

2018~2018 91 cases

Number of registered cases by region			Birth order		Meconium color		
	No. registrations	No. hospitals	First child	50	Normal		34
Hokkaido	5	2	Second child	32	Abnormal		16
Tohoku	7	2	Third child	7	Unknown		41
Kanto	21	12	Fourth child	0	Yellow stool		
Chubu and Hokuriku	18	5	Fifth child and later	1	Yes		54
Kinki	14	8	Unknown	1	No		11
Chugoku and Shikoku	8	7	Number of babies (1999~)		Unknown		26
Kyushu	18	6	Singleton	86	Neonatal jaundice		
Total	91	42	Twins or more	4	Yes		43
Age of parents			Unknown	1	No		24
	Mean	Standard deviation	Gender		Unknown		
Father	32.9	6.7	Male	31	Phototherapy		
Mother	31.3	5.4	Female	60	Yes		33
Maternal history during pregnancy			Unknown	0	No		33
	Febrile rash disease	0	Birth weight (g)		Unknown		
	Other	7	Maximum	3720	Pathological bleeding		
		0	Minimum	824	Yes		2
Family history (parents)			Mean	2785.9	(bleeding sites have duplications)	Intracranial	2
Biliary atresia	Parents	0	Standard deviation	420.7		Gastrointestinal	0
	Siblings	0	~1499	1		Subcutaneous	0
	Other	0	1500~1999	2		Other	0
Choledochal cyst	Parents	0	2000~2499	12		No	86
	Siblings	0	2500~2999	51		Unknown	3
	Other	0	3000~3499	20	Age at bleeding (days)		
Hepatitis	Parents	0	3500~3999	2	Maximum		77
	Siblings	0	4000~	0	Minimum		76
	Other	0	不明	3	Mean		76.5
Other		19	Gestational age (weeks)		Standard deviation		
			Maximum	41	Prothrombin activity (%)		
			Minimum	24	Maximum		131
			Mean	38.4	Minimum		2
			Standard deviation	2.3	Mean		78.4
			~29	2	Standard deviation		29.2
			30~31	0	≥70%		60
			32~33	0	≥40%<70%		18
			34~35	2	<40%		10
			36~37	13	PT-INR		
			38~39	51	Maximum		12.2
			40~41	20	Minimum		0.9
			42~	0	Mean		1.4
			Unknown	3	Standard deviation		1.4
Chromosomal abnormality					<1.15%		
Yes					≥1.15% <1.4%		
	Down症				≥1.4%		
	その他						
No					83		
Unknown					5		

Age at initial consult (days)		Name of associated congenital anomaly		Name of associated congenital anomaly (Cont.)	
Maximum	166	Heart		Urogenital	
Minimum	0	Tetralogy of Fallot	0	Polycystic kidney	0
Mean	46.7	Patent foramen ovale	0	Renal hypoplasia	0
Standard deviation	34.0	Patent ductus arteriosus	0	Hydronephrosis	0
<b>Age at hospitalization (days)</b>		VSD	0	Horseshoe kidney	0
Maximum	171	ASD	0	Double renal pelvis and ureter	0
Minimum	0	Dextrocardia	0	Hydrocele of the testis	0
Mean	49.0	Pulmonary stenosis	0	Cryptorchidism	0
Standard deviation	34.1	Other	3	Extrophy of bladder	0
<b>Prenatal diagnosis (1999~)</b>				Hypospadias	0
Yes	4	Vascular		Urethral stricture	0
No	85	Preduodenal portal vein	2	VUR	0
Unknown	2	Absence of the inferior vena cava	0	Other	4
<b>Screening</b>		Anomalous pulmonary venous return	0		
Yes	47	Other	3	Musculoskeletal	
No	43			Spina bifida	0
Unknown	1	Spleen	0	Costal malformation	0
<b>Stool color at admission</b>		Polysplenia	4	Other	0
Yellow	11	Accessory spleen	0		
Pale yellow	41	Asplenia	0	Limb and external anomaly	
Gray	38	Other	0	Accessory ear	0
Unknown	1			Cleft lip	0
<b>Total bilirubin immediately before surgery (mg/dl)</b>		Gastrointestinal		Cleft lip and palate	0
Maximum	18.7	Malrotation of intestine	0	Polysyndactyly	0
Minimum	3.4	Esophageal atresia	0	Clubfoot	0
Mean	8.7	Intestinal atresia	0	Other	0
Standard deviation	3.5	Anorectal malformation	0		
<b>Direct bilirubin immediately before</b>		Hirschsprung's disease	0	Central nervous system	
Maximum	14.2	Situs inversus	2	Hydrocephalus	0
Minimum	0.4	Annular pancreas	0	Microcephaly	0
Mean	5.6	Gastric rupture	0	Anencephaly	0
Standard deviation	2.5	Meconium peritonitis	0	Other	0
<b>Implemented tests</b>		Meckel's diverticulum	0		
Serum Lp-X	1	Cloacal exstrophy	0	Endocrine metabolic system	
Score test	2	Alagille syndrome	1	Cretinism	0
Ultrasound	88	Pancreaticobiliary maljunction	0	Other	0
ERCP	0	Intussusception	0		
Serum bile acid	58	Other	6	Other malformations	
Percutaneous liver biopsy	0			Diaphragmatic hernia	0
Duodenal juice analysis	26	Respiratory		Omphalocele	0
Hepatobiliary scintigraphy	47	Tracheal/bronchial stenosis	0	Gastroschisis	0
MRI (MRCP)	9	Cystic lung disease	0	Inguinal hernia	0
CT	8	Lobar emphysema	0	Umbilical hernia	0
Other	3	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	7	7	6	85.7	2	28.6
	Bile drainage surgery	31~45	18	18	12	66.7	9	50.0
	Liver transplant	46~60	19	19	13	68.4	7	36.8
	Exploratory laparotomy	61~70	14	14	10	71.4	9	64.3
No (all 1998 cases)		71~80	7	7	3	42.9	5	71.4
Unknown		81~90	9	9	3	33.3	4	44.4
<b>Biliary reconstruction</b>		91~120	12	12	5	41.7	6	50.0
	Hepaticenterostomy	121~150	2	2	2	100.0	2	100.0
	Portoenterostomy	151~	1	0	0		0	
	Other	Unknown	2	2	1	50.0	0	0.0
	Unknown		0					
Total			91	90	55	61.1	44	48.9

Age at time of surgery (days)	
Maximum	128
Minimum	21
Mean	63.2
Standard deviation	26.2

Surgical procedure		No. Bile drainage surgery	Jaundice disappeared (%)	Cholangitis (%)
No cholangitis prevention		84		
Simple Roux-en-Y anastomosis		84	50	59.5
	<20 cm	0	0	0
	≥20 cm <40 cm	42	27	64.3
	≥40 cm <60 cm	37	20	54.1
	≥60 cm	4	3	75.0
	Unknown	1	0	0.0
Porto/hepaticoduodenostomy		0	0	0
Other		0	0	0
Unknown		0	0	0
Cholangitis prevention		6	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		6	5	83.3
Hepatic portocholecystostomy or hepaticocholecystostomy		0	0	0
Other		0	0	0
Unknown		0	0	0
Cholangitis prevention unknown		0	0	0
Total		90		

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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	2	66.7	1	33.3	Not checked	80
I -cyst	11	11	10	90.9	4	36.4	Varices present	2
II	3	3	3	100.0	0	0.0	No varices	8
III	72	71	39	54.9	37	52.1	Unknown	1
Unknown	2	2	1	50.0	2	100.0	Total	91
Total	91	90	55	61.1	44	48.9	<b>Choleretics</b>	
Sub type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	Dehycol	
a1	18	18	12	66.7	8	44.4	UDCA	86
a2	7	6	3	50.0	1	16.7	Glucagon	8
b1	44	44	28	63.6	25	56.8	Secretin	0
b2	0	0	0		0		PGE2	0
c1	6	6	4	66.7	1	16.7	PGF2 $\alpha$	0
c2	7	7	1	14.3	2	28.6	Steroids	87
d	3	3	3	100.0	1	33.3	Taurine	0
Unknown	6	6	4	66.7	6	100.0	Phenobarbital	2
Total	91	90	55	61.1	44	48.9	Other	1
Type of porta hepatitis	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	(includes duplications)	
$\alpha$	4	4	4	100.0	0	0.0	<b>Bile drainage after first surgery</b>	
$\beta$	3	3	3	100.0	0	0.0	Yes	1989~1998 0
$\gamma$	3	2	2	100.0	0	0.0	No	0
$\mu$	8	8	4	50.0	3	37.5	Unknown	0
$\nu$	61	61	35	57.4	34	55.7	Total	0
o	4	4	2	50.0	1	25.0	<b>Jaundice after first surgery</b>	
Unknown	8	8	5	62.5	6	75.0	Disappeared	1989~1998 0
Total	91	90	55	61.1	44	48.9	Improved	0
<b>Gallbladder</b>							Unchanged	0
Yes							Worsened	0
No bile							Unknown	0
Yellow							Total	0
Pale yellow							<b>Jaundice re-elevation</b>	1989~1998 0
Colorless							No re-elevation	0
Other							Re-elevated after disappearance	0
Unknown							Re-elevated after improvement	0
Absent							Unknown	0
Unknown							Total	0
Total	91						Measures for re-elevation	1999~ 80
<b>Cystic structure in extrahepatic bile duct</b>							Monitor progress	2
Yes							Conservative treatment	19
Yellow							Re-do surgery	3
Pale yellow							Liver transplant	22
Colorless							Total	46
Other								
Unknown								
No								
Unknown								
Total	91							

Cholangitis		1989~1998	1999~	Postoperative complications				
Yes		0	44		Ileus		2	
	Mean timing of first time (weeks)		12.7		Peritonitis		2	
	Mean timing of second time (weeks)		17.4		Hemorrhage		5	
No		0	44		Pulmonary complications		0	
Unknown		0	2		Acute liver failure		0	
Total		0	90		Infection		3	
Reoperation within 6 months (1999~)				(Includes duplications)	Other		1	
	Yes		20	Age at liver transplant (days) (1999~)	25	After initial surgery		
	No		71	Maximum	409	360		
	Unknown		0	Minimum	139	0		
Total			91	Mean	244.1	163.6		
Reoperation		28		Standard deviation	71.6	88.3		
Liver transplant		22	Interim outcome at 6 months following the initial surgery (1999~)					
Hilar re-dissection		1		Death	2			
Hilar curettage		4		Living	88			
Endoscopic hilar curettage		0		Without jaundice	55			
Other		1		With jaundice	17			
Reoperations other than liver transplant (1999~)		6		Jaundice unknown	1			
Age (days)	Maximum	169	Age at death (days) within 6 months (1999~)					
	Minimum	64		Unknown	1			
	Mean	116.2	Total				91	
	Standard deviation	41.2						
Jaundice	Disappeared	3	Maximum				186	
	Improved	2	Minimum				152	
	Unchanged	0	Mean				169.0	
	Worsened	1	Standard deviation				24.0	
	Unknown	0						
Re-reoperation		5	Final outcome					
Liver transplant		2		Death	2	0	2	
Hilar re-dissection		0		Living	89	0	89	
Hilar curettage		0		Without jaundice	0	0	51	
Endoscopic hilar curettage		0		With jaundice	0	0	13	
Other		3		Unknown living	0	0	0	
Re-reoperations other than liver transplant (1999~)		3		Transplant living	0	0	25	
Age (days)	Maximum	320	Survival details unknown				0	0
	Minimum	184	Unknown				0	0
	Mean	252.0	Total				91	0
	Standard deviation	96.2						
Jaundice	Disappeared	2	Cause of death					
	Improved	0		Liver failure	1			
	Unchanged	0		Rupture of esophageal varices	0			
	Worsened	1		Pneumonia	0			
	Unknown	0		Peritonitis	0			
					Sepsis after cholangitis	0		
					Post-transplantation death	0		
					Other	2		
				Unknown	0			
			(Includes duplications)					

Screening (from 2012)					
By stool color card					
Yes: 42					
Before 1 month old	23	At 1 month old	26	After 1 month old	27
Observed age (days)		Observed age (days)		Observed age (days)	
Observed date not stated	4	Observed date not stated	8	Observed date not stated	4
Maximum	30	Maximum	60	Maximum	107
Minimum	3	Minimum	14	Minimum	32
Mean	13.3	Mean	33.9	Mean	58.5
Standard deviation	7.5	Standard deviation	9.3	Standard deviation	20.9
Number of the color					
1	0	1	1	1	2
2	6	2	8	2	15
3	7	3	6	3	7
4	8	4	9	4	2
5	0	5	0	5	0
6	0	6	1	6	0
7	1	7	0	7	0
Unknown	1	Unknown	1	Unknown	1
By urinary sulfated bile acid		By direct bilirubin		Other	
Yes	3	Yes	27	Yes	1
Abnormal	3	Abnormal	27		
Normal	0	Normal	0		
Unknown	0	Unknown	0		