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DUODENAL ATRESIA IN FINLAND FROM 2004 to 2017: PREVALENCE, MORTALITY AND ASSOCIATED ANOMALIES A population-based study

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Abstract:

Introduction

Duodenal atresia (DA) is the most common atresia of the small bowel. This study aims to assess the prevalence, mortality, and associated anomalies related to DA in Finland during 2004-2017.

Material and Methods

A Nationwide study based on registers maintained by the Finnish Institute for Health and Welfare and Statistics Finland containing data on all live births and stillbirths and terminations of pregnancy. The cases were identified based on the ICD-9 and 10 codes. Associated anomalies were classified based on the EUROCAT criteria; minor anomalies were excluded.

Results

There were 249 DA cases including 222 (89.2%) live births, 16 (6.4%) stillbirths, and 11 (4.4%) terminations. There was no significant change in the prevalence rates between 2004 and 2017. Live birth prevalence was 2.75/10000 and total prevalence 3.08/10000 births. One hundred (40.2%) cases were isolated, 67 (26.9%) had other major congenital anomalies and 83 (33.3%) were syndromic. There were no terminations in isolated DA. Most associated anomalies were cardiac (36.1%), followed by other gastrointestinal tract anomalies (23.7%) and limb deformities/defects (7.2%). Trisomy 21 was observed in 63 cases (25.3%). Neonatal mortality was 3.6% (n=8) and at one year 95.0% were alive. Both neonatal and infant mortality were associated with cardiac anomalies ($p<0.001$ and $p=0.001$ respectively). All neonatal deaths had associated cardiac defect(s).

Conclusions

The prevalence of DA in Finland remains stable and among the highest reported. DA is often associated with cardiac anomalies which portend high risk for mortality. Despite the burden of associated anomalies, overall survival is high.

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DUODENAL ATRESIA IN FINLAND FROM 2004 to 2017: PREVALENCE, MORTALITY AND ASSOCIATED ANOMALIES

A population-based study

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cardiac (36.1%), followed by other gastrointestinal tract anomalies (23.7%) and limb deformities/defects (7.2%). Trisomy 21 was observed in 63 cases (25.3%). Neonatal mortality was 3.6% (n=8) and at one year 95.0% were alive. Both neonatal and infant mortality were associated with cardiac anomalies (p<0.001 and p=0.001 respectively). All neonatal deaths had associated cardiac defect(s).

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The prevalence of DA in Finland remains stable and among the highest reported. DA is often associated with cardiac anomalies which portend high risk for mortality. Despite the burden of associated anomalies, overall survival is high.

Keywords: duodenal atresia, mortality, prevalence, associated anomalies

Introduction

Duodenal atresia and stenosis (DA) is characterized as a complete or partial obstruction of duodenum.[1,2] The prevalence of DA ranges from 0.7 to 2 per 10 000 births with regional differences occurring.[3-5] The prevalence of other small intestinal atresias reaches only 0.8/10 000 making DA the most common small bowel atresia.[5] Pathophysiology differs between small intestinal atresias. The most accredited theory for DA is the failure of recanalization during 8th to 10th gestational weeks.[4,5] For other SIAs, the etiology is thought to be vascular.[1,5]

DA is characterized as a continuum from mucosal web within an otherwise normal muscular wall to the total atresia of duodenum and duodenal mesenterium.[1,2,6] Location of the atresia is usually (70-85%) distal of papilla Vater, causing the main findings, which are “double bubble” and polyhydramnios in prenatal ultrasound as well as bilious vomits after birth.[3,6] Half of the children are diagnosed after birth as prenatal ultrasound detects approximately 44 to 55 % of the cases, sensitivity increasing with gestational weeks.[3,6-8]

Associated anomalies are seen in about 50% of the cases.[4,6] Trisomy 21 is diagnosed in approximately 30% of the DA cases, but reported prevalence varies from 16 to 46%.[5,8-10] From congenital structural anomalies, cardiac defects are the most common as represented approximately in 30% of the cases, and together with trisomy 21, the rate rises up to 60%.[11] Other co-occurring anomalies include other gastrointestinal anomalies like esophageal atresia or anorectal malformation as well as genitourinary malformations.[1,8] Associated intestinal atresias as well as biliary duct anomalies are rare.[4,6]

Nearly half of DA cases are born slightly preterm.[1,9,10] As the operative techniques and management for neonates and preterm infants have developed, survival rates up to 96% have

been reported.[8,12,13] Overall, 87% of the pregnancies with DA, have been reported to result in live births with induced abortion and spontaneous fetal loss rates both being around 6%.[5]

This nationwide study aims to investigate the prevalence, survival as well as examine other associated congenital malformations related to DA. We hypothesize that survival of newborns with DA is high, and mortality is associated with other co-morbidities and anomalies.

Materials and Methods

The data for this study were collected from the national, population-based registries which contain data on all live births, stillbirths and terminations of pregnancy due to fetal anomalies (TOPFA) in Finland. The data from the Finnish Register of Congenital Malformations, the Register of Induced Abortions, and the Medical Birth Register and Cause of Death Register were utilized for this study. All these registers are maintained by the THL Finnish Institute for Health and Welfare with the exception of Cause of Death Register, collected by Statistics Finland.

Notification of Malformation -forms received from the maternity and pediatric hospitals were used for data collection. The Finnish Register of Congenital Malformations also identifies cases with malformation code from other registers, and medical records are collected from the hospitals to confirm the final diagnoses. All the data collected were double-checked by a medical geneticist and only after that, the collected data with possible additional information (patient records etc.) was entered to the registers.

The International Classification of Diseases and Health Related Problems (ICD) revisions nine and ten (ICD-9 Atlanta modification, ICD-10) by the World Health Organization were used to code the diagnosis for DA and as well as for adjacent comorbidities and anomalies. Used ICD-10 codes were Q40.9, Q41, Q41.9, Q43.8 and Q45, and for ICD-9 751100 and 751560. Confirmation of the diagnosis was made with the written diagnosis, as recorded in the Finnish Register of Congenital Malformations. All cases born during a time period from January 1st 2004 to December 31st 2017 were included to the study. DA cases were categorized as isolated in cases with no other major congenital malformations, as cases with multiple congenital anomalies (MCA) in the presence of other major anomalies, or syndromic (SDR). EUROCAT guidelines were used for the classification of associated anomalies, syndromes and chromosomal abnormalities. These guides were also used for the exclusion of minor anomalies.[14,15]

Chi-square test or Fisher's exact test were utilized for categorical variables. The change in prevalence rates during the study period was evaluated with linear regression. All statistical tests were performed as two-sided, with a significance level set at $p<0.05$. The analyses were performed using SAS System, version 9.4 for Windows (SAS Institute Inc., Cary, North Carolina, United States). Birth, live birth, and total prevalence rates are given per 10.000, as defined by EUROCAT.[16]

Results

This register study identified 249 DA cases in Finland, including 222 (89.2%) live births, 16 (6.4%) stillbirths, and 11 (4.4%) terminations of pregnancy for fetal anomaly. The majority (58.9%) of all births were males (140 males, 98 females). The sex was not reported in TOPFA cases. The total prevalence was 3.08/10 000, live birth prevalence 2.75/10 000 and birth prevalence 2.95/10 000. The overall prevalence trend was very slightly decreasing, but statistically insignificantly, p=0.76 (Figure 1). The neonatal mortality was 3.6% (8 deaths) and at 1-year 211 (95%) were alive. Both neonatal and infant mortality were associated with cardiac anomalies (p<0.001 and p=0.001), respectively (Table 1). Neonatal deaths were observed in cases with Tetralogy of Fallot (n=3), anomalous pulmonary venous return (n=2), one case with ventricular septal defect (VSD), overriding aorta and right-sided aortic arch, and one case with pulmonary valve atresia and overriding aorta. There was only one case with isolated VSD, but that individual had other major congenital anomalies including pulmonary hypoplasia, esophageal atresia, anorectal malformation and sirenomelia. There were no neonatal deaths in those without cardiac anomalies.

There were 100 (40.2%) isolated cases, 66 (26.9%) had other major congenital anomalies (MCA), and 83 (33.3%) were syndromic (SDR). Heart defects were present in 90 (36.1%) cases, followed by other gastrointestinal tract anomalies in 59 (23.7%) and limb deformities/defects in 18 (7.2%) cases (Table 2). Among co-occurring gastrointestinal anomalies malrotation was the most common (n=30), followed by anorectal malformations (n=16) and esophageal atresias (n=12). In addition, there were four biliary atresias and four other small intestinal atresias and three patients had Hirschsprung disease. Horseshoe kidney was the most common urinary tract anomaly (n=6), followed by agenesis of kidney (n=4), ureteral obstruct anomalies (n=3) and polycystic kidneys (n=1). One case of bladder exstrophy and one case of double system were also present.

Chromosomal abnormalities were found in 77 (30.9%) cases of which Trisomy 21 was the most common and observed in 63 (25.3%) cases. Other diagnosed syndromes included Trisomy 18 (n=2), Williams syndrome (n=2), different gene translocations (n=4), and one case of each of the following syndromes: Sotos, Cornelia de Lange, Holt-Oram and CATCH 22. VACTERL-association (presence of at least three of the following malformations: vertebral defects, anorectal malformation, cardiac defects, tracheoesophageal fistula, renal anomalies, limb abnormalities) was present in 8 (3.2%) cases.

The mean maternal age was 30.1 years (SD 6.13). The mean gestational age at birth was 36.8 weeks (standard deviation (SD) 3.1 weeks). The mean birth weight was 2673 g (SD 794, range 360 – 4480 g). TOPFA rate among all cases was 4.4% (n=11) but in presence of MCA or SDR it rose to 10.9% and 5.3%, respectively. In isolated cases there were no terminations (Table 3).

Discussion

Based on this population-based study duodenal atresia carries a high risk of morbidity especially in terms of co-occurring congenital heart anomalies. Despite this burden, the overall survival is high (95%) due to developed care management. Still, mortality is clearly linked to congenital heart anomalies.

This register-based study comprising 249 DA cases from the years 2004-2017 observed a national total prevalence of 3.08/10 000 which is one of the highest reported. This finding may reflect our ability to identify not only live births, but also stillbirths and terminated cases. In previous EUROCAT studies, Morris et al. [17] observed a potentially increasing trend whereas Best et al.[5] found only regional differences in total prevalence which was 0.9/10 000 in both papers. The number for TOPFA 11 (4.4 %) in the current study was in line with a previous EUROCAT study where 6.2% of the families opted for the termination of pregnancy.[5] Nevertheless, this frequency is significantly lower than for gastroschisis in Finland[18] and isolated DA does not result to termination in Finland.

Previous prevalence rate for DA in Finland was reported 1.4/10 000 by Kyyrönen and Hemminki from the 1970s suggesting a slightly increasing prevalence. They reported mean maternal age of 28.7 years as compared to 30.1 years in the current study.[19] According to the Finnish Register of Congenital Malformations, the incidence of Trisomy 21 has risen from 1993 to 2013.[20,21] Therefore, we speculate the higher prevalence of DA reported here could be explained by increasing maternal age and simultaneously increasing risk of Trisomy 21.

DA is known to have a clear association with cardiac defects. We also found most structural anomalies to be cardiac. The rate in this study was 36% which is in line with Pijpers et al., who report cardiac defects in nearly one third of DA patients,[9,11] although rates as low as 12% in DA have previously been reported.[5] This apparent increase is likely due to the improvements in echocardiography technology.

Overall survival among DA patients is already very high, in developed countries over 90%.[3] In line with this, we observed a one-year survival of 95%. We found the presence of the heart defect to be a significant risk factor for both neonatal and infant mortality. During the neonatal period, there were eight deaths, mostly among those with complex cardiac anomalies, including Tetralogy of Fallot and anomalous pulmonary venous return and only one isolated ventricular septal defect. At the one-year time point nine out of 11 deaths had cardiac defect(s), $p<0.001$ for both. Grosfeld et al.[6] and Escobar et al.[10] reported deaths in DA with heart anomalies. In the study by Escobar et al. there were five early deaths (< 30 days postoperatively) all of them related to complex cardiac anomalies. Five out of ten late deaths (from 3 months to 14 years) were related to complex cardiac anomalies. Cardiac anomalies were present in 27% of their study population. In 1993 Grosfeld et

al. reported operative mortality of 5% and this was associated with cardiac defects, overall mortality rates were not reported. In the long term, the mortality was still mainly associated with cardiac problems.

As Keckler et al.[22] reported, congenital heart defects are a heterogenous group. Interestingly, these authorities found an increased incidence of cyanotic heart defects in DA patients without Trisomy 21. Unfortunately, they did not report mortality associated with these comorbidities. The incidence of cardiac anomalies in Trisomy 21 alone has been reported to be 60%, and in case of DA + Trisomy 21 the pooled percentage was calculated to be 51%.[9] Brantberg et al.[23] hypothesized that the reason behind the increased pre and postnatal mortality in DA to be bradycardia caused by vasovagal over-reactivity. This also occurred in infants with isolated DA and normal karyotype, so no relation to Trisomy 21 was observed. Mortality related to heart defect(s) was not examined. In general, according to the French study, infant mortality due to congenital heart diseases was 6.4%, for VSD excluded it was 8.5%, and 60% of the deaths occurred during the neonatal period. However, in their study, other concomitant congenital anomalies or syndromes were neither reported nor analyzed.[24]

Strengths and Limitations

This retrospective study is based on national registries with a demonstrably high quality of validated data and a total population coverage also including terminations due to fetal anomalies.[25-27] Thus, it gives a broad and an up to date view of phenomenon of interest. The limitations still include the limited number of cases. Also, this study relies only on this retrospective register data, but the diagnoses were confirmed by a medical geneticist. The different types of duodenal atresias could not be classified on the basis of the register data. Chromosomal abnormalities were observed in 31% of the cases. The total number of patients undergoing testing for genetic abnormalities remains unknown. However, this study is among the most extensive regarding case numbers among published reports on DA.

In conclusion, the prevalence of duodenal atresia in Finland is high with no significant change over study period, although clearly higher than in the to data from the 1970s. DA carries a high risk of cardiac anomalies which are a significant risk factor for both neonatal and infant mortality. Nevertheless, the overall survival is as high as 95%.

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Figure 1. Live birth prevalence of duodenal atresia

Table 1. Heart defect associated mortality in duodenal atresia. P-values for neonatal and infant mortalities $p<0.0001$ and $p=0.001$, respectively.

Table 2. Incidence of individual concurrent anomalies in 249 duodenal atresia cases organized by affected organ systems. GI=gastrointestinal, AWD=abdominal wall defects, VACTERL=vertebral, anal, cardiac, trachea-esophageal, renal, limb.

Table 3. Distribution of duodenal atresias in presence of other comorbidities among birth status and in case of termination of pregnancy for fetal anomaly (TOPFA). MCA=major congenital anomaly, SDR=syndromes

	Neonatal mortality n (%)		Infant mortality n (%)	
No heart defect (n=142)	0	p< 0.0001	2 (1.4%)	p=0.001
Heart defect (n=80)	8 (10.0%)		9 (11.3%)	
Total (n=222)	8 (3.6%)		11 (5.0%)	

Table 1. Heart defect associated mortality in duodenal atresia. P-values for neonatal and infant mortalities p<0.0001 and p=0.001, respectively.

Isolated % (n)	Nervous system % (n)	Heart % (n)	Cleft % (n)	Eye % (n)	Ear, face, and neck % (n)	GI % (n)	Respiratory % (n)	Urinary % (n)	Genital % (n)	AWD % (n)	Limb % (n)	
40.16 (100)	2.41 (6)	36.14 (90)	1.20 (3)	1.61 (4)	0.41 (1)	23.69 (59)	2.41 (6)	6.83 (17)	2.41 (6)	0,80 (2)	7.23 (18)	

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	Isolated % (n)	MCA % (n)	SDR % (n)
Live birth, n=222 (89.2%)	42.8 (95)	25.2 (56)	32.0 (71)
Stillbirth, n=16 (6.4%)	31.3 (5)	18.8 (3)	50.0 (8)
Terminated, n=11 (4.4%)	- (0)	63.6 (7)	36.4 (4)
All, n=249	40.2 (100)	26.9 (67)	33.3 (83)

Table 3. Distribution of duodenal atresias in presence of other comorbidities among birth status and in case of termination of pregnancy for fetal anomaly (TOPFA). MCA=major congenital anomaly, SDR=syndromes