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BILIARY FISTULAS

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Biliary fistulas are broadly classified into: external and internal fistulas. Internal biliary fistulas are further classified into, bilio-enteric, bilio-biliary, bilio-vacular, and bilio-bronchial. [1]
Both external and internal biliary fistulas can occur due to some disease (spontaneous or pathological) or trauma (either iatrogenic or external trauma). While external fistulas are more frequently post-traumatic, internal fistulas are more commonly spontaneous [1] ***A third etiology for biliary fistulas is the intentionally created fistulas for therapeutic purposes. This latter group will not be included in this article.***
Spontaneous fistulas may be associated with the following diseases: gallstones, peptic ulcer, bile duct cancer, hepatic abscess, hydatid cyst, amebic abscess, and rare diseases (T.B., mucomycosis, polyarteritis nodosa), [2, 3].
Iatrogenic fistulas commonly follow the following procedures: cholecystostomy, cholecystectomy, choledochotomy, biliary-intestinal anastomoses, drainage of intra-abdominal collections, interventional radiology, liver surgery, and endoscopic sphincterotomy [1].
Any part of the biliary tree can be involved in fistulas including: the gallbladder, common bile duct, common hepatic duct, cystic duct remnant, right or left hepatic ducts and intra-hepatic biliary radicles [4,1].
An "uncontrolled fistula" denotes fistula formation with intraperitoneal leakage and collection of bile. Meanwhile, a "controlled fistula" denotes a fistula with drainage to the exterior through the abdominal wall without any significant intra-abdominal collection [1].

ETIOLOGY OF EXTERNAL BILIARY FISTULAS

Cholecystostomy is now infrequently performed. A persistent cholecysto-cutaneous fistula after cholecystostomy is usually due to unrecognized retained gallstone impacted in the hartmann's pouch or less commonly due to unrecognized distal bile duct obstruction by retained stone or malignant obstruction[1]. Recently percutaneous CT-guided cholecystostomy has been increasingly practiced in those patients with acute cholecystitis who can't stand surgical intervention. Two factors have been reported to contribute to a higher incidence of a cholecysto-cutaneous fistula in these cases: retained stones in the anterior abdominal wall[5] and premature removal of the catheter [6]. The incidence of this complication, in these cases, varied from 4% (when cystic duct was not obstructed) to 40% (when it was obstructed) [7].

An external biliary fistula continuing through the drainage tube track after elective cholecystectomy almost always indicates surgical error [8]. The incidence of such fistulas becomes higher in association with difficult cholecystectomy. Causes of difficulty include: gangrenous gallbladder, pericholecystic abscess, intense inflammatory reaction and adhesions in the region of Calot's triangle [9], excessive bleeing due to coagulopathy or increased vascularity in portal hypertension [10].

Predisposing ductal anatomy

Certain anatomical factors predispose to the development of biliary fistulas after cholecystectomy. They include anomalies of the bile ducts and hepatic blood vessels, e.g. the presence of accessory or aberrant ducts or vessels. Pertinent bile duct anomalies include sectoral right hepatic duct [8]. Anomalies of cystic duct include insertion into: retroduodenal or intrapancreatic parts of common bile duct, into right hepatic duct or into the left hepatic duct [11].

Cholecysto-hepatic ducts (ducts of Luschka) are accessory channels which connect intrahepatic ducts and the gallbladder [12]. Inability to identify these ducts during surgery was reported to result in postoperative biliary leak and peritonitis [13]. Superficial intrahepatic bile ducts may also be injured during a difficult cholecystectomy, commonly in association with coagulation-related parenchymal necrosis in the bed region. These injuries may present by bile leakage [14], extrahepatic biloma, or intrahepatic biloma [15,16]. Removal of an intrahepatic gallbladder may also. be followed by a transient biliary leak caused by damage to tiny bile ducts in the liver around the gallbladder fossa [1].

Predisposing vascular anatomy

Injury to blood vessels may result in marked bleeding which predisposes to duct injury during attempts to stop it. These injuries may result from one of two factors: 1) anomalies of extra-hepatic blood vessels such as: caterpillar hump right hepatic artery [11], and 2) unusual large blood vessels which cross the gallbladder bed, e.g. a large branch of cystic artery that runs superficially across the bed and occurs in 4% of people [17], and a large tributary of the middle hepatic vein which was reported in 10% of people [18].

Predisposing pathology

The term "difficult cholecystectomy" is applied to a diversity of situations where laparoscopic cholecystectomy is associated with a higher risk of bile duct injuries or conversion to open surgery [19]. The more frequent situations that lead to this difficulty are: acute cholecystitis [1,20], significant bleeding in cases of portal hypertension and cirrhosis [10,21], and Type II Mirrizi syndrome [22]. laparoscopically, higher incidence of bile duct injuries, including fistula, is expected. This was reported to be as high as 1.3-5.5% [23,24].

Sites & mechanisms of bile duct injury

The more frequent sites of origin of these fistulas are: cystic duct stump (55-71%), common bile duct (23-24%), gallbladder bed (16%), intrahepatic ducts (6%), [25,26]. A leak from the cystic duct stump may occur from burn injury, pressure necrosis of a metal clip, but in most instances, from distal bile duct obstruction caused by a retained stone resulting in a build up of pressure with a resultant blow out of the cystic stump [1]. Injury to CBD may occur due to: misidentification of the common duct for the cystic duct resulting in complete transection of the common duct. The RHD or right sectoral duct may be injured due to an unrecognizcd.anomalous insertion of the CD [27]. Other less common mechanisms include thermal injury due to excessive use of the cautery or laser, and the application of excessive clips to control bleeding in the triangle of Calot [1].

Post-Choledochotomy Fistulas

Replaced by laparoscopic and endoscopic techniques, the classic open common duct exploration is now less frequently performed [28]. The most common cause of external biliary fistula after exploration of the common bile duct is residual bile duct gallstone [1]. Much less commonly is an overlooked malignant distal obstruction or injury of the duct by metal bougies passed through the papilla that may result in the creation of a false tract, particularly when there is a papillary stenosis. Recently, an incidence of 2.6% was reported for external biliary fistula after removal of T-tube [29]

Liver lnjury

Serious liver trauma is complicated by bilomas and biliary fistula in around 5% of instances [34]. Three mechanisms may be involved in the pathogenesis of an external biliary fistula after liver injury: 1) direct injury of the liver or bile ducts that is not properly dealt with during surgery, 2) secondary liver damage after repair of the injury, e.g. sequestration and infection of areas of liver necrosis, and 3) secondary to emergency liver resection [35].

Liver resection

Liver resection carried out for tumor may be followed by biliary fistula which may result from inadequate ligation of the bile ducts at the cut liver surface, or failure to secure the bile ducts at the hilus [36]. This is more likely following right hepatectomy where the anatomy of the right sectoral hepatic ducts is variable in the hilar region. Extended left hepatic lobectomy has also been associated with a high incidence of biliary fistula [37].

Operative injury to the biliary tract likely to result in fistulation is more common following resection of lesions involving or close to the hilar structures. It is also more likely to occur after resection of lesions involving the caudate lobe ducts since the anatomy of these ducts is significantly variable in the hilar region [36]. Hepatic cryotherapy which may be used either at the cut liver surface following hepatectomy or for ablation of deep intrahepatic lesions may be complicated by biliary fistula [38]. Also, the use of harmonic scalpel in resection was recently observed to be associated with a higher incidence of fistulas, which is about 24% [39]. It may be prudent to leave a drain in-situ following this procedure. The incidence of fistulas after liver resection for bile duct cancer was reported to be 7-8% [40].

Fistula after liver transplantation

The incidence of external biliary fistula after liver transplantation was reported to be 6.7% [41]. Biliary leak and fistula are a continuing source of morbidity and mortality following liver transplantation [42]. Most bile leaks after transplantation are anastomotic and occur in the early

postoperative period [43]. There may be leakage from the cystic duct stump, the exit of the T-tube, or from the cut surface of the reduced liver graft [44]. Late ischemic necrosis of an intra-hepatic bile duct may lead to biliary leakage [1]. Technical errors such as excessive tension on the biliary anastomosis, kinking, or redundancy of the bile duct, and poor blood supply to both donor and recipient common bile duct may result in biliary fistula [44].

Fistula after hydatid disease of the liver

Biliary fistula develops after operation for hydatid disease in three situations: 1) a communication between the cyst cavity and the biliary system which is missed at operation, 2) obstruction of the biliary tree by hydatid material, and 3) as a complication of liver resection which is rarely performed in hydatid disease [1]. Intrabiliary rupture is the most common complication of hepatic hydatid disease (HHD), and persistent biliary leakage is the most frequent complication after surgery for HHD [45]. Erosion of the biliary tree by an enlarging cyst may cause hydatid material to enter into the biliary tree or, conversely, bile may leak into the cyst [1]. Biliary leakage after hydatid resection was reported to occur in up to 26% of cases. This incidence is particularly higher in patients who have purulent and/or bilious cyst content, or pre-operative raised alkaline phosphatase and gamma glutamyl transferase levels. The majority of these fistulas persist for more than 10 days after surgery [46].

PREVENTION OF EXTERNAL BILIARY FISTULAS

During cholecystectomy

The role of operative cholangiography in the prevention of bile duct injuries is controversial. On one hand, it may provide information concerning unsuspected choledocholithiasis and anomalous anatomy [55,56]. On the other hand, there is no evidence that it may prevent major bile duct injury [57]. Accordingly, it was recently found that only 40% of the surgeons in the USA employ operative cholangiographv [58]. Operative cholangiography can be used selectively, when ductal anatomic variations or anomalies or bile duct injuries are suspected. Early conversion to laparotomy is favored if the ductal anatomy remains unclear [59].

During common bile duct exploration

As biliary fistula that follows exploration of the common bile duct or removal of a T-tube is almost always due to a residual bile duct gallstone, performing peroperative cholangiography and excluding retained stones before removal of a T-tube, is essential [1].

During surgery for liver injuries

Emergency partial hepatectomy for major liver trauma may result in an injury to the bile ducts at the confluence with an early biliary leak and, later, a biliary stricture [36]. Complete transection of the common bile duct requires immediate hepatico-jejunostomy, while lacerations of the main biliary channel may be sutured after placement of a T-tube [35].

During surgery for hydatid cyst

Several precautions are required to avoid the development of biliary fistula after hydatid surgery. Drainage of all cyst cavities, particularly multiloculated hydatid cysts, in order to ensure that if a biliary fistula develops, it will be controlled, particularly with the fact that these fistulas usually close spontaneously. Assessment of the biliary tree by ERCP should be performed prior to surgery in patients with a history of jaundice or cholangitis or in the presence of a large cyst located centrally and abutting the hilar structures, to avoid postoperative obstruction by hydatid material or the cyst [1]. In these cases, pre-operative sphincterotomy is recommended [60].

During liver transplantation

It has been reported, on basis of data from a large number of patients, that insertion of a T-tube into the biliary anastomosis in liver transplantation was associated with a higher incidence of biliary fistula [41].

During bilio-enteric anastomosis

Three factors are essential to avoid external fistula after bilio-enteric anastomosis: 1) identification of healthy bile duct mucosa proximally, 2) proper selection of the jejunal segment, and 3) direct mucosa to mucosa anastomosis [31].

PATHOPHYSIOLOGICAL CONSEQUENCES

Consequences of biliary fistula are mainly due to depletion of electrolytes and fluid, to the absence of bile from the gut and to the possibility of ascending exogenously acquired biliary infection. Total biliary loss for short periods of up to 3 weeks may not result in a serious depletion of electrolytes and fluid since the body is able to compensate for this loss. Long-term total external biliary fistula results in fluid and electrolyte disturbances if replacement therapy is not instituted. Sodium loss is usually in excess of chloride loss, leading to metabolic acidosis. The serum potassium level is initially lowered, but the accompanying fluid loss may lead to a decrease in plasma volume, low-output renal failure and hyperkalemia. Absence of bile from the gastrointestinal tract causes interference in the absorption of fat soluble vitamins A, D and K. Vitamin A and D deficiency is associated with long-term total biliary fistula and is rarely seen today, while vitamin K deficiency is evident: earlier. Clinically, patients with an external biliary fistula even in

the short term, feel unwell, weak and lethargic. In advanced and neglected cases, caloric and protein malnutrition results in gradual weight loss while the electrolyte changes may result in stupor and vasomotor collapse [1].

CLINICAL PRESENTATION

External biliary fistula usually presents as biliary discharge from the drain, the wound or drain track. However, some patients present with more serious presentations as peritonitis, intra-abdominal abscesses, abdominal distension with bile and colangiohepatitis. This occurs when the fistula is uncontrolled and the drainage of bile is inadequate. Although bile is considered sterile, septic complications soon occur in these cases. The presence of skin excoriation and digestion implies that activated digestive enzymes are present in the fistula effluent, since bile alone contains no digestive enzymes. Skin excoriation never occurs in pure biliary fistula [1].

One factor which is involved in the timing of presentation is the presence and adequacy of drainage instituted during surgery. In laparoscopic cholecystectomy, the majority of external fistulas do not present in the immediate postoperative period mainly because drains are usually not left in the abdomen and the patient leaves the hospital usually within 24—48 h [27]. The resulting uncontrolled biliary fistula becomes evident within days or weeks after the operation, presenting with abdominal pain, fever and jaundice and the demonstration of an intra-abdominal fluid collection which produces bile on puncture. Another factor is the mechanism by which the fistula resulted. Fistulas which are due to technical errors or direct injuries present earlier than those which result from a secondary pathology, e.g. tissue necrosis, ischemia or abscess [1].

MANAGEMENT OF EXTERNAL BILIARY FISULAS

The management of external biliary fistulas follows the following steps: [1] resuscitation and initial management, [2] drainage (converting an uncontrolled fistula into a controlled one), [3] definitive diagnosis of the anatomy of the fistula, [4] definitive treatment [1].

1) Resuscitation

Several factors dictate the vigor of resuscitation in patients with external biliary fistula, including: the general condition of the patient, duration of the fistula, the output of the fistula, the discharge of the fistula whether it is pure biliary or associated with other gastrointestinal secretion. Oxygen exchange and hemodynamic stability should be ensured. Volume-contraction alkalosis and electrolyte imbalance which occur in high output fistulas are corrected [61].

Local control

Wound care assumes a high priority because, if operation is necessary, it should not be done through a septic, indurated, cellulitic, and denuded abdominal wall. Simple "bagging" of fistulas may lead to skin closure over the fistula tract, and lead to abscess formation. Accurate records of fistula output are requisite to daily volume and electrolyte management. Local skin care is crucial and, consequently, many devices have been tried for the local management of fistula drainage. Preventing excoriation or superinfection of the skin surrounding a fistulous tract is paramount. Preparations useful in preventing skin maceration and breakdown include Karaya powder or-seal, ileostomy cement, and glycerine. Ion exchange resins also may be helpful in preventing excoriation. Stoma adhesive or similar products may be very effective [61].

Somatostatin

Use of somatostatin in biliary fistulas has been advocated, but proof of efficacy is scant. Somatostatin decreases biliary secretions and, when used in biliary fistulas, a 50% reduction in fistula volume has been noted. It may be stated that, somatostatin enhances closure in fistulas that would otherwise close spontaneously. In those fistulas that would not close without a specific intervention, somatostatin has limited value [62,61].

Nutritional management

Most biliary fistulas can, and should, be cared for with enteral therapy. A low-fat diet may assist in decreasing fistula drainage. In the presence of large bile fistulas or biliary obstruction, every attempt should be made to refeed bile on a regular basis. Clearly this will require postpyloric nasointestinal tube placement [61].

Antibiotics

Unless evidence of cholangitis, cellulitis, or abscess exists, antibiotic use should be reserved. However, in critically ill patients who are not good operative candidates, it seems prudent to attempt bile sterilization [61].

2) Drainage

Drainage means converting an uncontrrolled fistula into a controlled one. Two reasons place drainage as an early essential step in the management of an uncontrolled fistula which should precede definitive diagnosis and treatment. First, an intra-abdominal collection may predispose to serious septic complications unless promptly drained. Second, definitive repair should not be attempted at this early stage, since the involved bile duct(s) are collapsed, friable and are usually embedded within a severe local inflammatory reaction. Moreover, it is virtually impossible to expose healthy bile ducts for any form of long-lasting definitive repair, and such an attempt, which is

bound to fail, will render further operation more difficult. Drainage should entail inserting a tube into the site of collection. Percutaneous computerized tomography (CT) or ultrasound-guided drainage can be done for a subphrenic or infrahepatic fluid collection. In other occasions, reoperation is required for peritonitis or drainage of a major intra-abdominal collection. It is most important to ensure adequate drainage and that the fistula is indeed controlled. Biliary drainage is ideally carried out using a sealed drainage bag system. Drainage should, at least initially, be under a low pressure, closed suction system which is valuable in reducing the cavity of the intra-abdominal bile collection or abscess to a fistula track. Improvement of the clinical picture together with repeat ultrasound or CT studies should eventually demonstrate proper positioning of the drain and no residual collection or abscess. HIDA scintigraphy and tubography are helpful in this respect [1].

To date, a total of 25 cases of laparoscopic drainage of post-cholecystectomy bilomas have been reported in the literature. Two main advantages are claimed to laparoscopic drainage over non-surgical drainage methods: visual identification of the exact site and amount of bile that requires drainage, and the safe, optically-guided application of suction to ensure complete evacuation of the collection, particularly that in pockets and recesses. These minor collections are more prone to predispose to intractable infection [63].

3) Definitive diagnosis of the anatomy of the fistula

Following the establishment of a controlled fistula, various radiological investigations are then performed with the aim of assessing the origin of the fistula, the presence and extent of an injury to the extrahepatic biliary system, adequacy of drainage, and the presence of biliary-enteric bile flow. The anatomy of the entire intrahepatic and extrahepatic biliary tree should be demonstrated, and this is achieved by a variety of radiological studies. All contrast examinations should be covered with antibiotic prophylaxis to minimize the risk of a bacteremic episode [1].

Tube cholangiography should be performed routinely prior to removal of a cholecystostomy or choledochostomy tubes or tube drainage across biliary-enteric anastomoses. Whenever an obstruction to free biliary-enteric bile flow is present, it should be treated. Removal of such a tube in the presence of a distal stricture or a retained bile duct stone will invariably result in a persistent biliary fistula [1]. Fistulography is a simple and effective means of finding out whether biliary drainage is adequate and whether a fistulous cavity, has indeed converted to a fistulous track. The site and underlying cause of the biliary fistula can also be clearly demonstrated by fistulography [1].

Biliary fistulas should be investigated by ERCP. It is the most useful diagnostic and therapeutic tool in instances where there is a continuity of the extrahepatic biliary system, particularly following laparoscopic cholecystectomy and in liver transplanted patients. The site of the fistula, the etiology, as well as the status of the distal biliary tree can be ascertained by ERCP in more than 90% of cases. It can be performed in the absence of dilated common and intrahepatic ducts [61]. The value of ERCP is limited in fistulation arising at the hilus. If the biliary tree has been ligated or is not in continuity at the level of the common bile duct, ERCP likely will not reveal the fistula. Thus, when ERCP does not demonstrate the fistula, sphincteroromy and stenting are unlikely to be successful and an operation will likely be necessary [61]. In one series ERCP failed to visualize 16% of the fistulas arising within the liver (above the liver hilus) following blunt hepatic trauma [64].

Percutaneous transhepatic cholangiography provides excellent anatomic detail of the biliary tree and identification of the site of most biliary fistulas. It is the only diagnostic modality to diagnose iatrogenic damage to a right sectoral hepatic duct. In addition, it provides a route for decompression. The drawback is that PTC is unlikely to provide definitive management of a pathologic distal biliary tree [1,61].

HIDA Scan may be useful initial test when postoperative injury of the extrahepatic biliaiy tree is suspected. It has the advantage of being easy to perform and noninvasive in nature [61]. Though it may not supply accurate anatomical details, information regarding the presence of a fistula, its origin (liver or extrahepatic biliary system), adequacy of drainage (controlled or uncontrolled), and mode of biliary-enteric bile flow can all be clearly obtained [1].

CT-scan, MRI and Ultrasound will not demonstrate the precise fistula anatomy and, therefore, are not considered primary diagnostic modalities in the evaluation of a biliary fistula. Their utility, as is the case for gastrointestinal fistulas, is the localization of undrained collections, evaluation of the adequacy of collection drainage, and evaluation of tumor recurrence. When symptoms are persistent or sepsis occurs, these studies should be used liberally to localize treatable causes [61].

Magnetic resonance cholangiopancreatography (MRCP) can provide a better road map of the fistula than an ERCP [65]. It is the most accurate method for diagnosis of cystic duct leak after cholecystectomy [66].

4) Definitive treatment

The therapeutic decision

A controlled fistula is first evaluated for biliary-enteric continuity. Lack of this continuity, i.e. a total fistula, is a straight forward indication for surgery after a reasonable period of conservative treatment. When biliary-enteric continuity is present, the next factor to consider is distal obstruction. In absence of obstruction, conservative treatment results in spontaneous closure in most cases. If there is distal obstruction, some intervention is required to relieve it. An attempt is made to achieve this non-surgically. Non-surgical methods include endoscopic methods (sphincterotomy, alone or sphincterotomy and stenting) and percutaneously by balloon dilatation. Should these methods fail, surgery is indicated [1]. It is important to select the appropriate therapeutic approaches according to the setting. Resorting to surgery as a primary approach for therapy should not be the standard practice. On the other hand, strict adherence to a conservative approach, which employs non-surgical methods and excludes surgery, is associated with an obligatory 9% conversion to surgery at an advanced stage of the disease, together with a mortality rate of 3.5% [67].

Endoscopic stenting

Fistula closure may be facilitated by temporary placement of a stent across the fistulous opening in the bile duct thus excluding bile flow through the fistula. This method may be attempted in instances where there is an intact common bile duct above the fistula origin and the defect in the bile duct is not too large. It may be particularly helpful in instances of cystic stump fistula and may facilitate early closure [68]. Stenting may be achieved by endoscopic placement of an endoprosthesis or a nasobiliary tube with its tip above the origin of the fistula [67]. When a short period of stenting is anticipated, nasobiliary intubation is preferred since it enables follow-up cholangography. Damage to the papilla is minimal and a second endoscopic procedure is avoided. Using this method, some fistulas will close within 2 weeks [69]. On the other hand, a nasobiliary tube may be uncomfortable for the patient. It may dislodge and it may enhance metabolic acidosis resulting from bile loss if used for prolonged periods of time. In most instances a biliary endoprosthesis is used and is left in place usually for several weeks until fistula closure. Closure is verified by HIDA scan prior to removal of the stent [1]. The use of temporary stenting is limited in high complex hilar fistulas combined with stricture(s), particularly those with separation of a right and left ductal system. Some authors place an endoprosthesis across the stricture and leave it for months [70] and even for more than a year with periodic replacements [71]. This is not only unnecessary but it may be harmful, resulting in obstruction and septic complications. The endoprosthesis, if placed, should be removed when the fistula has closed. An 85-100% success has been reported for endoscopic stenting when it was performed for high fistulas that involved intrahepatic bile ducts after blunt hepatic trauma [72, 73]. Likely, a 100% rate of success was also reported in fistulas following hepatic resection when they arise from bile duct stumps [64]. Success rate for endoscopic management of biliary fistulas after hydatid resection (employing nasobiliary drainage) was reported to be 81% [74]. Endoscopic stenting for post-cholecystectomy fistulas was reported to be feasible in 84% of cases in general but only 40.6% of lesions of the common bile duct or the right antero-medial duct. The success rate was reported to be 97% [25]. Nasobiliary tube placement is effective in closing biliary fistulas after cholecystectomy. It is preferred to combine it with sphincterotomy in fistulas due to common bile duct injury to reduce the resistance to bile flow to a minimum [26].

Endoscopic sphincterotomy

Endoscopic sphincterotomy may be of value in fistulas that arise from the intrahepatic part of the biliary tree. One example is fistulas that follow excision of hydatid cysts [45]. In these cases, success rates as high as 100% have been reported with sphincterotomy alone [75]. The value of this approach is debatable. It has been shown that stenting is more effective than sphincteroromy alone in the resolution of biliary fistulas [76]. On the other hand, some authors believe, in light of the high rates of success reported recently, that sphincterotomy should be the first line of treatment [75]. Post-cholecystectomy fistulas have also been managed by sphincterotomy without stenting. A recently reported 100% success encourage the use of this method [77].

Other endoscopic maneuvers

Once an obstruction distal to the fistula has been diagnosed it should be dealt with since the fistula will not close spontaneously. Obstruction is usually caused by a retained stone or a stricture. Retained stones are usually removed by endoscopic means. Relief of a benign stricture can be achieved using balloon dilatation applied endoscopically. The patient is then treated conservatively and expectanlly either for fistula closure or for re-stricture which, once it occurs, is then treated operatively [1].

Percutaneous methods

Attempts to block the fistulous tract have been made by several investigators. This is particularly useful in fistulas with isolated intrahepatic bile ducts which are not communicating to the main biliary system. Several agents have been tried with variable rates of success, including ethanol [78]. More recently, biliary stents were placed percutaneously across the injured portion of the bile duct, employing a coaxial guidewire. Alternatively, an expandable metallic biliary endoprosthesis was inserted [79]. Results were encouraging. Percutaneous solutions for posthepatectomy fistulas from isolated ducts have been recently intreoduced. One of these is percutaneous selective

portal vein embolization to ablate the involved liver segment [80].

Surgery

Surgery, in the management of biliary fistula, may be required for 2 goals: 1) drainage for collections in uncontrolled fistulas, and 2) definitive treatment of the fistula in selected cases. The first goal has been discussed before. As a definitive therapy, surgery is indicated when: 1) thers is no bilio-enteric continuity, 2) non-surgical methods failed to relieve distal obstruction in fistulas with bilio-enteric continuity, 3) non-surgical methods can't be used to relieve the distal obstrruction, e.g. high ligature of a major bile duct, and 4) surgery is the primary line of treatment for an associated pathology, e.g. malignancy [1].

While surgery, indicated for drainage, can be considered as an urgent step that should be carried out promptly to avoid serious complications, attempting definitive surgery for biliary fistula should be postponed as much as possible. It is a mistake to think that immediate repair is a simple matter since exposure of healthy bile duct mucosa without a sufficiently dilated duct can be very demanding and indeed may be impossible. The incidence of external re-fistulaization, under these circumstances was reported to reach 67% [32]. A cautious approach is preferable since even ultimate closure of the fistula with the development of jaundice is usually associated with proximal ductal dilatation and easier subsequent repair [1]. A bilio-enteric anastomosis, performed 4 to 6 months after the initial surgery on a dilated common hepatic duct is more likely to succeed than an operation on a septic, hypoproteinemic patient with sodden, friable, non-dilated bile ducts. On the other hand, waiting for the ducts to dilate in a patient with a complete transection of the bile ducts with complete biliary diversion only leads to depletion of the bile acid pool, severe electrolyte derangement and nutritional failure, leading on to sepsis and death [65].

The role of surgery in different settings

Post-cholecytostomy biliary fistula Persistent biliary fistula is almost always caused by distal obstruction to the gallbladder outlet or to common bile duct. Surgical options include: removal of gallstones in the gall bladder, the bile duct or both, and surgical excision or bypass of the malignant obstruction unrecognized at the time of initial operation [1].

Post-cholecystectomy biliary fistula It is sometimes possible to carry out drainage through a mobilized and approximated Roux-en-Y loop of jejunum, the drainage tube being ked from the transected bile duct across the jejunm to the exterior. This procedure allows initial control and the almost certain necessity of re-operation for re- stenosis at a later date. If large quantities of bile, of the order of 100ml per day or more, drain in the postoperative period then there is usually a serious cause and indeed operative injury of a major bile duct should be suspected. Such patients may become generally ill and superimposed sepsis is common. The essential in management in this situation is not re operate rapidly. It is wiser to treat infection, nourish the patient and wait. If fistulography or cholangiography reveals continuity between the biliary system and the gastrointestinal tract, then a prolonged period of drainage may result in spontaneous closure of the fistula. Re-operation for definitive repair of a bile duct stricture should be delayed Occasionally, percutaneous transhepatic or endoscopic cholangiography will reveal a normal biliary ductal system. This unsual situation probably means that the external biliary fistula is due to injury to the subvesical duct in the gallbladder fossa. In this situation it is safe to treat the patient expectably since the biliary drainage will cease in due course without further sequelae. Recently, a success rate of 90% was reported after Roux-en-Y hepatico-jejunostomy for fistulas following cholecystectomy [81].

Biliary fistula after Choledochotomy The usual cause of potential or actual postcholedochotomy fistula is a retained stone in the bile duct distal to the T-tube although it rarely may be due to undetected malignant obstruction in the duct. Surgery is indicated when stone extraction fails by other methods, or a malignant obstruction is discovered [1].

Biliary fistula after drainage of intra-abdominal collections Biliary fistula may occur following the drainage of subhepatic, perihepatic or periductal collections of pus and bile or may be associated with the complication of severe acute necrotizing panceratitis. Irrespective of the cause, initial management is drainage of the related cavity, removal of slough, fluid replacement and nutrition. The use of appropriate antibiotics is an important element. The majority of such fistulas close spontanepouly within a month to six weeks without the need for further surgery. However, if the residual cavity remains inadequately drained or if there is still pancreatic or peripancreatic slough present, the fistula will not close spontaneously and further operative drainage is required [1].

Biliary fistula after hepatic surgery or liver injury The main problem is that of a fistula from an isolated biliary duct after repair of injury or resection. These fistulas are not likely to close spontaneously. Non-surgical (percutaneous) methods have been occasionally attempted to close these fistulas by injection [78] or ablation of the liver segment by selective portal embolization [80]. Surgical correction entails 3 options. Firstly, resection of the isolated segment of liver tissue may be carried out. This can be difficult and is seldom warranted. In rare instances the fistula may be identified at operation and simply oversewn [37]: alternatively a well developed fibrous fistulous tract can be anastomosed either to a prepared loop of jejunum or to the gallbladder Such an operation may produce a permanent

cure but late stenosis of the stricture may still be a problem for which secondary repair must be carried out [1]. Biliary fistulation after partial hepatectomy for tumours may occur from the liver surface. If biliary reconstruction, e.g. following liver resection for hilar cholangio-carcinoma, has been performed, leakage from the anastomosis or from bile ducts not identified at the time of surgery may occur. Such leakage is particularly prone to arise from caudate lobe ducts. Management is conservative and most of such fistulas will close in due course [36]. Biliary fistula after removal of a hydatid cyst is a difficult problem. Initial treatment should be conservative, but if the fistula fails to close then it is important to carry out fistulography and/or endospic retrograde cholangiography to determine that there is no biliary ductal obstruction. Re-operation is usually necessary, either to relieve biliary obstruction or to attempt to close biliary communications under vision. Such approaches can be combined with omentoplasty. The cavity is not drained but the operative area adjacent to it is drained postoperatively [46].

BILIO-ENTERIC FISTULAS

Cholecysto-duodenal fistula without gallstone ilues

Cholecystoduodenal fistula is the commonest type of bilioenteric fistula. It affects about 5% of all patients with cholelithiasis [82], and presents 68-77% of all bilio-enteric fistulas [2,61]. Two groups of factors predispose to cholecysto-duodenal fistulas. The first is repeated inflammatory episodes of the gallbladder which may gradually involve the adjacent duodenum. This process may less commonly involve the hepatic flexure of colon or the gastric wall. The second factor is impacted stone in the gallbladder neck during an episode of acute cholecystitis, which may result in decreased blood supply, gangrene which will fistulize with adjacent adherent wall of the duodenum with extrusion of the gallstone into its lumen [83].

Gallbladder calcification (porcelain gallbladder) may be rarely complicated by a cholecystoduodenal fistula [84]. The exact explanation for the occasional co-existence of cholecysto-duodenal fistula and cancer gallbladder is debatable. Cancer gallbladder may definitely result in fistula. On the other hand, a fistula may be the predisposing factor for cancer through reflux of the duodenal contents into the gallbladder [82].

Cholecystoduodenal fistulas may be asymptomatic. Less frequently, they are associated with non-specific symptoms or result in gallstone ileus. Non-specific symptoms are similar to those of chronic biliary disease. Rarely, symptoms of cholangitis may occur, e.g. fever, rigors, and jaundice [85,8]. Symptoms of impaired gastric emptying and acute upper gastro- intestinal hemorrhage can be a rare clinical presentation of biliary concrete impacted in the duodenum, secondary to cholecystoduodenal fistula, a condition referred to as Bouveret's syndrome [86].

Preoperative recognition of non obstructive cholecysto-duodenal fistula is difficult and operative accidental diagnosis is the usual [87]. Pneumobilia detected preoperatively by plain radiograph of the abdomen may give a suspicion of the presence of bilioenteric fistula but it is not diagnostic [8]. However, reflux of contrast material into the biliary tree during upper GIT barium or gastrografin study may be the diagnostic procedure of cholecytoduosdenal fistula [8]. Preoperative diagnosis of cholecystoduodenal fistula using abdominal ultrasonography needs experience [88]. A CT-scan based diagnosis is also difficult [89].

Elective surgery may never be necessary in a completely asymptomatic individuals and high-risk elderly patients with mild symptoms may require no treatment. When associated biliary disorders are present, conservative management with careful observation, endoscopic papillotomy and stone extraction from common bile duct, if present, may be tried [8]. However, in a patient with good general condition, cholecystectomy with careful dissection of the fistula, and repair of the duodenal defect has the best long term therapeutic result [90].

Cholecysto-duodenal fistula with gallstone ileus

Gallstone ileus is an unusual and peculiar complication of biliary lithiasis. Less than 1% of gallstones migrate into the gut, causing about 25% of non- strangulated small bowel obstructions in elderly population [91].

The clinical picture of gallstone ileus is not like that of any mechanical intestinal obstruction [92]. During migration of the stone, symptoms are minimal or absent, thus the onset is insidious, occasionally acute [91]. The stone can produce intermittent obstruction during its migration in the gastro-intestinal tract, however recurrent incomplete bowel obstruction can be present for weeks or months before complete obstruction [8].

Pneumobilia in plain abdominal radiograph may be found in 20-50% of patients with gallstone ileus [84,8]. Abdominal C.T scan and ultrasonography also may be helpful in diagnosis and detection of both the cholecystoduodenal fistula and the gall stone in the terminal ileum or in other part of G.I.T [91].

Gallstone ileus is a surgical emergency that should be approached without delay in hope that a suspected stone will pass [84]. Preoperatively, the patient should receive adequate fluid and electrolyte resuscitation as well as nasogastric decompression and preoperative antibiotics [91].

The terminal ileum is the site of obstruction in 70% of cases. Unless the obstructed segment is ischemic, or has perforated requiring a small bowel resection, the obstructing calculus can be manipulated proximally to a healthy dilated jejunum where a safe enterotomy and stone removal may be executed [84]. Jejunal impaction, often by stones larger than 4 cm, occurs approximately in 15% of cases and enterostomy may be made at that site or just proximal to it [91]. Duodenal obstruction, usually in the bulb, is known as Bouvret's Syndrome which occurs in 10% of cases with gallstone ileus and may be handled by duodenotomy and primary closure with tube duodenostomy or pyloroplasty. Occasionally it may be possible to manipulate it back into the stomach and the stone can be removed via a gastrotomy. Rarely a gastroenterostomy be necessary to protect a duodenotomy or severely traumatized duodenum at the site of impaction and to improve symptoms secondary to decreased gastric emptying [8]. In rare instances, the sigmoid colon is the site of obstruction. If the impacted stone cannot be manipulated proximally to a transverse colostomy, consideration should be given to exteriorization of the impacted segment [91,8].

Cholecysto-colic Fistula

Cholecysto-colic fistulas occur in up to 15% of bilio-enteric fistulas. They are heralded by a sudden change in bowel habit with multiple, loose stools and the development of fever, chills and other signs of cholangitis from colonic bacterial reflux into the biliary tract [94]. Cholangitis appears to be a more prominent feature of cholecysto-colic fistula when the opening of the fistula is narrow and liable to intermittent obstruction of bile flow, whereas diarrhea with a lesser incidence of fever and chills is more common with wider fistulas [95].

The plain abdominal film reveals air in the biliary tree in only 50% of cases [94]. Only when a barium enema is done, preferably with air contrast, the diagnosis becomes evident as both barium and air fill the gallbladder and extrahepatic bile ducts. Failure of a barium enema to demonstrate a cholecysto-colic fistula has been reported, but that is quite rare [61].

There is a controversy about the appropriate treatment for cholecysto-colic fistula. In the most circumstances, they should be dismantled because of the present risk of sepsis. If the diagnosis can be made preoperatively, suitable mechanical and antibiotic bowel preparation will reduce the chance of infectious complications of surgery, and permit primary closure of the large bowel. Cholecystectomy and, if indicated, common duct exploration should be carried out at the same time [8]. Bilio-colic fistula may very rarely occur after cholecystectomy between the cystic duct stump and colon. Only 3 cases were reported in literature up to 2002. The treatment of such type of rare fistula should by surgical excision of the cystic duct stump with primary closure of the colon [4].

Cholecysto-gastric fistula

Cholecytograstric fistulas form 4.9-6% of all bilioenteric fistulas [2,61]. They usually manifest by symptoms of severe biliary gastritis associated with those of biliary lithiasis. Gastric symptoms may be so severe with gastric erosions and massive haematemsis. Diagnosis can be made with certainty by ERCP. Upper GIT barium study with reflux of the contrast into the biliary tree may also demonstrate the fistula [96]. Treatment of these fistulas is essentially surgical, where cholecystectomy with repair of gastric wall defect in layers is the surgical procedure of choice [97].

Choledocho-duodenal fistula

A choledocho-duodenal fistula may evolve due to any of three etiological mechanisms: a duodenal pathology, a biliary pathology, or iatrogenic (endoscopic) injury. Duodenal causes of this fistula include: chronic duodenal ulcer, in 80% of cases [98], peri-ampullary carcinoma [99], duodenal tuberculosis [3]. Biliary causes include: impacted stones and cholangiocarcinoma [92]. Iatrogenic injury occurs during ERCP. The resulting fistula is called parapapillary fistula. It may involve the intramural part of the common bile duct (Type I) or the extramural part (Type II) [100].

An abdominal plain radiograph may show air in the biliary tree in 14-58% of cases. Upper GIT barium study is diagnostic in most cases by baruim reflux into the biliary tree through the fistula [92,98]. Upper GIT endocsopic examination together with biopsy is recommended to exclude neoplastic lesions. In addition, ERCP can help in detection of the fistula tract [99,92].

Recommended treatment of the choledochoduodenal fistula due to peptic ulcer is the treatment of the ulcer. There is good evidence that such fistula heals on medical therapy. However, the risk of biliary stricture exists after healing of the fistula [98,92]. If surgery is indicated for the ulcer disease, vagotomy and anterectomy, with Billroth II reconstruction, is recommended to avoid the area of the fistula. If there is associated cholangitis, or biliary obstruction, biliary-enteric amastomosis, via Roux-en-Y loop of Jejunum, is added [98,61,92]. Treatment of para-papillary fistula is controversial. While some authors recommend no treatment for the fistula, others prefer doing sphincterotomy. Anyway, sphincterotomy may be necessary for another indication, e.g. stone extraction [101].

BILIO-BILIARY FISTULA (MIRIZZI SYNDROME)

In 1948, Mirizzi described a syndrome which consists of 4 components: parallel course of the cystic duct to the common hepatic duct (CHD); impaction of stones in the cystic duct or neck of the gallbladder; mechanical obstruction of the CHD by the stones or secondary

inflammation and intermittent or constant jaundice and recurrent cholangitis [102,22]. A bilio-biliary fistula is only significant when it entails an abnormal communication between the gallbladder and the extra-hepatic main bile ducts. This occurs usually as an uncommon complication of severe inflammatory reaction to gallblader stones, i.e. Mirizzi syndrome (MS) [28,22,103]. However in very rare occasions this fistulous communication my occur secondary to gallbladder adenocarcinoma [104]. Because the first is much more common, the following discussion focuses on this condition. Mirizzi syndrome occurs in about 0.7-1.1% of all patients with cholelithiasis [22,105]. The consecutive studies of MS resulted in identification of 4 types of this syndrome: Type I (extemal compression of the CHD by a calculus impacted in the cystic duct), and Types II, III and IV (the calculus had eroded into the bile duct creating a cholecysto-choledochal fistula with variable degree of common bile duct obstruction, i.e. 1/3 of the duct in Type I, 2/3 in Type II, and full obstrtuction in Type III [106,28]. An amended pathological classification for Mirizzi syndrome was reported in a humble Egyptian study [107]

The clinical presentation of MS is that of Charcot's triad. The differential diagnosis varies according to its clinical manifestations. In absence of jaundice, identification of MS is difficult. On the other hand, jaundiced patients having MS with or without cholangitis should be differentiated from other causes of benign and malignant obstructive jaundice in general and specifically those associated with common hepatic duct obstruction [108,109]. Abdominal US has a sensitivity of 23-27% in diagnosing of MS as it may reveal bile duct dilatation in 81% of patients [22,110]. CT-scan probably has a higher sensitivity and specificity in this concern [111]. ERCP has a sensitivity of 55% in detecting MS. In addition to the diagnostic value, ERCP has the advantage of aiding surgical intervention by stone retrieval and biliary stenting. The latter facilitates identification of the CBD during operative dissection [22]. However, ERCP is limited by failure to canulate the CBD in 5-10% of cases and suboptimal study from incomplete contrast filling of the ducts due to tight strictures or intraductal debris together with its complications including sepsis and pancreatitis which can occur after ERCP. When ERCP is unsuccessful or difficult, PTC is a valuable alternative [112]. In MS, MRCP can be as good as ERCP in diagnosis and its ability to delineate details of biliary strictures and to detect the fistula. In addition, MRCP can differentiate a neoplastic mass from an inflammatory one which US or CT scan may not be capable of [111]. On the other hand, ERCP is recommended when an endoscopic therapeutic procedure is potentially required at the same time [22,108].

Because preoperative diagnosis of MS, especially the Type II, is often inconclusive [113], a high index of suspicion must be maintained. This is particularly true for patients with gallstones presenting with a history of jaundice [108]. The two objectives during surgery are: the safe completion of the cholecystectomy without inflicting injury to the bile duct, and the appropriate management of the cholecysto-choledochal fistula [22]. Several technical remarks facilitate dealing with MS. They include meticulous dissection at Calot's triangle and hepatoduodenal ligament, and adopting the fundus first approach to the gallbladder [22]. Timely performance of intraoperative cholangiography is essential to confirm the diagnosis, identify the full extent of the disease, assess the location and size of the fistula, exclude the presence of stones or strictures in the bile duct, and select an appropriate surgical procedure [114]. Decompressing the gallbladder in acute cholecystitis or when the gallbladder is distended and tense is also useful [113]. In Type 1 MS, a cholecystectomy is adequate. In the absence of CBD stones on pre or per-operative cholangiogram, stones impacted in the cystic duct or the neck of the gallbladder are milked back into the gallbladder which is then removed and the cystic duct oversewn [108]. However, the cystic duct is frequently occluded and obscured by inflammatory changes in the region of Calot's triangle and a subtotal cholecystectomy, fundus first dissection and leaving the neck of the gallbladder behind is a more prudent approach [22]. Routine CBD exploration is not necessary unless stones are noted pre-operatively or on intraopertive cholangiography and should be carried out only if the CBD is easily exposed, otherwise definitive management of CBD stenosis should be left till the inflammatory process has resolved. It was observed that bile duct stenosis generally resolves as inflammation subsides following cholecystectomy [22]. Surgical procedures for Type 2 MS depend on the severity of the fistula and any associated bile duct strictures. A small fistula may be closed primarily by interrupted stitches and a larger defect can be closed using a cuff of gallbladder remnant following subtotal cholecystectomy [22,109,110]. Well-vascularised flap from the gallbladder or cystic duct can be used for the closure of a large defect [115]. Routine biliary bypass of the choledochal fistula to the duodenum or jejunal loop is recommended to be used in MS type IV fistula where there is complete section and questionable vascularity of the CHD. Roux-en-Y hepaticojejunostomy is the procedure of choice [22,114,108]. Occasionally, tumours of the cystic duct and biliary tree may mimic MS [116]. However routine frozen section to detect gallbladder cancer in young and fit patients where radical resection at the same operation can be considered [22].

BILIO-VASCULAR FISTULAS (HEMOBILIA)

Common causes of the bilio-vascular communication that gives rise to hemobilia are: accidental or iatrogenic trauma [118], gallstones, tumors, inflammatory processes, and vascular disorders [119]. The incidence of hemobilia due to interventional procedures, e.g. liver biopsy [120], and transjugular intrahepatic portosystemic shunt, is increasing.

It is now as frequent as that of external trauma which used to be more common in the past[121, 122]. The risk of hemobilia following medical interventions is higher if vascular tissue e.g. hemangioma is punctured, if there is portal hypertension or when there is bleeding tendency [119]. Vascular disease is responsible for only 10% of cases of gross hemobilia [123]. Rupture of an aneurysm into the biliary tree is the main mechanism [124,125]. The frequency of true aneurysms of the hepatic artery is diminishing. This is mainly due to the decreasing incidence of mycotic aneurysms, though they are still reported with lesser frequency [126]. Atherosclerotic aneurysms are more common [127] and pseudoaneurysms, caused by iatrogenic injury to the hepatic or cystic artery are more frequently reported [128]. Macroscopic hemobilia due to gall-stones is rare and comprises only about 10% of all major hemobilia cases reported. It usually occurs when large stones erode the cystic artery [129]. Tumors play an insignificant role as a cause of hemobilia comprising only 6% of all cases of major hemobilia [119]. Giant hemangioma is the most frequent tumor to cause hemobilia [130].

The symptoms of hemobilia include gastrointestinal bleeding (melena in 90% of cases and hematemesis in 60%), biliary colic in 70% and jaundice in 60%. These events constitute the pathognomonic symptom triad of hemobilia [131]. Gastrointestinal bleeding in connection with biliary symptoms should always arouse suspicion of biliary tract hemorrhage, however minor hemobilia runs an entirely symptomless course, but it may occasionally show all the signs of solid clot formation [122]. Upper gastrointestinal endoscopy should be the first measure to be done in order to rule out other bleeding sources and blood flowing from the papilla of Vater can be visualised. Blood clots can sometimes be detected in the CBD by the ERCP[121.] Cholangiography, either postoperative through a T tube or by means of ERCP or PTC, may reveal a lesion by contrast filling of a cavity or by demonstration of clots which will appear as defects in the contrast [120]. Magnetic resonance imaging proved to be a useful tool that obviates the need for the invasive ERCP. MRI does not only show clots, as is the case with CT-scan, but it can also demonstrate active bleeding. Selective arteriography of the hepatic artery is the best way of verify a suspected diagnosis of hemobilia which will reveal the source of a major bleeding in most cases by displacement of the vessels around a liver mass, or by filling a true aneurysm.[131].Contrast material may be seen travelling down the hepatic duct, thus proving the existence of a communication with the biliary tract. Arteriography is of special value in discovering central liver lesions which may be difficult or impossible to be localized during exploratory laparotomy.(sandblom,2000). Moreover, it is both diagnostic and therapeutic [118].

The treatment of hemobilia depends on the cause of the hemorrhage [115]. Arterial embolization has become the preferred treatment [118]. Abnormal communications between the branches of the hepatic artery and the biliary tract could be selectively occluded by several substances [132,120]. Another approach is injection with sclerosant meterial during ERCP [126]. Several advantages make embolization preferable to surgery in the treatment of hemobilia. The risk of hepatic necrosis should he minimal as the obstruction can be limited to a segmental branch of the hepatic artery. Embolization may be repeated easily if hemobilia should recur. Ligature of the hepatic artery, or one of its branches, is nowadays reserved for cases where the abdomen has been opened for exploration or repair and when embolization is not possible or has been unsuccessful [119]. When an aneurysm only leaks, selective arterial embolization can be tried first, however if this step failed or the aneurysm ruptured into the biliary tract emergency operation with vascular reconstruction by excision and graffting should be attempted [131].

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