

Japanese Biliary Atresia Registry (JBAR) Sheet 1/3 [Follow-up Year 10]

Target years for follow-up year 1: 1989~2012

Total number of initial registrations in target years: 2883

Number of deaths: 414

Total number of registrations for follow-up year 25: 1884 (76.3 %)

Outcome with liver transplant		781	Outcome without liver transplant		1058	Unknown cases		45
Living		694	Living		1008	Living		8
Living without jaundice		621	Living without jaundice		862	Living without jaundice		5
Living with jaundice		43	Living with jaundice		124	Living with jaundice		1
Unknown		30	Unknown		22	Unknown		2
Death		9	Death		11	Death		0
Unknown		78	Unknown		39	Unknown		37
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		588	Minimum		505	Minimum		ND
Maximum		4117	Maximum		3470	Maximum		ND
Mean		2863.8	Mean		2054.9	Mean		ND
Standard deviation		1101.5	Standard deviation		1128.0	Standard deviation		ND
Cause of death			Cause of death			Cause of death		
Liver failure		1	Liver failure		6	Liver failure		0
Rupture of esophageal varices		0	Rupture of esophageal varices		2	Rupture of esophageal varices		0
Pneumonia		0	Pneumonia		0	Pneumonia		0
Peritonitis		0	Peritonitis		0	Peritonitis		0
Sepsis after cholangitis		0	Sepsis after cholangitis		1	Sepsis after cholangitis		0
Post-transplantation death		6	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		1	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 10]

Target years for follow-up year 1: 1989~2012

Total number of initial registrations in target years: 2883

Number of deaths: 414

Total number of registrations for follow-up year 25: 1884 (76.3 %)

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 cas								
Number of liver function tests (implemented 1000			Yes 262			Yes 67								
Total bilirubin (mg/dl)			(Duplications in treatment content)			(Duplications in treatment content)								
Minimum 0.1									No treatment 177			Hilar re-dissection 5		
Maximum 33.4									Treatment 85			Hilar curettage 3		
Mean 1.1									Endoscopic treatment or sclerotherapy 79			Endoscopic hilar curettage 0		
Standard deviation 2.0									Direct approach 1			Other 72		
									Shunt 2			Unknown 5		
ALT (IU/l)						None 1574								
Minimum 1.0						Unknown 198								
Maximum 2702.0														
Mean 49.7			None 487											
Standard deviation 101.8			Unknown 309											
γGTP (IU/l)			5. Hypersplenism in cases without liver transplan			8. Re-elevation of jaundice (except for unknown cases)								
Minimum 1.0			Yes 351			Yes 225								
Maximum 2482.0			(Duplications in treatment content)			No 1436								
Mean 87.8						No treatment 263			Unknown 178					
Standard deviation 142.1						Treatment 60			If yes					
						Partial splenic embolization 43			Before re-elevation					
ChE (IU/l)						Splenectomy 10			No jaundice 146					
Below normal range 201						Other 9			Mild jaundice 60					
Within normal range 686			Details of treatment unknown 0			Unknown 19								
Unknown 113			Use of treatment unknown 30			Cause of re-elevation (includes duplications)								
ALP (IU/l)			None 583			Ascending cholangitis 124								
Above normal range 89			Unknown 124			Other 86								
Within normal range 848			6. Other complications (except for unknown cases)			Unknown 23								
Unknown 63			Cholangitis 23			Final measures								
			Gastrointestinal bleeding other than varices 30			Monitor patient 48								
			Ascites 30			Conservative treatment 125								
			Fat soluble vitamin deficiency 16			Re-curative surgery 3								
			Intrahepatic cholelithiasis 2			Liver transplant 32								
			Pulmonary hypertension 6			Other 19								
			Hepatopulmonary syndrome 21			Unknown 10								
			Intestinal obstruction 133											
			Other 132											
			(Includes duplications)											

Japanese Biliary Atresia Registry (JBAR) Sheet 3/3 [Follow-up Year 10]

Target years for follow-up year 1: 1989~2012

Total number of initial registrations in target years: 2883

Number of deaths: 414

Total number of registrations for follow-up year 25: 1884 (76.3 %)

Outcome with liver transplant		781	Outcome without liver transplant		1058
9. Mental development			9. Mental development		
Excellent		22	Excellent		39
Normal		579	Normal		900
Low		50	Low		37
Unknown		130	Unknown		82
10-1. Physical development			10-1. Physical development		
Excellent		24	Excellent		35
Normal		601	Normal		911
Low		28	Low		21
Unknown		128	Unknown		91
10-2. Physical status			10-2. Physical status		
Height (cm)			Height (cm)		
	Minimum	120.0		Minimum	115.7
	Maximum	156.7		Maximum	162.4
	Mean	139.1		Mean	140.8
	Standard deviation	7.4		Standard deviation	8.3
Weight (kg)			Weight (kg)		
	Minimum	17.4		Minimum	18.4
	Maximum	63.8		Maximum	69.3
	Mean	33.1		Mean	35.6
	Standard deviation	7.2		Standard deviation	8.1
11. Menstruation (females)			11. Menstruation (females)		
Yes		23	Yes		23
No		239	No		325
Unknown		244	Unknown		321
12. Pregnancy (females)			12. Pregnancy (females)		
Yes		0	Yes		0
No		409	No		580
Unknown		97	Unknown		89
13. Restrictions in daily activity			13. Restrictions in daily activity		
Infant, preschool child		4	Infant, preschool child		2
Generally, no restrictions in school life, working life		604	Generally, no restrictions in school life, working life		906
Requires restrictions in school life, working life		43	Requires restrictions in school life, working life		65
Unknown		127	Unknown		80
14. Marriage			14. Marriage		
Yes		0	Yes		0
No		660	No		970
Unknown		121	Unknown		88
15. Employment			15. Employment		
Yes		0	Yes		0
No		661	No		968
Unknown		120	Unknown		90

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