



저작자표시-비영리-변경금지 2.0 대한민국

이용자는 아래의 조건을 따르는 경우에 한하여 자유롭게

- 이 저작물을 복제, 배포, 전송, 전시, 공연 및 방송할 수 있습니다.

다음과 같은 조건을 따라야 합니다:



저작자표시. 귀하는 원저작자를 표시하여야 합니다.



비영리. 귀하는 이 저작물을 영리 목적으로 이용할 수 없습니다.



변경금지. 귀하는 이 저작물을 개작, 변형 또는 가공할 수 없습니다.

- 귀하는, 이 저작물의 재이용이나 배포의 경우, 이 저작물에 적용된 이용허락조건을 명확하게 나타내어야 합니다.
- 저작권자로부터 별도의 허가를 받으면 이러한 조건들은 적용되지 않습니다.

저작권법에 따른 이용자의 권리는 위의 내용에 의하여 영향을 받지 않습니다.

이것은 [이용허락규약\(Legal Code\)](#)을 이해하기 쉽게 요약한 것입니다.

[Disclaimer](#)

의학석사 학위논문

**Epidemiology of Biliary Atresia in Korea**

한국의 담도폐쇄증의 역학연구

2017년 2월

서울대학교 대학원  
의과대학 임상외과학과  
이경재

**A thesis of the Master of Science in Medicine**

**한국의 담도폐쇄증의 역학연구**

**Epidemiology of Biliary Atresia in Korea**

**February 2017**

**The Department of Clinical Medical Science**

**Seoul National University Graduate School**

**Kyung Jae Lee**

## ABSTRACT

**Introduction:** Biliary atresia (BA) is the major cause of cholestasis and the leading indication for liver transplantation (LT). However, the incidence of BA in Korea has not been reported. The aim of this study was to investigate the incidence and clinical outcomes of BA in Korea.

**Methods:** We used the Korean universal health insurance database and extracted data regarding BA patients younger than 18 years of age admitted between 2011 and 2015. The incidence of BA was calculated by dividing the number of BA patients by the number of live births.

**Results:** Two hundred forty infants were newly diagnosed with BA. A total of 963 BA patients younger than 18 years of age were followed up for 5 years. The overall incidence of BA was 1.06 cases per 10,000 live births. The incidence of BA was 1.4 times higher for female patients than for male patients. Additionally, significant seasonal variation was observed; in particular, the incidence of BA was two times higher from June through August than from December through February.

Congenital anomalies were found in 38 out of 240 patients (15.8%). Congenital heart diseases were major associated congenital anomalies (6.3%). Several complications developed during the study period, including cholangitis (24.0%), varix (6.2%) and gastrointestinal bleeding (4.4%). Three hundred one of the 963 BA patients under 18 years of age (31.3%) received LT for BA.

**Conclusions:** The incidence of BA is higher in Korea than that in Western countries. We also report significant gender-associated differences and seasonal variation with respect to the incidence of BA.

**Keywords:** Biliary Atresia, Incidence, Liver Transplantation.

**Student number:** 2015-22252

# CONTENTS

Abstract .....	i
Contents .....	iii
List of tables and figures .....	iv
List of abbreviations .....	v
Introduction .....	1
Materials and Methods .....	2
Results .....	4
1. Incidence .....	4
2. Congenital anomalies .....	7
3. Complications .....	9
4. Liver transplantation .....	9
Discussion .....	11
References .....	14
Abstract in Korean .....	18

## LIST OF TABLES AND FIGURES

Table 1. Incidence and relative risk of biliary atresia by sex and year in Korea, 2011-2015 .....	5
Figure 1. Seasonal variation in the incidence of biliary atresia.....	6
Table 2. Frequency of congenital anomalies in biliary atresia patients .....	8
Table 3. Cases of liver transplantation in biliary atresia patients during 2011-2015 ...	10

## LIST OF ABBREVIATIONS

BA; Biliary atresia

LT; Liver transplantation

HIRA; Health insurance review and assessment service

KCD-7; Korean Classification of Disease, 7<sup>th</sup> Revision

EBC; Endoscopic bleeding control

HCC; Hepatocellular carcinoma

DDLT; Deceased donor LT

LDLT; Living donor LT



# INTRODUCTION

Biliary atresia (BA) is the major cause of neonatal cholestatic jaundice, and untreated BA can cause chronic liver disease, hepatic failure and death in children (1-11). BA is well established as the leading indication for liver transplantation (LT) in children in Korea. Furthermore, BA is the most common cause of LT in infants under 12 months (12).

The incidence of BA is slightly higher in Eastern countries (1.04 to 1.79/10,000 live births) than in Western countries (0.42 to 0.71/10,000) (1-3, 5-9, 13-21). This difference in incidence rates may be due to a combination of factors, including genetics, environmental factors and infectious pathogens (5, 7, 20). The incidence of BA in Korea has not been reported. The aim of this study was to investigate the incidence and clinical outcomes of BA in Korea.

# MATERIALS AND METHODS

## MATERIALS AND METHODS

### Data source

We used the Health Insurance Review and Assessment Service (HIRA) database based on the Korean universal health insurance system. We retrieved information about patient's age, sex, diagnosis and treatments, and diagnosis, which was presented by the Korean Classification of Disease, 7<sup>th</sup> Revision (KCD-7) code.

### Patient selection and definition of cases

We extracted the following information from the database: KCD-7, Q442 or Q443 BA status in patients under 18 years of age from January 2011 to December 2015. We defined two BA groups: the “newly diagnosed BA group”, which included patients with BA who received the Kasai operation (Q735-Q737) or LT (Q814) before their first birthday, and the “total BA group”, which included all BA patients between 0 and 18 years of age during the 5-year study period.

The newly diagnosed BA group was used to calculate the incidence of BA and the percentage of congenital anomalies because definite and suspected BA cases could not be discriminated using the KCD-7. The total BA group was used to calculate the percentages of complications and LT.

The incidence of BA was calculated by dividing newly diagnosed BA patients by the population of live births. We defined the date of diagnosis as the first day that a patient visited a medical facility for BA. Month of diagnosis was grouped into four seasons: winter (December-February), spring (March-May), summer (June-August) and autumn (September-November). Incidence is presented as the number of BA patients per 10,000 live births with a

95% confidence interval.

We investigated the percentage of congenital anomalies such as spleen anomalies, situs inversus, and heart disease in the newly diagnosed BA group. If different congenital anomalies were present in a single patient, each anomaly was counted separately. Percentages of complications and LT were calculated by dividing the number of applicable cases by the size of the total BA group. We also evaluated patients subjected to endoscopic variceal bleeding control.

For the total BA group, we determined the number of patients who underwent LT during the 5-year study period and the number of patients who had already undergone LT prior to 2011.

### **Statistics**

We analyzed the data using SAS Enterprise version 6.1 (SAS Institute, Cary, NC, USA). The 95% confidence interval of BA incidence was calculated using the Poisson distribution.

The seasonal, annual and gender variation in the occurrence of BA was measured using the Poisson regression test. We used linear regression to evaluate the linear trend in LT over time.

### **Ethical approval**

This study was exempt from Institutional Review Board review because it contains open data and does not include any personal identifiable information.

# RESULTS

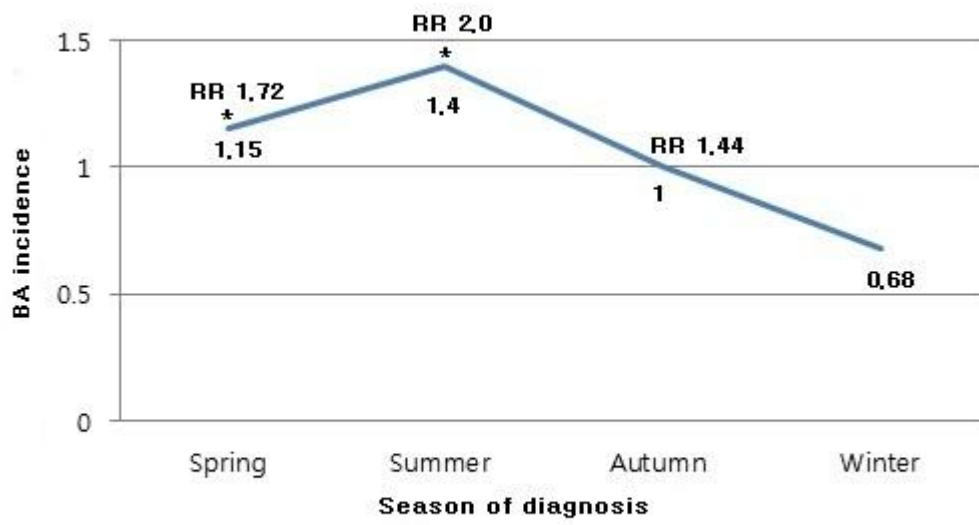
## **Incidence**

A total of 240 BA patients (137 females, 103 males) were born during the 5-year study period (56 in 2011, 57 in 2012, 55 in 2013, 42 in 2014, and 40 in 2015).

The incidence of BA was 1.06/10,000 [95% CI: 0.93-1.19], (females 1.28, males 0.92). The incidence of BA was 1.4 times higher for female than male patients, and this difference was statistically significant ( $p=0.02$ ) (Table 1). When the months were grouped into one of four seasons, the rate from June through August (summer) was two times higher than that from December through February (winter) ( $p<0.01$ ) (Figure 1). There was no statistically significant difference in annual incidence.

Table 1. Incidence and relative risk of biliary atresia by sex and year in Korea, 2011-2015

Variable	Number of BA patients	Incidence of BA per 10,000 live births	Relative risk	<i>P</i> value
<b>Sex</b>				
M	103	0.92	.	.
F	137	1.28	1.4	0.02
<b>Year</b>				
2011	56	1.19	1.4	0.1
2012	57	0.97	1.43	0.09
2013	55	1.26	1.38	0.16
2014	42	0.96	1.05	0.83
2015	40	0.91	.	.
Total	240	1.06		



**RR: Relative risk**

**\*  $P < 0,05$**

Figure 1. Seasonal variation in the incidence of biliary atresia

## **Congenital anomalies**

Congenital anomalies were found in 38/240 of patients in the newly diagnosed BA group (15.8%); specifically, this group included 2 patients with spleen anomalies (0.8%) and 1 patient with situs inversus (0.4%). Congenital heart diseases comprised major associated congenital anomalies (15 patients, 6.3%), which ranged from relatively minor disease (atrial septal defect, patent ductus arteriosus) to life threatening severe disease (hypoplastic left heart syndrome, pulmonary atresia). Two patients had more than two congenital heart diseases. Ten patients (4.2%) had biliary anomalies (choledochal cyst or duplication of the biliary duct), and 8 patients (3.3%) were diagnosed with intestinal anomalies (small bowel atresia, malrotation, tracheoesophageal fistula, Meckel's diverticulum). One patient with congenital cystic lung and another with congenital vesicoureteral reflux were reported (Table 2).

Table 2. Frequency of congenital anomalies in biliary atresia patients

<b>Congenital anomalies</b>	<b>N (%)</b>
<b>Congenital heart disease</b>	<b>15 (6.3)</b>
Atrial septal defect	10 (4.2)
Patent ductus arteriosus	1 (0.4)
Pulmonary stenosis	4 (1.3)
Pulmonary valve atresia	1 (0.4)
Hypoplastic left heart syndrome	1 (0.4)
<b>Biliary anomalies</b>	<b>10 (4.2)</b>
Choledochal cyst	3 (1.25)
Biliary duct duplication	5 (2.1)
Other congenital malformations of the gall bladder	2 (0.8)
<b>Gastrointestinal</b>	<b>8 (3.3)</b>
Esophageal atresia, tracheoesophageal fistula	2 (0.8)
Small bowel stenosis, small bowel atresia, malrotation, Meckel diverticulum	6 (2.5)



## **Complications**

The total BA group included 963 BA patients. Two hundred thirty-one patients (24%) experienced at least one episode of cholangitis, and their median age was 1 (Table 2). There were 60 patients (6.2%) who had esophageal and gastric varices, with a median age of 7. Gastrointestinal bleeding was reported in 42 patients (4.4%), and there were 44 patients (4.6%) who received endoscopic bleeding control (EBC). Three patients received repetitive EBC, with one patient who received EBC 6 times during a 5-year period.

Two patients developed hepatocellular carcinoma (HCC) before they turned 18 (at age 3 and 7) and two other patients showed secondary pulmonary hypertension (at age 4 and 13).

## **Liver transplantation**

One hundred twenty-four patients with BA underwent LT (including re-transplantation) during the 5-year study period, and the median age at LT was 1 year. Sixty-five out of these 124 patients (52.4%) received deceased donor LT (DDLTL); split LT was the preferred approach (34.0%), and 7 out of these 65 patients received re-DDLT (split DDLTL in 4 cases and total DDLTL in 3 cases). The remaining 59 out of the aforementioned 124 patients (47.6%) received living donor LT (LDLT); typically, the left lateral section was used (37.3%), and 2 out of these 59 patients received re-LDLT using the left lateral section (Table 3). The cases of deceased DDLTL (split DDLTL) increased ( $p=0.1$ ), while LDLTL decreased over time ( $p=0.72$ ), but there was no statistically significant difference. Fifty-three out of 115 patients (46.0%) received LT prior to 1 year of age. Eleven patients (8.7%) received re-LT, and 3 out of these 11 patients were less than 1 year of age.

Three hundred one of the 963 patients in the total BA group (31.3%) received LT, including 124 patients who first received LT during the 5-year study period and 177 patients who underwent LT prior to 2011.

Table 3. Cases of liver transplantation in biliary atresia patients during 2011-2015

	2011	2012	2013	2014	2015	N (%)
DDLT	10	6	16	14	19	65 (52.4)
Total liver	4	1	3	5	4	17 (13.7)
Split liver	6	5	13	9	15	48 (38.7)
LDLT	9	13	13	14	10	59 (47.6)
Lt lat.	8	11	12	10	8	49 (39.5)
Lt liver	0	2	0	3	2	7 (5.6)
Rt liver	1	0	1	1	0	3 (2.4)
Total	19	19	29	28	29	124 (100)

\*DDLT: Deceased donor liver transplantation †LDLT: Living donor liver transplantation

## DISCUSSION

This is the first report of BA incidence in Korea using the national health system database and the entire population of children born in the country. We identified the differences in BA incidence by gender and season, and we also reported the associated congenital anomalies and clinical outcomes of BA.

The incidence of BA in Korea (1.06/10,000) is similar to that in other East Asian countries, including Japan (1.0-1.1) (15, 17) and Taiwan (1.5-1.8) (7, 9), but higher than that in European countries such as France (0.5) (2), the UK (0.6) (3, 19), Scotland (0.7) [16], and Switzerland (0.6) (18)) as well as the US cities of Atlanta (0.7) (1) and New York (0.9) (5). Because our incidence calculations only included patients who underwent the Kasai operation or LT before their first birthday, we may not have accounted for certain patients who died before receiving either of these operations or ceased receiving treatment.

The etiology of BA is unknown. However, environmental factors, including infections, toxins and ischemia, are purported to cause this disease. Several studies describe seasonal and geographic variation in the incidence of BA (1, 2, 5, 7, 9, 16-19, 21, 22). In Atlanta, significant seasonal clustering of BA was reported, with rates three times higher from December through March than rates from April through July (1, 7, 19). We also reported the seasonal variation of BA, and these results are consistent with environmental factors such as viral infection. This epidemiologic observation suggests that further investigation into the timing of possible viral infection around the perinatal period and subsequent identification of BA is necessary.

However, other studies insist that genetic factors are important to the pathogenesis of BA, showing the ethnic and gender variations in the incidence of BA (1, 6, 7, 13, 17, 19, 23). In

our study, the incidence of BA was 1.4 times higher in females than in males; this finding is consistent with the results of many previous studies (1, 6, 7, 13, 17, 19, 23).

In this study, 15.8% out of 240 BA patients had associated congenital anomalies; similar findings have been obtained in other studies (with corresponding percentages of 21.5%, 20.8%, 19.6%, 14%, and 12.6% for the UK, Switzerland, Japan, the US, and Canada, respectively) (1, 3, 6, 15, 18, 20, 24). Congenital heart disease (6.3%) was the major associated anomaly in our study, much like in other countries such as Canada (7.5%) and the US (8.7%), followed by biliary anomalies (4.2%) and gastrointestinal anomalies (3.3%) (20, 24).

In our total BA group, two HCC patients (0.2%, 3 and 7 years of age) were identified. At King's College Hospital, 3 out of 387 BA patients (0.8%) developed histologically proven HCC, which was detected at a median age of 2.1 years (range 1.8-4.9 years); in Belgium, an infant who was affected by BA complicated by HCC when he was six months old was treated via LT, suggesting that HCC can develop at an early age in children with BA (25-27). Pulmonary hypertension was reported in 5 out of the 617 BA patients who underwent LDLT (0.5%); this condition is more common among patients who undergo LDLT than among BA patients with naïve livers because the former patients are LT candidates with end-stage liver disease (28, 29). We also found two secondary pulmonary hypertension patients (of 4 and 13 years of age) in the total BA group.

LT, including re-LT, was performed in 124 BA patients during the 5-year study period, and the median age at LT was 1 year. Other studies have found median ages for LT for BA of 1.1 years in Taiwan, 11-14 months in Canada, and 12 months in the UK and Switzerland, which are similar to the age calculated in our study (3, 7, 18, 20).

In a study of patients from Seoul National University Hospital in Korea, a total of 152

children received LT from 1992 to 2010; 92 of these patients (60.5%) had BA, and 33 (35.9%) patients received LT for BA before 1 year of age (12). Our study produced similar results: 53 out of 115 patients (46.0%) received LT before 1 year of age. In Asian countries, LDLT has been the main form of LT because of a shortage of deceased donors attributable to cultural and ethical concerns (12). However, the number of pediatric DDLT increased from 2006 to 2010 due to increased total deceased donation rates and the pediatric split graft policy (30). In our report, the number of cases involving DDLT (split DDLT) increased over the study period, whereas the number of cases involving LDLT decreased during this time. Three hundred one of the 963 patients (31.3%) received LT; this rate is slightly lower than that observed in the US (40.3%) but higher than the corresponding rates in Taiwan (26.0%) and Croatia (20.7%) (7, 8, 24).

Our study has some limitations. The HIRA database only contains five years (2011-2015) of data; thus, this investigation utilized relatively short-term data. The database did not provide information regarding mortality; therefore, we could not examine survival rates.

In conclusion, this is the first report describing the incidence and clinical outcomes of BA in Korea using the national health insurance system database. In Korea, BA incidence is relatively higher than that in other Western countries but is similar to that in other East Asian countries and shows statistically significant female dominance and seasonal clustering.

## REFERENCES

1. Yoon PW, Bresee JS, Olney RS, James LM, Khoury MJ. *Epidemiology of biliary atresia: a population-based study. Pediatrics* 1997; 99: 376-82.
2. Chardot C, Carton M, Spire-Bendelac N, Le Pommelet C, Golmard JL, Auvert B. *Epidemiology of biliary atresia in France: a national study 1986-96. J Hepatol* 1999; 31: 1006-13.
3. McKiernan PJ, Baker AJ, Kelly DA. *The frequency and outcome of biliary atresia in the UK and Ireland. Lancet* 2000; 355: 25-9.
4. Perlmutter DH, Shepherd RW. *Extrahepatic biliary atresia: a disease or a phenotype? Hepatology* 2002; 35: 1297-304.
5. Caton AR, Druschel CM, McNutt LA. *The epidemiology of extrahepatic biliary atresia in New York State, 1983-98. Paediatr Perinat Epidemiol* 2004; 18: 97-105.
6. Wada H, Muraji T, Yokoi A, Okamoto T, Sato S, Takamizawa S, Tsugawa J, Nishijima E. *Insignificant seasonal and geographical variation in incidence of biliary atresia in Japan: a regional survey of over 20 years. J Pediatr Surg* 2007; 42: 2090-2.
7. Tiao MM, Tsai SS, Kuo HW, Chen CL, Yang CY. *Epidemiological features of biliary atresia in Taiwan, a national study 1996-2003. J Gastroenterol Hepatol* 2008; 23: 62-6.
8. Grizelj R, Vukovic J, Novak M, Batinica S. *Biliary atresia: the Croatian experience 1992-2006. Eur J Pediatr* 2010; 169: 1529-34.
9. Lin YC, Chang MH, Liao SF, Wu JF, Ni YH, Tiao MM, Lai MW, Lee HC, Lin CC, Wu TC, Lau BH, Tsai TC, Yang YJ, Chen AC, Shih HH, Lee IH, Lee WC, Chen HL, Hsu HY, Chiou ST. *Decreasing rate of biliary atresia in Taiwan: a survey, 2004-2009.*

- Pediatrics* 2011; 128: e530-6.
10. Utterson EC, Shepherd RW, Sokol RJ, Bucuvalas J, Magee JC, McDiarmid SV, Anand R. *Biliary atresia: clinical profiles, risk factors, and outcomes of 755 patients listed for liver transplantation. J Pediatr* 2005; 147: 180-5.
  11. McDiarmid SV, Anand R, Lindblad AS. *Studies of Pediatric Liver Transplantation: 2002 update. An overview of demographics, indications, timing, and immunosuppressive practices in pediatric liver transplantation in the United States and Canada. Pediatr Transplant* 2004; 8: 284-94.
  12. Byun J, Yi NJ, Lee JM, Suh SW, Yoo T, Choi Y, Ko JS, Seo JK, Kim H, Lee HW, Kim HY, Lee KW, Jung SE, Lee SC, Park KW, Suh KS. *Long term outcomes of pediatric liver transplantation according to age. J Korean Med Sci* 2014; 29: 320-7.
  13. Balistreri WF, Grand R, Hoofnagle JH, Suchy FJ, Ryckman FC, Perlmutter DH, Sokol RJ. *Biliary atresia: current concepts and research directions. Summary of a symposium. Hepatology* 1996; 23: 1682-92.
  14. Davenport M. *Biliary atresia: outcome and management. Indian J Pediatr* 2006; 73: 825-8.
  15. Nio M, Ohi R, Miyano T, Saeki M, Shiraki K, Tanaka K. *Five- and 10-year survival rates after surgery for biliary atresia: a report from the Japanese Biliary Atresia Registry. J Pediatr Surg* 2003; 38: 997-1000.
  16. Houwen RH, Kerremans, II, van Steensel-Moll HA, van Romunde LK, Bijleveld CM, Schweizer P. *Time-space distribution of extrahepatic biliary atresia in The Netherlands and West Germany. Z Kinderchir* 1988; 43: 68-71.
  17. Nakamizo M, Toyabe S, Kubota M, Komata O, Suzuki H, Akazawa K. *Seasonality in the incidence of biliary atresia in Japan. Acta Paediatr* 2006; 95: 509-10.

18. Wildhaber BE, Majno P, Mayr J, Zachariou Z, Hohlfeld J, Schwoebel M, Kistler W, Meuli M, Le Coultre C, Mentha G, Belli D, Chardot C. *Biliary atresia: Swiss national study, 1994-2004. J Pediatr Gastroenterol Nutr* 2008; 46: 299-307.
19. Livesey E, Cortina Borja M, Sharif K, Alizai N, McClean P, Kelly D, Hadzic N, Davenport M. *Epidemiology of biliary atresia in England and Wales (1999-2006). Arch Dis Child Fetal Neonatal Ed* 2009; 94: F451-5.
20. Guttman OR, Roberts EA, Schreiber RA, Barker CC, Ng VL. *Biliary atresia with associated structural malformations in Canadian infants. Liver Int* 2011; 31: 1485-93.
21. Jimenez-Rivera C, Jolin-Dahel KS, Fortinsky KJ, Gozdyra P, Benchimol EI. *International incidence and outcomes of biliary atresia. J Pediatr Gastroenterol Nutr* 2013; 56: 344-54.
22. Tayler R, Barclay AR, Rogers P, McIntyre K, Russell RK, Devadason D, Bisset WM, Ling SC, McGrogan P. *Scottish outcomes for extra hepatic biliary atresia post-rationalisation of services. Arch Dis Child* 2013; 98: 381-3.
23. Smith BM, Laberge JM, Schreiber R, Weber AM, Blanchard H. *Familial biliary atresia in three siblings including twins. J Pediatr Surg* 1991; 26: 1331-3.
24. Shneider BL, Brown MB, Haber B, Whittington PF, Schwarz K, Squires R, Bezerra J, Shepherd R, Rosenthal P, Hoofnagle JH, Sokol RJ. *A multicenter study of the outcome of biliary atresia in the United States, 1997 to 2000. J Pediatr* 2006; 148: 467-74.
25. Hol L, van den Bos IC, Hussain SM, Zondervan PE, de Man RA. *Hepatocellular carcinoma complicating biliary atresia after Kasai portoenterostomy. Eur J Gastroenterol Hepatol* 2008; 20: 227-31.
26. Hadzic N, Quaglia A, Portmann B, Paramalingam S, Heaton ND, Rela M, Mieli-Vergani G, Davenport M. *Hepatocellular carcinoma in biliary atresia: King's College*



- Hospital experience. J Pediatr* 2011; 159: 617-22 e1.
27. Brunati A, Feruzi Z, Sokal E, Smets F, Fervaille C, Gosseye S, Clapuyt P, de Ville de Goyet J, Reding R. *Early occurrence of hepatocellular carcinoma in biliary atresia treated by liver transplantation. Pediatr Transplant* 2007; 11: 117-9.
  28. Shirouzu Y, Kasahara M, Takada Y, Taira K, Sakamoto S, Uryuhara K, Ogawa K, Doi H, Egawa H, Tanaka K. *Development of pulmonary hypertension in 5 patients after pediatric living-donor liver transplantation: de novo or secondary? Liver Transpl* 2006; 12: 870-5.
  29. Soh H, Hasegawa T, Sasaki T, Azuma T, Okada A, Mushiake S, Kogaki S, Matsushita T, Harada T. *Pulmonary hypertension associated with postoperative biliary atresia: report of two cases. J Pediatr Surg* 1999; 34: 1779-81.
  30. Kim JM, Kim KM, Yi NJ, Choe YH, Kim MS, Suh KS, Kim SI, Lee SK, Lee SG. *Pediatric liver transplantation outcomes in Korea. J Korean Med Sci* 2013; 28: 42-7.

## 국문 초록

**서론:** 담도폐쇄증은 담즙정체의 주요 원인이자 간이식의 주요 적응증이 되는 질환이지만 현재까지 한국의 담도폐쇄증의 발병률은 보고되지 않았다. 본 연구의 목적은 한국의 담도폐쇄증의 발병률과 임상결과에 대하여 조사하는 것이다.

**방법:** 한국의 범국민 건강보험 데이터베이스를 이용하여 2011년부터 2015년까지 담도폐쇄증으로 입원한 18세 이하 환자의 데이터를 추출하였다.

**결과:** 총 240명의 담도폐쇄증 환자가 새롭게 진단 되었다. 총 963명의 18세 이하의 담도폐쇄증 환자가 5년 동안 추적 관찰을 받았다. 담도폐쇄증의 발병률은 10,000명 출생당 1.06건으로 확인되었다. 여성에서의 발병률은 남성보다 1.4배 높았다. 또한 통계적으로 유의한 계절별 차이도 확인되었는데 6월부터 8월(여름)까지의 발병률이 12월부터 2월(겨울)까지의 발병률보다 2배 높았다. 선천성기형은 240명의 담도폐쇄증 환자 중 38명(15.8%)에서 확인되었다. 선천성 심기형이 주요 관련 선천성기형(5.8%) 이었다. 담도염(24%), 정맥류(6.2%), 위장관 출혈(4.4%) 등의 여러 합병증들이 발생하였다. 18세 이전의 담도폐쇄증 환자 963명 중 301명(31.3%)이 간이식을 시행 받

았다.

**결론:** 한국의 담도폐쇄증의 발병률은 서양의 발병률보다 높다.

본 연구에서는 통계적으로 유의한 성별 및 계절별 발병률의 차이를 보고한다.

**주요어:** 담도폐쇄증, 발병률, 간이식

**학 번:** 2015-22252