

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

Target years for follow-up year 1: 1989~2017

Total number of initial registrations in target years: 3411

Number of deaths: 282

Total number of registrations for follow-up year 25: 2288 (73.1 %)

Outcome with liver transplant		822	Outcome without liver transplant		1435	Unknown cases		31
Living		746	Living		1396	Living		9
Living without jaundice		682	Living without jaundice		1238	Living without jaundice		7
Living with jaundice		28	Living with jaundice		121	Living with jaundice		0
Unknown		36	Unknown		37	Unknown		2
Death		12	Death		17	Death		1
Unknown		64	Unknown		22	Unknown		21
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		578	Minimum		516	Minimum		ND
Maximum		2209	Maximum		2116	Maximum		ND
Mean		1383.6	Mean		1314.9	Mean		ND
Standard deviation		583.0	Standard deviation		506.4	Standard deviation		ND
Cause of death			Cause of death			Cause of death		
Liver failure		2	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: 1989~2017

Total number of initial registrations in target years: 3411

Number of deaths: 282

Total number of registrations for follow-up year 25: 2288 (73.1 %)

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 cas								
Number of liver function tests (implemented 1126			Yes 392			Yes 114								
Total bilirubin (mg/dl)			(Duplications in treatment content)			(Duplications in treatment content)								
Minimum 0.1									No treatment 228			Hilar re-dissection 17		
Maximum 73.3									Treatment 153			Hilar curettage 6		
Mean 1.0									Endoscopic treatment or sclerotherapy 140			Endoscopic hilar curettage 0		
Standard deviation 3.0									Direct approach 2			Other 111		
									Shunt 0			Unknown 2		
ALT (IU/l)						None 1975								
Minimum 0.1						Unknown 168								
Maximum 585.0														
Mean 52.1			None 659											
Standard deviation 54.7			Unknown 384											
γGTP (IU/l)			5. Hypersplenism in cases without liver transplan			8. Re-elevation of jaundice (except for unknown cases)								
Minimum 8.0			Yes 410			Yes 339								
Maximum 1028.0			(Duplications in treatment content)			No 1766								
Mean 100.2						No treatment 334			Unknown 152					
Standard deviation 121.4						Treatment 73			If yes					
						Partial splenic embolization 46			Before re-elevation					
ChE (IU/l)						Splenectomy 8			No jaundice 210					
Below normal range 226						Other 15			Mild jaundice 86					
Within normal range 769			Details of treatment unknown 4			Unknown 19								
Unknown 131			Use of treatment unknown 27			Cause of re-elevation (includes duplications)								
ALP (IU/l)			None 911			Ascending cholangitis 211								
Above normal range 81			Unknown 114			Other 102								
Within normal range 985						Unknown 28								
Unknown 60			6. Other complications (except for unknown cases)			Final measures								
			Cholangitis 28			Monitor patient 47								
			Gastrointestinal bleeding other than varices 51			Conservative treatment 187								
			Ascites 56			Re-curative surgery 5								
			Fat soluble vitamin deficiency 26			Liver transplant 72								
			Intrahepatic cholelithiasis 5			Other 25								
			Pulmonary hypertension 4			Unknown 14								
			Hepatopulmonary syndrome 21											
			Intestinal obstruction 266											
			Other 174											
			(Includes duplications)											

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

Target years for follow-up year 1: 1989~2017

Total number of initial registrations in target years: 3411

Number of deaths: 282

Total number of registrations for follow-up year 25: 2288 (73.1 %)

Outcome with liver transplant		822	Outcome without liver transplant		1435
9. Mental development			9. Mental development		
Excellent		24	Excellent		56
Normal		611	Normal		1245
Low		46	Low		50
Unknown		141	Unknown		84
10-1. Physical development			10-1. Physical development		
Excellent		16	Excellent		50
Normal		634	Normal		1261
Low		31	Low		43
Unknown		141	Unknown		81
10-2. Physical status			10-2. Physical status		
Height (cm)			Height (cm)		
	Minimum	89.6		Minimum	92.0
	Maximum	120.7		Maximum	125.9
	Mean	108.8		Mean	109.1
	Standard deviation	4.9		Standard deviation	5.7
Weight (kg)			Weight (kg)		
	Minimum	12.6		Minimum	12.0
	Maximum	25.7		Maximum	26.3
	Mean	18.7		Mean	18.5
	Standard deviation	2.2		Standard deviation	2.5
13. Restrictions in daily activity			13. Restrictions in daily activity		
Infant, preschool child		366	Infant, preschool child		572
Generally, no restrictions in school life, working life		324	Generally, no restrictions in school life, working life		497
Requires restrictions in school life, working		14	Requires restrictions in school life, working l		27
Unknown		117	Unknown		338

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