

Japanese Biliary Atresia Registry (JBAR) Sheet 1/3 [Follow-up Year 25]

Target years for follow-up year 1: 1989~1997

Total number of initial registrations in target years: 1163

Number of deaths: 299

Total number of registrations for follow-up year 25: 577 (66.8 %)

Outcome with liver transplant		228	Outcome without liver transplant		268	Unknown cases		81
Living		176	Living		213	Living		2
Living without jaundice		152	Living without jaundice		166	Living without jaundice		1
Living with jaundice		17	Living with jaundice		36	Living with jaundice		0
Unknown		7	Unknown		11	Unknown		1
Death		4	Death		4	Death		0
Unknown		48	Unknown		51	Unknown		79
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		655	Minimum		5008	Minimum		ND
Maximum		9312	Maximum		9630	Maximum		ND
Mean		4705.5	Mean		8137.0	Mean		ND
Standard deviation		3620.2	Standard deviation		2119.1	Standard deviation		ND
Cause of death			Cause of death			Cause of death		
Liver failure		1	Liver failure		2	Liver failure		0
Rupture of esophageal varices		0	Rupture of esophageal varices		0	Rupture of esophageal varices		0
Pneumonia		0	Pneumonia		0	Pneumonia		0
Peritonitis		0	Peritonitis		0	Peritonitis		0
Sepsis after cholangitis		0	Sepsis after cholangitis		0	Sepsis after cholangitis		0
Post-transplantation death		3	Post-transplantation death		0	Post-transplantation death		0
Cause of death unknown		0	Cause of death unknown		0	Cause of death unknown		0
Other		0	Other		2	Other		0
(Includes duplications)			(Includes duplications)			(Includes duplications)		
NA		0	NA		0	NA		0

Japanese Biliary Atresia Registry (JBAR) Sheet 2/3 [Follow-up Year 25]

Target years for follow-up year 1: 1989~1997

Total number of initial registrations in target years: 1163

Number of deaths: 299

Total number of registrations for follow-up year 25: 577 (66.8 %)

3. Native liver survivors: liver function tests (since 1998)			4. Esophageal varices in cases without liver transplant			7. Surgeries other than liver transplant (except for unknown cases)		
Number of liver function tests (implemented) 206			Yes 41			Yes 21		
Total bilirubin (mg/dl)			(Duplications in treatment content)	No treatment 31		(Duplications in treatment content)	Hilar re-dissection 2	
Minimum 0.2				Treatment 9			Hilar curettage 0	
Maximum 31.0				Endoscopic treatment or sclerotherapy 7			Endoscopic hilar curetta 0	
Mean 1.5				Direct approach 0			Other 25	
Standard deviation 2.9				Shunt 0			Unknown 1	
ALT (IU/l)				Other 1			None 363	
Minimum 2.0			Unknown 1		Unknown 112			
Maximum 288.0			Details of treatment unknown 1		8. Re-elevation of jaundice (except for unknown cases)			
Mean 37.4			None 84		Yes 80			
Standard deviation 35.7			Unknown 143		No 302			
γGTP (IU/l)			5. Hypersplenism in cases without liver transplantation			Unknown 114		
Minimum 4.0			Yes 63			If yes		
Maximum 1710.0			No treatment 48		Before re-elevation			
Mean 118.3			Treatment 9		No jaundice 48			
Standard deviation 188.5			Partial splenic embolization 5		Mild jaundice 29			
ChE (IU/l)			Splenectomy 4		Unknown 3			
Below normal range 72			Other 2		Cause of re-elevation (includes duplications)			
Within normal range 110			Details of treatment unknown 0		Ascending cholangitis 41			
Unknown 24			Use of treatment unknown 6		Other 31			
ALP (IU/l)			None 127		Unknown 10			
Above normal range 71			Unknown 78		Final measures			
Within normal range 120			6. Other complications (except for unknown cases)			Monitor patient 19		
Unknown 15			Cholangitis 11		Conservative treatment 45			
			Gastrointestinal bleeding other than varices 7		Re-curative surgery 1			
			Ascites 6		Liver transplant 14			
			Fat soluble vitamin deficiency 2		Other 9			
			Intrahepatic cholelithiasis 8		(includes duplications) Unknown 4			
			Pulmonary hypertension 5					
			Hepatopulmonary syndrome 2					
			Intestinal obstruction 59					
			Other 43					
			(Includes duplications)					

Japanese Biliary Atresia Registry (JBAR) Sheet 3/3 [Follow-up Year 25]

Target years for follow-up year 1: 1989~1997

Total number of initial registrations in target years: 1163

Number of deaths: 299

Total number of registrations for follow-up year 25: 577 (66.8 %)

Outcome with liver transplant		228	Outcome without liver transplant		268
9. Mental development			9. Mental development		
Excellent		6	Excellent		22
Normal		148	Normal		180
Low		13	Low		7
Unknown		61	Unknown		59
10-1. Physical development			10-1. Physical development		
Excellent		3	Excellent		17
Normal		152	Normal		187
Low		5	Low		2
Unknown		68	Unknown		62
10-2. Physical status			10-2. Physical status		
Height (cm)			Height (cm)		
	Minimum	141.4		Minimum	144.1
	Maximum	181.0		Maximum	186.0
	Mean	160.5		Mean	162.4
	Standard deviation	8.9		Standard deviation	8.5
Weight (kg)			Weight (kg)		
	Minimum	29.0		Minimum	35.8
	Maximum	80.0		Maximum	123.7
	Mean	53.4		Mean	58.6
	Standard deviation	9.0		Standard deviation	10.8
11. Menstruation (females)			11. Menstruation (females)		
Yes		65	Yes		101
No		0	No		2
Unknown		80	Unknown		72
12. Pregnancy (females)			12. Pregnancy (females)		
Yes		18	Yes		23
No		65	No		97
Unknown		62	Unknown		55
13. Restrictions in daily activity			13. Restrictions in daily activity		
Infant, preschool child		0	Infant, preschool child		0
Generally, no restrictions in school life, working life		147	Generally, no restrictions in school life, working life		201
Requires restrictions in school life, working life		10	Requires restrictions in school life, working life		17
Unknown		70	Unknown		48
14. Marriage			14. Marriage		
Yes		34	Yes		46
No		106	No		120
Unknown		88	Unknown		102
15. Employment			15. Employment		
Yes		115	Yes		146
No		21	No		27
Unknown		92	Unknown		95

NB 10.2 Physical status is an aggregation of data collected from JBAR2016.