

## Japanese Biliary Atresia Registry (JBAR) Sheet 1/3 [Follow-up Year 30 ]

Target years for follow-up year 1: 1989~1992

Total number of initial registrations in target years: 508

Number of deaths: 162

Total number of registrations for follow-up year 25: 199 (57.5 %)

Outcome with liver transplant		70	Outcome without liver transplant		88	Unknown cases		41
Living		53	Living		69	Living		1
Living without jaundice		45	Living without jaundice		52	Living without jaundice		1
Living with jaundice		7	Living with jaundice		13	Living with jaundice		0
Unknown		1	Unknown		4	Unknown		0
Death		3	Death		1	Death		0
Unknown		14	Unknown		18	Unknown		40
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		7321	Minimum		10988	Minimum		ND
Maximum		11389	Maximum		10988	Maximum		ND
Mean		8896	Mean		10988	Mean		ND
Standard deviation		-	Standard deviation		-	Standard deviation		ND
Cause of death			Cause of death			Cause of death		
Liver failure		2	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		2	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		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 30 ]

Target years for follow-up year 1: 1989~1992

Total number of initial registrations in target years: 508

Number of deaths: 162

Total number of registrations for follow-up year 25: 199 (57.5 %)

3. Native liver survivors: liver function tests (since 1988)		4. Esophageal varices in cases without liver transplantation		7. Surgeries other than liver transplant (except for unknown cases)			
Number of liver function tests (implemented) 63		Yes	11	Yes	10		
Total bilirubin (mg/dl)		(Duplications in treatment content)	No treatment	(Duplications in treatment content)	Hilar re-dissection	0	
Minimum	0.4		Treatment		3	Hilar curettage	0
Maximum	28.5		Endoscopic treatment or sclerotherapy		3	Endoscopic hilar curettage	0
Mean	1.8		Direct approach		0	Other	10
Standard deviation	4.1		Shunt		0	Unknown	1
ALT (IU/l)			Other		0	None	108
Minimum	7.0		Unknown		0	Unknown	40
Maximum	381.0	Details of treatment unknown 2		8. Re-elevation of jaundice (except for unknown cases)			
Mean	45.5	None	27		Yes	29	
Standard deviation	56.4	Unknown	50		No	91	
yGTP (IU/l)		5. Hypersplenism in cases without liver transplantation			Unknown	38	
Minimum	11.0	Yes	15	If yes			
Maximum	796.0	(Duplications in treatment content)	No treatment	Before re-elevation	No jaundice	19	
Mean	125.1		Treatment	4	Cause of re-elevation (includes duplications)	Mild jaundice	8
Standard deviation	160.2		Partial splenic embolization	1		Unknown	2
ChE (IU/l)			Splenectomy	2		Ascending cholangitis	13
Below normal range	23		Other	2		Other	13
Within normal range	33		Details of treatment unknown	0		Unknown	1
Unknown	7		Use of treatment unknown	3		Final measures	Monitor patient
ALP (IU/l)		None	46	Conservative treatment		14	
Above normal range	16	Unknown	27	Re-curative surgery	0		
Within normal range	42	6. Other complications (except for unknown cases)		Liver transplant	3		
Unknown	5	Cholangitis	3	Other	4		
		Gastrointestinal bleeding other than varices	2	(includes duplications)	Unknown	1	
		Ascites	3				
		Fat soluble vitamin deficiency	1				
		Intrahepatic cholelithiasis	2				
		Pulmonary hypertension	0				
		Hepatopulmonary syndrome	0				
		Intestinal obstruction	15				
		Other	19				
		(Includes duplications)					

# Japanese Biliary Atresia Registry (JBAR) Sheet 3/3 [Follow-up Year 30 ]

Target years for follow-up year 1: 1989~1992

Total number of initial registrations in target years: 508

Number of deaths: 162

Total number of registrations for follow-up year 25: 199 (57.5 %)

Outcome with liver transplant		70	Outcome without liver transplant		88
<b>9. Mental development</b>			<b>9. Mental development</b>		
Excellent		1	Excellent		4
Normal		47	Normal		56
Low		7	Low		2
Unknown		15	Unknown		26
<b>10-1. Physical development</b>			<b>10-1. Physical development</b>		
Excellent		0	Excellent		2
Normal		52	Normal		58
Low		2	Low		0
Unknown		16	Unknown		28
<b>10-2. Physical status</b>			<b>10-2. Physical status</b>		
Height (cm)			Height (cm)		
	Minimum	144.4		Minimum	148.0
	Maximum	177.3		Maximum	173.0
	Mean	159.0		Mean	161.9
	Standard deviation	8.5		Standard deviation	7.2
Weight (kg)		0.0	Weight (kg)		0.0
	Minimum	37.7		Minimum	42.1
	Maximum	79.9		Maximum	88.5
	Mean	53.1		Mean	60.0
	Standard deviation	10.0		Standard deviation	11.3
<b>11. Menstruation (females)</b>			<b>11. Menstruation (females)</b>		
Yes		23	Yes		33
No		0	No		0
Unknown		21	Unknown		23
<b>12. Pregnancy (females)</b>			<b>12. Pregnancy (females)</b>		
Yes		9	Yes		18
No		19	No		19
Unknown		16	Unknown		19
<b>13. Restrictions in daily activity</b>			<b>13. Restrictions in daily activity</b>		
Infant, preschool child		0	Infant, preschool child		0
Generally, no restrictions in school life, working life		42	Generally, no restrictions in school life, working life		58
Requires restrictions in school life, working		8	Requires restrictions in school life, working l		3
Unknown		20	Unknown		26
<b>14. Marriage</b>			<b>14. Marriage</b>		
Yes		17	Yes		23
No		25	No		24
Unknown		28	Unknown		41
<b>15. Employment</b>			<b>15. Employment</b>		
Yes		32	Yes		48
No		9	No		3
Unknown		29	Unknown		37

注)「10-2.身体状況」についてはJBAR2016より収集しているデータの集計です。