

# Japanese Biliary Atresia Registry (JBAR) Sheet 1/3 [Follow-up Year 5 ]

Target years for follow-up year 1: 1989~2013

Total number of initial registrations in target years: 2972

Number of deaths: 283

Total number of registrations for follow-up year 25: 1997 (74.3 %)

Outcome with liver transplant		696	Outcome without liver transplant		1274	Unknown cases		27
Living		637	Living		1238	Living		9
Living without jaundice		580	Living without jaundice		1090	Living without jaundice		7
Living with jaundice		26	Living with jaundice		113	Living with jaundice		0
Unknown		31	Unknown		35	Unknown		2
Death		11	Death		17	Death		1
Unknown		48	Unknown		19	Unknown		17
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		578	Minimum		516	Minimum		ND
Maximum		2209	Maximum		5689	Maximum		ND
Mean		1398.9	Mean		2535.9	Mean		ND
Standard deviation		608.9	Standard deviation		1636.1	Standard deviation		ND
Cause of death			Cause of death			Cause of death		
Liver failure		1	Liver failure		8	Liver failure		0
Rupture of esophageal varices		0	Rupture of esophageal varices		1	Rupture of esophageal varices		0
Pneumonia		2	Pneumonia		1	Pneumonia		0
Peritonitis		0	Peritonitis		1	Peritonitis		0
Sepsis after cholangitis		1	Sepsis after cholangitis		0	Sepsis after cholangitis		0
Post-transplantation death		4	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		3	Other		0
(Includes duplications)			(Includes duplications)			(Includes duplications)		
NA		0	NA		0	NA		1

# Japanese Biliary Atresia Registry (JBAR) Sheet 2/3 [Follow-up Year 5 ]

Target years for follow-up year 1: 1989~2013

Total number of initial registrations in target years: 2972

Number of deaths: 283

Total number of registrations for follow-up year 25: 1997 (74.3 %)

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) 967		Yes	352	Yes	100		
Total bilirubin (mg/dl)		(Duplications in treatment content)	No treatment	202	Hilar re-dissection	17	
Minimum	0.1		Treatment	139	Hilar curettage	5	
Maximum	73.3		Endoscopic treatment or sclerotherapy	127	Endoscopic hilar curettage	0	
Mean	1.0		Direct approach	2	Other	93	
Standard deviation	3.2		Shunt	0	Unknown	2	
ALT (IU/l)			Other	8	None	1727	
Minimum	0.0	Unknown	4	Unknown	143		
Maximum	585.0	Details of treatment unknown	11	8. Re-elevation of jaundice (except for unknown cases)			
Mean	52.9	None	586	Yes	301		
Standard deviation	54.9	Unknown	336	No	1543		
γGTP (IU/l)		5. Hypersplenism in cases without liver transplan		Unknown	126		
Minimum	8.0	Yes	375	If yes			
Maximum	1028.0	(Duplications in treatment content)	No treatment	307	Before	No jaundice	177
Mean	104.0		Treatment	68	re-elevation	Mild jaundice	82
Standard deviation	125.2		Partial splenic embolization	44	Cause of re-elevation (includes duplications)	Unknown	18
ChE (IU/l)			Splenectomy	6		Ascending cholangitis	190
Below normal range	193		Other	15		Other	89
Within normal range	661		Details of treatment unknown	3		Unknown	27
Unknown	113	Use of treatment unknown	21	Final measures	Monitor patient	39	
ALP (IU/l)		None	804	(includes duplications)	Conservative treatment	162	
Above normal range	70	Unknown	95		Re-curative surgery	4	
Within normal range	849	6. Other complications (except for unknown cases)			Liver transplant	69	
Unknown	48	Cholangitis	26		Other	22	
		Gastrointestinal bleeding other than varices	49		Unknown	14	
		Ascites	54				
		Fat soluble vitamin deficiency	25				
		Intrahepatic cholelithiasis	5				
		Pulmonary hypertension	4				
		Hepatopulmonary syndrome	20				
		Intestinal obstruction	214				
		Other	153				
		(Includes duplications)					

# Japanese Biliary Atresia Registry (JBAR) Sheet 3/3 [Follow-up Year 5 ]

Target years for follow-up year 1: 1989~2013

Total number of initial registrations in target years: 2972

Number of deaths: 283

Total number of registrations for follow-up year 25: 1997 (74.3 %)

Outcome with liver transplant		696	Outcome without liver transplant		1274
<b>9. Mental development</b>			<b>9. Mental development</b>		
Excellent		21	Excellent		51
Normal		524	Normal		1113
Low		43	Low		39
Unknown		108	Unknown		71
<b>10-1. Physical development</b>			<b>10-1. Physical development</b>		
Excellent		15	Excellent		45
Normal		543	Normal		1127
Low		28	Low		35
Unknown		110	Unknown		67
<b>10-2. Physical status</b>			<b>10-2. Physical status</b>		
Height (cm)			Height (cm)		
	Minimum	89.6		Minimum	93.4
	Maximum	120.7		Maximum	125.9
	Mean	108.7		Mean	109.1
	Standard deviation	5.2		Standard deviation	5.6
Weight (kg)			Weight (kg)		
	Minimum	13.6		Minimum	13.9
	Maximum	25.7		Maximum	24.1
	Mean	18.7		Mean	18.5
	Standard deviation	2.3		Standard deviation	2.5
<b>13. Restrictions in daily activity</b>			<b>13. Restrictions in daily activity</b>		
Infant, preschool child		316	Infant, preschool child		479
Generally, no restrictions in school life, working life		267	Generally, no restrictions in school life, working life		435
Requires restrictions in school life, working		13	Requires restrictions in school life, working l		25
Unknown		100	Unknown		335

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