

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

Target years for follow-up year 1: 1989~2002

Total number of initial registrations in target years: 1806

Number of deaths: 355

Total number of registrations for follow-up year 25: 998 (68.8 %)

Outcome with liver transplant		443	Outcome without liver transplant		473	Unknown cases		82
Living		355	Living		415	Living		4
Living without jaundice		294	Living without jaundice		342	Living without jaundice		4
Living with jaundice		43	Living with jaundice		58	Living with jaundice		0
Unknown		18	Unknown		15	Unknown		0
Death		3	Death		2	Death		0
Unknown		85	Unknown		56	Unknown		78
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		6292	Minimum		6341	Minimum		ND
Maximum		7447	Maximum		7542	Maximum		ND
Mean		7043.3	Mean		6941.5	Mean		ND
Standard deviation		651.3	Standard deviation		849.2	Standard deviation		ND
Cause of death			Cause of death			Cause of death		
Liver failure		3	Liver failure		1	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: 1989~2002

Total number of initial registrations in target years: 1806

Number of deaths: 355

Total number of registrations for follow-up year 25: 998 (68.8 %)

3. Native liver survivors: liver function tests (since 1989)			4. Esophageal varices in cases without liver transplant			7. Surgeries other than liver transplant (except for unknown cases)								
Number of liver function tests (implemented) 419			Yes 97			Yes 39								
Total bilirubin (mg/dl)			(Duplications in treatment content)			(Duplications in treatment content)								
Minimum 0.1									No treatment 79			Hilar re-dissection 4		
Maximum 13.8									Treatment 18			Hilar curettage 0		
Mean 1.1									Endoscopic treatment or sclerotherapy 19			Endoscopic hilar curettage 0		
Standard deviation 1.1									Direct approach 0			Other 38		
ALT (IU/l)			Shunt 0			Unknown 3								
Minimum 6.0			Other 0			None 707								
Maximum 1980.0			Unknown 1			Unknown 170								
Mean 43.7			Details of treatment unknown 0			8. Re-elevation of jaundice (except for unknown cases)								
Standard deviation 110.5			None 194			Yes 144								
γGTP (IU/l)			Unknown 182			No 610								
Minimum 3.0			5. Hypersplenism in cases without liver transplant			Unknown 162								
Maximum 1650.0			Yes 121			If yes								
Mean 97.1			(Duplications in treatment content)			Before re-elevation								
Standard deviation 166.3						No treatment 100			No jaundice 93					
ChE (IU/l)						Treatment 14			Mild jaundice 35					
Below normal range 111						Partial splenic embolization 7			Unknown 16					
Within normal range 265						Splenectomy 9			Ascending cholangitis 76					
Unknown 43			Other 0			Other 62								
ALP (IU/l)			Details of treatment unknown 0			Unknown 11								
Above normal range 108			Use of treatment unknown 7			Final measures								
Within normal range 279			None 244			Monitor patient 31								
Unknown 32			Unknown 108			Conservative treatment 78								
			6. Other complications (except for unknown cases)			Re-curative surgery 4								
			Cholangitis 19			Liver transplant 13								
			Gastrointestinal bleeding other than varices 14			Other 22								
			Ascites 10			(includes duplications) Unknown 6								
			Fat soluble vitamin deficiency 2											
			Intrahepatic cholelithiasis 8											
			Pulmonary hypertension 4											
			Hepatopulmonary syndrome 2											
			Intestinal obstruction 95											
			Other 76											
			(Includes duplications)											

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

Target years for follow-up year 1: 1989~2002

Total number of initial registrations in target years: 1806

Number of deaths: 355

Total number of registrations for follow-up year 25: 998 (68.8 %)

Outcome with liver transplant		443	Outcome without liver transplant		473
9. Mental development			9. Mental development		
Excellent		20	Excellent		44
Normal		303	Normal		346
Low		18	Low		14
Unknown		102	Unknown		69
10-1. Physical development			10-1. Physical development		
Excellent		11	Excellent		33
Normal		316	Normal		358
Low		11	Low		7
Unknown		105	Unknown		75
10-2. Physical status			10-2. Physical status		
Height (cm)			Height (cm)		
	Minimum	133.7		Minimum	144.5
	Maximum	178.2		Maximum	186.1
	Mean	158.4		Mean	162.3
	Standard deviation	8.0		Standard deviation	8.6
Weight (kg)			Weight (kg)		
	Minimum	36.6		Minimum	38.0
	Maximum	82.3		Maximum	103.8
	Mean	53.3		Mean	56.9
	Standard deviation	8.7		Standard deviation	10.5
11. Menstruation (females)			11. Menstruation (females)		
Yes		130	Yes		156
No		3	No		4
Unknown		163	Unknown		158
12. Pregnancy (females)			12. Pregnancy (females)		
Yes		8	Yes		10
No		169	No		200
Unknown		119	Unknown		108
13. Restrictions in daily activity			13. Restrictions in daily activity		
Infant, preschool child		0	Infant, preschool child		0
Generally, no restrictions in school life, working life		298	Generally, no restrictions in school life, working life		380
Requires restrictions in school life, working life		30	Requires restrictions in school life, working life		26
Unknown		114	Unknown		65
14. Marriage			14. Marriage		
Yes		4	Yes		10
No		270	No		319
Unknown		169	Unknown		144
15. Employment			15. Employment		
Yes		127	Yes		140
No		145	No		185
Unknown		171	Unknown		148

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