

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

Target years for follow-up year 1: 1989~2007

Total number of initial registrations in target years: 2334

Number of deaths: 383

Total number of registrations for follow-up year 25: 1384 (70.9 %)

Outcome with liver transplant		592	Outcome without liver transplant		749	Unknown cases		43
Living		500	Living		703	Living		4
Living without jaundice		427	Living without jaundice		575	Living without jaundice		4
Living with jaundice		55	Living with jaundice		107	Living with jaundice		0
Unknown		18	Unknown		21	Unknown		0
Death		11	Death		6	Death		1
Unknown		81	Unknown		40	Unknown		38
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		2281	Minimum		126	Minimum		4406
Maximum		5864	Maximum		5798	Maximum		4406
Mean		4807.1	Mean		3763.4	Mean		4406.0
Standard deviation		1106.8	Standard deviation		2515.6	Standard deviation		-
Cause of death			Cause of death			Cause of death		
Liver failure		0	Liver failure		2	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		2	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: 1989~2007

Total number of initial registrations in target years: 2334

Number of deaths: 383

Total number of registrations for follow-up year 25: 1384 (70.9 %)

3. Native liver survivors: liver function tests (since follow-up year 1)			4. Esophageal varices in cases without liver transplant			7. Surgeries other than liver transplant (except for unknown cases)								
Number of liver function tests (implemented) 691			Yes 177			Yes 53								
Total bilirubin (mg/dl)			(Duplications in treatment content)			(Duplications in treatment content)								
Minimum 0.1									No treatment 134			Hilar re-dissection 2		
Maximum 71.0									Treatment 43			Hilar curettage 3		
Mean 1.2									Endoscopic treatment or sclerotherapy 40			Endoscopic hilar curettage 0		
Standard deviation 2.9									Direct approach 0			Other 56		
ALT (IU/l)			Shunt 0			Unknown 1								
Minimum 1.0			Other 3			None 1121								
Maximum 571.0			Unknown 2			Unknown 167								
Mean 38.9			Details of treatment unknown 0			8. Re-elevation of jaundice (except for unknown cases)								
Standard deviation 46.1			None 344			Yes 201								
None 344			Unknown 228			No 979								
Unknown 228			5. Hypersplenism in cases without liver transplant			Unknown 161								
Yes 223			Yes 223			If yes								
Minimum 5.0			No treatment 163			Before re-elevation								
Maximum 1092.0			Treatment 58			No jaundice 124								
Mean 84.0			Partial splenic embolization 29			Mild jaundice 66								
Standard deviation 116.6			Splenuctomy 10			Unknown 11								
ChE (IU/l)			Other 8			Cause of re-elevation (includes duplications)								
Below normal range 154			Details of treatment unknown 1			Ascending cholangitis 102								
Within normal range 461			Use of treatment unknown 18			Other 79								
Unknown 76			None 424			Unknown 24								
Above normal range 123			Unknown 102			Final measures								
Within normal range 521			6. Other complications (except for unknown cases)			Monitor patient 63								
Unknown 47			Cholangitis 19			Conservative treatment 105								
			Gastrointestinal bleeding other than varices 23			Re-curative surgery 1								
			Ascites 14			Liver transplant 22								
			Fat soluble vitamin deficiency 8			Other 17								
			Intrahepatic cholelithiasis 8			(includes duplications) Unknown 7								
			Pulmonary hypertension 10											
			Hepatopulmonary syndrome 14											
			Intestinal obstruction 129											
			Other 92											
			(Includes duplications)											

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

Target years for follow-up year 1: 1989~2007

Total number of initial registrations in target years: 2334

Number of deaths: 383

Total number of registrations for follow-up year 25: 1384 (70.9 %)

Outcome with liver transplant		592	Outcome without liver transplant		749
9. Mental development			9. Mental development		
Excellent		24	Excellent		43
Normal		429	Normal		600
Low		32	Low		22
Unknown		107	Unknown		84
10-1. Physical development			10-1. Physical development		
Excellent		18	Excellent		49
Normal		455	Normal		603
Low		11	Low		11
Unknown		108	Unknown		86
10-2. Physical status			10-2. Physical status		
Height (cm)			Height (cm)		
	Minimum	122.8		Minimum	121.0
	Maximum	178.8		Maximum	179.5
	Mean	157.5		Mean	159.7
	Standard deviation	8.4		Standard deviation	8.1
Weight (kg)			Weight (kg)		
	Minimum	29.4		Minimum	21.1
	Maximum	86.9		Maximum	92.0
	Mean	50.9		Mean	52.4
	Standard deviation	9.2		Standard deviation	8.8
11. Menstruation (females)			11. Menstruation (females)		
Yes		185	Yes		226
No		8	No		16
Unknown		193	Unknown		228
12. Pregnancy (females)			12. Pregnancy (females)		
Yes		3	Yes		1
No		292	No		386
Unknown		91	Unknown		83
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		453	Generally, no restrictions in school life, working life		640
Requires restrictions in school life, working life		38	Requires restrictions in school life, working life		37
Unknown		100	Unknown		72
14. Marriage			14. Marriage		
Yes		2	Yes		2
No		483	No		654
Unknown		107	Unknown		93
15. Employment			15. Employment		
Yes		4	Yes		8
No		477	No		639
Unknown		111	Unknown		102

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