

Japanese Biliary Atresia Registry (JBAR) Sheet 1/3 [Follow-up Year 15]

Target years for follow-up year 1: 1989~2003

Total number of initial registrations in target years: 1907

Number of deaths: 347

Total number of registrations for follow-up year 25: 1134 (72.7 %)

Outcome with liver transplant		470	Outcome without liver transplant		627	Unknown cases		37
Living		389	Living		586	Living		3
Living without jaundice		331	Living without jaundice		477	Living without jaundice		3
Living with jaundice		44	Living with jaundice		92	Living with jaundice		0
Unknown		14	Unknown		17	Unknown		0
Death		11	Death		5	Death		1
Unknown		70	Unknown		36	Unknown		33
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		2281	Minimum		2126	Minimum		4406
Maximum		5864	Maximum		5798	Maximum		4406
Mean		4807.1	Mean		4672.8	Mean		4406.0
Standard deviation		1106.8	Standard deviation		1710.2	Standard deviation		-
Cause of death			Cause of death			Cause of death		
Liver failure		0	Liver failure		1	Liver failure		1
Rupture of esophageal varices		0	Rupture of esophageal varices		1	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		6	Post-transplantation death		0	Post-transplantation death		0
Cause of death unknown		2	Cause of death unknown		1	Cause of death unknown		0
Other		2	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 15]

Target years for follow-up year 1: 1989~2003

Total number of initial registrations in target years: 1907

Number of deaths: 347

Total number of registrations for follow-up year 25: 1134 (72.7 %)

3. Native liver survivors: liver function tests (sing			4. Esophageal varices in cases without liver tran			7. Surgeries other than liver transplant (except for unknown cases)		
Number of liver function tests (implemented) 575			Yes	151		Yes	46	
Total bilirubin (mg/dl)			(Duplications in treatment content)	No treatment	113	(Duplications in treatment content)	Hilar re-dissection	1
Minimum	0.1			Treatment	38		Hilar curettage	3
Maximum	71.0			scopic treatment or sclerotherapy	36		Endoscopic hilar curettage	0
Mean	1.2			Direct approach	0		Other	48
Standard deviation	3.1			Shunt	0		Unknown	0
Other				Other	3		None	902
ALT (IU/l)			Unknown	1	Unknown	149	8. Re-elevation of jaundice (except for unknown cases)	
Minimum	1.0		Details of treatment unknown 0					
Maximum	571.0		None	281		Yes	168	
Mean	37.7		Unknown	195		No	788	
Standard deviation	43.2		5. Hypersplenism in cases without liver transplan			Unknown	141	
yGTP (IU/l)			Yes	187				
Minimum	5.0		(Duplications in treatment content)	No treatment	138	If yes		
Maximum	847.0			Treatment	56	Before	No jaundice	105
Mean	81.0			Partial splenic embolization	28	re-elevation	Mild jaundice	57
Standard deviation	106.3			Splenectomy	9		Unknown	6
ChE (IU/l)				Other	7	Cause of	Ascending cholangitis	88
Below normal range	122			Details of treatment unknown	1	re-elevation	Other	64
Within normal range	394		Use of treatment unknown	8	(includes duplications)	Unknown	19	
Unknown	59		6. Other complications (except for unknown cases)			Final measures	Monitor patient	46
ALP (IU/l)			None	355		(includes duplications)	Conservative treatment	93
Above normal range	97		Unknown	85			Re-curative surgery	1
Within normal range	438		Cholangitis				Liver transplant	17
Unknown	40		Gastrointestinal bleeding other than varices				Other	16
			Ascites				Unknown	4
			Fat soluble vitamin deficiency					
			Intrahepatic cholelithiasis					
			Pulmonary hypertension					
			Hepatopulmonary syndrome					
			Intestinal obstruction					
			Other					
			(Includes duplications)					

Japanese Biliary Atresia Registry (JBAR) Sheet 3/3 [Follow-up Year 15]

Target years for follow-up year 1: 1989~2003

Total number of initial registrations in target years: 1907

Number of deaths: 347

Total number of registrations for follow-up year 25: 1134 (72.7 %)

Outcome with liver transplant		470	Outcome without liver transplant		627
9. Mental development			9. Mental development		
Excellent		21	Excellent		35
Normal		336	Normal		502
Low		24	Low		15
Unknown		89	Unknown		75
10-1. Physical development			10-1. Physical development		
Excellent		14	Excellent		38
Normal		357	Normal		503
Low		9	Low		9
Unknown		90	Unknown		77
10-2. Physical status			10-2. Physical status		
Height (cm)			Height (cm)		
	Minimum	122.8		Minimum	121.0
	Maximum	176.0		Maximum	179.5
	Mean	157.8		Mean	159.4
	Standard deviation	8.3		Standard deviation	9.0
Weight (kg)			Weight (kg)		
	Minimum	29.4		Minimum	21.1
	Maximum	73.3		Maximum	92.0
	Mean	50.5		Mean	51.6
	Standard deviation	8.1		Standard deviation	9.1
11. Menstruation (females)			11. Menstruation (females)		
Yes		138	Yes		188
No		7	No		15
Unknown		160	Unknown		196
12. Pregnancy (females)			12. Pregnancy (females)		
Yes		3	Yes		1
No		220	No		319
Unknown		82	Unknown		79
13. Restrictions in daily activity			13. Restrictions in daily activity		
Infant, preschool child		1	Infant, preschool child		0
Generally, no restrictions in school life, working life		352	Generally, no restrictions in school life, working life		528
Requires restrictions in school life, working		32	Requires restrictions in school life, working l		35
Unknown		85	Unknown		64
14. Marriage			14. Marriage		
Yes		2	Yes		2
No		377	No		540
Unknown		91	Unknown		85
15. Employment			15. Employment		
Yes		4	Yes		8
No		369	No		525
Unknown		97	Unknown		94

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