

Japanese Biliary Atresia Registry (JBAR) Summary Sheet 2016~

2016

2016~2016 81 cases

Number of registered cases by region			Birth order		Meconium color		
	No. registrations	No. hospitals	First child	39	Normal		41
Hokkaido	1	1	Second child	24	Abnormal		7
Tohoku	9	4	Third child	13	Unknown		33
Kanto	26	14	Fourth child	2	<b>Yellow stool</b>		
Chubu and Hokuriku	12	6	Fifth child and later	2	Yes		44
Kinki	17	9	Unknown	1	No		9
Chugoku and Shikoku	6	4	<b>Number of babies (1999~)</b>		Unknown		28
Kyushu	10	3	Singleton	80	<b>Neonatal jaundice</b>		
Total	81	41	Twins or more	0	Yes		42
<b>Age of parents</b>		Mean	Standard deviation	Unknown	1	No	25
Father	33.1	6.3	<b>Gender</b>		Unknown		
Mother	31.4	5.2	Male	37	<b>Phototherapy</b>		
Maternal history during pregnancy			Female	44	Yes		31
	Febrile rash disease	2	Unknown	0	No		37
	Other	15	<b>Birth weight (g)</b>		Unknown		13
<b>Family history (parents)</b>			Maximum	4250	<b>Pathological bleeding</b>		
Biliary atresia	Parents	0	Minimum	1200	(bleeding sites have duplications)	Yes	3
	Siblings	0	Mean	2897.8		Intracranial	3
	Other	0	Standard deviation	492.9		Gastrointestinal	0
Choledochal cyst	Parents	0	~1499	2		Subcutaneous	0
	Siblings	0	1500~1999	1		Other	1
	Other	1	2000~2499	9		No	76
Hepatitis	Parents	1	2500~2999	31	Unknown		2
	Siblings	0	3000~3499	31	<b>Age at bleeding (days)</b>		
	Other	0	3500~3999	3	Maximum		73
Other		5	4000~	1	Minimum		54
			Unknown	3	Mean		63.0
			<b>Gestational age (weeks)</b>		Standard deviation		9.5
			Maximum	41	<b>Prothrombin activity (%)</b>		
			Minimum	29	<b>No. cases with data: 78 cases</b>		
			Mean	38.4	Maximum		145
			Standard deviation	2.1	Minimum		8
			~29	1	Mean		86.9
			30~31	2	Standard deviation		32.1
			32~33	0	≥70%		62
			34~35	2	≥40%<70%		7
			36~37	10	<40%		9
			38~39	42	<b>PT-INR</b>		
			40~41	22	<b>No. cases with data: 80 cases</b>		
			42~	0	Maximum		10.0
			Unknown	3	Minimum		0.8
			<b>Chromosomal abnormality</b>		Mean		1.4
			Yes		Standard deviation		1.4
					<1.15%		59
			Down's syndrome		≥1.15% <1.4%		10
			Other		≥1.4%		11
			No		<b>Hepaplastin test (%)</b>		
			Unknown		<b>No. cases with data: 24 cases</b>		
					Maximum		98
					Minimum		10
					Mean		58.8
					Standard deviation		27.4

Age at initial consult (days)		Name of associated congenital anomaly		Name of associated congenital anomaly (Cont.)	
Maximum	141	Heart		Urogenital	
Minimum	0	Tetralogy of Fallot	0	Polycystic kidney	0
Mean	42.3	Patent foramen ovale	0	Renal hypoplasia	0
Standard deviation	31.9	Patent ductus arteriosus	0	Hydronephrosis	1
<b>Age at hospitalization (days)</b>		VSD	0	Horseshoe kidney	0
Maximum	187	ASD	0	Double renal pelvis and ureter	1
Minimum	0	Dextrocardia	0	Hydrocele of the testis	0
Mean	44.9	Pulmonary stenosis	0	Cryptorchidism	0
Standard deviation	33.4	Other	3	Exstrophy of bladder	0
<b>Prenatal diagnosis (1999~)</b>				Hypospadias	0
Yes	6	Vascular		Urethral stricture	0
No	75	Preduodenal portal vein	1	VUR	0
Unknown	0	Absence of the inferior vena cava	0	Other	2
<b>Screening</b>		Anomalous pulmonary venous return	0		
Yes	44	Other	1	Musculoskeletal	
No	37			Spina bifida	0
Unknown	0	Spleen		Costal malformation	0
<b>Stool color at admission</b>		Polysplenia	1	Other	0
Yellow	7	Accessory spleen	0		
Pale yellow	34	Asplenia	0	Limb and external anomaly	
Gray	40	Other	0	Accessory ear	0
Unknown	0			Cleft lip	0
<b>Total bilirubin immediately before surgery (mg/dl)</b>		Gastrointestinal		Cleft lip and palate	0
Maximum	22.6	Malrotation of intestine	1	Polysyndactyly	0
Minimum	3.3	Esophageal atresia	0	Clubfoot	0
Mean	8.4	Intestinal atresia	0	Other	1
Standard deviation	3.2	Anorectal malformation	0		
<b>Direct bilirubin immediately before surgery (mg/dl)</b>		Hirschsprung's disease	0	Central nervous system	
Maximum	15.7	Situs inversus	1	Hydrocephalus	0
Minimum	1.3	Annular pancreas	0	Microcephaly	0
Mean	5.4	Gastric rupture	0	Anencephaly	0
Standard deviation	2.2	Meconium peritonitis	0	Other	0
<b>Implemented tests</b>		Meckel's diverticulum	0		
Serum Lp-X	0	Cloacal exstrophy	0	Endocrine metabolic system	
Score test	1	Alagille syndrome	0	Cretinism	0
Ultrasound	77	Pancreaticobiliary maljunction	0	Other	0
ERCP	0	Intussusception	0		
Serum bile acid	56	Other	1	Other malformations	0
Percutaneous liver biopsy	1			Diaphragmatic hernia	0
Duodenal juice analysis	38	Respiratory		Omphalocele	0
Hepatobiliary scintigraphy	51	Tracheal/bronchial stenosis	0	Gastroschisis	0
MRI (MRCP)	3	Cystic lung disease	0	Inguinal hernia	0
CT	6	Lobar emphysema	0	Umbilical hernia	0
Other	8	Pulmonary hypoplasia	0	Other	0
Multiple answers allowed		Other	0		

Surgery implemented		Age at time of surgery (days)	Total No.	Jaundice disappeared (%)	Cholangitis (%)		
Yes		~30	12	8	66.7	5	41.7
	Bile drainage surgery	31~45	18	11	61.1	9	50.0
	Liver transplant	46~60	22	12	54.5	5	22.7
	Exploratory laparotomy	61~70	11	6	54.5	6	54.5
No (all ~ 1998 cases)		71~80	7	4	57.1	4	57.1
Unknown		81~90	2	0	0.0	0	0.0
<b>Biliary reconstruction</b>		91~120	6	2	33.3	1	16.7
Hepaticoenterostomy		121~150	3	0	0.0	1	33.3
Portoenterostomy		151~	0	0		0	
Other		Unknown	0	0		0	
Unknown			0				
		<b>Total</b>	<b>81</b>	<b>43</b>	<b>53.1</b>	<b>31</b>	<b>38.3</b>

Age at time of surgery (days)	
Maximum	149
Minimum	16
Mean	57.0
Standard deviation	27.6

Surgical procedure	Total No.	Jaundice disappeared (%)	Cholangitis (%)		
No cholangitis prevention	69				
Simple Roux-en-Y anastomosis	69	38	55.1	29	42.0
<20 cm	1	0	0.0	0	0.0
≥20 cm <40 cm	21	14	66.7	12	57.1
≥40 cm <60 cm	44	21	47.7	16	36.4
≥60 cm	3	3	100.0	1	33.3
Unknown	0	0		0	
Porto/hepaticoduodenostomy	0	0		0	
Other	0	0		0	
Unknown	0	0		0	
Cholangitis prevention	8				
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	8	5	62.5	2	25.0
Hepatic portocholecystostomy or hepaticocholecystostomy	0	0		0	
Other	0	0		0	
Unknown	0	0		0	
Cholangitis prevention unknown	0	0		0	
<b>Total</b>	<b>77</b>				

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Main type	Total No.	Jaundice disappeared	(%)	Cholangitis	(%)	Esophageal varices checked before or during surgery		
I	2	0	0.0	0	0.0	Not checked	76	
I-cyst	6	6	100.0	3	50.0	Varices present	1	
II	0	0		0		No varices	3	
III	73	37	50.7	28	38.4	Unknown	1	
Unknown	0	0		0		<b>Total</b>	81	
<b>Total</b>	81	43	53.1	31	38.3	<b>Choleretics</b>		
Sub type	Total No.	Jaundice disappeared	(%)	Cholangitis	(%)	Dehycol	0	
a1	13	10	76.9	3	23.1	Urso	76	
a2	7	3	42.9	2	28.6	Glucagon	9	
b1	51	25	49.0	25	49.0	Secretin	0	
b2	0	0		0		PGE2	0	
c1	5	4	80.0	1	20.0	PGF2α	0	
c2	3	0	0.0	0	0.0	Steroids	75	
d	1	0	0.0	0	0.0	Taurine	3	
Unknown	1	1	100.0	0	0.0	Phenobarbital	4	
<b>Total</b>	81	43	53.1	31	38.3	Other	3	
Type of porta hepatitis	Total No.	Jaundice disappeared	(%)	Cholangitis	(%)	(includes duplications)		
α	2	1	50.0	1	50.0	<b>Bile drainage after first surgery</b>		
β	4	2	50.0	1	25.0	Yes	71	
γ	0	0		0		No	6	
μ	4	3	75.0	2	50.0	Unknown	0	
ν	63	36	57.1	26	41.3	<b>Total</b>	77	
ο	7	0	0.0	1	14.3	<b>Jaundice after first surgery</b>		
Unknown	1	1	100.0	0	0.0	Disappeared	43	
<b>Total</b>	81	43	53.1	31	38.3	Improved	22	
<b>Gallbladder</b>							Unchanged	5
Yes	79					Worsened	7	
Color of content	No bile	32				Unknown	0	
	Yellow	2				<b>Total</b>	77	
	Pale yellow	1				<b>Jaundice re-elevation</b>		
	Colorless	39				No re-elevation	34	
	Other	3				Re-elevated after disappearance	9	
	Unknown	2				Re-elevated after improvement	17	
Absent	2					Unknown	5	
Unknown	0					<b>Total</b>	65	
<b>Total</b>	81					<b>Measures for re-elevation</b>		
<b>Cystic structure in extrahepatic bile duct</b>							Monitor progress	1
Yes	7					Conservative treatment	9	
Color of content	Yellow	2				Re-do surgery	6	
	Pale yellow	2				Liver transplant	25	
	Colorless	3				<b>Total</b>	41	
	Other	0						
	Unknown	0						
No	73							
Unknown	1							
<b>Total</b>	81							

<b>Cholangitis</b>	1989~1998	1999~	<b>Age at liver transplant (days) (1999~)</b>	31	<b>After initial surgery</b>
Yes	466	31	Maximum	379	336
	Mean timing of first time	10.2 weeks	Minimum	0	0
	Mean timing of second time	14.5 weeks	Mean	220.6	158.3
No	747	46	Standard deviation	74.1	75.9
Unknown	45	0	<b>Reoperations other than liver transplant (1999~)</b>		8
Total	1258	77	Age (days)	Maximum	371
<b>Postoperative complications</b>				Minimum	41
	Ileus	1		Mean	137.9
	Peritonitis	2		Standard deviation	108.7
	Hemorrhage	5	Jaundice	Disappeared	6
	Pulmonary complications	0		Improved	0
	Acute liver failure	1		Unchanged	1
(Includes duplications)	Infection	4		Worsened	0
	Other	1		Unknown	1
<b>Interim outcome at 6 months following the initial surgery (1999~)</b>			<b>Re-reoperations other than liver transplant (1999~)</b>		
Death	0		Age (days)	Maximum	229
Living	80			Minimum	217
	Without jaundice	40		Mean	223.0
	With jaundice	15		Standard deviation	8.5
	Jaundice unknown	2	Jaundice	Disappeared	1
	Post-transplant	23		Improved	0
Unknown	1			Unchanged	0
Total	81			Worsened	0
<b>Age at death (days) within 6 months (1999~)</b>				Unknown	1
	Maximum	0	<b>Final outcome</b>		
	Minimum	0	Death	1	
	Mean		Living	75	
	Standard deviation			Without jaundice	41
<b>Reoperation within 6 months (1999~)</b>				With jaundice	3
	Yes	29		Unknown living	0
	No	51		Transplant living	31
	Unknown	1		Survival details unknown	0
Total		81	Unknown	5	
<b>Reoperation</b>	37		Total	81	81
	Liver transplant	29	<b>Cause of death</b>		
	Hilar re-dissection	0		Liver failure	1
	Hilar curettage	4		Rupture of esophageal varices	1
	Endoscopic hilar curettage	0		Pneumonia	0
	Other	4		Peritonitis	0
<b>Re-reoperation</b>	4			Sepsis after cholangitis	0
	Liver transplant	2		Post-transplantation death	0
	Hilar re-dissection	0		Other	1
	Hilar curettage	0		Unknown	0
	Endoscopic hilar curettage	0	<b>(Includes duplications)</b>		
	Other	2			

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<b>Screening (from 2012)</b>					
By stool color card					
Yes: 39					
Before 1 month old	29	At 1 month old	23	After 1 month old	15
Observed age (days)		Observed age (days)		Observed age (days)	
Observed date not stated	8	Observed date not stated	9	Observed date not stated	2
Maximum	28	Maximum	50	Maximum	85
Minimum	1	Minimum	29	Minimum	34
Mean	15.5	Mean	33.1	Mean	54.2
Standard deviation	8.8	Standard deviation	5.4	Standard deviation	12.5
Number of the color		Number		Number	
1	3	1	2	1	2
2	4	2	3	2	6
3	6	3	6	3	6
4	9	4	7	4	0
5	1	5	0	5	0
6	0	6	0	6	0
7	0	7	0	7	0
Unknown	6	Unknown	5	Unknown	1
<b>By urinary sulfated bile acid</b>		<b>By direct bilirubin</b>		<b>Other</b>	
Yes	6	Yes	19	Yes	1
Abnormal	4	Abnormal	19		
Normal	2	Normal	0		
Unknown	0	Unknown	0		