

Japanese Biliary Atresia Registry (JBAR) Summary Sheet

2022~

2022 年

2022~2022 70 cases

Number of registered cases by region			Birth order		Meconium color		
	No. registrations	No. hospitals	First child	39	Normal	37	
Hokkaido	1	1	Second child	21	Abnormal	5	
Tohoku	4	1	Third child	9	Unknown	28	
Kanto	26	16	Fourth child	0	Yellow stool		
Chubu and Hokuriku	15	6	Fifth child and later	0	Yes	43	
Kinki	13	6	Unknown	1	No	11	
Chugoku and Shikoku	4	2	Number of babies (1999~)		Unknown	16	
Kyushu	7	5	Singleton	65	Neonatal jaundice		
Total	70	37	Twins or more	0	Yes	39	
Age of parents			Unknown	5	No	27	
	Mean	Standard deviation	Gender		Unknown	4	
Father	32.5	5.0	Male	28	Phototherapy		
Mother	31.2	4.8	Female	42	Yes	27	
Maternal history during pregnancy			Unknown	0	No	37	
	Febrile rash disease	3	Birth weight (g)		Unknown	6	
	Other	4	Maximum	3900	Pathological bleeding		
		0	Minimum	834	Yes	3	
Family history (parents)			Mean	2900.5	(bleeding sites have duplications)	Intracranial	0
Biliary atresia	Parents	0	Standard deviation	482.6		Gastrointestinal	1
	Siblings	0	~1499	1		Subcutaneous	0
	Other	0	1500~1999	2		Other	2
Choledochal cyst	Parents	0	2000~2499	3		No	67
	Siblings	0	2500~2999	34	Age at bleeding (days)		
	Other	0	3000~3499	24	Maximum	7	
Hepatitis	Parents	0	3500~3999	6	Minimum	5	
	Siblings	0	4000~	0	Mean	6.3	
	Other	0	不明	0	Standard deviation	1.2	
Other		1	Gestational age (weeks)		Prothrombin activity (%)		
			Maximum	42	Maximum	240	
			Minimum	28	Minimum	7	
			Mean	38.5	Mean	95.2	
			Standard deviation	1.9	Standard deviation	33.7	
			~29	1	≥70%	54	
			30~31	0	≥40%<70%	8	
			32~33	1	<40%	4	
			34~35	1	PT-INR		
			36~37	7	Maximum	6.5	
			38~39	41	Minimum	0.8	
			40~41	18	Mean	1.3	
			42~	1	Standard deviation	0.7	
			Unknown	0	<1.15%	38	
Chromosomal abnormality					≥1.15% <1.4%	14	
	Yes	0			≥1.4%	15	
	Down症	0					
	その他	0					
	No	65					
	Unknown	5					

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	0	Polycystic kidney	0
Mean	42.1	Patent foramen ovale	1	Renal hypoplasia	1
Standard deviation	54.7	Patent ductus arteriosus	0	Hydronephrosis	0
Age at hospitalization (days)		VSD	5	Horseshoe kidney	0
Maximum	387	ASD	0	Double renal pelvis and ureter	0
Minimum	0	Dextrocardia	0	Hydrocele of the testis	0
Mean	44.3	Pulmonary stenosis	1	Cryptorchidism	0
Standard deviation	54.4	Other	2	Extrophy of bladder	0
Prenatal diagnosis (1999~)				Hypospadias	0
Yes	5	Vascular		Urethral stricture	0
No	62	Preduodenal portal vein	1	VUR	1
Unknown	3	Absence of the inferior vena cava	0	Other	0
Screening		Anomalous pulmonary venous return	0		
Yes	33	Other	0	Musculoskeletal	
No	37			Spina bifida	0
Unknown	0	Spleen	0	Costal malformation	0
Stool color at admission		Polysplenia	0	Other	0
Yellow	7	Accessory spleen	0		
Pale yellow	34	Asplenia	0	Limb and external anomaly	
Gray	10	Other	0	Accessory ear	0
Unknown	19			Cleft lip	0
Total bilirubin immediately before		Gastrointestinal		Cleft lip and palate	0
Maximum	32.4	Malrotation of intestine	2	Polysyndactyly	0
Minimum	2.9	Esophageal atresia	0	Clubfoot	0
Mean	8.7	Intestinal atresia	0	Other	0
Standard deviation	4.8	Anorectal malformation	1		
Direct bilirubin immediately before		Hirschsprung's disease	0	Central nervous system	
Maximum	23.6	Situs inversus	0	Hydrocephalus	0
Minimum	1.7	Annular pancreas	0	Microcephaly	0
Mean	5.6	Gastric rupture	0	Anencephaly	0
Standard deviation	3.8	Meconium peritonitis	0	Other	0
Implemented tests		Meckel's diverticulum	0		
Serum Lp-X	0	Cloacal exstrophy	0	Endocrine metabolic system	
Score test	0	Alagille syndrome	0	Cretinism	0
Ultrasound	69	Pancreaticobiliary maljunction	0	Other	1
ERCP	0	Intussusception	0		
Serum bile acid	42	Other	1	Other malformations	
Percutaneous liver biopsy	0			Diaphragmatic hernia	0
Duodenal juice analysis	20	Respiratory		Omphalocele	0
Hepatobiliary scintigraphy	41	Tracheal/bronchial stenosis	0	Gastroschisis	0
MRI (MRCP)	6	Cystic lung disease	0	Inguinal hernia	0
CT	2	Lobar emphysema	0	Umbilical hernia	1
Other	1	Pulmonary hypoplasia	0	Other	1
Multiple answers allowed		Other	0		

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Surgery implemented		Age at time of surgery (days)	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)
Yes		~30	16	16	11	68.8	8	50.0
	Bile drainage surgery	31~45	19	19	13	68.4	9	47.4
	Liver transplant	46~60	11	11	9	81.8	5	45.5
	Exploratory laparotomy	61~70	8	8	7	87.5	8	100.0
		71~80	3	3	3	100.0	2	66.7
No (all ~ 1998 cases)		81~90	2	2	1	50.0	0	0.0
Unknown								
Biliary reconstruction			8	7	4	57.1	4	57.1
	Hepaticoenterostomy	121~150	3	3	0	0.0	3	100.0
	Portoenterostomy	151~	0	0	0		0	
	Other	Unknown	0	0	0		0	
	Unknown		0	0	0		0	
Age at time of surgery (days)			70	69	48	69.6	39	56.5
	Maximum	148						
	Minimum	16						
	Mean	54.9						
	Standard deviation	31.2						

Surgical procedure		No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)
No cholangitis prevention		53				
Cholangitis prevention	Simple Roux-en-Y anastomosis	49	32	65.3	24	49.0
	<20 cm	1	1	100.0	0	0.0
	≥20 cm <40 cm	25	15	60.0	12	48.0
	≥40 cm <60 cm	23	16	69.6	12	52.2
	≥60 cm	0	0		0	
	Unknown	0	0		0	
	Porto/hepaticoduodenostomy	0	0		0	
	Other	1	1	100.0	1	100.0
	Unknown	3	1	33.3	2	66.7
	Cholangitis prevention	16	0	0.0	0	0.0
Suruga II method	0	0		0		
Double Roux-en-Y reconstruction	0	0		0		
Outer end jejunostomy	0	0		0		
Roux-en-Y reconstruction with intestinal valve	12	10	83.3	8	66.7	
Hepatic portocholecystostomy or hepaticocholecystostomy	0	0		0		
Other	4	4	100.0	4	100.0	
Unknown	0	0		0		
Cholangitis prevention unknown	0	0		0		
Total	69	0	0		0	

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Basio Type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	Esophageal varices checked before or during surgery	
I	3	3	3	100.0	1	33.3	Not checked	67
I -cyst	6	6	5	83.3	3	50.0	Varices present	0
II	0	0	0		0		No varices	2
III	61	60	40	66.7	35	58.3	Unknown	1
Unknown	0	0	0		0		Total	70
Total	70	69	48	69.6	39	56.5	Choleretics	
Sub type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)		
a1	7	7	5	71.4	6	85.7	Dehycol	0
a2	1	1	0	0.0	0	0.0	UDCA	68
b1	49	48	34	70.8	26	54.2	Glucagon	5
b2	2	2	1	50.0	1	50.0	Secretin	0
c1	8	8	5	62.5	4	50.0	PGE2	0
c2	0	0	0		0		PGF2 α	0
d	2	2	2	100.0	1	50.0	Steroids	61
Unknown	1	1	1	100.0	1	100.0	Taurine	6
Total	70	69	48	69.6	39	56.5	Phenobarbital	3
							Other	0
Type of porta hepatic	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	(includes duplications)	
α	2	2	2	100.0	1	50.0	Bile drainage after first surgery	
β	6	6	5	83.3	3	50.0	1989~1998	1999~
γ	7	7	5	71.4	4	57.1	Yes	0
μ	3	3	2	66.7	3	100.0	No	0
ν	49	48	33	68.8	28	58.3	Unknown	0
\omicron	2	2	0	0.0	0	0.0	Total	69
Unknown	1	1	1	100.0	0	0.0	Jaundice after first surgery	
Total	70	69	48	69.6	39	56.5	Disappeared	0
							Improved	0
							Unchanged	0
							Worsened	0
							Unknown	0
							Total	69
							Jaundice re-elevation	
							1989~1998	1999~
							No re-elevation	0
							Re-elevated after disappearance	0
							Re-elevated after improvement	0
							Unknown	0
							Total	59
							Measures for re-elevation	
							Monitor progress	5
							Conservative treatment	10
							Re-do surgery	9
							Liver transplant	12
							Total	36
Gallbladder								
Yes	68							
No bile	32							
Yellow	4							
Pale yellow	2							
Colorless	19							
Other	1							
Unknown	9							
Absent	2							
Unknown	0							
Total	70							
Cystic structure in extrahepatic bile duct								
Yes	13							
Yellow	2							
Pale yellow	3							
Colorless	5							
Other	1							
Unknown	2							
No	56							
Unknown	1							
Total	70							

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Cholangitis		1989~1998	1999~	Postoperative complications		
Yes		0	39		Ileus	5
	Mean timing of first time (weeks)	9.5			Peritonitis	2
	Mean timing of second time (weeks)	14.1			Hemorrhage	1
No		0	29		Pulmonary complications	1
Unknown		0	1		Acute liver failure	0
Total		0	69		Infection	2
Reoperation within 6 months (1999~)				(Includes duplications)	Other	6
	Yes	16		Age at liver transplant (days) (1999~)	13	After initial surgery
	No	48		Maximum	357	282
	Unknown	6		Minimum	169	0
Total		70		Mean	242.4	188.7
Reoperation			21	Standard deviation	54.2	58.0
	Liver transplant	10		Interim outcome at 6 months following the initial surgery (1999~)		
	Hilar re-dissection	4		Death	0	
	Hilar curettage	5		Living	66	
	Endoscopic hilar curettage	0		Without jaundice	45	
	Other	2		With jaundice	10	
Reoperations other than liver transplant (1999~)			11	Jaundice unknown	2	
Age (days)	Maximum	353		Post-transplant	9	
	Minimum	69		Unknown	4	
	Mean	166.8		Total	70	
	Standard deviation	82.6		Age at death (days) within 6 months (1999~)		
Jaundice	Disappeared	3		Maximum	-	
	Improved	3		Minimum	-	
	Unchanged	3		Mean	-	
	Worsened	1		Standard deviation	-	
	Unknown	1		Final outcome		
Re-reoperation			3	Death	1	1989~1998
	Liver transplant	3		Living	68	0
	Hilar re-dissection	0		Without jaundice	0	68
	Hilar curettage	0		With jaundice	0	44
	Endoscopic hilar curettage	0		Unknown living	0	8
	Other	0		Transplant living	0	1
Re-reoperations other than liver transplant (1999~)			0	Survival details unknown	0	15
Age (days)	Maximum	-		Unknown	1	0
	Minimum	-		Total	70	0
	Mean	-		Cause of death		
	Standard deviation	-		Liver failure	0	
Jaundice	Disappeared	0		Rupture of esophageal varices	0	
	Improved	0		Pneumonia	0	
	Unchanged	0		Peritonitis	0	
	Worsened	0		Sepsis after cholangitis	0	
	Unknown	0		Post-transplantation death	1	
				Other	0	
			Unknown	0		
				(Includes duplications)		

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Screening (from 2012)					
By stool color card					
Yes: 31					
Before 1 month old	22	At 1 month old	21	After 1 month old	9
Observed age (days)		Observed age (days)		Observed age (days)	
Observed date not stated	0	Observed date not stated	2	Observed date not stated	0
Maximum	29	Maximum	43	Maximum	96
Minimum	2	Minimum	24	Minimum	36
Mean	13.8	Mean	31.6	Mean	56.2
Standard deviation	7.8	Standard deviation	4.7	Standard deviation	22.2
Number of the color					
1	1	1	3	1	0
2	3	2	7	2	6
3	2	3	3	3	2
4	2	4	7	4	1
5	0	5	0	5	0
6	0	6	0	6	0
7	0	7	0	7	0
Unknown	14	Unknown	0	Unknown	0
By urinary sulfated bile acid		By direct bilirubin		Other	
Yes		Yes		Yes	
Abnormal	1	Abnormal	15		1
Normal	0	Normal	4		
Unknown	0	Unknown	0		
			11		