

Japanese Biliary Atresia Registry (JBAR) Summary Sheet

1989~2022 only type III

1989~2022

3310 cases

(1999~2022

2224 cases)

Number of registered cases by region			Birth order		Meconium color	
	No. registrations	No. hospitals	First child	1565	Normal	1451
Hokkaido	86	3	Second child	1139	Abnormal	442
Tohoku	286	13	Third child	413	Unknown	1417
Kanto	1133	41	Fourth child	69	Yellow stool	
Chubu and Hokuriku	549	24	Fifth child and later	34	Yes	1878
Kinki	615	17	Unknown	90	No	685
Chugoku and Shikoku	267	20	Number of babies (1999~)		Unknown	747
Kyushu	374	15	Singleton	2088	Neonatal jaundice	
Total	3310	133	Twins or more	58	Yes	1971
Age of parents			Unknown	77	No	900
	Mean	Standard deviation	Gender		Unknown	439
Father	32.5	6.0	Male	1274	Phototherapy	
Mother	30.1	5.1	Female	2034	Yes	1229
Maternal history during pregnancy			Unknown	2	No	1644
	Febrile rash disease	42	Birth weight (g)		Unknown	437
	Other	640	Maximum	4573	Pathological bleeding	
		0	Minimum	834	Yes	293
Family history (parents)			Mean	2899.3	Intracranial	156
Biliary atresia	Parents	1	Standard deviation	450.2	(bleeding sites have duplications)	Gastrointestinal
	Siblings	3	~1499	34		Subcutaneous
	Other	4	1500~1999	65		Other
Choledochal cyst	Parents	5	2000~2499	390	No	2900
	Siblings	1	2500~2999	1428	Unknown	117
	Other	2	3000~3499	1113	Age at bleeding (days)	
Hepatitis	Parents	46	3500~3999	211	Maximum	134
	Siblings	4	4000~	24	Minimum	0
	Other	33	不明	45	Mean	52.8
Other		270	Gestational age (weeks)		Standard deviation	27.4
			Maximum	42	Prothrombin activity (%)	
			Minimum	26	Maximum	240
			Mean	38.6	Minimum	2
			Standard deviation	1.8	Mean	85.4
			~29	10	Standard deviation	32.0
			30~31	19	≥70%	438
			32~33	32	≥40%<70%	86
			34~35	82	<40%	55
			36~37	465	PT-INR	
			38~39	1558	Maximum	21.0
			40~41	1029	Minimum	0.2
			42~	21	Mean	1.5
			Unknown	94	Standard deviation	1.6
Chromosomal abnormality					<1.15%	385
Yes					≥1.15% <1.4%	102
	Down症				≥1.4%	102
	その他					
No						2588
Unknown						710

Age at initial consult (days)		Name of associated congenital anomaly		Name of associated congenital anomaly (Cont.)	
Maximum	387	Heart		Urogenital	
Minimum	0	Tetralogy of Fallot	7	Polycystic kidney	3
Mean	52.7	Patent foramen ovale	3	Renal hypoplasia	4
Standard deviation	32.5	Patent ductus arteriosus	17	Hydronephrosis	8
<b>Age at hospitalization (days)</b>		VSD	31	Horseshoe kidney	1
Maximum	435	ASD	24	Double renal pelvis and ureter	2
Minimum	0	Dextrocardia	5	Hydrocele of the testis	12
Mean	54.9	Pulmonary stenosis	16	Cryptorchidism	2
Standard deviation	33.0	Other	48	Extrophy of bladder	0
<b>Prenatal diagnosis (1999~)</b>				Hypospadias	1
Yes	52	Vascular		Urethral stricture	0
No	2122	Preduodenal portal vein	45	VUR	3
Unknown	50	Absence of the inferior vena cava	24	Other	18
<b>Screening</b>		Anomalous pulmonary venous return	1		
Yes	554	Other	14	Musculoskeletal	
No	2625			Spina bifida	2
Unknown	131	Spleen	0	Costal malformation	0
<b>Stool color at admission</b>		Polysplenia	64	Other	6
Yellow	155	Accessory spleen	15		
Pale yellow	1004	Asplenia	5	Limb and external anomaly	
Gray	1960	Other	0	Accessory ear	1
Unknown	191			Cleft lip	0
<b>Total bilirubin immediately before surgery (mg/dl)</b>		Gastrointestinal		Cleft lip and palate	9
Maximum	58.2	Malrotation of intestine	56	Polysyndactyly	8
Minimum	2.9	Esophageal atresia	5	Clubfoot	5
Mean	9.3	Intestinal atresia	17	Other	10
Standard deviation	3.7	Anorectal malformation	16		
<b>Direct bilirubin immediately before surgery</b>		Hirschsprung's disease	0	Central nervous system	
Maximum	40.5	Situs inversus	40	Hydrocephalus	3
Minimum	0.9	Annular pancreas	2	Microcephaly	0
Mean	6.4	Gastric rupture	0	Anencephaly	0
Standard deviation	2.7	Meconium peritonitis	13	Other	10
<b>Implemented tests</b>		Meckel's diverticulum	14		
Serum Lp-X	1379	Cloacal exstrophy	0	Endocrine metabolic system	
Score test	895	Alagille syndrome	11	Cretinism	1
Ultrasound	3137	Pancreaticobiliary maljunction	2	Other	3
ERCP	94	Intussusception	2		
Serum bile acid	2096	Other	26	Other malformations	
Percutaneous liver biopsy	89			Diaphragmatic hernia	1
Duodenal juice analysis	1803	Respiratory		Omphalocele	1
Hepatobiliary scintigraphy	2180	Tracheal/bronchial stenosis	1	Gastroschisis	1
MRI (MRCP)	222	Cystic lung disease	2	Inguinal hernia	66
CT	279	Lobar emphysema	0	Umbilical hernia	79
Other	117	Pulmonary hypoplasia	2	Other	13
<b>Multiple answers allowed</b>		Other	4		

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Surgery implemented		Age at time of surgery (days)	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)		
Yes		~30	208	205	139	67.8	79	38.5		
	Bile drainage surgery	31~45	530	530	325	61.3	205	38.7		
	Liver transplant	46~60	806	803	518	64.5	347	43.2		
	Exploratory laparotomy	61~70	502	502	309	61.6	215	42.8		
No	(all ~ 1998 cases)	71~80	467	464	282	60.8	205	44.2		
Unknown		81~90	279	278	153	55.0	121	43.5		
<b>Biliary reconstruction</b>		91~120	349	343	183	53.4	150	43.7		
	Hepaticoenterostomy	121~150	111	106	37	34.9	39	36.8		
	Portoenterostomy	151~	50	35	6	17.1	19	54.3		
	Other	Unknown	8	8	6	75.0	3	37.5		
	Unknown		12	Total	3310	3274	1958	59.8	1383	42.2
<b>Age at time of surgery (days)</b>										
	Maximum		303							
	Minimum		7							
	Mean		66.0							
	Standard deviation		27.9							
<b>Surgical procedure</b>		No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)				
No cholangitis prevention		2404								
No cholangitis prevention	Simple Roux-en-Y anastomosis	2376	1428	60.1	1038	43.7				
	<20 cm	23	12	52.2	10	43.5				
	≥20 cm <40 cm	761	450	59.1	346	45.5				
	≥40 cm <60 cm	1342	822	61.3	573	42.7				
	≥60 cm	222	130	58.6	97	43.7				
	Unknown	28	14	50.0	12	42.9				
	Porto/hepaticoduodenostomy	10	9	90.0	3	30.0				
	Other	4	1	25.0	1	25.0				
	Unknown	14	7	50.0	3	21.4				
	Cholangitis prevention	861	0	0.0	0	0.0				
Cholangitis prevention	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	593	360	60.7	213	35.9				
	Hepatic portocholecystostomy or hepaticocholecystostomy	5	3	60.0	0	0.0				
	Other	42	23	54.8	20	47.6				
	Unknown	4	4	100.0	2	50.0				
Cholangitis prevention unknown	9	0	66.7	5	55.6					
Total	3274									



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Cholangitis		1989~1998	1999~	Postoperative complications		
Yes		401	982		Ileus	151
	Mean timing of first time (weeks)		11.2		Peritonitis	62
	Mean timing of second time (weeks)		19.0		Hemorrhage	131
No		626	1116		Pulmonary complications	38
Unknown		55	94		Acute liver failure	47
Total		1082	2192		Infection	203
Reoperation within 6 months (1999~)				(Includes duplications)	Other	211
	Yes	530		Age at liver transplant (days) (1999~)	660	After initial surgery
	No	1510		Maximum	5630	5574
	Unknown	184		Minimum	83	0
Total		2224		Mean	323.7	249.6
Reoperation		1185		Standard deviation	359.6	364.0
	Liver transplant	539		Interim outcome at 6 months following the initial surgery (1999~)		
	Hilar re-dissection	395		Death	34	
	Hilar curettage	132		Living	2172	
	Endoscopic hilar curettage	11			Without jaundice	1262
	Other	108			With jaundice	560
Reoperations other than liver transplant (1999~)			333		Jaundice unknown	55
Age (days)	Maximum	865		Unknown	18	
	Minimum	28		Total	2224	
	Mean	139.8		Age at death (days) within 6 months (1999~)		
	Standard deviation	95.6		Maximum	330	
Jaundice	Disappeared	128		Minimum	48	
	Improved	72		Mean	174.1	
	Unchanged	67		Standard deviation	69.6	
	Worsened	52		Final outcome		
	Unknown	14		1989~1998		
Re-reoperation		259		Death	176	97
	Liver transplant	133		Living	3112	982
	Hilar re-dissection	64			Without jaundice	563
	Hilar curettage	13			With jaundice	224
	Endoscopic hilar curettage	6			Unknown living	86
	Other	43			Transplant living	101
Re-reoperations other than liver transplant (1999~)			53		Survival details unknown	8
Age (days)	Maximum	771		Unknown	22	7
	Minimum	75		Total	3310	1086
	Mean	243.2		Cause of death		
	Standard deviation	158.3		Liver failure	77	
Jaundice	Disappeared	23		Rupture of esophageal varices	8	
	Improved	4		Pneumonia	7	
	Unchanged	11		Peritonitis	2	
	Worsened	10		Sepsis after cholangitis	9	
	Unknown	5		Post-transplantation death	38	
				Other	63	
			Unknown	5		
				(Includes duplications)		

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Screening (from 2012)					
By stool color card					
Yes: 368					
Before 1 month old	230	At 1 month old	237	After 1 month old	187
Observed age (days)		Observed age (days)		Observed age (days)	
Observed date not stated	67	Observed date not stated	71	Observed date not stated	33
Maximum	64	Maximum	87	Maximum	147
Minimum	0	Minimum	14	Minimum	30
Mean	14.8	Mean	33.5	Mean	58.1
Standard deviation	9.5	Standard deviation	7.9	Standard deviation	20.8
Number of the color		Number of the color		Number of the color	
1	7	1	14	1	23
2	40	2	57	2	87
3	48	3	60	3	43
4	69	4	77	4	18
5	5	5	1	5	1
6	0	6	1	6	0
7	5	7	0	7	1
Unknown	56	Unknown	26	Unknown	14
By urinary sulfated bile acid		By direct bilirubin		Other	
Yes	30	Yes	248	Yes	10
Abnormal	28	Abnormal	224		
Normal	2	Normal	4		
Unknown	1	Unknown	20		