

PREMATURITY AND BILIARY ATRESIA: DOES IT MATTER?

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Aim of Study: The diagnosis of biliary atresia (BA) remains challenging and delay can lead to significant morbidity with time to surgery a key factor in determining outcome. Prematurity may impact on outcome potentially delaying diagnosis. We sought to assess whether the premature BA infants (PBA) have a delayed time to surgery and as such, worse outcomes.

Methods: Review of a single-centre prospectively-maintained database. Prematurity was defined as delivery <37/40 gestation. PBA was compared with date-matched controls (2:1). Primary outcomes were clearance of jaundice (<20µmol/L) and native liver survival. Non-parametric univariate analysis was undertaken with a $P \leq 0.05$ considered statistically significant. Data are quoted as median(range) unless indicated.

Results: 21(female n=14,67%), infants were treated in the period Jan. 1988 – Dec. 2016. Median gestation was 33(29-36)wks. and birth weight was 1930(948-4230)g. Twin pregnancy (n=10) was the leading cause for prematurity and significantly higher than the controls (48% vs. 0%; $P<0.0001$). Maternal co-morbidity was high (52%) including pre-eclampsia (19%) and diabetes (14%). Syndromic BASM (n=7) (33% vs 7.5%; $P=0.01$) was more frequently in the PBA cohort.

Liver biopsy was performed in 18(90%) patients (all diagnostic) at a mean of 70(8-266)days. Delayed diagnosis (i.e. >50 days) was seen in 12(60%).

Primary surgery was Kasai portoenterostomy (n=20) at median age 63.5 (16-188) days [vs 56 days in controls $P=0.06$] and liver transplantation (n=1). There were Type 3(n=20) and 2(n=1).

There was increased (but non-significant) clearance of jaundice) in PBA (n=12/19 (63%) vs 18/38 (47%); $P=0.19$). Native liver survival (to 5 years) (FIGURE) was unchanged ($P = 0.22$).

Conclusions:

- PBA infants do as well as term infants – *despite* delayed diagnosis and higher frequency of syndromic form.
- Twinning appears to be the key feature.

