

# Japanese Biliary Atresia Registry (JBAR) Sheet 1/3 [Follow-up Year 20 ]

Target years for follow-up year 1: 1989~1998

Total number of initial registrations in target years: 1288

Number of deaths: 301

Total number of registrations for follow-up year 25: 718 (72.7 %)

Outcome with liver transplant		300	Outcome without liver transplant		354	Unknown cases		64
Living		234	Living		304	Living		3
Living without jaundice		185	Living without jaundice		242	Living without jaundice		3
Living with jaundice		34	Living with jaundice		48	Living with jaundice		0
Unknown		15	Unknown		14	Unknown		0
Death		3	Death		1	Death		0
Unknown		63	Unknown		49	Unknown		61
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		6292	Minimum		6341	Minimum		ND
Maximum		7447	Maximum		6341	Maximum		ND
Mean		7043.3	Mean		6341.0	Mean		ND
Standard deviation		651.3	Standard deviation		-	Standard deviation		ND
Cause of death			Cause of death			Cause of death		
Liver failure		3	Liver failure		0	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		0	Post-transplantation death		0	Post-transplantation death		0
Cause of death unknown		0	Cause of death unknown		1	Cause of death unknown		0
Other		0	Other		0	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 20 ]

Target years for follow-up year 1: 1989~1998

Total number of initial registrations in target years: 1288

Number of deaths: 301

Total number of registrations for follow-up year 25: 718 (72.7 %)

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 case			
Number of liver function tests (implemented) 307		Yes	75	Yes	30		
Total bilirubin (mg/dl)		(Duplications in treatment content)	No treatment	(Duplications in treatment content)	Hilar re-dissection	4	
Minimum	0.1		Treatment		12	Hilar curettage	0
Maximum	13.8		Endoscopic treatment or sclerotherapy		12	Endoscopic hilar curettage	0
Mean	1.1		Direct approach		0	Other	28
Standard deviation	1.2		Shunt		0	Unknown	3
ALT (IU/l)			Other		0	None	491
Minimum	0.0	Unknown	1	Unknown	133		
Maximum	1980.0	Details of treatment unknown 0		8. Re-elevation of jaundice (except for unknown cases)			
Mean	47.1	None	140	Yes	112		
Standard deviation	127.5	Unknown	139	No	413		
γGTP (IU/l)		5. Hypersplenism in cases without liver transplan		Unknown	129		
Minimum	3.0	Yes	89	If yes			
Maximum	1650.0	(Duplications in treatment content)	No treatment	Before	No jaundice	72	
Mean	102.1		Treatment	11	re-elevation	Mild jaundice	30
Standard deviation	169.5		Partial splenic embolization	6	Cause of re-elevation (includes duplications)	Unknown	10
ChE (IU/l)			Splenectomy	6		Ascending cholangitis	62
Below normal range	80		Other	0		Other	48
Within normal range	195		Details of treatment unknown	0		Unknown	7
Unknown	32	Use of treatment unknown	3	Final measures	Monitor patient	24	
ALP (IU/l)		None	177	(includes duplications)	Conservative treatment	65	
Above normal range	79	Unknown	88		Re-curative surgery	3	
Within normal range	207	6. Other complications (except for unknown cases)			Liver transplant	7	
Unknown	21	Cholangitis	13		Other	19	
		Gastrointestinal bleeding other than varices	11		Unknown	3	
		Ascites	6				
		Fat soluble vitamin deficiency	2				
		Intrahepatic cholelithiasis	4				
		Pulmonary hypertension	3				
		Hepatopulmonary syndrome	2				
		Intestinal obstruction	73				
		Other	52				
		(Includes duplications)					

# Japanese Biliary Atresia Registry (JBAR) Sheet 3/3 [Follow-up Year 20 ]

Target years for follow-up year 1: 1989~1998

Total number of initial registrations in target years: 1288

Number of deaths: 301

Total number of registrations for follow-up year 25: 718 (72.7 %)

<b>Outcome with liver transplant</b>		300	<b>Outcome without liver transplant</b>		354
<b>9. Mental development</b>			<b>9. Mental development</b>		
Excellent	16		Excellent	26	
Normal	195		Normal	259	
Low	12		Low	10	
Unknown	77		Unknown	59	
<b>10-1. Physical development</b>			<b>10-1. Physical development</b>		
Excellent	7		Excellent	19	
Normal	206		Normal	265	
Low	9		Low	5	
Unknown	78		Unknown	65	
<b>10-2. Physical status</b>			<b>10-2. Physical status</b>		
Height (cm)			Height (cm)		
	Minimum	145.0		Minimum	144.5
	Maximum	175.1		Maximum	186.0
	Mean	159.1		Mean	162.0
	Standard deviation	7.8		Standard deviation	8.4
Weight (kg)			Weight (kg)		
	Minimum	38.0		Minimum	40.7
	Maximum	79.6		Maximum	95.0
	Mean	54.2		Mean	55.9
	Standard deviation	7.4		Standard deviation	9.1
<b>11. Menstruation (females)</b>			<b>11. Menstruation (females)</b>		
Yes	82		Yes	110	
No	2		No	1	
Unknown	111		Unknown	125	
<b>12. Pregnancy (females)</b>			<b>12. Pregnancy (females)</b>		
Yes	5		Yes	6	
No	107		No	140	
Unknown	83		Unknown	90	
<b>13. Restrictions in daily activity</b>			<b>13. Restrictions in daily activity</b>		
Infant, preschool child	0		Infant, preschool child	0	
Generally, no restrictions in school life, working life	195		Generally, no restrictions in school life, working life	278	
Requires restrictions in school life, working	24		Requires restrictions in school life, working l	19	
Unknown	81		Unknown	57	
<b>14. Marriage</b>			<b>14. Marriage</b>		
Yes	3		Yes	8	
No	175		No	226	
Unknown	122		Unknown	120	
<b>15. Employment</b>			<b>15. Employment</b>		
Yes	79		Yes	107	
No	95		No	123	
Unknown	126		Unknown	124	

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