

2023~2023 83 cases

Number of registered cases by region			Birth order		Meconium color		
	No. registrations	No. hospitals	First child	35	Normal	49	
Hokkaido	4	2	Second child	28	Abnormal	9	
Tohoku	5	1	Third child	14	Unknown	25	
Kanto	35	14	Fourth child	2	Yellow stool		
Chubu and Hokuriku	19	6	Fifth child and later	1	Yes	58	
Kinki	15	6	Unknown	3	No	13	
Chugoku and Shikoku	0	0	Number of babies (1999~)		Unknown	12	
Kyushu	5	4	Singleton	80	Neonatal jaundice		
Total	83	33	Twins or more	0	Yes	44	
			Unknown	1	No	31	
Age of parents			Gender		Unknown		
	Mean	Standard deviation	Male	30	Phototherapy		
Father	34.3	6.2	Female	53	Yes	27	
Mother	32.8	4.2	Unknown	0	No	49	
Maternal history during pregnancy			Birth weight (g)		Unknown		
	Febrile rash disease	1	Maximum	4005	Pathological bleeding		
	Other	12	Minimum	1232	Yes	3	
		0	Mean	2870.4	(bleeding sites have duplications)	Intracranial	0
Family history (parents)			Standard deviation	489.0		Gastrointestinal	1
Biliary atresia	Parents	0	~1499	3		Subcutaneous	1
	Siblings	0	1500~1999	1		Other	1
	Other	0	2000~2499	13	No	80	
Choledochal cyst	Parents	0	2500~2999	33	Unknown	0	
	Siblings	0	3000~3499	22	Age at bleeding (days)		
	Other	0	3500~3999	7	Maximum	93	
Hepatitis	Parents	0	4000~	1	Minimum	15	
	Siblings	0	不明	3	Mean	57.0	
	Other	0	Gestational age (weeks)		Standard deviation	39.3	
		1	Maximum	41	Prothrombin activity (%)		
			Minimum	31	Maximum	171	
			Mean	38.4	Minimum	21	
			Standard deviation	1.5	Mean	95.4	
			~29	1	Standard deviation	26.9	
			30~31	1	≥70%	68	
			32~33	0	≥40%<70%	9	
			34~35	1	<40%	2	
			36~37	19	PT-INR		
			38~39	42	Maximum	10.0	
			40~41	17	Minimum	0.7	
			42~	0	Mean	1.3	
			Unknown	2	Standard deviation	1.0	
Chromosomal abnormality					<1.15%	44	
	Yes				≥1.15% <1.4%	15	
		Down症			≥1.4%	21	
		その他					
	No					76	
	Unknown					7	

Age at initial consult (days)		Name of associated congenital anomaly		Name of associated congenital anomaly (Cont.)	
Maximum	192	Heart		Urogenital	
Minimum	0	Tetralogy of Fallot	1	Polycystic kidney	0
Mean	48.3	Patent foramen ovale	3	Renal hypoplasia	0
Standard deviation	40.2	Patent ductus arteriosus	0	Hydronephrosis	0
Age at hospitalization (days)		VSD	3	Horseshoe kidney	0
Maximum	198	ASD	1	Double renal pelvis and ureter	0
Minimum	0	Dextrocardia	0	Hydrocele of the testis	0
Mean	49.3	Pulmonary stenosis	2	Cryptorchidism	0
Standard deviation	41.0	Other	2	Extrophy of bladder	0
Prenatal diagnosis (1999~)				Hypospadias	0
Yes	5	Vascular		Urethral stricture	0
No	78	Preduodenal portal vein	1	VUR	0
Unknown	0	Absence of the inferior vena cava	0	Other	0
Screening		Anomalous pulmonary venous return	0		
Yes	37	Other	0	Musculoskeletal	
No	41			Spina bifida	0
Unknown	5	Spleen	0	Costal malformation	0
Stool color at admission		Polysplenia	1	Other	3
Yellow	6	Accessory spleen	0		
Pale yellow	37	Asplenia	1	Limb and external anomaly	
Gray	37	Other	0	Accessory ear	0
Unknown	3			Cleft lip	0
Total bilirubin immediately before surgery (mg/dl)		Gastrointestinal		Cleft lip and palate	0
Maximum	19.9	Malrotation of intestine	4	Polysyndactyly	0
Minimum	3.7	Esophageal atresia	0	Clubfoot	0
Mean	8.1	Intestinal atresia	0	Other	0
Standard deviation	3.1	Anorectal malformation	1		
Direct bilirubin immediately before		Hirschsprung's disease	0	Central nervous system	
Maximum	13.2	Situs inversus	2	Hydrocephalus	0
Minimum	1.6	Annular pancreas	0	Microcephaly	0
Mean	5.4	Gastric rupture	0	Anencephaly	0
Standard deviation	2.4	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	79	Pancreaticobiliary maljunction	0	Other	0
ERCP	0	Intussusception	0		
Serum bile acid	57	Other	2	Other malformations	
Percutaneous liver biopsy	1			Diaphragmatic hernia	0
Duodenal juice analysis	27	Respiratory		Omphalocele	0
Hepatobiliary scintigraphy	50	Tracheal/bronchial stenosis	0	Gastroschisis	0
MRI (MRCP)	6	Cystic lung disease	0	Inguinal hernia	3
CT	5	Lobar emphysema	0	Umbilical hernia	5
Other	0	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		83	~30	15	14	10	5	35.7
	Bile drainage surgery	81	31~45	18	18	11	9	50.0
	Liver transplant	1	46~60	22	22	13	7	31.8
	Exploratory laparotomy	1	61~70	3	3	2	1	33.3
No (all 1996 cases)		0	71~80	6	6	5	3	50.0
Unknown		0	81~90	6	6	6	3	50.0
Biliary reconstruction		0	91~120	5	5	5	5	100.0
	Hepaticoenterostomy	3	121~150	5	5	0	2	40.0
	Portoenterostomy	77	151~	3	2	0	1	50.0
	Other	1	Unknown	0	0	0	0	
	Unknown	0						
Total				83	81	52	36	44.4

Age at time of surgery (days)	
	0
Maximum	202
Minimum	12
Mean	60.6
Standard deviation	35.8

Surgical procedure		No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)
No cholangitis prevention		66				
Simple Roux-en-Y anastomosis		66	40	60.6	32	48.5
	<20 cm	1	1	100.0	1	100.0
	≥20 cm <40 cm	35	22	62.9	18	51.4
	≥40 cm <60 cm	30	17	56.7	13	43.3
	≥60 cm	0	0		0	
	Unknown	0	0		0	
Porto/hepaticoduodenostomy	0	0		0		
Other	0	0		0		
Unknown	0	0		0		
Cholangitis prevention		15	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	3	25.0
Hepatic portocholecystostomy or hepaticocholecystostomy		0	0		0	
Other		3	2	66.7	1	33.3
Unknown		0	0		0	
Cholangitis prevention unknown		0	0		0	
Total		81				

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Basic Type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	Esophageal varices checked before or during surgery		
I	4	4	3	75.0	3	75.0	Not checked	76	
I -cyst	4	4	4	100.0	1	25.0	Varices present	0	
II	1	1	1	100.0	0	0.0	No varices	4	
III	72	71	43	60.6	32	45.1	Unknown	3	
Unknown	2	1	1	100.0	0	0.0	Total	83	
Total	83	81	52	64.2	36	44.4	Choleretics		
Sub type	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)			
a1	16	16	9	56.3	7	43.8	Dehycol	0	
a2	3	3	3	100.0	2	66.7	UDCA	78	
b1	46	45	31	68.9	21	46.7	Glucagon	8	
b2	2	2	2	100.0	1	50.0	Secretin	0	
c1	11	11	5	45.5	5	45.5	PGE2	0	
c2	2	2	0	0.0	0	0.0	PGF2 α	0	
d	0	0	0	#DIV/0!	0	#DIV/0!	Steroids	73	
Unknown	3	2	2	100.0	0	0.0	Taurine	7	
Total	83	81	52	64.2	36	44.4	Phenobarbital	0	
Type of porta hepatitis	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	Other	3	
α	1	1	1	100.0	0	0.0	(includes duplications)		
β	8	8	7	87.5	4	50.0	Bile drainage after first surgery		
γ	1	1	1	100.0	1	100.0	Yes	1989~1998	1999~
μ	5	5	2	40.0	4	80.0	No	0	8
ν	63	62	38	61.3	26	41.9	Unknown	0	0
Total	83	81	52	64.2	36	44.4	Total	0	81
Type of porta hepatitis	Total No.	No. Bile drainage surgery	Jaundice disappeared	(%)	Cholangitis	(%)	Jaundice after first surgery		
α	1	1	1	100.0	0	0.0	Disappeared	0	52
β	8	8	7	87.5	4	50.0	Improved	0	15
γ	1	1	1	100.0	1	100.0	Unchanged	0	7
μ	5	5	2	40.0	4	80.0	Worsened	0	7
ν	63	62	38	61.3	26	41.9	Unknown	0	0
Total	83	81	52	64.2	36	44.4	Total	0	81
Gallbladder							Jaundice re-elevation		
Yes							1989~1998		
No bile							1999~		
Yellow							0		
Pale yellow							4		
Colorless							33		
Other							2		
Unknown							5		
Absent							6		
Unknown							4		
Total							83		
Cystic structure in extrahepatic bile duct							Measures for re-elevation		
Yes							Monitor progress		
Yellow							10		
Pale yellow							7		
Colorless							7		
Other							3		
Unknown							24		
No							Total		
Unknown							44		
Total							83		

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Cholangitis		1989~1998	1999~	Postoperative complications	
Yes		0	36	Ileus	2
	Mean timing of first time (weeks)		10.7	Peritonitis	2
	Mean timing of second time (weeks)		16.2	Hemorrhage	1
No		0	45	Pulmonary complications	1
Unknown		0	0	Acute liver failure	0
Total		0	81	Infection	4
Reoperation within 6 months (1999~)			(Includes duplications)	Other	2
Yes		18	18	Age at liver transplant (days) (1999~)	20
	No	64	64	Maximum	460
	Unknown	1	1	Minimum	161
Total		83	83	Mean	259.1
Reoperation		25	25	Standard deviation	81.6
Liver transplant		16	16	After initial surgery	371
Hilar re-dissection		2	2	Minimum	161
Hilar curettage		3	3	Mean	259.1
Endoscopic hilar curettage		0	0	Standard deviation	81.6
Other		3	3	Minimum outcome at 6 months following the initial surgery (1989~)	89.3
Reoperations other than liver transplant (1999~)		9	9	Death	0
Age (days)	Maximum	119	119	Living	82
	Minimum	60	60	Without jaundice	43
	Mean	86.1	86.1	With jaundice	23
	Standard deviation	22.1	22.1	Jaundice unknown	1
Jaundice	Disappeared	4	4	Post-transplant	15
	Improved	2	2	Unknown	1
	Unchanged	2	2	Total	83
	Worsened	1	1	Age at death (days) within 6 months (1999~)	
	Unknown	0	0	Maximum	-
Re-reoperation		4	4	Minimum	-
Liver transplant		3	3	Mean	-
Hilar re-dissection		0	0	Standard deviation	-
Hilar curettage		0	0	Final outcome	
Endoscopic hilar curettage		0	0	Death	1
Other		1	1	Living	79
Re-reoperations other than liver transplant (1999~)		1	1	Without jaundice	0
Age (days)	Maximum	369	369	With jaundice	0
	Minimum	369	369	Unknown living	0
	Mean	369.0	369.0	Transplant living	0
	Standard deviation	-	-	Survival details unknown	0
Jaundice	Disappeared	1	1	Cause of death	
	Improved	0	0	Liver failure	0
	Unchanged	0	0	Rupture of esophageal varices	0
	Worsened	0	0	Pneumonia	0
	Unknown	0	0	Peritonitis	0
				Sepsis after cholangitis	0
			Post-transplantation death	0	
			Other	1	
			Unknown	0	
(Includes duplications)					
				1989~1998	1999~
				0	1
				0	79
				0	44
				0	10
				0	0
				0	25
				0	0
				3	0
				83	0
				0	3
				0	0
				0	0
				0	0
				0	0
				0	0
				1	0
				0	0

Screening (from 2012)					
By stool color card					
Yes: 36					
Before 1 month old	19	At 1 month old	26	After 1 month old	15
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	54	Maximum	145
Minimum	0	Minimum	9	Minimum	32
Mean	11.6	Mean	29.9	Mean	68.8
Standard deviation	9.3	Standard deviation	8.8	Standard deviation	38.1
Number of the color					
1	0	1	0	1	2
2	7	2	5	2	7
3	1	3	6	3	1
4	4	4	9	4	2
5	0	5	1	5	0
6	0	6	0	6	0
7	0	7	0	7	0
Unknown	7	Unknown	5	Unknown	3
By urinary sulfated bile acid		By direct bilirubin		Other	
Yes	1	Yes	9	Yes	0
Abnormal	1	Abnormal	3		
Normal	0	Normal	0		
Unknown	0	Unknown	6		