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Biliary Atresia: A Report from the King Abdulaziz University Hospital in Jeddah, Kingdom of Saudi Arabia

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Abstract. Biliary atresia is under misconception notions or the rarity of the problem is unknown in Saudi Arabia. An experience at King Abdulaziz University Hospital, Jeddah, Saudi Arabia, reported in the management of biliary atresia. A retrospective study of cholestasis, surgically explored between January 1997 and December 2010. The data collected included age at presentation and operation, sex, diagnostic methodology, operative procedures, histopathology reports and post-operative results. Twenty-seven cases of biliary atresia; 17 males and 10 females; mean age at presentation was 62 days; mean age for surgical exploration was 75 days. Twenty-four cases of Type A variety; and three of Type B. Last follow-up, 16 (59.25%) live patients; 2 developed esophageal varices; 3 underwent liver transplants; 6 recur jaundice and liver functions deteriorate after initial improvement among the longest survivors. 2 patients with esophageal varieces; 1 patient underwent a liver transplant. 11 (40.7%) deaths caused mainly by severe cholangitis in the early post-operative period and liver failure afterwards. Concluding, state of liver is important for a successful procedure and esophageal varieces has no relation with the length of survival. Biliary atresia is not rare and the lack of publications is due to a delay in the diagnosis and insufficient reporting.

Keywords: Biliary atresia, western part of Kingdom of Saudi Arabia

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Introduction

Biliary atresia (BA) is a serious cause of obstructive jaundice in neonates. In this condition, the bile ductules are stenosed or obliterated and bile flow is prevented leading to stagnation of bile within the liver cells, with a variable degree of liver fibrosis. The incidence of this anomaly ranges between 1 in 10,000 to 1 in 17,000 live births with a slight female preponderance[1-3]. The etiology of this condition is not clearly known but several assumptions have been postulated. These include a viral infection, an autoimmune reaction, and failure of remodeling and canalization of the biliary ducts. However, some researchers believe that there is enough evidence to conclude that BA occurs as a result of an intense immune response to an inflammatory process, involving the biliary ducts in a genetically susceptible patient[4-6].

Three distinctive types of BA are known to occur. The most frequently seen is the type A, where the entire extra hepatic system is obliterated. Type B, in which the distal parts (that is, the gallbladder, cystic and common bile ducts) are patent, and type C, where the hepatic ducts are patent[7]. The Japanese Association of Pediatric Surgeons has another system for classification, which is more or less similar[8].

The clinical presentation of this condition is a progressive jaundice accompanied with alcoholic stool within few days after birth, and a failure to thrive. Similar manifestations are presented by other conditions like neonatal hepatitis, inborn errors of metabolism, inspissated bile syndrome and biliary hypoplasia, which make clear differentiation and preoperative diagnosis difficult[9]. Biliary atresia is commonly suggested by the presence of high direct serum bilirubin levels, absent or contracted gallbladder, and extra hepatic ducts by sonographic examination of the abdomen[10-13]. In addition to the failure to demonstrate bile flow into the gut by radioisotope scanning of the liver using technetium 99m Tc-labeled diisopropyl iminodiacetic acid (DISIDA)[14-16]. Investigations like liver biopsy and magnetic resonance cholangiopancreatography (MRCP) may be more sensitive; however, laparotomy and laparoscopy remain the most accurate methods of diagnosis. Delayed diagnosis and late referrals often lead to progressive liver damage and fibrosis which are the main causes of poor prognosis[17,18].

Biliary atresia is generally managed by drainage of the bile at the porta hepatis or by liver transplant. Several techniques describe to

reestablish bile flow from the porta hepatis. The most popular one is the Kasai procedure reported in 1957 and its subsequent modification (Fig. 1), which results in nearly one third of the cases being successful. The rest of the cases either deteriorate or show no improvement, unless those that can be salvaged by a liver transplant[19,20]. Liver transplants can be carried out as a primary procedure depending on the degree of fibrosis and feasibility of transplant services. It has also shown marked improvement in the 5 and 10 year survival rates and has become an integral part of the management of BA[21,22].

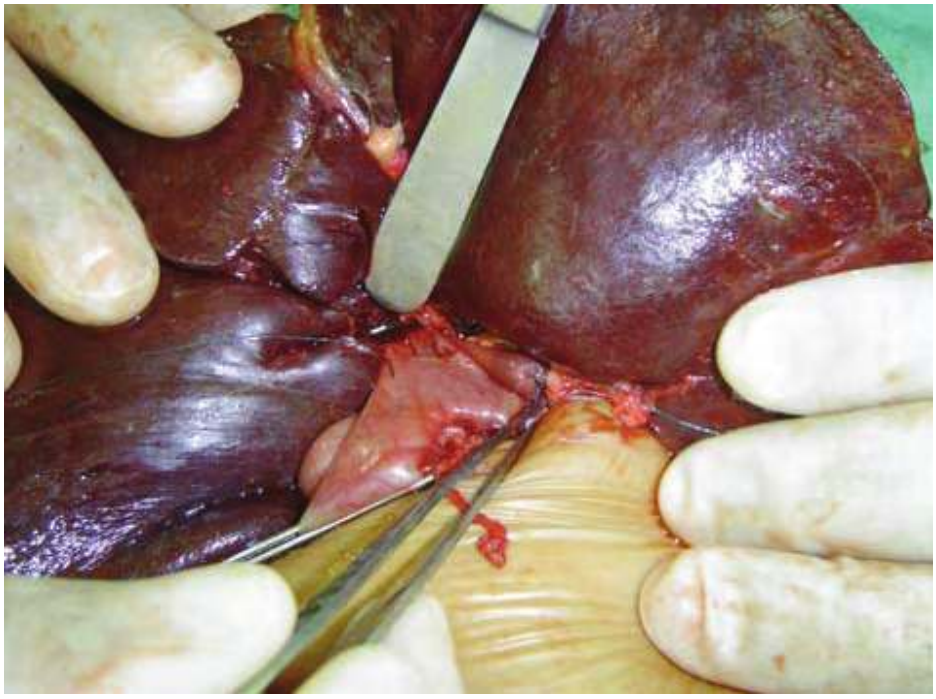

Fig. 1. Portoenterostomy (modified Kasai) in a case with minimal liver fibrosis operated after the age of 3 months.

Materials and Methods

A retrospective data analysis was carried out from the medical records of patients diagnosed and managed for BA at our hospital, the King Abdulaziz University Hospital, Jeddah, KSA. All patients with BA between January 1997 and December 2008 were included. Data collection included age at the onset of symptoms and referral to our hospital, sex of the patient, types of investigations, age at which the operation was performed, type of procedure carried out, and the results of their follow up.

Patients were divided into two groups: Group 1 operated at an age between 65-90 days, while Group 2 was those operated at the age between 90-130 days.

Results

Out of 29 cases reviewed, 27 (93.1%) were found to have BA; the other 2 had biliary hypoplasia and were excluded from the analysis. Among those included were 17 males and 10 females of ages at presentation between 32-135 days (mean age of 62 days). The basis for surgical exploration included, a high serum direct bilirubin level (more than 2/3 of total), elevated liver enzymes, non-visualization or contracted gallbladder. In addition, extra hepatic ducts by ultrasound and the absence of tracer in the small bowel by DISIDA. Since the introduction of MRCP to our hospital, this investigation and pre-operative liver biopsies were carried out in 3 patients in addition to previous workup, and all were found to be positive for BA at exploration.

Group 1 consisted of 11 patients while Group 2 covered 16 patients. All patients were surgically explored under antibiotic cover (metronidazole with a third generation cephalosporin). At exploration, variable degree of liver fibrosis was seen (fig 2). All patients underwent Tru-cut liver biopsy and an intraoperative cholangiogram whenever the gall bladder was seen.

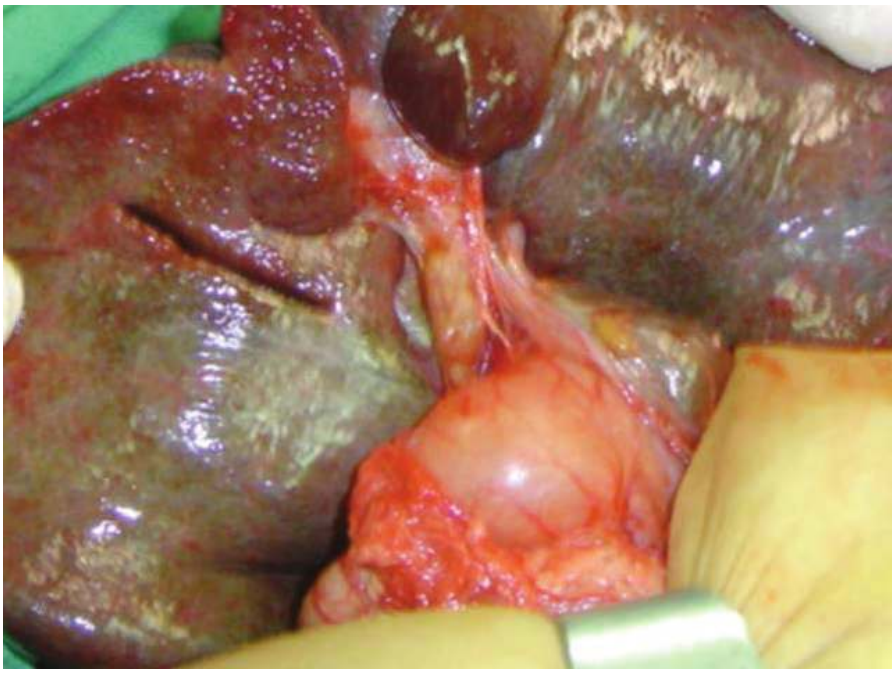

Fig. 2. A case of extra hepatic biliary atresia, with moderate degree of liver fibrosis.

Three cases were found to be type B (11.1%); hence, subjected to hepatic porto cholecystostomy, whereas 24 patients were found to be type A (88.9%), and they had undergone the Roux- en- Y hepatic porto jejunostomy (modified Kasai). No case of type C was seen. All patients, except two, had a smooth post-operative recovery. Mixed greenish stool was seen in 16 patients between the 7^{th} and 12^{th} post-operative days, which subsequently diminished in nine cases. There was no color change in the stool of 11 patients. The mean hospital stay was 16 days. Out of the 27 cases, 16 patients (59.25%) were alive and 11 cases were lost (40.7%). The main cause of death was generalized sepsis in the early post-operative period (5 cases) and progressive liver failure (6 cases) afterwards. Three of the survivors were from Group 2; 5 patients were jaundice free with normal liver functions, 2 patients developed esophageal varices (treated by multiple sclerotherapy), 3 patients had liver transplants at another institution. In six patients, however, jaundice recurs and liver functions deteriorate after initial improvement and considered for transplant. The mean survival age was found to be 6.3 years. The longest survival was observed in the 2 cases with esophageal varices, (11 and 9 years) and in one patient who underwent a liver transplant (8 years). A summary of our results is shown in Table 1.

Table 1. Summary of results obtained after management of biliary atresia at KAUH.

Patient Characteristics	No. (Percentages)
Established Bile Flow with normal liver functions	5 (18.5%)
Esophageal Varices	2 (7.4%)
Liver Transplant	3 (11.1%)
Deterioration after initial improvement	6 (22.2%)
Deaths	11 (40.7%)
Total Patients	27 (100%)

Discussion

Biliary diseases in children are infrequent and can be associated with high morbidity and mortality rates if an accurate diagnosis is not made, and adequate treatment is not provided at the appropriate time. Surgical and non-surgical conditions can be associated with similar clinical manifestations, laboratory and radiographic findings, which can make accurate diagnosis difficult. The right treatment for each of these clinical entities is different and, when adopted can significantly reduce morbidity and mortality from these diseases[23-26]. In BA, the surgical management

aims at restoring bile flow from the porta hepatis into the small bowel or by liver replacement.

Several attempts to reestablish bile flow have been attempted since 1953 with success noted only in few cases[27,28]. In 1957, Kasai reported the Roux- en- Y hepaticoporto enterostomy in a case with minimal liver fibrosis operated after the age of 3 months. Modification of this procedure omits stoma placement and remains the standard treatment of BA. Following the Kasai procedure, a great reduction in the early mortality was noticed, and primary biliary drainage was achieved in nearly 60-80% of the cases within 10-14 days. However, half of these patients could go on to develop progressive liver damage, leaving only about one third with a good bile drainage[29]. There are several factors which determine the success of this procedure. One of these is the age at which the operation is done. Better results have been observed among patients who are less than 3 months of age. Other factors for success are the degree of liver fibrosis, the presence of bile efflux from the porta hepatis after dissection, and the size of the bile ductules on histological examination of the portal plate. Sharma and Roy in two different reports showed that there is no significant difference between survival and age at which the operation is performed. They concluded that advanced histological findings in the form of greater fibrosis and ductal plate malformations even in the younger age group are associated with a poor prognosis and consider BA as an emergency condition[31-33]. The 10 year survival after the Kasai procedure has steadily increased and currently ranges from about 40-60% with a subsequent reduction to 20-30% at the age of 20 years[34,35]. Complications like portal hypertension and bleeding esophageal varices may develop after this procedure, particularly in those associated with severe liver damage and fibrosis[36-38].

Patients who do not respond or deteriorate after the Kasai procedure may be salvaged by a liver transplant. Hence, liver transplant is an integral part of the rescue therapy for BA and accounts for nearly half of all pediatric liver transplants. The dramatic improvement in survival (overall 85%) after liver transplant with advances in immunosuppressant therapy, has raised the question of conducting a primary liver transplant. However, it remains a debatable topic among pediatric surgeons as nearly half of the patients achieve adequate liver function after hepatic portoenterostomy which supports the delay of liver transplants in addition to the difficulty in obtaining suitable donors[39-41]. The 5 years

survival rate after liver transplant reached up to 78% in a Japanese series of 1400 patients, but reduced to 53% at 10 years, which is approximately the survival rate following the Kasai procedure[8]. In another study, De Vries from the Netherlands, recommended early operative listing of patients who had not improved after portoenterostomy for liver transplant in order to avoid the high pre-transplant mortality associated with sepsis[42].

In this review, it was found that the incidence of BA to be slightly higher in males than females (63% and 47%, respectively). The accuracy of our investigation procedures was 93%, although in only 3 cases, the addition of MRCP and liver biopsy to the other investigations rendered diagnosis to be more accurate. More than half of our cases were operated after 3 months of age; among them 2 patients were the longest survivors, indicating that survival may relate to the degree of liver pathology at the time of the procedure and not due to the advanced age. This observation is similar to the reports of Sharma and Roy. This study also found that complications like esophageal varices after the Kasai procedure does not affect the survival. In this series, it also noted that infection is the main cause of death in the early post-operative period, with the occurrence of liver failure later on. Liver transplant was carried out in 3 patients, but a favorable outcome was achieved only in 1 of the 3 patients. Additionally, none of the patients who underwent the transplant reached 10 years of age.

There are only few reports from Saudi Arabia about BA, all from the Central and Eastern parts. In 2 articles, a total of 29 cases of BA were reported without mentioning of the outcome[43,44]. Another case report was published in association with multiple congenital anomalies and hyaline membrane disease[45]. This report on BA is probably the first to come from the Western region of Saudi Arabia with some details regarding management and outcome. The reasons for this inadequate reporting could probably be due to the delay in diagnosis and untimely referrals to larger centers which is similar to a report from Brazil[46].

Conclusion

The exact incidence of BA in KSA is unknown; however, the condition is not rare. Limited publications and data may be attributed to delayed diagnosis of the condition and inadequate reporting. However,

facilities for investigations are available in large centers in KSA, and our results of management coincide with other globally reported cases.

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انسداد القناة الصفراوية الخلفي: تقرير من مستشفى جامعة الملك عبدالعزيز في جدة ، المملكةالعربية السعودية

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جدة – المملكة العربية السعودية

المستخلص. إن ضمور القنوات الصفراوية الخلقي يعتبر من الأمراض الصعبة والنادرة، وعلى الرغم من توفر العلاج لهذا المرض في مستشفيات المملكة إلا أن ما تم نشره عن هذا المرض ونتائج علاجه في مستشفيات المملكة يعتبر قليلا، قياسا بالأمراض الجراحية الأخرى لدى الأطفال. ويعود ذلك إما للتأخر في التشخيص اوالتقصير في النشر في هذه الدراسة نستعرض تجربة المستشفى الجامعي بجدة في علاج هذا المرض. فقد قمنا بمراجعة سجلات المواليد المصابين بالصفراء الاحتقاني خلال الفترة من يناير ١٩٩٧ إلى ديسمبر ٢٠١٠م واستخلاص حالات ضمور القنوات الصفراوية، وشملت الدراسة: جنس المولود، عمر المريض عند الإحالة وعند الجراحة، طرق التشخيص، نوع العملية الجراحية، المضاعفات بعد الجراحة والحالة العامة للمتعافين. كان عدد الذكور ١٧ وعدد الإناث ١٠ ومعدل العمر عند التشخيص والإحالة ٦٢ يوما وعند الجراحة ٧٥ يوما. تمت العمليات الجراحية في ١١ طفل في عمر أقل من ٩٠ يوما و ١٦ طفل في عمر أكثر من ٩٠ يوم. كان عدد المصابين بهذا المرض ٢٧ مريضا، ٢٤ منهم مصابا بالنوع أو ثلاثة بالنوع ب. كان عدد من تبقوا على قيد الحياة ١٦ مريض (٥٩,٢٥٪

بمعدل عمر ٦,٣ سنة، اثنان منهم أصيبوا بدوالي المريء، ثلاثة أجريت لهم زراعة كبد ولا يزال ستة مرضى في حالة غير مستقرة بسبب تدهور وظائف الكبد وعودة ألصفاري ويحتاجون لعمليات زراعة كبد. لوحظ أن من الأطول عمرا أولئك اللذين حصل لديهم دوالي المريء وواحد ممن أجريت له زراعة الكبد. و كان عدد الوفيات ١١ (٤٠,٧٪) بسبب الالتهابات الحادة في القنوات الصفراوية خلال الأيام الأولى بعد العملية، وتدهور وظائف الكبد لاحقا. نستنتج من ذلك أن حالة الكبد عند إجراء العملية من عوامل الاستجابة، وأن حصول دوالي المريء ليس له تأثير في عمر المريض بعد الجراحة كما أن هذا المرض ليس نادرا وعلاجه متوفر بالمملكة.