

Japanese Biliary Atresia Registry (JBAR) Sheet 1/3 [Follow-up Year 25]

Target years for follow-up year 1: 1989~1993

Total number of initial registrations in target years: 622

Number of deaths: 183

Total number of registrations for follow-up year 25: 324 (73.8 %)

Outcome with liver transplant		118	Outcome without liver transplant		159	Unknown cases		47
Living		90	Living		125	Living		1
Living without jaundice		76	Living without jaundice		92	Living without jaundice		0
Living with jaundice		7	Living with jaundice		25	Living with jaundice		0
Unknown		7	Unknown		8	Unknown		1
Death		4	Death		2	Death		0
Unknown		24	Unknown		32	Unknown		46
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		655	Minimum		5008	Minimum		ND
Maximum		9312	Maximum		8716	Maximum		ND
Mean		4705.5	Mean		6862.0	Mean		ND
Standard deviation		3620.2	Standard deviation		2622.0	Standard deviation		ND
Cause of death			Cause of death			Cause of death		
Liver failure		1	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		3	Post-transplantation death		0	Post-transplantation death		0
Cause of death unknown		0	Cause of death unknown		0	Cause of death unknown		0
Other		0	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 25]

Target years for follow-up year 1: 1989~1993

Total number of initial registrations in target years: 622

Number of deaths: 183

Total number of registrations for follow-up year 25: 324 (73.8 %)

3. Native liver survivors: liver function tests			4. Esophageal varices in cases without liver trar			7. Surgeries other than liver transplant (except for unknown cas		
Number of liver function tests (implemer 121)			Yes 26			Yes 13		
Total bilirubin (mg/dl)			(Duplications in treatment content)	No treatment 20		(Duplications in treatment content)	Hilar re-dissection 2	
Minimum 0.3				Treatment 6			Hilar curettage 0	
Maximum 17.3				scopic treatment or scleroththerapy 6			Endoscopic hilar curetta 0	
Mean 1.4				Direct approach 0			Other 14	
Standard deviation 1.7				Shunt 0			Unknown 1	
ALT (IU/l)				Other 0			None 202	
Minimum 6.0			Unknown 0		Unknown 62			
Maximum 288.0			Details of treatment unknown 0		8. Re-elevation of jaundice (except for unknown cases)			
Mean 38.6			None 53			Yes 49		
Standard deviation 38.4			Unknown 80			No 159		
γGTP (IU/l)			5. Hypersplenism in cases without liver transplar			Unknown 69		
Minimum 4.0			Yes 39			If yes		
Maximum 1710.0			No treatment 31			Before		
Mean 126.4			Treatment 5			re-elevation		
Standard deviation 216.8			Partial splenic embolization 3			Cause of		
ChE (IU/l)			(Duplications in treatment content)			re-elevation		
Below normal range 42			Splenuctomy 2			(includes duplications)		
Within normal range 65			Other 1			Final measures		
Unknown 14			Details of treatment unknown 0			Monitor patient 9		
ALP (IU/l)			Use of treatment unknown 3			Conservative treatment 25		
Above normal range 37			None 75			Re-curative surgery 1		
Within normal range 76			Unknown 45			Liver transplant 11		
Unknown 8			6. Other complications (except for unknown cases)			Other 7		
			Cholangitis 7			(includes duplications)		
			Gastrointestinal bleeding other than varices 6			Unknown 3		
			Ascites 3					
			Fat soluble vitamin deficiency 2					
			Intrahepatic cholelithiasis 6					
			Pulmonary hypertension 3					
			Hepatopulmonary syndrome 2					
			Intestinal obstruction 37					
			Other 27					
			(Includes duplications)					

Japanese Biliary Atresia Registry (JBAR) Sheet 3/3 [Follow-up Year 25]

Target years for follow-up year 1: 1989~1993

Total number of initial registrations in target years: 622

Number of deaths: 183

Total number of registrations for follow-up year 25: 324 (73.8 %)

Outcome with liver transplant		118	Outcome without liver transplant		159
9. Mental development			9. Mental development		
Excellent		2	Excellent		11
Normal		76	Normal		108
Low		6	Low		4
Unknown		34	Unknown		36
10-1. Physical development			10-1. Physical development		
Excellent		1	Excellent		8
Normal		77	Normal		111
Low		3	Low		2
Unknown		37	Unknown		38
10-2. Physical status			10-2. Physical status		
Height (cm)			Height (cm)		
	Minimum	141.4		Minimum	147.0
	Maximum	181.0		Maximum	180.5
	Mean	159.4		Mean	162.2
	Standard deviation	8.6		Standard deviation	8.0
Weight (kg)			Weight (kg)		
	Minimum	29.0		Minimum	38.9
	Maximum	73.4		Maximum	123.7
	Mean	52.3		Mean	58.8
	Standard deviation	8.5		Standard deviation	12.0
11. Menstruation (females)			11. Menstruation (females)		
Yes		36	Yes		66
No		0	No		2
Unknown		41	Unknown		42
12. Pregnancy (females)			12. Pregnancy (females)		
Yes		9	Yes		13
No		38	No		64
Unknown		30	Unknown		33
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		70	Generally, no restrictions in school life, working life		116
Requires restrictions in school life, working		9	Requires restrictions in school life, working l		12
Unknown		39	Unknown		31
14. Marriage			14. Marriage		
Yes		16	Yes		27
No		58	No		74
Unknown		44	Unknown		58
15. Employment			15. Employment		
Yes		53	Yes		87
No		13	No		14
Unknown		52	Unknown		58

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