

Japanese Biliary Atresia Registry (JBAR) Summary Sheet

1989~

2022

1989~2022

3862 cases

(1999~2022

2573 cases)

Number of registered cases by region			Birth order		Meconium color		
	No. registrations	No. hospitals	First child	1825	Normal		1703
Hokkaido	107	3	Second child	1334	Abnormal		545
Tohoku	325	13	Third child	472	Unknown		1614
Kanto	1353	41	Fourth child	82	Yellow stool		
Chubu and Hokuriku	639	24	Fifth child and later	39	Yes		2204
Kinki	695	17	Unknown	110	No		803
Chugoku and Shikoku	307	20	Number of babies (1999*)		Unknown		855
Kyushu	436	15	Singleton	2422	Neonatal jaundice		
Total	3862	133	Twins or more	65	Yes		2337
Age of parents			Unknown	85	No		1030
	Mean	Standard deviation	Gender		Unknown		495
Father	32.4	6.0	Male	1431	Phototherapy		
Mother	30.0	5.1	Female	2429	Yes		1447
Maternal history during pregnancy			Unknown	2	No		1914
	Febrile rash disease	45	Birth weight (g)		Unknown		501
	Other	725	Maximum	4940	Pathological bleeding		
		0	Minimum	824	Yes		332
Family history (parents)			Mean	2901.4	Intracranial		169
Biliary atresia	Parents	1	Standard deviation	452.4	(bleeding sites have duplications)		100
	Siblings	3	~1499	39	Subcutaneous		32
	Other	4	1500~1999	77	Other		57
Choledochal cyst	Parents	8	2000~2499	452	No		3388
	Siblings	3	2500~2999	1668	Unknown		142
	Other	3	3000~3499	1285	Age at bleeding (days)		
Hepatitis	Parents	53	3500~3999	254	Maximum		134
	Siblings	7	4000~	31	Minimum		0
	Other	36	不明	56	Mean		52.1
Other		308	Gestational age (weeks)		Standard deviation		27.1
			Maximum	42	Prothrombin activity (%)		
			Minimum	24	Maximum		240
			Mean	38.6	Minimum		2
			Standard deviation	1.8	Mean		85.9
			~29	13	Standard deviation		31.0
			30~31	19	≥70%		503
			32~33	41	≥40%<70%		101
			34~35	95	<40%		55
			36~37	554	PT-INR		
			38~39	1821	Maximum		21.0
			40~41	1181	Minimum		0.2
			42~	27	Mean		1.4
			Unknown	111	Standard deviation		1.5
Chromosomal abnormality					<1.15%		437
Yes					≥1.15% <1.4%		124
	Down症				≥1.4%		108
	その他						
No							3000
Unknown							848

1989~2022

3862 cases

(1999~2022

2573 cases)

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	8	Polycystic kidney	4
Mean	51.2	Patent foramen ovale	3	Renal hypoplasia	5
Standard deviation	33.7	Patent ductus arteriosus	19	Hydronephrosis	12
Age at hospitalization (days)		VSD	44	Horseshoe kidney	1
Maximum	435	ASD	30	Double renal pelvis and ureter	4
Minimum	0	Dextrocardia	7	Hydrocele of the testis	15
Mean	53.6	Pulmonary stenosis	26	Cryptorchidism	2
Standard deviation	34.3	Other	76	Extrophy of bladder	0
Prenatal diagnosis (1999~)				Hypospadias	2
Yes	139	Vascular		Urethral stricture	0
No	2377	Preduodenal portal vein	53	VUR	5
Unknown	57	Absence of the inferior vena cava	27	Other	22
Screening		Anomalous pulmonary venous return	6		
Yes	640	Other	14	Musculoskeletal	
No	3071			Spina bifida	5
Unknown	151	Spleen	0	Costal malformation	0
Stool color at admission		Polysplenia	80	Other	6
Yellow	212	Accessory spleen	16		
Pale yellow	1172	Asplenia	5	Limb and external anomaly	
Gray	2245	Other	0	Accessory ear	2
Unknown	233			Cleft lip	0
Total bilirubin immediately before surgery (mg/dl)		Gastrointestinal		Cleft lip and palate	13
Maximum	58.2	Malrotation of intestine	76	Polysyndactyly	8
Minimum	1.4	Esophageal atresia	6	Clubfoot	6
Mean	9.3	Intestinal atresia	21	Other	13
Standard deviation	3.8	Anorectal malformation	18		
Direct bilirubin immediately before surgery (mg/dl)		Hirschsprung' s disease	0	Central nervous system	
Maximum	40.5	Situs inversus	48	Hydrocephalus	3
Minimum	0.4	Annular pancreas	4	Microcephaly	0
Mean	6.3	Gastric rupture	0	Anencephaly	0
Standard deviation	2.8	Meconium peritonitis	14	Other	11
Implemented tests		Meckel' s diverticulum	16		
Serum Lp-X	1601	Cloacal exstrophy	0	Endocrine metabolic system	
Score test	1025	Alagille syndrome	11	Cretinism	1
Ultrasound	3636	Pancreaticobiliary maljunction	2	Other	3
ERCP	115	Intussusception	2		
Serum bile acid	2424	Other	33	Other malformations	
Percutaneous liver biopsy	102			Diaphragmatic hernia	1
Duodenal juice analysis	2051	Respiratory		Omphalocele	1
Hepatobiliary scintigraphy	2534	Tracheal/bronchial stenosis	2	Gastroschisis	2
MRI (MRCP)	276	Cystic lung disease	3	Inguinal hernia	79
CT	322	Lobar emphysema	0	Umbilical hernia	90
Other	150	Pulmonary hypoplasia	2	Other	15
Multiple answers allowed		Other	5		

1989~2022

3862 cases

(1999~2022 2573 cases)

Surgery implemented		Age at time of surgery (days)	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)
Yes	3857	~30	299	296	211	71.3	106	35.8
	Bile drainage surgery	31~45	636	635	401	63.1	246	38.7
	Liver transplant	46~60	919	916	600	65.5	390	42.6
	Exploratory laparotomy	61~70	583	583	376	64.5	243	41.7
No	(all ~ 1998 cases)	71~80	505	502	312	62.2	222	44.2
Unknown	0	81~90	314	312	176	56.4	134	42.9
Biliary reconstruction		91~120	391	385	214	55.6	166	43.1
	Hepaticoenterostomy	121~150	131	126	49	38.9	51	40.5
	Portoenterostomy	151~	69	49	10	20.4	25	51.0
	Other	Unknown	10	10	7	70.0	3	30.0
	Unknown		14					
Total			3857	3814	2356	61.8	1586	41.6
Age at time of surgery (days)		0						
	Maximum	303						
	Minimum	4						
	Mean	65.3						
	Standard deviation	29.1						
Surgical procedure		No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)		
No cholangitis prevention		2834						
Simple Roux-en-Y anastomosis		2793	1736	62.2	1197	42.9		
	<20 cm	26	13	50.0	10	38.5		
	≥20 cm <40 cm	921	568	61.7	415	45.1		
	≥40 cm <60 cm	1551	984	63.4	650	41.9		
	≥60 cm	259	153	59.1	109	42.1		
	Unknown	36	18	50.0	13	36.1		
Porto/hepaticoduodenostomy		16	15	93.8	4	25.0		
Other		9	3	33.3	3	33.3		
Unknown		16	8	50.0	4	25.0		
Cholangitis prevention		967	0	0.0	0	0.0		
Suruga II method		162	90	55.6	74	45.7		
Double Roux-en-Y reconstruction		10	7	70.0	4	40.0		
Outer end jejunostomy		68	35	51.5	31	45.6		
Roux-en-Y reconstruction with intestinal valve		669	418	62.5	238	35.6		
Hepatic portocholecystostomy or hepaticocholecystostomy		5	3	60.0	0	0.0		
Other		49	29	59.2	23	46.9		
Unknown		4	4	100.0	2	50.0		
Cholangitis prevention unknown		13	0	0.0	6	46.2		
Total		3814						

1989~2022

3862 cases

(1999~2022 2573 cases)

Basic type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	Esophageal varices checked before or during surgery	
I	139	139	99	71.2	57	41.0	Not checked	2780
I -cyst	303	302	238	78.8	116	38.4	Varices present	115
II	72	71	48	67.6	21	29.6	No varices	784
III	3310	3274	1958	59.8	1383	42.2	Unknown	178
Unknown	33	28	13	46.4	9	32.1	Total	3857
Total	3857	3814	2356	61.8	1586	41.6	Choleretics	
Sub type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)		
a1	501	492	315	64.0	204	41.5	Dehydrocol	1324
a2	137	134	80	59.7	42	31.3	UDCA	3380
b1	2278	2264	1384	61.1	976	43.1	Glucagon	725
b2	105	104	49	47.1	39	37.5	Secretin	124
c1	340	340	226	66.5	146	42.9	PGE2	228
c2	107	101	41	40.6	29	28.7	PGF2 α	51
d	177	176	132	75.0	69	39.2	Steroids	3352
Unknown	212	203	129	63.5	81	39.9	Taurine	336
Total	3857	3814	2356	61.8	1586	41.6	Phenobarbital	263
							Other	157
Type of porta hepatis	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	(includes duplications)	
α	109	109	99	90.8	35	32.1	Bile drainage after first surgery	
β	200	200	151	75.5	86	43.0	1989~1998	1999~
γ	187	184	112	60.9	71	38.6	Yes	1119
μ	391	386	229	59.3	164	42.5	No	124
ν	2641	2624	1604	61.1	1124	42.8	Unknown	38
Total	3857	3814	2356	61.8	1586	41.6	Total	1281
								2533
							Jaundice after first surgery	
							1989~1998	1999~
							Disappeared	781
							Improved	218
							Unchanged	71
							Worsened	42
							Unknown	169
							Total	1281
								2533
							Jaundice re-elevation	
							1989~1998	1999~
							No re-elevation	586
							Re-elevated after disappearance	182
							Re-elevated after improvement	181
							Unknown	50
							Total	999
								2223
							Measures for re-elevation	
							Monitor progress	96
							Conservative treatment	609
							Re-do surgery	280
							Liver transplant	452
							Total	1437
Gallbladder								
Yes	3581							
No bile	1473							
Yellow	82							
Pale yellow	153							
Colorless	1541							
Other	102							
Unknown	229							
Absent	206							
Unknown	70							
Total	3857							
Cystic structure in extrahepatic bile duct								
Yes	493							
Yellow	80							
Pale yellow	122							
Colorless	173							
Other	42							
Unknown	76							
No	3256							
Unknown	108							
Total	3857							

1989~2022

3862 cases

(1999~2022 2573 cases)

Cholangitis		1989~1998	1999~	Postoperative complications		
Yes		470	1116	Ileus		171
	Mean timing of first time (weeks)		10.9	Peritonitis		72
	Mean timing of second time (weeks)		18.5	Hemorrhage		142
No		746	1305	Pulmonary complications		43
Unknown		65	112	Acute liver failure		49
Total		1281	2533	Infection		231
Reoperation within 6 months (1999~)				(Includes duplications)	Other	246
	Yes		600	Age at liver transplant (days) (1999~)	715	After initial surgery
	No		1747	Maximum	5630	5574
	Unknown		226	Minimum	61	0
Total			2573	Mean	332.8	257.6
Reoperation				Standard deviation	404.5	409.5
		1318		Interim outcome at 6 months following the initial surgery (1999~)		
	Liver transplant	576		Death	45	
	Hilar re-dissection	450		Living	2503	
	Hilar curettage	149		Without jaundice	1514	
	Endoscopic hilar curettage	13		With jaundice	603	
	Other	130		Jaundice unknown	64	
Reoperations other than liver transplant (1999~)				Post-transplant	322	
Age (days)	Maximum	865		Unknown	25	
	Minimum	28		Total	2573	
	Mean	140.9		Age at death (days) within 6 months (1999~)		
	Standard deviation	96.4		Maximum	330	
Jaundice	Disappeared	165		Minimum	48	
	Improved	84		Mean	171.9	
	Unchanged	71		Standard deviation	69.4	
	Worsened	55		Final outcome		
	Unknown	17		1989~1998		
Re-reoperation				1999~		
		292		Death	217	120
	Liver transplant	148		Living	3616	1160
	Hilar re-dissection	72		Without jaundice	706	1465
	Hilar curettage	15		With jaundice	241	239
	Endoscopic hilar curettage	8		Unknown living	97	26
	Other	49		Transplant living	106	723
Re-reoperations other than liver transplant (1999~)				Survival details unknown	10	3
Age (days)	Maximum	0		Unknown	29	9
	Minimum	75		Total	3862	1289
	Mean	243.0		Cause of death		
	Standard deviation	152.9		Liver failure	85	
Jaundice	Disappeared	28		Rupture of esophageal varices	8	
	Improved	5		Pneumonia	8	
	Unchanged	12		Peritonitis	4	
	Worsened	10		Sepsis after cholangitis	13	
	Unknown	5		Post-transplantation death	41	
				Other	88	
				Unknown	7	
				(Includes duplications)		

6

1989~2022

3862 cases

(1999~2022 2573 cases)

Screening (from 2012)					
By stool color card					
Yes: 412					
Before 1 month old	265	At 1 month old	260	After 1 month old	198
Observed age (days)		Observed age (days)		Observed age (days)	
Observed date not stated	73	Observed date not stated	76	Observed date not stated	36
Maximum	64	Maximum	87	Maximum	147
Minimum	0	Minimum	12	Minimum	30
Mean	14.2	Mean	33.3	Mean	57.9
Standard deviation	9.1	Standard deviation	7.9	Standard deviation	20.9
Number of the color					
1	7	1	15	1	25
2	45	2	63	2	90
3	58	3	66	3	46
4	85	4	84	4	20
5	5	5	2	5	1
6	0	6	1	6	0
7	5	7	0	7	2
Unknown	60	Unknown	27	Unknown	14
By urinary sulfated bile acid		By direct bilirubin		Other	
Yes	32	Yes	287	Yes	11
Abnormal	29	Abnormal	259		
Normal	2	Normal	4		
Unknown	2	Unknown	24		