

Japanese Biliary Atresia Registry (JBAR) Summary Sheet

1989~2018 only type III

1989~2018 2968 cases

(1999~2018 1884 cases)

Number of registered cases by region			Birth order		Meconium color		
	No. registrations	No. hospitals	First child	1393	Normal		1286
Hokkaido	79	3	Second child	1037	Abnormal		412
Tohoku	263	13	Third child	364	Unknown		1270
Kanto	999	40	Fourth child	62	Yellow stool		
Chubu and Hokuriku	494	22	Fifth child and later	30	Yes		1666
Kinki	562	17	Unknown	82	No		640
Chugoku and Shikoku	239	20	Number of babies (1999~)		Unknown		662
Kyushu	332	14	Singleton	1774	Neonatal jaundice		
Total	2968	129	Twins or more	51	Yes		1797
			Unknown	59	No		764
Age of parents			Gender		Unknown		407
	Mean	Standard deviation	Male	1139	Phototherapy		
Father	32.3	6.0	Female	1826	Yes		1108
Mother	29.9	5.0	Unknown	3	No		1462
Maternal history during pregnancy			Birth weight (g)		Unknown		398
	Febrile rash disease	35	Maximum	4573	Pathological bleeding		
	Other	607	Minimum	928	Yes		278
		0	Mean	2900.3	(bleeding sites have duplications)	Intracranial	148
Family history (parents)			Standard deviation	449.7	Gastrointestinal		84
Biliary atresia	Parents	0	~1499	28	Subcutaneous		26
	Siblings	3	1500~1999	60	Other		44
	Other	4	2000~2499	359	No		2573
Choledochal cyst	Parents	4	2500~2999	1266	Unknown		117
	Siblings	1	3000~3499	998	Age at bleeding (days)		
	Other	2	3500~3999	189	Maximum		134
Hepatitis	Parents	44	4000~	23	Minimum		0
	Siblings	4	不明	45	Mean		52.9
	Other	33	Gestational age (weeks)		Standard deviation		26.7
Other		250	Maximum	42	Prothrombin activity (%)		
			Minimum	26	Maximum		181
			Mean	38.6	Minimum		2
			Standard deviation	1.8	Mean		82.1
			~29	8	Standard deviation		31.6
			30~31	17	≥70%		184
			32~33	27	≥40%<70%		37
			34~35	76	<40%		29
			36~37	418	PT-INR		
			38~39	1379	Maximum		13.4
			40~41	931	Minimum		0.8
			42~	20	Mean		1.5
			Unknown	92	Standard deviation		1.5
Chromosomal abnormality					<1.15%		170
	Yes			11	≥1.15% <1.4%		39
	Down症			0	≥1.4%		44
	その他			12			
	No			2258			
	Unknown			699			

1989~2018

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1884 case

Age at initial consult (days)		Name of associated congenital anomaly		Name of associated congenital anomaly (Cont.)	
Maximum	410	Heart		Urogenital	
Minimum	0	Tetralogy of Fallot	5	Polycystic kidney	3
Mean	53.9	Patent foramen ovale	2	Renal hypoplasia	3
Standard deviation	32.4	Patent ductus arteriosus	17	Hydronephrosis	8
Age at hospitalization (days)			VSD	Horseshoe kidney	1
Maximum	435		ASD	Double renal pelvis and ureter	2
Minimum	0	Dextrocardia	5	Hydrocele of the testis	11
Mean	56.1	Pulmonary stenosis	12	Cryptorchidism	2
Standard deviation	33.0	Other	45	Extrophy of bladder	0
Prenatal diagnosis (1999~)				Hypospadias	1
Yes	36	Vascular		Urethral stricture	0
No	1804	Preduodenal portal vein	39	VUR	1
Unknown	44	Absence of the inferior vena cava	19	Other	19
Screening		Anomalous pulmonary venous return	1		
Yes	385	Other	12	Musculoskeletal	
No	2455			Spina bifida	2
Unknown	128	Spleen	0	Costal malformation	0
Stool color at admission		Polysplenia	55	Other	5
Yellow	125	Accessory spleen	15		
Pale yellow	847	Asplenia	4	Limb and external anomaly	
Gray	1849	Other	0	Accessory ear	1
Unknown	147			Cleft lip	0
Total bilirubin immediately before surgery (mg/dl)		Gastrointestinal		Cleft lip and palate	6
Maximum	58.2	Malrotation of intestine	43	Polysyndactyly	6
Minimum	2.9	Esophageal atresia	3	Clubfoot	5
Mean	9.4	Intestinal atresia	11	Other	13
Standard deviation	3.7	Anorectal malformation	14		
Direct bilirubin immediately before surgery		Hirschsprung's disease	0	Central nervous system	
Maximum	40.5	Situs inversus	33	Hydrocephalus	3
Minimum	1.4	Annular pancreas	2	Microcephaly	0
Mean	6.5	Gastric rupture	0	Anencephaly	0
Standard deviation	2.7	Meconium peritonitis	12	Other	10
Implemented tests		Meckel's diverticulum	14		
Serum Lp-X	1377	Cloacal exstrophy	0	Endocrine metabolic system	
Score test	894	Alagille syndrome	9	Cretinism	1
Ultrasound	2814	Pancreaticobiliary maljunction	2	Other	2
ERCP	94	Intussusception	2		
Serum bile acid	1883	Other	35	Other malformations	
Percutaneous liver biopsy	77			Diaphragmatic hernia	1
Duodenal juice analysis	1694	Respiratory		Omphalocele	1
Hepatobiliary scintigraphy	1951	Tracheal/bronchial stenosis	1	Gastroschisis	1
MRI (MRCP)	203	Cystic lung disease	2	Inguinal hernia	65
CT	258	Lobar emphysema	0	Umbilical hernia	78
Other	114	Pulmonary hypoplasia	2	Other	12
Multiple answers allowed		Other	4		

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Surgery implemented		Age at time of surgery (days)	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	
Yes		2968	~30	157	154	105	68.2	61	39.6
	Bile drainage surgery	2933	31~45	448	448	270	60.3	171	38.2
	Liver transplant	14	46~60	725	722	465	64.4	307	42.5
	Exploratory laparotomy	21	61~70	462	462	280	60.6	190	41.1
	Unknown	0	71~80	431	428	257	60.0	184	43.0
No (all 1998 cases)		0	81~90	263	262	142	54.2	111	42.4
Biliary reconstruction		0	91~120	320	315	170	54.0	138	43.8
	Hepaticoenterostomy	26	121~150	103	98	35	35.7	35	35.7
	Portoenterostomy	2893	151~	49	34	7	20.6	18	52.9
	Other	2	Unknown	10	10	7	70.0	3	30.0
	Unknown	12	Total	2968	2933	1738	59.3	1218	41.5

Age at time of surgery (days)	
	0
Maximum	419
Minimum	7
Mean	67.1
Standard deviation	28.6

Surgical procedure		No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)
2106						
No cholangitis prevention						
Simple Roux-en-Y anastomosis		2084	1244	59.7	901	43.2
	<20 cm	13	7	53.8	5	38.5
	≥20 cm <40 cm	624	367	58.8	289	46.3
	≥40 cm <60 cm	1203	731	60.8	502	41.7
	≥60 cm	218	127	58.3	95	43.6
	Unknown	26	12	46.2	10	38.5
	Porto/hepaticoduodenostomy	10	9	90.0	3	30.0
	Other	4	1	25.0	1	25.0
	Unknown	8	6	75.0	0	0.0
Cholangitis prevention		819	0	0.0	0	0.0
	Suruga II method	145	79	54.5	67	46.2
	Double Roux-en-Y reconstruction	10	7	70.0	4	40.0
	Outer end jejunostomy	62	31	50.0	27	43.5
	Roux-en-Y reconstruction with intestinal valve	554	329	59.4	192	34.7
	Hepatic portocholecystostomy or hepaticocholecystostomy	5	3	60.0	0	0.0
	Other	39	20	51.3	17	43.6
	Unknown	4	4	100.0	2	50.0
Cholangitis prevention unknown		8	0	0.0	4	50.0
Total		2933				

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Basic Type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	Esophageal varices checked before or during surgery	
I	0	0	0		0		Not checked	2092
I -cyst	0	0	0		0		Varices present	102
II	0	0	0		0		No varices	631
III	2968	2933	1738	59.3	1218	41.5	Unknown	143
Unknown	0	0	0		0		Total	2968
Total	2968	2933	1738	59.3	1218	41.5	Choleretics	
Sub type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)		
a1	417	408	259	63.5	167	40.9	Dehycol	1155
a2	121	118	70	59.3	37	31.4	UDCA	2599
b1	1824	1812	1072	59.2	778	42.9	Glucagon	599
b2	92	91	42	46.2	36	39.6	Secretin	108
c1	220	220	133	60.5	95	43.2	PGE2	198
c2	94	88	33	37.5	26	29.5	PGF2 α	44
d	136	135	97	71.9	51	37.8	Steroids	2628
Unknown	64	61	32	52.5	28	45.9	Taurine	267
Total	2968	2933	1738	59.3	1218	41.5	Phenobarbital	216
							Other	126
Type of porta hepatitis	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	(includes duplications)	
α	4	4	2	50.0	0	0.0	Bile drainage after first surgery	
β	33	33	24	72.7	13	39.4	Yes	940
γ	118	116	69	59.5	44	37.9	No	111
μ	341	336	195	58.0	141	42.0	Unknown	29
ν	2294	2278	1378	60.5	962	42.2	Total	1080
o	122	114	41	36.0	38	33.3	Jaundice after first surgery	
Unknown	56	52	29	55.8	20	38.5	Disappeared	629
Total	2968	2933	1738	59.3	1218	41.5	Improved	200
							Unchanged	67
							Worsened	40
							Unknown	144
							Total	1080
							Jaundice re-elevation	
							No re-elevation	470
							Re-elevated after disappearance	152
							Re-elevated after improvement	164
							Unknown	43
							Total	829
							Measures for re-elevation	
							Monitor progress	67
							Conservative treatment	509
							Re-do surgery	202
							Liver transplant	352
							Total	1130
Gallbladder								
Yes 2750								
No bile 1229								
Yellow 17								
Pale yellow 60								
Colorless 1204								
Other 73								
Unknown 167								
Absent 170								
Unknown 48								
Total 2968								
Cystic structure in extrahepatic bile duct								
Yes 129								
Yellow 6								
Pale yellow 22								
Colorless 72								
Other 14								
Unknown 15								
No 2761								
Unknown 78								
Total 2968								

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Cholangitis		1989~1998	1999~	Postoperative complications					
Yes		400	818	(Includes duplications)	Ileus	133			
	Mean timing of first time (weeks)		11.3		Peritonitis	57			
	Mean timing of second time (weeks)		19.4		Hemorrhage	126			
No	625	945	Pulmonary complications		37				
Unknown	55	90	Acute liver failure		45				
Total	1080	1853	Infection		185				
Reoperation within 6 months (1999~)					Other	182			
	Yes	438	Age at liver transplant (days) (1999~)		Maximum	5630	After initial surgery	5574	
	No	1277			Minimum	83		0	
	Unknown	169			Mean	331.6		256.0	
Total	1884		Standard deviation	384.8		389.8			
Reoperation	1054		Interim outcome at 6 months following the initial surgery (1999~)						
Liver transplant	464		Death	31					
Hilar re-dissection	366		Living	1842					
Hilar curettage	114			Without jaundice	1054				
Endoscopic hilar curettage	11		With jaundice	497					
Other	99		Jaundice unknown	50					
Reoperations other than liver transplant (1999~)			Post-transplant	241					
Age (days)	Maximum	865	Total				1884		
	Minimum	28	Age at death (days) within 6 months (1999~)						
	Mean	140.2	Maximum	330					
	Standard deviation	101.4	Minimum	48					
Jaundice	Disappeared	100	Mean	171.7					
	Improved	61	Standard deviation	72.6					
	Unchanged	60	Final outcome						
	Worsened	45	1989~1998				1999~		
Unknown	11	Death				167	96	71	
Re-reoperation	231	Living				2783	981	1802	
Liver transplant	112	Without jaundice				563	1010		
Hilar re-dissection	63	With jaundice				224	202		
Hilar curettage	13	Unknown living				86	19		
Endoscopic hilar curettage	6	Transplant living				100	569		
Other	37	Survival details unknown				8	2		
Re-reoperations other than liver transplant (1999~)			Unknown				18	7	11
Age (days)	Maximum	771	Total				2968	1084	1884
	Minimum	75	Cause of death						
	Mean	243.2	Liver failure				74		
	Standard deviation	168.5	Rupture of esophageal varices				8		
Jaundice	Disappeared	18	Pneumonia				7		
	Improved	4	Peritonitis				2		
	Unchanged	11	Sepsis after cholangitis				9		
	Worsened	10	Post-transplantation death				34		
	Unknown	3	Other				61		
			Unknown				5		
			(Includes duplications)						

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Screening (from 2012)					
By stool color card					
Yes: 212					
Before 1 month old	132	At 1 month old	136	After 1 month old	122
Observed age (days)		Observed age (days)		Observed age (days)	
Observed date not stated	52	Observed date not stated	50	Observed date not stated	29
Maximum	37	Maximum	61	Maximum	147
Minimum	0	Minimum	14	Minimum	31
Mean	13.7	Mean	32.9	Mean	58.8
Standard deviation	8.6	Standard deviation	6.2	Standard deviation	21.7
Number of the color		Number of the color		Number of the color	
1	5	1	5	1	15
2	22	2	25	2	46
3	32	3	37	3	35
4	44	4	45	4	13
5	2	5	1	5	0
6	0	6	1	6	0
7	4	7	0	7	1
Unknown	23	Unknown	22	Unknown	12
By urinary sulfated bile acid		By direct bilirubin		Other	
Yes	25	Yes	168	Yes	9
Abnormal	23	Abnormal	163		
Normal	2	Normal	3		
Unknown	0	Unknown	2		