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Biliary Atresia Outcome in Egypt: A Descriptive Study

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Abstract

Biliary atresia is a rare disease that affects children and neonates, nowadays the current management of BA is portoenterostomy with or without liver transplantation, in this study we described the outcome of BA in single center .

We included 48 patients in this study with type 3 BA and measured the incidence of clearance of jaundice and cholangitis the incidence of clearance of jaundice was 43% and cholangitis was 20% also the mean age was 74.6±21.8 days which is quiet high.

We concluded that early diagnosis and centralization is the key point for getting best outcome for patients of BA.

Keywords: *Biliary Atresia, Porto entersostomy, Jaundice clearance, cholangitis.*

Introduction

BA is an obstructive cholangiopathy, an inflammatory process that affects both intra and extra hepatic bile ducts causing obliteration of the bile ducts ends up to cirrhosis and liver cell failure if left untreated(1).

Although it is an uncommon disease occurring in 1/10000- 1/15000 live births world wide it It is rare but with obvious geographical variation as in Europe and North American for example it has an incidence of 1 in 15–,20,000 live births and by contrast can be seen in up to 1 in 5000 live births in Taiwan and presumably mainland China. It is not a homogenous, uniform disease and within the umbrella term are several variants which have separate and distinct causes and different outcomes. These make comparison of treatment options difficult but not impossible(2).

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The etiology of BA I unknown but thought to be multifactorial, currently there is 3 main types of BA; cystic type of BA, isolated BA and syndromic BA which is thought to be congenital in nature(3).

The first porto enterostomy was done by Prof Moro Kasai 1953 by chance while he was dissecting in the porta hepatis and noted bleeding then he put the dudenum for hemostatsis then the patients stool got coloured since then many modifications has been done to this operation and became the first line of treatment of BA with or without liver transplantation(4).

There are many factors that affects the outcome of Kasai procedure, the most important factor is patients age at time of operation, the second is the type of BA the best in prognosis is cystic type of BA while the worst is the syndromic BA, attacks of cholangitis also affects the outcome also adjuvant therapy may play a role in improving the outcome(3).

The prognosis of BA has dramatically changed in the last decades: before the Kasai operation most BA patients died, while nowadays with the sequential treatment with Kasai operation, with or without liver transplantation, BA patient survival is close to 90%. Early diagnosis is very important since the chances of success of the Kasai procedure decrease with time(5).

In Egypt and due to limitation of liver transplant in patient with BA below one year of age we have to do our best to increase the success rate of the porto- enterostomy in patient with BA including both clearance of jaundice and the percentage of native liver survival

Patients and Method

48 patients were included in this trial we included all cases of BA type 3 after investigations were done including Lab investigations which showed direct hyper bilirubinemia, elevated GGt and Alkaline phosphatase,ultrasound showed absent or non contractile gall bladder, liver biopsy showed marked inflammation and bile duct proliferation .

All cases were operated and managed at Cairo University Children Hospital (CUPSH) and after referral to the surgical department the case is admitted 48 hours before surgery and preoperative labs were done, blood crossmatching and preoperative antibiotics were given .

On the OR patients were anasthesized supine with endotracheal intubation and sonar guided CVL .

The infant was positioned supine to ensure a non obstructed view for cholangiography,which was not routinely required .surgical exploration commenced through an extended kocher`s incision centered to the right ..this incision could be extended across the midline if biliary reconstruction was required . he the left upper quadrant was examined to determine splenic anatomy .

Liver consistency was noted and the porta hepatis inspected.. Cholangiography has a distinct role in this patients.

If the gallbladder had a lumen, diluted contrast material (diatrizoate Hypaque) was injected to determine the continuity of the biliary tree between the liver and duodenum. Complete extrahepatic duct patency was confirmed when contrast appears distally in the duodenum and proximally in the liver radicles. The size of the ducts is assessed by cholangiography to determine whether biliary hypoplasia or obstruction to bile flow from inspissation was present. If the contrast material moved only distally into the duodenum and not into the liver, gently occluding the distal common bile duct while reinjecting the gallbladder to encourage proximal ductal filling, although this maneuver rarely succeeded. If extrahepatic biliary patency is demonstrated, regardless of whether the ducts were of normal caliber or diminutive, a liver biopsy is performed and the incision closed.

If ductal patency was not confirmed, portal dissection and biliary reconstruction was performed (the Kasai procedure).

According delivery of the liver some surgeons preferred to deliver the both lobes of the liver from the abdominal cavity after dividing both triangular ligaments, some preferred to deliver the left lobe only and minority prefer not to deliver either lobes and doing the portoenterostomy insitu.

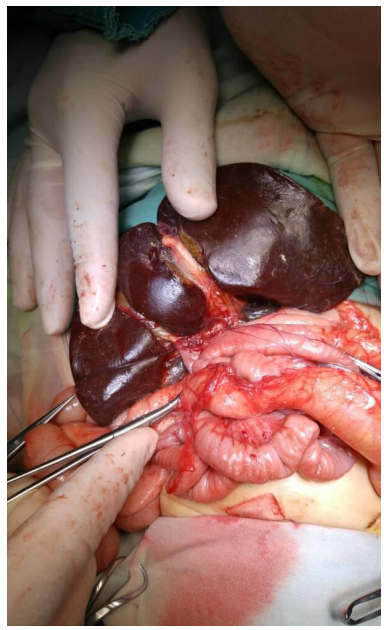

Figure 1: Liver delivery before dissection

The fibrous common duct was ligated distally with nonabsorbable suture and transected. Gentle traction applied to the remnant facilitates dissection toward the porta, where a cone of fibrous tissue anterior to the bifurcating portal vein is encountered.

When there was discontinuity of the biliary tree, exploration of the tissue overlying the portal vein bifurcation will almost invariably reveal the fibrous cone. Placing a suture in this fibrous tissue will facilitate establishing the plane above the portal vein.

Once dissection was complete, fine stay sutures were placed at the lateral margins of the fibrous cone. While maintaining moderate tension, the fibrous cone was sharply transected,

The resected biliary structures with attached gallbladder remnant are sent for pathologist to measure

the size of the biliary ductules for later prognostic evaluation.

Recently we standardized our technique by making wide portal plate extending from Rex fossa to the gall bladder fossa (innominate fossa) for allowing maximum drainage of bile through the porto-enterostomy .

The porto-enterostomy was done retro colic through an incision in the transverse mesocolon just to the right of the middle colic artery.

The prto-enterostomy was done end to side by making an incision of the anti-mesentric border of the roux limb 1-2 cm of its distal end to avoid blind pouch and then the anastomosis is done using parachuting technique for the posterior wall which allows better vision.

After completing the porto-enterostomy the roux limb was sutured to the glisson`s capsule and transverse meso colon to decrease the tension on anastomosis.

Drain was usually inserted and removed on the 5th day post operative.

Regional anaesthesia (TAP block) is injected at the lateral border of rectus sheath to decrease postoperative pain, the dose was calculated through the anesthesia team.

Abdomen was closed in 2 layers and Subcuticular closure for the skin was done.

Postoperative Management: Patient was kept on the ICU on nasal oxygen if needed with proper analgesia, Iv fluids and post operative CBC was done routinely to detect any need for blood transfusion.

Gradual feeding was introduced once intestinal motility was regained

Patient was followed up through CBC with differential and CRP to detect early sepsis and upgrade antibiotic line accordingly.

Discharge: Patients was discharged when they reached their full feeding with no fever and no laboratory signs of sepsis.

Outpatient follow up: After discharge Patients were followed up regularly at the hepatology clinic for follow up.

At day 12 post operative, 1,3 and 6 months postoperative.

General medical examination was done, jaundice is detected recording of any complication such as cholangitis, etc.

The following data were recorded: Hepato splenomegaly, Presence of jaundice, Colour of stool, Fever and cholangitis.

Results

Concerning our patients the range of age was **(30-118)** with a mean age was **74.6±21.8** . 25 were males (52%) while 23 were females (48%)

In our study 25 cases were male (52.1%) and 23 cases were female (47.9 %)

In our study 21 cases (43.8%) were jaundice free within the first 6 months postoperative .

27 cases **(56.3%)** were still jaundiced at 6 months postoperative . 35 cases (72.9%) has coloured stool while only in 13 cases (27.1%) clay coloured stool have persisted.

The median preoperative bilirubin was 9.8 mg/dl while the median bilirubin after 6 months were 5.1 mg/dl.

Discusson

Although our study seems to be intermediate in number of cases and this limits the presence of statistical significance of the data but this happened however this happened due to relatively short duration of the study which was only 2 years, Davenport made his trial over 11 years and the START trial was done over 6 years[(1)].

Concerning age in our study the mean age was74.6±21.8 (30-118), 14 cases were less than 60 days (29.2%),23 cases were 60-90 days (47.9%) and 11 cases were 90-120 days (22.9%).

By comparing the mean age with the mean age of the similar studies that discuss the use of steroids the mean age of this study is the oldest age, in the study of Tyraskis and Davenport 2016 the mean age was reaching 46 days (12-70) days[(1)]. while In the START trial done be Bezzera 2014 the mean age was 69 days[(6)], also at the study of Escobar 2006 the mean age was 43 days[(7)], the mean age of the study of Petersen 2008 was 62 days[(8)]. Chung 2008 also at has a mean age of 70 days[(9)].

From the previous data we can conclude that the youngest age was at Davenport study (45 days (12-70)) and we think that this could be achieved by the centralization system that the NHS provide to allow early detection and management of those cases and due increased awareness of this disease among pediatrician[1].

On the other hand in EGYPT and due to lack of awareness of this disease among pediatrician and due to lack of ability of differentiation between BA and physiological jaundice there is a delay in the management of these cases, In our study we excluded the cases aged above 120 days due to controversy about benefit of porto-enterostomy for those patients and if we didn't exclude those cases the mean age would be much higher .

Conclusion

For achieving best outcome for patients of BA two main factors should be present the first is early diagnosis and management of biliary atresia either by screening or increasing awareness of this disease between pediatricians the second factor is centralization which should increase surgical success.

Ethical Clearance: from ethical committee cairo university 2015 ethical comitte approval number I-111015.

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Conflicts of Interests: Nil.

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