DIFFERENTIAL DIAGNOSTICAL AND POLICY CONSIDERATIONS IN THE SURGICAL MANAGEMENT OF BILIARY TRACT ATRESIA IN NEWBORNS

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Congenital biliary tract atresias are rare abnormalities, occurring once in 20 000—30 000 births (Moore, Smith). However, on account of the insufficient differentiation of the hepatic parenchyma and due to the immaturity of hemopoiesis, the icterus as a symptom is frequently encountered in childhood and particularly in newborns. Our personal experience in this respect dates back 8 years ago and comprises 18 children doubtful for congenital anomalies of the biliary tracts, admitted and treated at the childrens' surgical department (Chair of Surgical diseases at the Higher Medical Institute, Varna), of which the diagnosis was confirmed in 12 — all undergoing surgery. In the remainder, transient conditions of mechanical icterus were involved, controlled by conservative (bile driving) therapy. The newborns with certain blood or isoimmune affections are not included in the latter group, and were left for treatment in the Children's Clinic or Maternity homes. The comprehensive and detailed clarification of all differential-diagnostical features and difficulties confronted in the course of work is not the aim of the present report and that is why we will limit ourselves with brief references to the most frequently met with issues in consulting icterus forms among newborns, suspected for congenital anomaly.

In the children with icterus gravis or erythroblastosis fetalis, we usually established icterus occurring in the very first hours after the birth, strongly impaired general condition and early enlargement of the spleen. As a rule, the stools were coloured.

Infectious hepatitis among the newborns was recorded very rarely, by way of exception in the early weeks of life. In these children, the general condition was invariably disturbed to a lesser extent, the liver and spleen were always enlarged, and the stools — initially normal, and subsequently — acholic.

Septic icterus was met with rather frequently. In this group of children a basic affection was always manifested, conditioning the exclusively heavy state of the child. The spleen and liver were similarly always enlarged, and the stools — normally tinged.

Luetic icterus was observed only in one case. The stools of the child were normally tinged.

The children with hemolytic icterus have been usually severely involved, with presence of pronounced anemia and the stools — normally tinged.

In one of the children consulted, we established galactosuria with icterus occurring in the second postbirth month. The child was with impaired general condition, with liver and spleen — enlarged and stools — with normal color. Evidence were found also for moderately pronounced ascites in the abdomen.

In some of the children physiological icterus with protracted course was the diagnosis established, and even up to the 2nd month, we failed to find out the cause for the condition.

The syndrome of Krigler—Nadjar or Idiopathic bilirubinemia, conditioned by ferment insufficiency of glucoronil-transferase, with absence or greatly reduced potential of indirect bilirubin binding to glucuronic acid, was observed in one child with intrahepatic cholostasis.

In immature infants, in the first post-natal days, not infrequently icterus was noted, caused by the toxic damage of erythrocytes with certain medicaments (vitamin K, etc). In the latter group the so-called biliary thrombi, resolving spontaneously after conservative treatment, were observed rather more frequently.

There are literature reports on rare cases of icterus in hypertrophic pyloric stenosis, due to the compression of the biliary tracts by the hypertrophic pylorus. No such case was referred to us.

In general outline, the range of etiological factors, leading to jaundice in the newborns, is very wide and comprises a number of parenchymal, blood, isoimmune and surgical affections, confronting the children's surgeon with serious diagnostical difficulties. Usually, children with protracted jaundice are being referred to the surgical department. The assumption is universally accepted that icterus in the early post-natal period has no relationship whatsoever with the congenital anomalies of the biliary tracts, and this group of children is referred most frequently to pediatrists or infectionists. The children with congenital atresiae of the biliary tracts at birth and within several days thereafter, are usually completely normal, the meconium is coloured and the initial clinical manifestations are hardly differentiated from the so-called physiological icterus of the newborn child. However, if the icterus persists after the third month of life, with the stools becoming colorless and the liver enlarged, despite that the general condition of the newborn is not particularly impaired, the problem is seriously posed concerning the presence of a mechanical obstruction somewhere along the course of the biliary tracts (mucous valves, concrements, congested bile, compression from outside, congenital atresia and the like).

Most of the children undergoing operation at our unit were hospitalized during the second month of life and only two were admitted at the age of 3 months. In 6 of the children, during the early months of pregnancy, the mothers sustained various infectious conditions.

The biliary tract abnormalities were presented in the following variants according to the classification of G. A. Bairov: 1. Complete aplasia of the biliary tracts — one case; 2. Intrahepatal atresia — two children in whom the operation disclosed once atresia of the common bile duct and a second time — atresia of the gall-bladder, ductus cysticus and of the common bile duct; 3. extrahepatal atresiae — in 9 of the children. Three of them were with low atresia in the region of the choledochus. External compression

of the biliary tracts by scars, neoplasms or hyperplastic lymph nodes was not encountered. In all the cildren the exact diagnosis was established on the operative table, resorting to intraoperative cholangiography in some of them, wherever and whenever practicable. As evident from the variants in our case material, a comparatively low percentage of the forms appear to lend favourably to radical surgical management. For the timebeing, no more than 15—20% of the children with biliary tract anomalies are considered operable. During the preoperative and postoperative periods, we adhered to the following policy: the requirements in biliary tract operations in newborns have been invariably equaled to those in extensive laparotomies, implying intense preoperative preparation and after-treatment. The preoperative preparation usually covered the following four basic requirements: sanation of the respiratory ways, emptying of the stomach, adequate water-electrolyte substitution, regulation of body temperature. In the postoperative period, parallel to the above measures, of no less importance appears to be sparing and restoring the liver therapy, vitamin K_1, struggle with shock and intestinal atony, prophylaxis of the pulmonary complications and infection. Treatment in the early stages was invariably commenced with conservative means — duodenal probe, intravenous administration of acholin, magnesium sulphuricum 25%, per os or with probe, diluted with a small quantity of mother's milk, 2—3 times daily, etc. The optimal term for performing the intervention was considered the time up to the second month of life, regardless of the fact that owing to the late reference of the children, some of the operations were made after the above term. Those operated later, usually ended lethally from irreversible hepatic damage, intercurrent infections or hemorrhages on the basis of portal hypertension. It is assumed that without operation, depending on the form of the anomaly, the children could have survived up to the 7—8 month of life and even to 2—3rd year. Svensson, published a case report as a rare exception, surviving up to 9 years.

Of all the children undergoing operation, a permanent outcome was attained only in two, operated in early terms, with normally developed intrahepatic biliary tracts and presence of part of the extrahepatic ways, thus permitting for bileduodenal anastomoses. In one of the cases, the anastomosis was made with the gall-bladder, and in the second case — with the anterior part of the choledochus. In all the remaining children, we had to limit ourselves with performing the palliative by-pass anastomoses between the liver and duodenum or stomach. In this respect, we have performed technicalwise, three different types of interventions, namely: 1) Anastomosis between the left hepatic lobe and the stomach. 2) Hepatoduodenoanastomosis in the area of porta hepatis and 3) Hepatoduodeno- or hepatogastroanastomosis, combined with cannulation of the liver parenchyma with a polyethylene tube. In two of the children, a personal method was applied, consisting of cannulation of the two hepatic lobes by means of thick cat gut, threaded at 5—6 points through the full thickness of the liver, and converged in a single bundle, penetrating the anastomosis between the duodenum and liver in the area of porta hepatis. In both children visible improvement of the general condition ensued, coloration, although temporary, of the stools, and reduction of the liver size to a certain extent. Anyway, the children died within 7—8 months after the operation from progress-

ing hepatic insufficiency. Very suitable for anastomosis, especially with the duodenum, was the bed of the aplastic gall bladder, where usually a small circular, cicatricial edge of fibrous tissue is found, permitting stable fixation of the liver, without undesirable bleeding. Upon performing the tunnelization of the liver by means of cat gut threads passing through its full thickness from all the directions towards the bed, we did not come across hemorrhages worth noticing. To avoid the cutting of the cat gut knot overlying the liver surface, the latter was enveloped in a button of small omentum pieces.

In conclusion, we would like to make some practical inferences concerning the operative treatment of congenital atresias of the biliary tracts, namely:

1. All newborns with physiological icterus, running a protracted course after the third week, should be mandatorily considered as doubtful for the presence of congenital anomalies of the biliary tracts.

2. From 0 to 2 months of life is to be accepted as the optimal time for operation.

3. The preoperative preparation and the postoperative cares should be similar to those required for extensive surgical interventions and laparotomies.

4. In timely performed palliative hepatodigestive anastomoses, the general condition of the children is improved, and regardless of their nonradicality, such interventions should be invariably resorted to.

5. In operable forms (operable in terms of radical interventions), the children tolerate very well the surgical intervention and, according to our personal observations, sustain more rarely than adults postoperative biliary fistulae as a complication. In our series a similar complication was not observed in any of the cases.

6. Biliary-hepatic surgery in newborns with biliary tract anomalies, unfortunately, is still hardly in a position to solve definitely the problem of their radical surgical management. Probably, the final solution of the issue would follow the solution of the question concerning the transplantation of parenchymal organs.

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ДИФФЕРЕНЦИАЛЬНО-ДИАГНОСТИЧЕСКИЕ И ТАКТИЧЕСКИЕ ЗАМЕЧАНИЯ ПРИ ХИРУРГИЧЕСКОМ ЛЕЧЕНИИ АТРЕЗИЙ ЖЕЛЧНЫХ ПУТЕЙ У НОВОРОЖДЕННЫХ

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РЕЗЮМЕ

На основе 8-летнего опыта лечения и диагностики врожденных атрезий желчных путей у новорожденных, по поводу чего было оперировано 18 детей, автор делает ряд важных для практики замечаний относительно наиболее часто встречающихся во время консультаций форм желтухи у новорожденных, породивших сомнение на врожеденные аномалии, а именно: злокачественная желтуха, гемолитическая болезнь новорожденных, инфекционная желтуха, септическая желтуха, галактозурия, затяжная физиологическая желтуха, синдром Криглера—Наджара и др. Аномалии желчных протоков у оперированных детей были представлены в следующих вариантах: 1. Полная аплазия желчных протоков — один случай; 2. Интрагепатальная атрезия — два случая; 3. Экстрагепатальные атрезии — у 9 детей. Оптимальный срок проведения операции автор рекомендует не позднее второго месяца с момента рождения. Операции были главным образом трех видов: 1. Анастомоз между левой долей печени и желудком. 2. Гепатодуоденоанастомозы в области порта гепатис. 3. Гепато-дуодено или гепато-гастроанастомозы, в сочетании с канюлирование паренхима печени полиэтиленовой трубочкой. Автор применил на двух детях собственную методику, состоящую в канюлировании двух долей печени, посредством толстых ниток кетгута, проведенных через несколько участков сквозь всю толщу печени и собранных в один пучок, впадающий в анастомоз между дуоденумом и печенью в области порта гепатис. Полученные автором результаты позволяют ему сделать вывод, что несмотря на нерадикальность большинства операций при атрезиях желчных протоков у новорожденных, эти операции все же обязательны.