila Sherlock, 2002)
- Irritable bowel disease (10%)
- Assisted with typhoid career state to Hepato Biliary cancer according to New York Cancer Association
- More common in persons with Ulcerative Colitis than with general population (10%)

- Liver Fluke infestations (20%) due to
  - √ Clonorchis sinesis (parasite usually common in Hongkong, China, Japan.Korea by eating raw fish containing the larval stage of flukes (Yamada, 2009)
  - √ Opisthorchis viverrini common in Thailand, Laos, Western Malaysia –induces DNA changes and Mutation through the production of carcinogens, free radicals and stimulation of cellular proliferation of intra hepatic duct epithelium
  - √ Increased Nitrate levels in body fluids (Yamada Tadataka,(2009)
- After removal of Gall stones –unknown (Yamada Tadataka, (2009)
- Congenital fibro poly cystic conditions (Sheila Sherlock, 2002)
  - $\sqrt{}$  Congenital hepatic fibrosis
  - √ Cystic dilatation (Caroli's syndrome)
  - √ Choledocal cyst (10%) due to untreated anomalously high pancreatic duct-Bile duct junction (Fledman,M., Friedman, S.L. and M.H. Sleisenger(2002)
  - √ Polycystic liver (Sheila Sherlock, 2002)
  - √ von Meyenburg complex (Fledman,M., Friedman, S.L. and M.H. Sleisenger(2002)

# Pathophysiology (Sheila Sherlock, 2002)

The confluence of cystic duct with main hepatic duct or the right and left main hepatic duct at the porta hepatics are common sites of origin. The tumor extends to liver. It causes complete obstruction of extra hepatic bile ducts with intra hepatic biliary dilatation and enlargement of the liver. The gall bladder is collapsed and flaccid. If the tumor is restricted to one hepatic duct, biliary obstruction is incomplete and jaundice absent. The lobe of the liver drained by obstructed duct atrophies. The other duct hypertrophies.

In the common bile duct, the tumor presents as a firm nodule or plaque which causes an annular stricture which may ulcerate. It spreads along the bile duct and through its wall. It also involves peritoneum, abdominal lymph nodes, diaphragm, liver and gall bladder. The tumor encircles the bile duct.

Blood vessel invasion is rare and extra abdominal spread is unusual. Compression of the portal vein can lead to lobar atrophy. Histologically the tumor is usually mucus secreting adeno carcinoma with cuboidal or columnar epithelium and abundant fibrous stroma.

Spread along neural sheaths may be noted. Metastatic nodules distributed irregularly through out liver and radiates into hepatic tissue or a spongy friable mass within the lumen of the duct. Bile production is not seen. The tumor cells provoke a variable desmoplastic reaction and presents with collagenized stroma (Sheila Sherlock, 2002)

# Clinical Features (Sheila Sherlock, 2002)

- Usually deep jaundice is present followed by pruritis. There is a point of distinction from primary biliary cirrhosis where itching usually comes first.
- Jaundice may be delayed if only one main duct is involved, so there is an increased Serum Bilirubin
- Pain in the epigastric region in one third of patients is seen.
- There may be diarrhea/ steatorrhea, weakness and weight loss
- It is associated with ulcerative colitis and long standing cholestasis due to sclerosing cholangitis
- They are afebrile at terminal stage
- The liver may be large, smooth, extending 5to 12 cm below costal margin.
- Spleen may not be palpable
- Ascites is unusual (Sheila Sherlock, 2002)

# Investigations (Sheila Sherlock, 2002)

- Serum Biochemical findings
  - √ Increased Serum Bilirubin
  - √ Increased Alkaline Phosphatase
  - √ Increased Y Glut amyl Trans Peptide levels
- Serum Mitochondrial Antibody test Negative
- Alpha feto Protein not increased
- Feces may be pale, fatty, positive occult blood
- Glycosuria will be present

- Severe Anemia may be present
- WBC shows increased Leukocyte Count and Increased Polymorphs
- Cytology taken at Hepatic biopsy shows large bile duct obstruction
- Attempt for ERCP/ Percutaneous Drainage confirm bile duct obstruction
- Scanning
- USG shows Intra Hepatic Bile duct, echogenic tumor at hilum and extension in and around bile duct (80%)
- CT scan shows Intra Hepatic Biliary dilatation, isodensed tumor
- Enhanced CT- Hilar Cholangio Carcinoma high density map bile duct dilatation detects tumor (40-70%)
- Helical CT-detects Cholangio carcinoma tumor size as small as 15 mm in diameter, in 90% cases
- MR imaging useful to confirm bile duct stones, strictures (90%)
- Endoscopic Percutaneous Cholangio Graphy (ERCP)
- ERCP trans papillary forceps biopsy obstruction at hilum
- Endoscopic ultra sound to assess the extend of tumor at the lower end of the bile duct

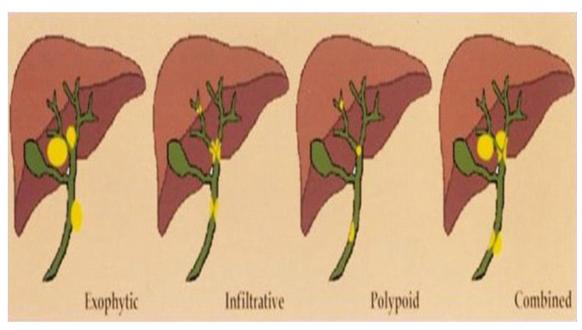
- Percutaneous obstruction is blunt on nipple like dilated intra hepatic bile duct accurately
- Demonstrates filling defects of the mid bile duct due to a polypoid carcinoma

# Prognosis (Sheila Sherlock, 2002)

Fatal but the tumor is slow growing and metastizes late so survival seems to the long. Mean survival 14.4 months to 5.6 years. The tumor kills by its site, making it inoperable rather than by its malignancy. Death is due to Hepato cellular failure and infection usually suppurative cholangitis and septicemia. Massive invasion of the liver by tumor or extra hepatic metastases rarely causes death. Prognosis depends on the site of tumor. Those distally placed are more likely to be resectable than those at the hilum. The histologically differentiated ones do better than the undifferentiated. Polypoid cancers have the best prognosis.

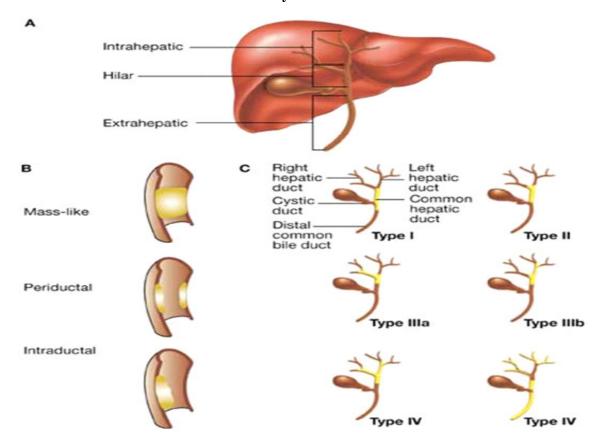
# Staging (Sheila Sherlock, 2002)

Low Common Bile duct lesions are resectable and angiography, venography are needed to exclude vascular invasion. Hilar Cholangio Carcinomas are problematic especially when secondary hepatic duct is involved in both hepatic lobes, angiography confirms with encasement of main portal vein, hepatic



Courtesy: Amit Sunny Mittal, Baylor college of Medicine www.bcm.edu/osa/radilogy club

# Bismuth's Cholangio carcinoma classification Courtesy: Tushar Patel



artery the lesion is resectable. A palliative procedure is needed. Lesion is resectable when the tumor is limited to hepatic duct bifurcation; affects one lobe of liver, only obstructs portal vein or hepatic artery.

Stage I – III Resectable tumor depends on angiographic findings

Stage IV Bilateral involvement of secondary hepatic duct – incurable disease

(Feldman, Friedman, 2002)

# Treatment of Klatskin's Tumor

http://www.surgery.usc.edu/divisions/tumor/pancreasdiseases/web%20pages/BILIARY%20SYSTEM/cholangiocarcinoma.html

# Surgery

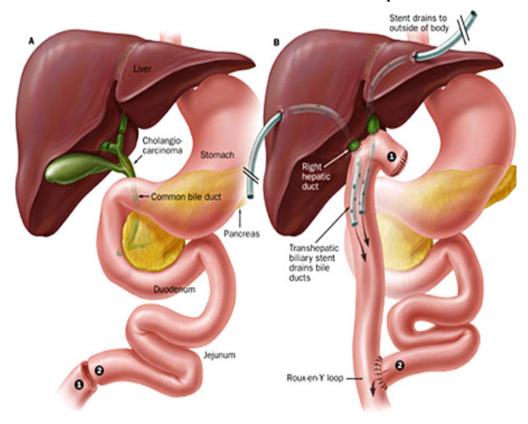
The liver is made up of two lobes: a right lobe and a left lobe. One of the two lobes can be safely removed at surgery. Klatskin's tumors often invade the blood vessels called hepatic artery and the portal vein that supply blood flow to the liver. The goal of staging prior to surgery is to assess whether the blood vessels of the liver are free of the tumor. The location of proximal bile duct tumors sometimes makes this evaluation difficult and often the final decision regarding surgery is made at the time of exploratory surgery.

Complete removal of the tumor is the only effective and potentially curative treatment for cancers of the upper bile duct. The treatment usually requires a surgical procedure to remove the tumor in the bile duct together with one side of the liver due to the high frequency with which the tumor invades blood vessels of the liver.

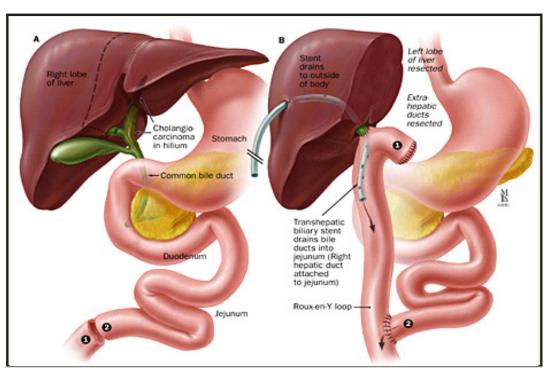
# A. Klatskin's tumors are removable if:

http://www.surgery.usc.edu/divisions/tumor/pancreasdiseases/web%20pages/BILIARY%20SYSTEM/cholangiocarcinoma.html

# A, B. Surgical technique for Bilateral Hepato Jejunostomy with Roux-en-Y Anastamosis for the Removal of an Extra Hepatic Tumor



A, B. Surgical Technique for Unilateral Hepato Jejunostomy with Roux-en-Y Anastamosis and Left Hepatic Lobectomy



# Cholangiocarcinoma in distal common bile duct causing obstruction Duodenom jejunostomy Pancreaticojejunostomy Pytorus Pancreaticojejunostomy

# A, B. Surgical technique for bilateral Hepato Jejunostomy with Roux-en-Y and the removal of an extra hepatic tumor

Courtesy: images from qlum.blog.hexun.com

Blood supply to one side of the liver is not affected by the tumor

Klatskin's tumors are closely associated with liver and as they grow invasion into the blood vessels that supply blood to the liver is often found. If the blood supply to one side of the liver is free of tumor then the portion of the liver invaded by the tumor can be removed.

The bile duct to one side of the liver is free of tumor

Klatskin's tumors affect the portion of the bile duct in the liver. One or both side of the bile ducts in the liver may be affected.

# B.Unresectable Klatskin's tumor:

Klatskin's tumor is unresectable if it invades the blood supply to both sides of the liver and/or the hepatic duct to the both sides of the liver therapy treatments is used. In general these tumors respond poorly to treatment. For Tumors of the middle third of Extra Hepatic duct, surgical options include Resection of the Mass possible primary End-to-End Bile duct Anastamosis (for early Small tumors) or Hepato Jejunostomy (If Large portion of Extra hepatic ducts should be removed)

Proximal tumors may be resectable by local or major liver surgery including Excision of the whole Bifurcation of the Common Bile Duct, Lobectomy, and Bilateral Hepato Jejunostomy. The liver may need to be split back to vena cava.

Early diagnosis of a peripheral Cholangio carcinoma is unusual and the tumor carries the poor prognosis. Resection is rarely possible, and the results of radiation therapy and chemotherapy are disappointing. For inoperable cases, biliary drainage must be established, usually be an endoscopic or radio graphic approach (Feldman, Friedman, 2002)

# Palliative surgical procedures

Intubation of stricture with the use of hepatic tubes.

Anastamosis of jejunum to III segment duct in the left lobe which is usually accessible despite the hilar tumor.

# Non Surgical Procedures

In patients who are with surgery or with unresectable tumors—placed Endo Prosthesis across the stricture either by Endoscopic or Percutaneous routes

# Endoscopic stent

- 1. Complication
- i. Cholangitis (7%)
- ii. III day mortality (10-28%)

Mean Survival Rate – 20 weeks

Percutaneous Trans Hepatic Endo Prosthesis insertion.

# Complication

- i. High risk for Puncture of Liver
- ii. Risk of bleeding
- iii. Bile leakage

Internal Radio Therapy -192 Iridium wire or Radium needle.

Implant with biliary drainage.

Cytotoxic drugs are ineffective.

External radio therapy –beneficial.

Symptomatic treatment for chronic cholestasis.

# Hepatic Transplantation

- a. Recurrence of tumour is unusual.
- b. Given poor results (retrospective studies).

# Post Operative Complications (reference)

# Major

- 1. Pneumonia
- 2. Hepatic insufficiency
- 3. Intra abdominal fluid collection
- 4. Intra abdominal bleeding
- 5. Myocardial infarction

Minor

- 6. Biliary leakage
- 7. Pleural effusion
- 8. Atelectasis
- 9. Urinary tract infection
- 10. Wound infection

# Case history

47 years old, Mr Velusamy a land lord hailing from Coimbatore, a father of three sons, was admitted into Gastroenterology ward with the complaints of progressive abdominal distention over a period of one month, with pain in right upper quadrant. The pain was significant especially after consumption of food, decreased appetite, easy fatigability, loss of weight, 2 kg in a month and predominantly severe abdominal pain for 3 days prior to the admission. He was smoker and had consumed alcohol for one year after the death of his first wife. He weighs at 58kg. His BMI was 23.

# Family history

Mr Velusamy's sons are studying in schools and colleges. They sustain from the income out of mortgage with their lands. He had a strong family history of Carcinoma. His father and uncle both died of lung and oral cancer respectively. is Hihi His wife died of breast cancer who was his first cousin.

# Physical findings and laboratory investigations

Mr Velusamy had normal body build, moderately nourished with reduced activities. He was anxious, looking pale and the sclera was dark yellow in color. His abdominal girth measuring 98cm confirmed distention due to ascitis and thrill on ballottement. Ascitic fluid Protein was 3.37g/dl saying Hypoalbuminemia in diuretic stage but his Serum Protein levels were within normal limits. He had stent fixation markings and scar due to radiation therapy on his abdomen. His Alkaline Phosphatase was 549 U/L indicating an obstruction of biliary system. Paracentesis confirmed infection with 610 cells/ cu mm of total cell count. But, there was no significant change in the vital signs.

Ascitic fluid cytology report revealed that reactive mesothelial proliferation with the changes probably

due to previous radiation therapy. But no malignant cells were seen on the smear studied. Adenosine Deaminase Activity (ADA) of Ascitic fluid was within normal limits (8.7 U/L). Ultrasonograph of Abdomen showed no evidence of mass growth, mesothelial cells present in ascetic fluid.

Color Doppler study of Abdomen revealed that mass in Right Lobe of Liver extending to hila, stent in situ in common bile duct, mild intra hepatic biliary rigidity dilatation in left lobe, severe ascitis, normal inferior vena cava, hepatic vein and portal vein. He was diagnosed as Hilar Cholangio Carcinoma i.e., Klatskin's Tumor with Stenting and Post Radiation Therapy with Reactive Mesothelial Proliferation.

# Past medical history

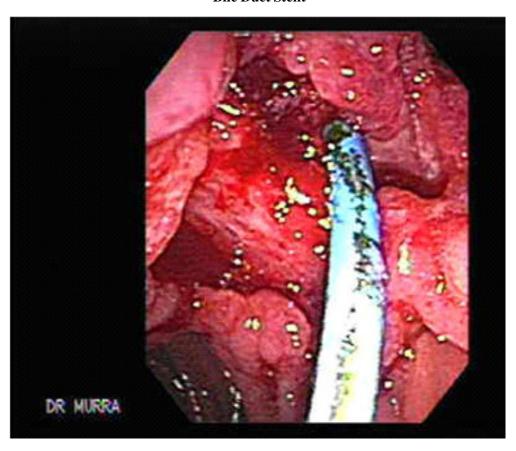
One year ago, Mr Velusamy had presented with yellowish sclera, loss of appetite and abdominal distention. He had taken native treatment with Keelanezhi herbal leaves. After, 6 months, he had

another episode with the same symptoms with increased

Serum Bilirubin and Alkaline Phosphatase and was diagnosed as Obstructive Jaundice. Based on USG abdomen he was diagnosed as Hilar Cholangio Carcinoma. He was referred to Hepato Biliary Unit CMCH, Vellore for Surgical Management. There he was found have Liver Secondaries (Unresectable Tumor) since the growth extended to the under surface of the liver. He had been advised for Palliative Stenting followed by Radiation Therapy.

At Coimbatore, Endoscopic Retrograde Cholangio Pancreatography (ERCP) was attempted twice and precut was extended. Both pancreatic duct and bile duct were delineated and amount of bile flow was seen. It was difficult to negotiate the cannula up the bile duct suggested a long malignant stricture. Guide wire manipulation induced bleeding possibly due to tumor and hence the procedure was abandoned. It was planned to do Percutaneous Transhepatic Biliary Dilatation (PTBD) followed by external drainage.

# **Bile Duct Stent**



Courtesy: images from Google search engine

PTBD was done with right lateral approach. Pig tail catheter was introduced and self retaining explainable Zilver stent 8 cm was placed in the right hepatic duct and crossing the stricture into duct with internal and external drainage. His condition had

symptomatically improved. Readmitted for PTBD assisted stenting after 2 months with icterus. USG Abdomen revealed Collapsed Left Ductal system. PTBD done on the Left side showed purulent material possibly due to underlying Cholangitis. Left duct was closed and right duct stent was present. Mr Velusamy was doing well so he was discharged with stent in Right duct system and referred to medical oncology. Radiation therapy was given at a Regional Cancer Centre for five sittings.

Treatment and nursing (Including Nursing Process)

- Pain related to underlying pathology as manifested by abdominal pain in right hypochondriac region after consumption of food
- Fluid volume excess related to extra vascular (ascitis) increased portal venous pressure, aldosterone imbalance as manifested by increased abdominal girth, ballottement and taut abdomen
- 3. Imbalanced nutrition less than body requirements related to decreased appetite as manifested lack of interest in consumption of food, inadequate food intake and loss of weight
- Activity intolerance related to generalized weakness as manifested by verbal report of fatigue
- 5. High risk for impaired skin integrity related to increasing abdominal girth, taut shiny skin
- 6. High risk for infection related to diagnostic abdominal paracentesis

After 5 months, Mr Velusamy had presented with the above mentioned symptoms and hospitalized and suspected to have Veno Occlusive Disease. Therapeutic paracentesis was done twice as 300 to 400 ml per day. Liver enzymes were given along with symptomatic treatment. His abdominal girth had reduced from 98 to 95 cm. He experienced reduction in abdominal pain.

There were no dietary restrictions other than bland diet. During hospitalization, he was motivated to eat small, frequent food. He was given chance to prefer food items with a standard schedule. As far as possible, pleasant environment was provided. There was no change in his weight during stay. He was given reference to the dietitian before discharge. His wife was included in dietary counseling.

His fluid intake was reduced to 1.5 lit per day. He had positive balance of 100 to 200 ml. Salt intake was reduced to 4 gm per day. No diuretics were prescribed. He was given the maximum possible bed rest. His finger nails were trimmed and provided comfortable bedding. His vital signs were within normal limits. His activities were planned and clubbed together. His activities were gradually increased as he could tolerate. He was advised not to exert much but to monitor and control his activities. His WBC was within normal range. He had no other sign of infection. He was protected from other patients and health team who had infections.

Mr Velusamy and his wife were explained about the reason for recurrence of ascitis as it is due to radiation therapy 6 months ago, had caused mesothelial cell proliferation in the Liver Obstruction in the Biliary system. The stent placed would take care of biliary drainage. He was discharged because he was symptomatically better. But he needed regular USG abdomen and follow up once in 2 months which will help to evaluate stent placement periodically. It is mandatory to monitor Serum Bilirubin, Serum Protein, and Complete blood count. He was asked to monitor for ascitis and restrict sodium and water as advised during hospital stay. The client and his family were explained about poor prognosis.

# **Summary**

Mr Velusamy experienced reduction in abdominal pain with normal bland diet, adequate bed rest and limited activities. His ascitis had reduced so he was discharged because prognosis was very poor. His nutritional status had improved. He was able to do his work within his capabilities. He had maintained normal skin integrity and had no signs of infection, the client and his family members were explained well about the prognosis.

## Conclusion

Klatskin's tumor is the slow growing tumor. The carcinoma of liver when the origin is from bile duct.

It is called as Klatskin's tumor or Cholangio carcinoma. Mr Velusamy was discharged and instructed to come for follow up after 3 weeks. He is regularly coming for follow up.

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