

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

Target years for follow-up year 1: 1989~2021

Total number of initial registrations in target years: 3792

Number of deaths: 254

追跡1年目登録総数 : 2641例 (74.6 %)

Outcome with liver transplant		738	Outcome without liver transplant		1894	Unknown cases		9
Living		692	Living		1820	Living		4
Living without jaundice		614	Living without jaundice		1582	Living without jaundice		3
Living with jaundice		29	Living with jaundice		204	Living with jaundice		1
Unknown		49	Unknown		34	Unknown		0
Death		15	Death		68	Death		0
Unknown		31	Unknown		6	Unknown		5
Age at death (days)			Age at death (days)			Age at death (days)		
Minimum		387	Minimum		248	Minimum		ND
Maximum		801	Maximum		920	Maximum		ND
Mean		594.7	Mean		492.8	Mean		ND
Standard deviation		129.0	Standard deviation		157.9	Standard deviation		ND
Cause of death			Cause of death			Cause of death		
Liver failure		0	Liver failure		8	Liver failure		0
Rupture of esophageal varices		0	Rupture of esophageal varices		1	Rupture of esophageal varices		0
Pneumonia		1	Pneumonia		1	Pneumonia		0
Peritonitis		0	Peritonitis		0	Peritonitis		0
Sepsis after cholangitis		0	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		2	Cause of death unknown		0
Other		10	Other		51	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 1]

Target years for follow-up year 1: 1989~2021

Total number of initial registrations in target years: 3792

Number of deaths: 254

追跡1年目登録総数 : 2641例 (74.6 %)

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 1196			Yes 330			Yes 201								
Total bilirubin (mg/dl)			(Duplications in treatment content)			(Duplications in treatment content)								
Minimum 0.1									No treatment 185			Hilar re-dissection 72		
Maximum 61.0									Treatment 132			Hilar curettage 21		
Mean 1.0									Endoscopic treatment or sclerotherapy 122			Endoscopic hilar curettage 4		
Standard deviation 3.0									Direct approach 0			Other 125		
									Shunt 1			Unknown 8		
ALT (IU/l)						None 2311								
Minimum 2.9						Unknown 121								
Maximum 786.0														
Mean 67.7			None 727											
Standard deviation 69.4			Unknown 837											
γGTP (IU/l)			5. Hypersplenism in cases without liver transplan			8. Re-elevation of jaundice (except for unknown cases)								
Minimum 7.0			Yes 319			Yes 598								
Maximum 1892.0			(Duplications in treatment content)			No 1917								
Mean 165.6						No treatment 294			Unknown 117					
Standard deviation 196.3						Treatment 5			If yes					
						Partial splenic embolization 1			Before re-elevation					
ChE (IU/l)						Splenectomy 0			No jaundice 230					
Below normal range 203						Other 9			Mild jaundice 149					
Within normal range 822			Details of treatment unknown 1			Unknown 23								
Unknown 171			Use of treatment unknown 21			Cause of re-elevation (includes duplications)								
ALP (IU/l)			None 1401			Ascending cholangitis 408								
Above normal range 54			Unknown 174			Other 108								
Within normal range 1078			6. Other complications (except for unknown cases)			Unknown 86								
Unknown 64			Cholangitis 36			Final measures								
			Gastrointestinal bleeding other than varices 90			Monitor patient 51								
			Ascites 173			Conservative treatment 339								
			Fat soluble vitamin deficiency 51			Re-curative surgery 32								
			Intrahepatic cholelithiasis 3			Liver transplant 126								
			Pulmonary hypertension 0			Other 44								
			Hepatopulmonary syndrome 4			Unknown 19								
			Intestinal obstruction 364											
			Other 242											
			(Includes duplications)											

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

Target years for follow-up year 1: 1989~2021

Total number of initial registrations in target years: 3792

Number of deaths: 254

追跡1年目登録総数 : 2641例 (74.6 %)

Outcome with liver transplant		738	Outcome without liver transplant		1894
9. Mental development			9. Mental development		
Excellent		9	Excellent		38
Normal		558	Normal		1637
Low		29	Low		72
Unknown		142	Unknown		147
10-1. Physical development			10-1. Physical development		
Excellent		8	Excellent		38
Normal		543	Normal		1615
Low		54	Low		119
Unknown		133	Unknown		122
10-2. Physical status			10-2. Physical status		
Height (cm)			Height (cm)		
	Minimum	67.5		Minimum	70.5
	Maximum	83.4		Maximum	77.4
	Mean	71.9		Mean	73.9
	Standard deviation	5.1		Standard deviation	2.4
Weight (kg)			Weight (kg)		
	Minimum	7.3		Minimum	7.8
	Maximum	12.2		Maximum	11.9
	Mean	8.9		Mean	9.5
	Standard deviation	1.6		Standard deviation	1.3

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