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Biliary atresia: service delivery and outcomes

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Biliary atresia is a complex disorder dependent on multidisciplinary management. A series of comprehensive national audits in the United Kingdom and France exposed a clear relationship between center volume and clinical outcomes. Different models were adopted in each country in an attempt to improve results. In the United Kingdom, the management of biliary atresia was centralized to three specialist units in 1999, whereas in France, a strategy of decentralized management with closer inter-unit cooperation was adopted in 1997. Both policy changes led to improved outcomes for infants with biliary atresia, but only centralization improved the overall results of Kasai portoenterostomy. Other countries have adopted alternative systems of audit based on voluntary registries, but the impact of these on clinical outcomes at a national level remains unknown. The utility of monitoring tools in assessing performance in biliary atresia, the importance of risk stratification, and the need for standardized definitions of outcome are highlighted.

This review focuses on clinical outcomes in biliary atresia (BA), particularly in relation to public policy and health care infrastructure. In the United Kingdom, the management of BA was centralized in 1999. The process by which this was achieved and its consequences are described together with results of alternative models of delivery of care in other countries, notably France and the United States. The utility and importance of monitoring outcomes in BA are highlighted.

Clinical management of BA

BA is a rare congenital obliterative cholangiopathy of unknown etiology.[1] Reasonably accurate incidence figures from various countries are shown in Table 1.[2-12] Although most western countries have a similar incidence of BA (approximately 1 in 14,000 to 1 in 19,000 live births), the condition is more common in the far East. The highest recorded incidence is in French Polynesia, where it is estimated that up to 1 in 3400 births is affected.[3]

Untreated infants die from secondary biliary cirrhosis and liver failure within the first few years of life.[13] In the late 1950s, Morio Kasai, a Japanese surgeon, reported the presence of patent microscopic biliary channels at the porta hepatis in young infants with BA.[14] He demonstrated that exposure of these channels by radical excision of atretic extrahepatic biliary remnants could result in effective drainage of bile, especially if the operation was performed before 8 weeks of age. The Kasai portoenterostomy operation is now accepted as the standard primary operation for BA in the vast majority of cases. The success of the procedure, as defined by complete clearance of jaundice (plasma bilirubin <20 μmol/L = 1.2 mg/dL), is related to several factors including age at presentation, extent of extrahepatic duct atresia (type of BA), and the presence of the BA splenic malformation syndrome.[15] Children who clear their jaundice completely and remain anicteric for the first 3 years of life have about an 80% chance of reaching adulthood with their native liver, ie, without a liver transplant. Infants who fail to clear their jaundice after portoenterostomy and

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Table 1 Incidence of biliary atresia in various countries

Country	Incidence per 100,000 live births	Reference
Australia (Victoria)	6.96	2
France	5.12	3
French Polynesia	29.4	3
Hawaii	10.6	4
Japan	10.4	5
Netherlands	5.04	6
Sweden	7.06	7
Taiwan	14.6	8
United Kingdom & Eire	5.99	9
United States: Texas	6.5	10
Georgia	7.3	11
New York State	8.5	12

those who, despite an initially successful Kasai procedure, develop severe complications of chronic liver disease require liver transplantation.

BA is the most common indication for liver transplantation in children, accounting for more than half of all cases in large pediatric series. Most such children require a transplant in the first few years of life. Techniques such as split-liver grafting and living-related liver transplantation have minimized the risk of these small children dying on the waiting list. Thus, the combination of Kasai portoenterostomy and liver transplantation has transformed a disease that was almost invariably fatal in the 1960s into one with a current overall 5-year survival in experienced centers of around 90%. Long-term studies have shown a relatively good quality of life in BA survivors after portoenterostomy alone[16,17] and after liver transplantation.[18]

Centralized management in the UK

Initial clearance of jaundice after Kasai portoenterostomy is a critical step in avoiding the need for liver transplantation and its attendant risks. In the UK, a series of three national studies demonstrated that the outcome of infants with BA was markedly affected by the experience of the center.[9,19,20] The last of these studies precipitated a major policy change in the management of infants with BA in the UK.

The first UK survey was conducted between 1980 and 1982 (Table 2).[19] This showed a significant difference in jaundice-free survival rates between the high-volume center treating more than 5 cases per year and the 15 low-volume centers treating fewer cases. A similar survey of all infants with BA in UK and Eire conducted between 1993 and 1995 again demonstrated that outcome was related to center volume and experience.[9] In this second study, survival without liver transplantation (native liver survival) and overall survival were both significantly greater in the 2 centers managing more than 5 cases per year than in the 13 centers treating fewer cases. After taking into account age at surgery, gestational age, and presence of the BA splenic malformation syndrome, stepwise multivariate regression showed that center size was the only significant independent factor predictive of overall survival.

There was widespread debate within the profession and the media about the dissemination and interpretation of these findings.[21,22] However, in 1999, the Department of Health decided that the management of BA in England and Wales would be centralized and limited to three supraregional pediatric liver units in the south (London), midlands (Birmingham), and north (Leeds) of England. Scotland has an independent body regulating these issues. Some pediatric surgeons objected to this decision.[22] Nevertheless, the decision was implemented, and, most importantly, subsequent outcomes were critically audited. Thus, the third national survey of infants with BA was published (Table 2).[20] This showed that, within the three centers, 57% of infants cleared their jaundice after Kasai portoenterostomy. Significantly, there was no difference in outcome between the three centers; all three had uniformly good results equivalent to those previously reported from centers treating five or more cases annually. Estimated actuarial 4-year survival

Table 2 Summary of three consecutive national surveys of biliary atresia in the UK

Author	Survey period (duration)	No. of infants with BA	No. of infants undergoing a portoenterostomy-type procedure	Median age at operation (days)	Duration of follow-up (months)	Center/volume effect and jaundice free outcome		
McClement et al.[19]	1980 to 1982 (UK, 3 year)	114	107	63	mean 27	>5 cases/year	(n = 1)	: 43%
						2 to 5 cases/year	(n = 15)	: 29%
						≤1 case/year		: 11%
McKiernan et al.[9]	1993 to 1995 (UK & Eire, 2 year)	93	91	54	median 42	>5 cases/year	(n = 2)	: 62%
						2 to 5 cases/year	(n = 5)	: 58%
						≤1 case/year	(n = 8)	: 17%
Davenport et al.[20]	1999 to 2002 (England & Wales, 3.5 year)	148	142	54	median 25	>5 cases/year	(n = 3)	: 57%