



# Medical



Mr Mark Davenport

## Where have we come from? Where are we going to?

**Biliary atresia from different perspectives.**  
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Most readers of this article will know that biliary atresia arrives without fanfare, innocuous and inconspicuous. “Only a touch of jaundice, most babies have that” or “probably due to breast feeding, it should fade soon” are all too frequently heard. But then, as the colour persists and perhaps someone picks up on the lack of faecal colouring and pigmentation, so alarms sound and referrals are made.

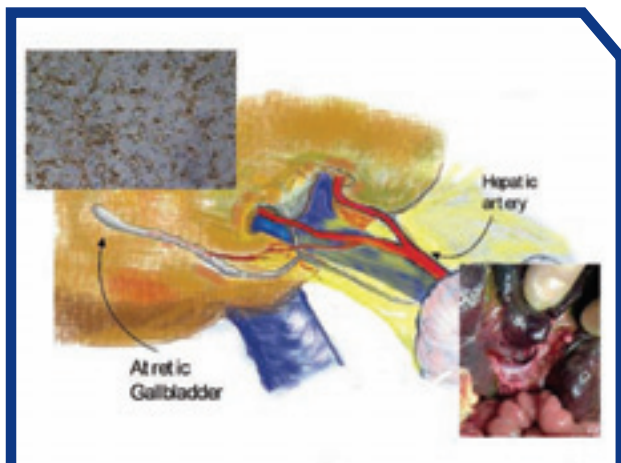
Perhaps it is when the hospital paediatricians start to sound serious, and you get to see, finally, a consultant does the realisation come that there is a clearly a problem; although even then you secretly can't believe it as he or she looks so well, feeds without a fuss and is putting on weight beautifully. Once the

diagnosis is suggested then the likeliest outcome is a referral to one of the three paediatric liver centres for advice and further management, culminating in an admission to try and prove the diagnosis one way or the other. Of course, no-one up to this point has discussed treatment, although maybe the Internet has offered glimpses or insights. For most babies this means surgery to try and restore what has been missing all along — the flow of bile. The operation (named after its Japanese innovator Morio Kasai) in a fortunate few (about half will clear their jaundice to normal levels) is all that is ever required, but in the rest is but a stepping stone to transplantation.

So how do we in the UK compare with the rest of the world? Two conferences devoted to biliary atresia

were held this year, one in Hannover in Germany and the other in Washington DC, in the USA. The first conference was the first opportunity to look at the how centres in Europe were performing since the turn of the millennium. The variability in outcome across Europe came across very early.

Table 1 shows two rough measures of outcome: the first column — time to the Kasai operation gives an indication of how good the pick-up rate or the diagnostic process is, the second column shows how good the overall treatment package (Kasai and transplant) is — the survival rate. Even countries with apparently excellent state-funded health systems (e.g. Denmark) reported survival rates of only 60%.



**Figure 1**  
Biliary atresia with inserts showing fibrosis of liver and appearance after removal of damaged bile ducts



**Figure 2**  
Biliary atresia — a worldwide problem

**Table 1.**  
**European Measures of Outcome in Biliary Atresia**

	Median Age at Kasai	Survival Overall
Italy *	72 days	?
Finland	69 days	70%
Austria	?	75%
Netherlands	?	75%
Switzerland	68 days	91%
Denmark	62 days	75%
Poland	61 days	90%
Czech Republic	59 days	76%
France	57 days	87%
UK	55 days	91%
* 5 leading centres		

What this type of meeting showed me is actually just how good the NHS is in picking up babies with this condition and getting them to surgery — certainly we have the shortest time in Europe to the Kasai operation.

In England and Wales, the care of these children is very centralised with both treatments limited to three large centres (there are more than 50 in Italy!) which I am sure focuses attention and concentrates the mind. This was done at the end of the 1990s because two previous surveys of outcome had shown that smaller centres were not performing as well as the larger centres. Now, our overall national survival rate is amongst the best in Europe. Still, even this should not blind us to the fact that biliary atresia is still a precarious life-threatening condition and that we still have some way to go before we approach 100% survival.

The American conference devoted some of its time to ways that other centres have tried to improve the pick-up rate, for instance by screening all babies in the first few weeks of life, rather than wait for symptoms to arise. In some parts of Japan and in Taiwan they have adopted a system of stool colour charts incorporated into their “baby books”. This highlights to the parents that pale, or white stools are not normal and should be checked out. Unfortunately, while there were improvements in getting these babies into hospital earlier, their overall time to Kasai surgery was still far greater than we see here in the UK, which has no screening programme.

The Americans have also adopted at least a way of working together to share research resources, generously encouraged and “pump-primed” by their own central government research

funding organisation. Unfortunately, we in Europe have not been able to match this and our own basic research lags somewhat by comparison.

Some original research was also reported looking at perhaps why the disease is able to damage so much of the internal bile duct system, and continues to do so even after a degree of bile drainage is restored after surgery. At both conferences, we took the opportunity of presenting our Kings/Leeds trial results from the use of steroids after the Kasai operation. This at least was original research from the UK (funded by CLDF incidentally) and is the first controlled trial of such medication in this condition anywhere in the world. We showed that although the dose of steroid used did not significantly reduce the need for transplant it did reduce the level of jaundice following surgery.

### So what of the future?

It is clear that given the rarity of the disease scientists need to pool resources to enable worthwhile research and this implies a greater degree of collaboration across national boundaries. We would hope that the two recently set up research networks (EFBAR — the European Federation for Biliary Atresia Research and BARC — Biliary Atresia Research Consortium in North America) will go some way to facilitate this.