A diagnostic score for biliary atresia

To the Editor:
We read with great interest the paper by El-Guindi et al. [1], evaluating a new diagnostic score for biliary atresia (BA). BA is a destructive inflammatory obliterator cholangiopathy of unknown origin that affects intra- and extrahepatic bile ducts. Success of surgical correction, in form of a Kasai portoenterostomy amongst other factors, depends on early diagnosis and timely intervention. Presently, there are no non-invasive diagnostic methods that can clearly identify infants with BA from other causes of neonatal cholestasis (NC). The current paradigm to exclude BA in any cholestatic infant hence requires evidence to prove patency of the extrahepatic bile duct [2–5]. To date, many research groups have tried to improve the differential diagnosis [6] but none has yet reached sensitivity and specificity as high as in the present study. We are convinced that elements of the proposed scoring may help in the pre-selection of infants to undergo imaging of the extrahepatic bile duct. However we are concerned that the study cohort of patients investigated might be skewed, thus limiting the diagnostic power of this score.

(1) Entry criteria did not describe if there were children with drug- or total parenteral nutrition-associated cholestasis or premature infants in the study population.

(2) There was no information whether there were children with the syndromic form of BA, with biliary stones, dilatation of bile ducts or choledochal cysts in the study cohort.

(3) Important and common causes of NC, such as alpha-1 antitrypsin deficiency, cystic fibrosis, bile acid synthetic disorders, tyrosinemia type 1, galactosemia and hypopituitarism were not identified.

(4) Normal or low values of gamma-glutamyl transeptidase (GGT) in a child with NC imply a defect of bile excretion at the canicular level. So it was not surprising that in the group of non-BA patients the level of GGT was lower than in the BA group. There was no information about the type of the progressive familial intrahepatic cholestasis (PFIC I or II) and whether the multi-drug resistance protein 3 (MDR3) was examined.

A variety of diseases can exactly mimic all separate features of BA and only systematic evaluation of the entire spectrum will lead to a reliable diagnosis. Only innovative tools, such as next generation sequencing, may possibly change this and contribute to a differential diagnosis in the future.

Easy diagnosis of BA has been on the agenda for decades. But, despite all efforts, as long as we still do not understand the aetiology and triggering factors of BA we remain at risk of missing the correct diagnosis for a child with NC just based on non-invasive diagnostics.

Conflict of interest
The authors declared that they do not have anything to disclosure regarding funding or conflict of interest with respect to this manuscript.

References

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Reply to: “A diagnostic score for biliary atresia”

To the Editor:
We appreciate the interest of Pfister et al. in our recent study [1] and would like to comment on the issues raised in their letter.

Although the concerns that were raised by Pfister et al. are interesting, none is related to the applicability of the BA score as the recruitment in our study was consecutive for all cases...