

2020~2020 100 cases

Number of registered cases by region			Birth order		Meconium color		
	No. registrations	No. hospitals	First child	49	Normal	50	
Hokkaido	3	2	Second child	32	Abnormal	9	
Tohoku	3	1	Third child	14	Unknown	41	
Kanto	51	23	Fourth child	1	Yellow stool		
Chubu and Hokuriku	15	4	Fifth child and later	3	Yes	74	
Kinki	9	6	Unknown	1	No	6	
Chugoku and Shikoku	10	8	Number of babies (1999~)		Unknown	20	
Kyushu	9	4	Singleton	96	Neonatal jaundice		
Total	100	48	Twins or more	3	Yes	46	
Age of parents			Unknown	1	No	45	
	Mean	Standard deviation	Gender		Unknown		
Father	34.7	6.4	Male	32	Phototherapy		
Mother	32.1	4.7	Female	68	Yes	41	
Maternal history during pregnancy			Unknown	0	No	53	
	Febrile rash disease	3	Birth weight (g)		Unknown		
	Other	12	Maximum	3844	Pathological bleeding		
		0	Minimum	1855	Yes	4	
Family history (parents)			Mean	2850.6	(bleeding sites have duplications)	Intracranial	2
Biliary atresia	Parents	0	Standard deviation	402.7		Gastrointestinal	0
	Siblings	0	~1499	0		Subcutaneous	1
	Other	0	1500~1999	3		Other	1
Choledochal cyst	Parents	1	2000~2499	13		No	96
	Siblings	0	2500~2999	50	Unknown	0	
	Other	0	3000~3499	29	Age at bleeding (days)		
Hepatitis	Parents	1	3500~3999	5	Maximum	74	
	Siblings	0	4000~	0	Minimum	69	
	Other	0	不明	0	Mean	71.3	
Other		8	Gestational age (weeks)		Standard deviation		
			Maximum	41	Prothrombin activity (%)		
			Minimum	28	Maximum	172	
			Mean	38.5	Minimum	4	
			Standard deviation	1.7	Mean	88.9	
			~29	1	Standard deviation	30.5	
			30~31	0	≥70%	76	
			32~33	0	≥40%<70%	13	
			34~35	2	<40%	6	
			36~37	18	PT-INR		
			38~39	50	Maximum	21.0	
			40~41	29	Minimum	0.8	
			42~	0	Mean	1.6	
			Unknown	0	Standard deviation	2.6	
Chromosomal abnormality					<1.15%		75
	Yes				≥1.15% <1.4%		12
			Down症	0	≥1.4%		11
			その他	0			
	No						95
	Unknown						5

Age at initial consult (days)		Name of associated congenital anomaly		Name of associated congenital anomaly (Cont.)	
Maximum	135	Heart		Urogenital	
Minimum	0	Tetralogy of Fallot	1	Polycystic kidney	0
Mean	36.0	Patent foramen ovale	0	Renal hypoplasia	0
Standard deviation	31.2	Patent ductus arteriosus	0	Hydronephrosis	0
Age at hospitalization (days)		VSD	1	Horseshoe kidney	0
Maximum	140	ASD	0	Double renal pelvis and ureter	0
Minimum	0	Dextrocardia	0	Hydrocele of the testis	0
Mean	38.4	Pulmonary stenosis	0	Cryptorchidism	0
Standard deviation	30.5	Other	3	Extrophy of bladder	0
Prenatal diagnosis (1999~)				Hypospadias	0
Yes	9	Vascular		Urethral stricture	0
No	89	Preduodenal portal vein	3	VUR	1
Unknown	2	Absence of the inferior vena cava	0	Other	2
Screening		Anomalous pulmonary venous return	0		
Yes	49	Other	2	Musculoskeletal	
No	51			Spina bifida	0
Unknown	0	Spleen	0	Costal malformation	0
Stool color at admission		Polysplenia	3	Other	0
Yellow	14	Accessory spleen	0		
Pale yellow	42	Asplenia	0	Limb and external anomaly	
Gray	42	Other	0	Accessory ear	0
Unknown	2			Cleft lip	0
Total bilirubin immediately before surgery (mg/dl)		Gastrointestinal		Cleft lip and palate	0
Maximum	18.4	Malrotation of intestine	3	Polysyndactyly	0
Minimum	3.1	Esophageal atresia	1	Clubfoot	0
Mean	7.9	Intestinal atresia	0	Other	0
Standard deviation	2.8	Anorectal malformation	1		
Direct bilirubin immediately before		Hirschsprung's disease	0	Central nervous system	
Maximum	13.6	Situs inversus	2	Hydrocephalus	0
Minimum	1.8	Annular pancreas	0	Microcephaly	0
Mean	5.3	Gastric rupture	0	Anencephaly	0
Standard deviation	2.2	Meconium peritonitis	0	Other	1
Implemented tests		Meckel's diverticulum	0		
Serum Lp-X	2	Cloacal exstrophy	0	Endocrine metabolic system	
Score test	2	Alagille syndrome	0	Cretinism	0
Ultrasound	89	Pancreaticobiliary maljunction	0	Other	0
ERCP	0	Intussusception	0		
Serum bile acid	64	Other	3	Other malformations	
Percutaneous liver biopsy	2			Diaphragmatic hernia	0
Duodenal juice analysis	28	Respiratory		Omphalocele	0
Hepatobiliary scintigraphy	78	Tracheal/bronchial stenosis	0	Gastroschisis	0
MRI (MRCP)	7	Cystic lung disease	0	Inguinal hernia	0
CT	14	Lobar emphysema	0	Umbilical hernia	0
Other	3	Pulmonary hypoplasia	0	Other	0
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	15	93.8	5	31.3
	Bile drainage surgery	31~45	31	31	24	77.4	13	41.9
	Liver transplant	46~60	24	24	17	70.8	11	45.8
	Exploratory laparotomy	61~70	8	8	4	50.0	5	62.5
No (all 1998 cases)		71~80	11	11	8	72.7	8	72.7
Unknown		81~90	2	2	0	0.0	1	50.0
Biliary reconstruction		91~120	6	6	1	16.7	3	50.0
	Hepaticenterostomy	121~150	2	2	0	0.0	0	0.0
	Portoenterostomy	151~	0	0	0		0	
	Other	Unknown	0	0	0		0	
	Unknown		0	0	0		0	
		Total	100	100	69	69.0	46	46.0

Age at time of surgery (days)	
Maximum	146
Minimum	10
Mean	52.3
Standard deviation	25.7

Surgical procedure		No. Bile drainage surgery	Jaundice disappeared (%)	Cholangitis (%)
No cholangitis prevention		90		
Simple Roux-en-Y anastomosis		90	62	68.9
	<20 cm	1	0	0.0
	≥20 cm <40 cm	49	34	69.4
	≥40 cm <60 cm	38	27	71.1
	≥60 cm	1	0	0.0
	Unknown	1	1	100.0
	Porto/hepaticoduodenostomy	0	0	0
	Other	0	0	0
	Unknown	0	0	0
Cholangitis prevention		10	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	10	7	70.0
	Hepatic portocholecystostomy or hepaticocholecystostomy	0	0	0
	Other	0	0	0
	Unknown	0	0	0
Cholangitis prevention unknown		0	0	0
Total		100		

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Basic Type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	Esophageal varices checked before or during surgery		
I	3	3	3	100.0	0	0.0	Not checked		94
I -cyst	9	9	8	88.9	4	44.4	Varices present		0
II	1	1	0	0.0	0	0.0	No varices		6
III	87	87	58	66.7	42	48.3	Unknown		0
Unknown	0	0	0		0		Total		100
Total	100	100	69	69.0	46	46.0	Choleretics		
Sub type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)			
							Dehycol		0
a1	11	11	8	72.7	4	36.4	UDCA		99
a2	3	3	2	66.7	1	33.3	Glucagon		8
b1	64	64	41	64.1	31	48.4	Secretin		1
b2	1	1	0	0.0	0	0.0	PGE2		0
c1	9	9	9	100.0	4	44.4	PGF2 α		0
c2	5	5	3	60.0	2	40.0	Steroids		96
d	4	4	4	100.0	2	50.0	Taurine		10
Unknown	3	3	2	66.7	2	66.7	Phenobarbital		5
Total	100	100	69	69.0	46	46.0	Other		2
Type of porta hepatitis	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	(includes duplications)		
α	3	3	3	100.0	1	33.3	Bile drainage after first surgery 1989~1998 1999~		
β	7	7	6	85.7	2	28.6	Yes		0 91
γ	2	2	1	50.0	1	50.0	No		0 9
μ	10	10	6	60.0	7	70.0	Unknown		0 0
ν	68	68	47	69.1	33	48.5	Total 0 100		
ω	5	5	2	40.0	0	0.0	Jaundice after first surgery 1989~1998 1999~		
Unknown	5	5	4	80.0	2	40.0	Disappeared		0 69
Total	100	100	69	69.0	46	46.0	Improved		0 20
Gallbladder							Unchanged 0 6		
Yes 94							Worsened 0 4		
No bile 42							Unknown 0 1		
Yellow 3							Total 0 100		
Pale yellow 2							Jaundice re-elevation 1989~1998 1999~		
Colorless 41							No re-elevation 0 50		
Other 3							Re-elevated after disappearance 0 16		
Unknown 3							Re-elevated after improvement 0 19		
Absent 6							Unknown 0 4		
Unknown 0							Total 0 89		
Total 100							Measures for re-elevation		
Cystic structure in extrahepatic bile duct							Monitor progress 4		
Yes 14							Conservative treatment 8		
Yellow 0							Re-do surgery 9		
Pale yellow 2							Liver transplant 18		
Colorless 7							Total 39		
Other 5									
Unknown 0									
No 86									
Unknown 0									
Total 100									

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Cholangitis		1989~1998	1999~	Postoperative complications			
Yes		0	46		Ileus		2
	Mean timing of first time (weeks)		11.2		Peritonitis		0
	Mean timing of second time (weeks)		17.1		Hemorrhage		0
No		0	54		Pulmonary complications		0
Unknown		0	0		Acute liver failure		1
Total		0	100		Infection		3
Reoperation within 6 months (1999~)				(Includes duplications)	Other		8
	Yes		25	Age at liver transplant (days) (1999~)	28	After initial surgery	
	No		73	Maximum	388		337
	Unknown		2	Minimum	141		0
Total			100	Mean	244.7		175.6
Reoperation		37		Standard deviation	63.5		77.3
Liver transplant			23	Interim outcome at 6 months following the initial surgery (1999~)			
Hilar re-dissection			7	Death		1	
Hilar curettage			5	Living		98	
Endoscopic hilar curettage			0		Without jaundice		67
Other			2		With jaundice		15
Reoperations other than liver transplant (1999~)			14		Jaundice unknown		0
Age (days)	Maximum		172		Post-transplant		16
	Minimum		62		Unknown		1
	Mean		113.1		Total		100
	Standard deviation		32.8	Age at death (days) within 6 months (1999~)			
Jaundice	Disappeared		9		Maximum		164
	Improved		2		Minimum		164
	Unchanged		0		Mean		164.0
	Worsened		3		Standard deviation		#DIV/0!
	Unknown		0	Final outcome			
Re-reoperation		8		Death		2	
Liver transplant			5	Living		97	
Hilar re-dissection			0		Without jaundice		61
Hilar curettage			1		With jaundice		6
Endoscopic hilar curettage			0		Unknown living		0
Other			2		Transplant living		30
Re-reoperations other than liver transplant (1999~)			3		Survival details unknown		0
Age (days)	Maximum		350		Unknown		1
	Minimum		119		Total		100
	Mean		248.0	Cause of death			
	Standard deviation		117.8		Liver failure		1
Jaundice	Disappeared		1		Rupture of esophageal varices		0
	Improved		0		Pneumonia		0
	Unchanged		1		Peritonitis		0
	Worsened		0		Sepsis after cholangitis		0
	Unknown		1		Post-transplantation death		1
					Other		0
				Unknown		0	
				(Includes duplications)			

Screening (from 2012)					
By stool color card					
Yes: 44					
Before 1 month old	31	At 1 month old	33	After 1 month old	14
Observed age (days)		Observed age (days)		Observed age (days)	
Observed date not stated	1	Observed date not stated	5	Observed date not stated	2
Maximum	26	Maximum	65	Maximum	65
Minimum	0	Minimum	12	Minimum	30
Mean	14.5	Mean	32.1	Mean	51.7
Standard deviation	7.3	Standard deviation	8	Standard deviation	12
Number of the color					
1	0	1	1	1	1
2	7	2	12	2	9
3	9	3	13	3	2
4	13	4	6	4	1
5	1	5	0	5	0
6	0	6	0	6	0
7	0	7	0	7	0
Unknown	1	Unknown	1	Unknown	1
By urinary sulfated bile acid		By direct bilirubin		Other	
Yes	1	Yes	28	Yes	1
Abnormal	1	Abnormal	27		
Normal	0	Normal	0		
Unknown	0	Unknown	1		