

Japanese Biliary Atresia Registry (JBAR) Summary Sheet

1989~

2018

1989~2018

3483 cases

(1999~2018

2195 cases)

Number of registered cases by region			Birth order		Meconium color		
	No. registrations	No. hospitals	First child	1635	Normal		1520
Hokkaido	99	3	Second child	1219	Abnormal		508
Tohoku	300	13	Third child	419	Unknown		1455
Kanto	1203	40	Fourth child	75	Yellow stool		
Chubu and Hokuriku	576	22	Fifth child and later	34	Yes		1962
Kinki	636	17	Unknown	101	No		752
Chugoku and Shikoku	276	20	Number of babies (1999~)		Unknown		769
Kyushu	393	14	Singleton	2070	Neonatal jaundice		
Total	3483	129	Twins or more	58	Yes		2141
Age of parents			Unknown	67	No		884
	Mean	Standard deviation	Gender		Unknown		458
Father	32.3	6.0	Male	1290	Phototherapy		
Mother	29.9	5.0	Female	2190	Yes		1310
Maternal history during pregnancy			Unknown	3	No		1713
	Febriile rash disease	38	Birth weight (g)		Unknown		460
	Other	686	Maximum	4940	Pathological bleeding		
		0	Minimum	824	Yes		317
Family history (parents)			Mean	2903.6	(bleeding sites have duplications)	Intracranial	162
Biliary atresia	Parents	0	Standard deviation	452.1		Gastrointestinal	97
	Siblings	3	~1499	32		Subcutaneous	30
	Other	4	1500~1999	72		Other	53
Choledochal cyst	Parents	6	2000~2499	413		No	3024
	Siblings	3	2500~2999	1490	Unknown		142
	Other	3	3000~3499	1161	Age at bleeding (days)		
Hepatitis	Parents	51	3500~3999	229	Maximum		134
	Siblings	7	4000~	30	Minimum		0
	Other	36	不明	56	Mean		52.1
Other		287	Gestational age (weeks)		Standard deviation		26.4
			Maximum	42	Prothrombin activity (%)		
			Minimum	24	Maximum		181
			Mean	38.6	Minimum		2
			Standard deviation	1.8	Mean		83.0
			~29	11	Standard deviation		30.6
			30~31	17	≥70%		217
			32~33	34	≥40%<70%		48
			34~35	88	<40%		29
			36~37	498	PT-INR		
			38~39	1622	Maximum		13.4
			40~41	1078	Minimum		0.8
			42~	26	Mean		1.4
			Unknown	109	Standard deviation		1.4
Chromosomal abnormality					<1.15%		198
Yes					≥1.15% <1.4%		53
	Down症				≥1.4%		46
	その他						
No							2636
Unknown							834

1989~2018

3483 cases

(1999~2018

2195 cases)

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	6	Polycystic kidney	4
Mean	52.5	Patent foramen ovale	2	Renal hypoplasia	3
Standard deviation	33.7	Patent ductus arteriosus	19	Hydronephrosis	12
Age at hospitalization (days)					
Maximum	435	VSD	34	Horseshoe kidney	1
		ASD	26	Double renal pelvis and ureter	4
Minimum	0	Dextrocardia	7	Hydrocele of the testis	14
Mean	54.9	Pulmonary stenosis	22	Cryptorchidism	2
Standard deviation	34.4	Other	70	Extrophy of bladder	0
Prenatal diagnosis (1999~)				Hypospadias	2
Yes	107	Vascular		Urethral stricture	0
No	2038	Preduodenal portal vein	46	VUR	2
Unknown	50	Absence of the inferior vena cava	21	Other	23
Screening		Anomalous pulmonary venous return	2		
Yes	457	Other	17	Musculoskeletal	
No	2878			Spina bifida	4
Unknown	148	Spleen	0	Costal malformation	0
Stool color at admission		Polysplenia	70	Other	6
Yellow	175	Accessory spleen	16		
Pale yellow	1003	Asplenia	4	Limb and external anomaly	
Gray	2127	Other	0	Accessory ear	2
Unknown	178			Cleft lip	0
Total bilirubin immediately before surgery (mg/dl)		Gastrointestinal		Cleft lip and palate	10
Maximum	58.2	Malrotation of intestine	62	Polysyndactyly	6
Minimum	2.1	Esophageal atresia	3	Clubfoot	6
Mean	9.4	Intestinal atresia	13	Other	16
Standard deviation	3.8	Anorectal malformation	15		
Direct bilirubin immediately before surgery (mg/dl)		Hirschsprung' s disease	0	Central nervous system	
Maximum	40.5	Situs inversus	41	Hydrocephalus	3
Minimum	0.4	Annular pancreas	3	Microcephaly	0
Mean	6.4	Gastric rupture	0	Anencephaly	0
Standard deviation	2.8	Meconium peritonitis	13	Other	11
Implemented tests		Meckel' s diverticulum	16		
Serum Lp-X	1598	Cloacal exstrophy	0	Endocrine metabolic system	
Score test	1023	Alagille syndrome	9	Cretinism	1
Ultrasound	3279	Pancreaticobiliary maljunction	2	Other	2
ERCP	115	Intussusception	2		
Serum bile acid	2181	Other	47	Other malformations	
Percutaneous liver biopsy	89			Diaphragmatic hernia	1
Duodenal juice analysis	1930	Respiratory		Omphalocele	1
Hepatobiliary scintigraphy	2283	Tracheal/bronchial stenosis	2	Gastroschisis	1
MRI (MRCP)	255	Cystic lung disease	2	Inguinal hernia	78
CT	299	Lobar emphysema	0	Umbilical hernia	89
Other	146	Pulmonary hypoplasia	2	Other	14
Multiple answers allowed		Other	5		

1989~2018

3483 cases

(1999~2018 2195 cases)

Surgery implemented		Age at time of surgery (days)	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	
Yes		~30	234	231	166	71.9	81	35.1	
	Bile drainage surgery	31~45	542	541	338	62.5	208	38.4	
	Liver transplant	46~60	833	830	543	65.4	349	42.0	
	Exploratory laparotomy	61~70	540	540	343	63.5	216	40.0	
No (all ~ 1996 cases)		5	71~80	471	468	288	61.5	202	43.2
Unknown		0	81~90	295	294	164	55.8	123	41.8
Biliary reconstruction		0	91~120	361	356	200	56.2	152	42.7
	Hepaticoenterostomy	196	121~150	123	118	47	39.8	47	39.8
	Portoenterostomy	3212	151~	67	48	11	22.9	24	50.0
	Other	16	Unknown	12	12	8	66.7	3	25.0
	Unknown	14	Total	3478	3438	2108	61.3	1405	40.9

Age at time of surgery (days)		
	0	
	Maximum	419
	Minimum	4
	Mean	66.5
	Standard deviation	29.7

Surgical procedure		No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)
No cholangitis prevention		2506				
Simple Roux-en-Y anastomosis		2473	1529	61.8	1049	42.4
	<20 cm	15	8	53.3	5	33.3
	≥20 cm <40 cm	766	468	61.1	350	45.7
	≥40 cm <60 cm	1402	885	63.1	575	41.0
	≥60 cm	257	152	59.1	108	42.0
	Unknown	33	16	48.5	11	33.3
Porto/hepaticoduodenostomy		16	15	93.8	4	25.0
Other		8	2	25.0	2	25.0
Unknown		9	7	77.8	0	0.0
Cholangitis prevention		920	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		626	384	61.3	215	34.3
Hepatic portocholecystostomy or hepaticocholecystostomy		5	3	60.0	0	0.0
Other		45	25	55.6	19	42.2
Unknown		4	4	100.0	2	50.0
Cholangitis prevention unknown		12	0	7	5	41.7
Total		3438				

1989~2018

3483 cases

(1999~2018 2195 cases)

Cholangitis		1989~1998	1999~	Postoperative complications			
Yes		469	936		Ileus	152	
	Mean timing of first time (weeks)		11.1		Peritonitis	67	
	Mean timing of second time (weeks)		18.7		Hemorrhage	136	
No		745	1115		Pulmonary complications	41	
Unknown		66	107		Acute liver failure	47	
Total		1280	2158		Infection	210	
Reoperation within 6 months (1999~)				(Includes duplications)	Other	213	
	Yes	497		Age at liver transplant (days) (1999~)	615	After initial surgery	
	No	1491		Maximum	5630	5574	
	Unknown	207		Minimum	61	0	
Total		2195		Mean	334.0	257.6	
Reoperation		1174		Standard deviation	387.1	392.6	
Liver transplant		500		Interim outcome at 6 months following the initial surgery (1999~)			
Hilar re-dissection		417		Death	41		
Hilar curettage		126		Living	2136		
Endoscopic hilar curettage		13		Without jaundice	1278		
Other		118		With jaundice	535		
Reoperations other than liver transplant (1999~)		324		Jaundice unknown	58		
Age (days)	Maximum	865		Post-transplant	266		
	Minimum	28		Unknown	18		
	Mean	141.5		Total	2195		
	Standard deviation	101.9		Age at death (days) within 6 months (1999~)			
Jaundice	Disappeared	129		Maximum	330		
	Improved	70		Minimum	48		
	Unchanged	64		Mean	171.5		
	Worsened	47		Standard deviation	71.4		
	Unknown	14		Final outcome			
Re-reoperation		260		Death	207	1989~1998	1999~
Liver transplant		124		Living	3252	120	87
Hilar re-dissection		71		Without jaundice	1159	2093	
Hilar curettage		14		With jaundice	706	1236	
Endoscopic hilar curettage		8		Unknown living	241	215	
Other		43		Transplant living	97	24	
Re-reoperations other than liver transplant (1999~)		52		Survival details unknown	105	616	
Age (days)	Maximum	771		Unknown	24	10	2
	Minimum	75		Total	3483	9	15
	Mean	245.5		Cause of death			
	Standard deviation	161.9		Liver failure	82		
Jaundice	Disappeared	23		Rupture of esophageal varices	8		
	Improved	5		Pneumonia	8		
	Unchanged	11		Peritonitis	4		
	Worsened	10		Sepsis after cholangitis	13		
	Unknown	3		Post-transplantation death	38		
				Other	86		
			Unknown	6			
				(Includes duplications)			

Screening (from 2012)					
By stool color card					
Yes: 243					
Before 1 month old	152	At 1 month old	151	After 1 month old	131
Observed age (days)		Observed age (days)		Observed age (days)	
Observed date not stated	56	Observed date not stated	54	Observed date not stated	32
Maximum	37	Maximum	61	Maximum	147
Minimum	0	Minimum	14	Minimum	31
Mean	13.4	Mean	32.8	Mean	59.2
Standard deviation	8.1	Standard deviation	6.3	Standard deviation	22.1
Number of the color					
1	5	1	5	1	17
2	27	2	31	2	48
3	38	3	39	3	37
4	52	4	50	4	15
5	2	5	2	5	0
6	0	6	1	6	0
7	4	7	0	7	2
Unknown	24	Unknown	23	Unknown	12
By urinary sulfated bile acid		By direct bilirubin		Other	
Yes	27	Yes	199	Yes	9
Abnormal	24	Abnormal	193		
Normal	2	Normal	3		
Unknown	1	Unknown	3		