

# Japanese Biliary Atresia Registry (JBAR) Sheet 1/3 [Follow-up Year 10 ]

Target years for follow-up year 1: 1989~2008

Total number of initial registrations in target years: 2438

Number of deaths: 377

Total number of registrations for follow-up year 25: 1603 (77.8 %)

Outcome with liver transplant		649	Outcome without liver transplant		914	Unknown cases		40
Living		579	Living		875	Living		8
Living without jaundice		512	Living without jaundice		740	Living without jaundice		5
Living with jaundice		41	Living with jaundice		114	Living with jaundice		1
Unknown		26	Unknown		21	Unknown		2
Death		8	Death		11	Death		0
Unknown		62	Unknown		28	Unknown		32
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		588	Minimum		505	Minimum		ND
Maximum		4117	Maximum		3470	Maximum		ND
Mean		2869.7	Mean		2054.9	Mean		ND
Standard deviation		1189.6	Standard deviation		1128.0	Standard deviation		ND
Cause of death			Cause of death			Cause of death		
Liver failure		1	Liver failure		6	Liver failure		0
Rupture of esophageal varices		0	Rupture of esophageal varices		2	Rupture of esophageal varices		0
Pneumonia		0	Pneumonia		0	Pneumonia		0
Peritonitis		0	Peritonitis		0	Peritonitis		0
Sepsis after cholangitis		0	Sepsis after cholangitis		1	Sepsis after cholangitis		0
Post-transplantation death		5	Post-transplantation death		0	Post-transplantation death		0
Cause of death unknown		0	Cause of death unknown		1	Cause of death unknown		0
Other		2	Other		1	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 10 ]

Target years for follow-up year 1: 1989~2008

Total number of initial registrations in target years: 2438

Number of deaths: 377

Total number of registrations for follow-up year 25: 1603 (77.8 %)

3. Native liver survivors: liver function tests (sin)		4. Esophageal varices in cases without liver tran		7. Surgeries other than liver transplant (except for unknown cases)			
Number of liver function tests (implemented) 866		Yes	238	Yes	61		
Total bilirubin (mg/dl)		(Duplications in treatment content)	No treatment	159	Hilar re-dissection	4	
Minimum	0.1		Treatment	79	Hilar curettage	3	
Maximum	33.4		Endoscopic treatment or sclerotherapy	73	Endoscopic hilar curettage	0	
Mean	1.1		Direct approach	1	Other	62	
Standard deviation	2.0		Shunt	2	Unknown	4	
Other			Other	5	None	1338	
ALT (IU/l)		Unknown	0	Unknown	164		
Minimum	1.0	Details of treatment unknown 0		8. Re-elevation of jaundice (except for unknown cases)			
Maximum	2702.0	None	416	Yes	194		
Mean	52.1	Unknown	260	No	1224		
Standard deviation	107.4	5. Hypersplenism in cases without liver transplan		Unknown	145		
γGTP (IU/l)		Yes	312	If yes			
Minimum	1.0	(Duplications in treatment content)	No treatment	236	Before	No jaundice	123
Maximum	1389.0		Treatment	56	re-elevation	Mild jaundice	56
Mean	92.1		Partial splenic embolization	40	Cause of re-elevation (includes duplications)	Unknown	15
Standard deviation	126.0		Splenectomy	9		Ascending cholangitis	107
ChE (IU/l)			Other	9		Other	77
Below normal range	179		Details of treatment unknown	0		Unknown	18
Within normal range	590	Use of treatment unknown	21	Final measures			
Unknown	97	None	497	Monitor patient	42		
ALP (IU/l)		Unknown	105	Conservative treatment	105		
Above normal range	76	6. Other complications (except for unknown cases)		Re-curative surgery	3		
Within normal range	739	Cholangitis	20	Liver transplant	30		
Unknown	51	Gastrointestinal bleeding other than varices	29	Other	19		
		Ascites	28	Unknown	7		
		Fat soluble vitamin deficiency	16	(includes duplications)			
		Intrahepatic cholelithiasis	2				
		Pulmonary hypertension	4				
		Hepatopulmonary syndrome	20				
		Intestinal obstruction	105				
		Other	113				
		(Includes duplications)					

# Japanese Biliary Atresia Registry (JBAR) Sheet 3/3 [Follow-up Year 10 ]

Target years for follow-up year 1: 1989~2008

Total number of initial registrations in target years: 2438

Number of deaths: 377

Total number of registrations for follow-up year 25: 1603 (77.8 %)

Outcome with liver transplant		649	Outcome without liver transplant		914
<b>9. Mental development</b>			<b>9. Mental development</b>		
Excellent		19	Excellent		35
Normal		485	Normal		779
Low		39	Low		29
Unknown		106	Unknown		71
<b>10-1. Physical development</b>			<b>10-1. Physical development</b>		
Excellent		21	Excellent		31
Normal		504	Normal		788
Low		20	Low		17
Unknown		104	Unknown		78
<b>10-2. Physical status</b>			<b>10-2. Physical status</b>		
Height (cm)			Height (cm)		
	Minimum	120.0		Minimum	115.7
	Maximum	156.7		Maximum	157.3
	Mean	138.7		Mean	139.9
	Standard deviation	7.8		Standard deviation	8.3
Weight (kg)			Weight (kg)		
	Minimum	17.4		Minimum	18.4
	Maximum	63.8		Maximum	66.3
	Mean	32.5		Mean	34.6
	Standard deviation	7.5		Standard deviation	7.8
<b>11. Menstruation (females)</b>			<b>11. Menstruation (females)</b>		
Yes		18	Yes		19
No		209	No		284
Unknown		189	Unknown		270
<b>12. Pregnancy (females)</b>			<b>12. Pregnancy (females)</b>		
Yes		0	Yes		0
No		338	No		497
Unknown		78	Unknown		76
<b>13. Restrictions in daily activity</b>			<b>13. Restrictions in daily activity</b>		
Infant, preschool child		4	Infant, preschool child		1
Generally, no restrictions in school life, working life		504	Generally, no restrictions in school life, working life		784
Requires restrictions in school life, working		36	Requires restrictions in school life, working l		61
Unknown		105	Unknown		68
<b>14. Marriage</b>			<b>14. Marriage</b>		
Yes		0	Yes		0
No		548	No		834
Unknown		101	Unknown		80
<b>15. Employment</b>			<b>15. Employment</b>		
Yes		0	Yes		0
No		549	No		832
Unknown		100	Unknown		82

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