

Epidemiology of small intestinal atresia in Europe: a register-based study

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SUMMARY

Background The epidemiology of congenital small intestinal atresia (SIA) has not been well studied. This study describes the presence of additional anomalies, pregnancy outcomes, total prevalence and association with maternal age in SIA cases in Europe.

Methods Cases of SIA delivered during January 1990 to December 2006 notified to 20 EUROCAT registers formed the population-based case series. Prevalence over time was estimated using multilevel Poisson regression, and heterogeneity between registers was evaluated from the random component of the intercept.

Results In total 1133 SIA cases were reported among 5126, 164 registered births. Of 1044 singleton cases, 215 (20.6%) cases were associated with a chromosomal anomaly. Of 829 singleton SIA cases with normal karyotype, 221 (26.7%) were associated with other structural anomalies. Considering cases with normal karyotype, the total prevalence per 10 000 births was 1.6 (95% CI 1.5 to 1.7) for SIA, 0.9 (95% CI 0.8 to 1.0) for duodenal atresia and 0.7 (95% CI 0.7 to 0.8) for jejunoileal atresia (JIA). There was no significant trend in SIA, duodenal atresia or JIA prevalence over time (RR=1.0, 95% credible interval (CrI): 1.0 to 1.0 for each), but SIA and duodenal atresia prevalence varied by geographical location ($p=0.03$ and $p=0.04$, respectively). There was weak evidence of an increased risk of SIA in mothers aged less than 20 years compared with mothers aged 20 to 29 years (RR=1.3, 95% CrI: 1.0 to 1.8).

Conclusion This study found no evidence of a temporal trend in the prevalence of SIA, duodenal atresia or JIA, although SIA and duodenal atresia prevalence varied significantly between registers.

INTRODUCTION

Small intestinal atresia (SIA) is a congenital anomaly characterised by the abnormal closure, discontinuity or narrowing of the duodenum, jejunum or ileum.¹ Duodenal atresia occurs when recanalisation, which takes place at the end of the second month of the embryonic period, of the bowel is unsuccessful.² Atresia or stenosis of the jejunum or ileum (jejunoileal atresia, JIA) is caused by vascular accidents leading to an obstruction of the blood supply to the small intestine.³ The different aetiologies suggest these subtypes should be considered separately.

What is known about the topic

There are few population-based studies investigating the prevalence of small intestinal atresia. Chromosomal anomalies, particularly trisomy 21, are more commonly associated with duodenal atresia than jejunoileal atresia. There is some evidence to suggest a higher risk of small intestinal atresia in younger mothers.

What this study adds

There was no evidence of an increase in prevalence of small intestinal atresia, duodenal atresia or jejunoileal atresia over time in Europe. Spontaneous fetal loss was higher in pregnancies affected by duodenal atresia than by jejunoileal atresia, and this was not explained by the presence of additional anomalies. There was a suggestion that younger mothers (<20 years) had an increased risk of small intestinal atresia compared with mothers aged 20–29 years.

Few studies have investigated the prevalence of SIA. Of those that have, the reported prevalence rates range from 1.3 to 2.9 per 10 000 live births.^{4,5} Estimates of the prevalence of duodenal atresia range from 0.7 to 1.8 per 10 000 total births.^{6,7} Estimates of the prevalence of JIA, for which there are very few, range from 1 in 330 to 0.8 per 10 000 live births.^{6,8}

Maternal age may explain some of the variation in SIA prevalence, with previous studies suggesting a possible U-shaped relationship^{5,7} or a higher risk in younger mothers.⁹ Although these associations were not significant, it is possible that the studies had insufficient power to investigate this relationship. Gastroschisis, a congenital anomaly of the abdominal wall that may share a similar aetiology to JIA, has an increased prevalence in younger mothers,^{10,11} but whether a similar association exists for JIA is not known. Previous studies have identified increasing gastroschisis rates in the UK,^{12,13} but not for the rest of Europe.^{11,14} Differences in prevalence according to geographical location have not been considered in SIA or SIA subtypes.

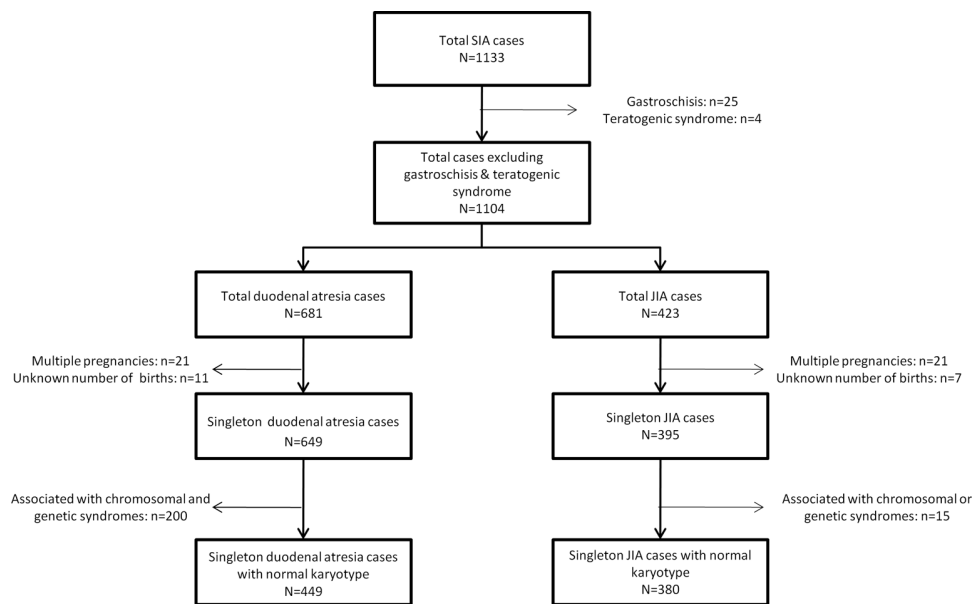


Figure 1 Flow chart showing derivation of study sample. JIA, jejunoileal atresia; SIA, small intestinal atresia.

The aim of this study is to describe the presence of additional anomalies, pregnancy outcome and prevalence of SIA and SIA subtypes in Europe during 1990–2006 using high-quality population-based register data. Prevalence over time and according to geographical location and the influence of maternal age were also investigated.

METHODS

The European Surveillance of Congenital Anomalies (EUROCAT)¹⁵ is a collaborative network of population-based congenital anomaly registers. Forty registers in 20 countries use multiple sources to collect data on anomalies occurring in spontaneous fetal losses ≥ 20 weeks' gestation, terminations of pregnancy for congenital anomaly following prenatal diagnosis and live births. The network surveys about 1.7 million births in Europe, representing almost 31% of the European birth population.¹⁶ Cases are coded using the WHO International Classification of Disease version 9 or 10 (ICD 9 or ICD 10) and exclude minor anomalies; further details of data collection are available on the EUROCAT website.¹⁷

All EUROCAT registers were invited to participate in the study. Cases of SIA with a delivery date between 1 January 1990 and 31 December 2006 notified to the 20 registers that agreed to participate, formed this population-based case series. Denominator and maternal age data were provided by EUROCAT.¹⁷

Inclusion and exclusion criteria

SIA cases included all cases of congenital absence, atresia and stenosis of the duodenum (ICD 10, Q41.0), jejunum (Q41.1) or ileum (Q41.2). Cases occurring in multiple pregnancies or associated with a chromosomal anomaly, teratogenic syndrome or gastroschisis were excluded from the analysis. All other cases were included and were coded as isolated or associated with other anomalies. Cases occurring alone or with malrotation or microcolon were coded as isolated. Cases occurring alongside any other major anomaly, as defined by EUROCAT,¹⁷ were classified as associated. In cases with multiple atresias, the type of atresia was classified as the highest level of obstruction.

Table 1 Most common associated structural anomalies* †

Structural anomaly	Duodenal atresia N (%)‡	JIA N (%)§
Cardiac anomalies	60 (12.3)	25 (6.6)
Ventricular septal defect	22 (4.9)	8 (2.1)
Atrial septal defect	20 (4.5)	12 (3.2)
Atrioventricular septal defect	8 (1.8)	2 (0.5)
Tetralogy of fallot	7 (1.6)	2 (0.5)
Cleft lip with or without palate	6 (1.3)	2 (0.5)
Digestive system (other than SIA)	39 (8.0)	10 (2.6)
Oesophageal atresia with or without fistula	17 (3.8)	1 (0.3)
Anorectal atresia and stenosis	17 (3.8)	4 (1.00)
Atresia of bile ducts	5 (1.1)	1 (0.3)
Urinary system	29 (6.5)	9 (2.4)
Congenital hydronephrosis	5 (1.1)	2 (0.5)
Limb	31 (6.9)	5 (1.3)
Limb reduction	9 (2.0)	1 (0.3)
Upper limb	7 (1.6)	–

*Cases with chromosomal or genetic syndromes, gastroschisis or multiple (or unknown) births were excluded from this analysis.

†Cases with two or more additional anomalies may appear more than once.

‡Percentage of total duodenal atresia cases.

§Percentage of total JIA cases.

JIA, jejunoileal atresia; SIA, small intestinal atresia.

Analysis

Total prevalence rates for SIA, duodenal atresia and JIA in each register were calculated as the number of cases (whether ending in fetal loss, termination of pregnancy for fetal anomaly, or live birth) per 10 000 total births; 95% CIs were derived from the binomial distribution.

Superior estimates for the risk of spontaneous fetal loss, which accounted for terminations of pregnancy using case censorship, were calculated for SIA using survival analysis. Kaplan–Meier curves were fitted to model spontaneous fetal loss according to SIA subtype, and a log-rank test was performed. To adjust for the presence of associated anomalies on survival, a Cox-proportional hazards regression model was fitted.

As the registers survey distinct geographical areas and contributed data for different time periods, a simple analysis of

Table 2 Total number of cases,* births and prevalence of SIA according to SIA subtype, by EUROCAT registry 1990–2006

Register	Time period	Total births	% of mothers <20	SIA		Duodenal atresia		JIA	
				No of cases	Rate per 10 000 births (95% CI)	No of cases	Rate per 10 000 births (95% CI)	No of cases	Rate per 10 000; births (95% CI)
Antwerp	1990–2006	236 873	2.3	35	1.5 (1.0 to 2.1)	22	0.9 (0.6 to 1.4)	13	0.6 (0.3 to 0.9)
Barcelona	1992–2006	196 160	1.8	18	0.9 (0.5 to 1.5)	8	0.4 (0.2 to 0.8)	10	0.5 (0.2 to 0.9)
Dublin	1990–2009	354 403	5.7	46	1.3 (1.0 to 1.7)	22	0.6 (0.4 to 0.9)	24	0.7 (0.4 to 1.0)
Emilia Romagna	1990–2006	471 367	1.5	89	1.9 (1.5 to 2.3)	44	0.9 (0.7 to 1.3)	45	1.0 (0.7 to 1.3)
EMSY† (UK)	1998–2006	549 515	8.8	86	1.6 (1.3 to 1.9)	53	1.0 (0.7 to 1.3)	33	0.6 (0.4 to 0.8)
Malta	1990–2006	771 54	4.6	9	1.2 (0.5 to 2.2)	4	0.5 (0.1 to 1.3)	5	0.7 (0.2 to 1.5)
North of England	2000–2006	214 037	10.7	48	2.2 (1.7 to 3.0)	28	1.3 (0.9 to 1.9)	20	0.9 (0.6 to 1.4)
Odense	1990–2006	955 99	2.1	18	1.9 (1.1 to 3.0)	9	0.9 (0.4 to 1.8)	9	0.9 (0.4 to 1.8)
Paris	1990–2006	619 431	0.9	160	2.6 (2.2 to 3.0)	95	1.6 (1.2 to 1.9)	65	1.1 (0.8 to 1.3)
Ile de la Reunion	2002–2006	73 023	–	16	2.2 (1.3 to 3.6)	5	0.7 (0.2 to 1.6)	11	1.5 (0.8 to 2.7)
South-East Ireland	1997–2006	62 197	5.4	5	0.8 (0.3 to 1.9)	3	0.5 (– to 1.4)	2	0.3 (0.0 to 1.2)
South Portugal	1990–2006	244 661	7.5	24	1.0 (0.6 to 1.5)	7	0.3 (0.1 to 0.6)	17	0.7 (0.4 to 1.1)
Saxony Anhalt	1990–2006	217 140	6.4	30	1.4 (0.9 to 2.0)	19	0.9 (0.5 to 1.4)	11	0.5 (0.3 to 0.9)
Strasbourg	1990–2004	204 328	3.2	21	1.0 (0.6 to 1.6)	13	0.6 (0.3 to 1.1)	8	0.4 (0.2 to 0.8)
Styria	1990–2006	198 781	4.7	20	1.0 (0.6 to 1.6)	9	0.5 (0.2 to 0.9)	11	0.6 (0.3 to 1.0)
Thames Valley	1996–2006	140 203	4.6	19	1.4 (0.8 to 2.1)	13	0.9 (0.5 to 1.6)	6	0.4 (0.2 to 0.9)
Tuscany	1990–2006	413 762	1.3	36	0.9 (0.6 to 1.2)	17	0.4 (0.2 to 0.7)	19	0.5 (0.2 to 0.7)
Vaud	1990–2006	127 534	1.3	11	0.9 (0.4 to 1.5)	4	0.3 (– to 0.8)	7	0.6 (0.2 to 1.1)
Wales	1998–2006	288 877	10.0	57	2.0 (1.5 to 2.6)	27	0.9 (0.6 to 1.4)	30	1.0 (0.7 to 1.5)
Wessex	1994–2006	341 119	6.3	81	2.6 (1.9 to 3.0)	47	1.4 (1.0 to 1.8)	34	1.0 (0.7 to 1.4)
Total	1990–2006	5 126 164	4.6	829	1.6 (1.5 to 1.7)	449	0.9 (0.8 to 1.0)	380	0.7 (0.7 to 0.8)

*Cases with chromosomal or genetic syndromes, gastroschisis or multiple (or unknown) births were excluded from this analysis.

†East Midlands and South Yorkshire Congenital Anomaly Register.

EMSY, East Midlands and South Yorkshire; EUROCAT, The European Surveillance of Congenital Anomalies; JIA, jejunoileal atresia; SIA, small intestinal atresia.

prevalence over time may have underestimated the standard errors and introduced confounding.¹⁸ Therefore, the prevalence of SIA, duodenal atresia and JIA were modelled using a multilevel approach. The number of cases per year were nested within register and modelled by Poisson regression with a random intercept, an offset equal to the log of the expected cases, and year as a continuous predictor. Heterogeneity between registers was evaluated from the random component of the intercept. Inter-regional differences in trends were tested through the incorporation of additional random effects, and variation terms were added to check for compliance with the Poisson distribution, but neither was necessary. A categorical variable was introduced to distinguish between the registers in the UK and the rest of Europe (including Ireland) and identify differences in prevalence. UK data were modelled separately to examine any specific trends.

To investigate associations between maternal age at delivery and SIA prevalence, all models were refitted to include age, which was categorised into three groups: <20, 20 to 29 and ≥30 years. The study period was divided into four groups: 1990 to 1993, 1994 to 1997, 1998 to 2001 and 2002 to 2006. The Ile de la Reunion (France) register was excluded from this analysis due to missing maternal age information, as were data from 1993 to 1994 for South Portugal and 1990 for Saxony-Anhalt (Germany). Interactions between study period and maternal age were explored by adding cross-product terms.

Multilevel model parameters were estimated using a random walk (Metropolis–Hastings) Markov Chain Monte Carlo (MCMC) algorithm. Assuming diffuse uniform priors, the procedure was run for a burn-in sample of 500 observations, and an analysis sample of 1 000 000 thinned by 10 (numbers guided by Raftery–Lewis calculations);¹⁹ 95% credible intervals (CrI) were obtained from the posterior distribution for each parameter.

Statistical analyses were performed using Stata version 11 (for descriptive analysis) and MLwiN 2.14 (for multilevel analysis). $p < 0.05$ was considered statistically significant.

RESULTS

Figure 1 shows the flow of study cases: 1133 cases of SIA were notified to the 20 EUROCAT registers between 1990 and 2006. Four cases were associated with a teratogenic syndrome and 25 with gastroschisis. Of the remaining 1104 cases, 681 had duodenal atresia and 423 had JIA (214 jejunal and 209 ileal). Five cases had multiple atresias and were classed as duodenal atresia. Forty-one twin pregnancies and one triplet pregnancy had SIA including 20 twins and one triplet with duodenal atresia and 21 twins with JIA, including two from the same pregnancy. Plurality data was missing for 18 cases so these were excluded from the analysis.

Associated anomalies

Of the 1044 singleton SIA cases, 215 (20.6%) were associated with a chromosomal or genetic syndrome, including 200 (30.8%) with duodenal atresia and 15 (3.8%) with JIA; 173 (16.6%) had Down syndrome, which was associated with duodenal atresia in 170 (16.3%) cases and JIA in three cases. There were 30 syndromic cases, three each had Patau syndrome, Edward syndrome and triploidy, 10 had other chromosomal anomalies and 13 other syndromes. There were 11 cases with VATER (V-vertebrae anomalies, A-anal atresia, TE-tracheoesophageal fistula, R-renal anomalies) association.

Of the remaining 829 singleton SIA cases with normal karyotype, 221 (26.7%) were associated with other structural anomalies. The prevalence of associated structural anomalies was significantly higher among cases of duodenal atresia ($n=149$ (33.2%)) than cases of JIA ($n=72$ (18.9%)) ($p < 0.001$).

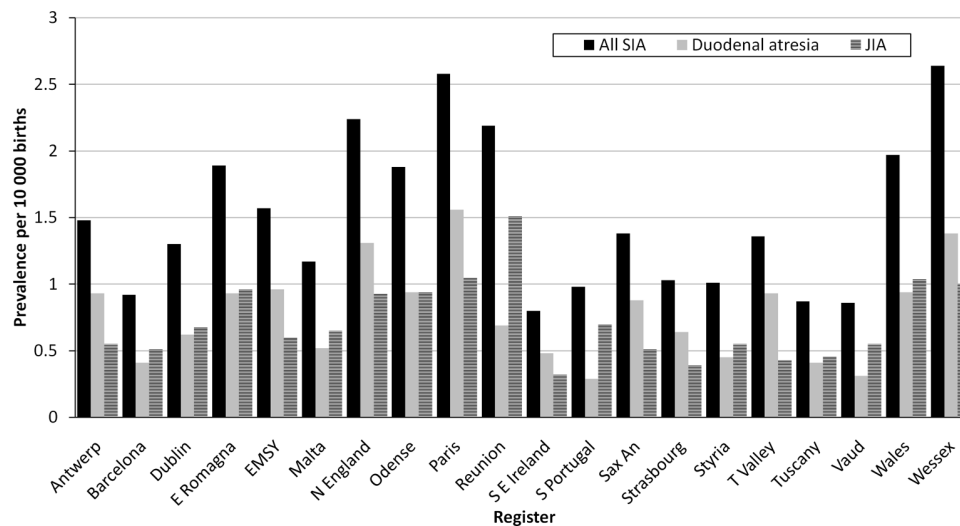


Figure 2 Total prevalence according to register. EMSY, East Midlands and South Yorkshire. JIA, jejunioleal atresia; SIA, small intestinal atresia.

Table 1 shows the most common associated anomalies by SIA subtype. Cardiovascular anomalies were most frequently observed, occurring in 85 (10.3%) cases and were more common in duodenal atresia cases than JIA cases (12.3% and 6.6% respectively, $p=0.004$).

Pregnancy outcome

Of the 829 SIA singleton cases, 754 (91.0%) resulted in live birth, 37 (4.5%) in spontaneous fetal loss (≥ 20 weeks' gestation) and 38 (4.6%) in termination of pregnancy. Of the 449 duodenal atresia cases, 391 (87.1%) resulted in live birth, 30 (6.7%) in spontaneous fetal loss and 28 (6.2%) in termination of pregnancy. Of the 380 JIA cases, 363 (95.5%) resulted in live birth, seven (1.8%) in spontaneous fetal loss and 10 (2.6%) in termination of pregnancy. The total probability of a spontaneous fetal loss (as estimated by survival analysis) was 6.5% in all SIA cases, varying significantly between duodenal atresia (9.7%) and JIA (2.5%) ($p<0.001$). This variation was not explained by differences in the number of associated anomalies ($p=0.11$).

Total prevalence

Table 2 shows the number of cases and total prevalence of SIA and of each subtype by register. Between 1990 and 2006, there were 5 126 164 registered births, giving an overall SIA prevalence of 1.6 (95% CI 1.5 to 1.7) per 10 000 births. The total prevalence of duodenal atresia and JIA was 0.9 (95% CI 0.8 to 1.0) and 0.7 (95% CI 0.7 to 0.8) per 10 000 births, respectively.

SIA prevalence varied significantly between registers ($p=0.03$), from 0.8 (95% CI 0.3 to 1.9) per 10 000 births in South-East Ireland to 2.6 (95% CI 1.9 to 3.0) per 10 000 births in Wessex (UK) (figure 2). Duodenal atresia prevalence also varied significantly between regions ($p=0.04$), ranging from 0.3 (95% CI 0.3 to 0.6) per 10 000 in South Portugal to 1.6 (95% CI 1.2 to 1.9) in Paris (figure 2). JIA prevalence ranged from 0.3 (95% CI <0.1 to 1.2) per 10 000 births in South-East Ireland to 1.1 (95% CI 0.8 to 1.3) per 10 000 births in Paris (France), but the overall difference between regions was not statistically significant ($p=0.16$). There was a borderline significant increased risk of duodenal atresia in the UK compared with the rest of Europe (RR=1.6, 95% CrI: 1.0 to 2.3, $p=0.08$). This relationship was not apparent for SIA or JIA (RR=1.3, 95% CrI: 0.9 to 1.9, $p=0.10$ and RR=1.16, 95% CrI: 0.8 to 1.6; $p=0.39$, respectively).

The multilevel analysis found no significant change in the registry-adjusted prevalence of SIA (RR per year=1.0, 95% CrI: 1.0 to 1.0; $p=0.94$), duodenal atresia (RR=1.0, 95% CrI: 1.0 to 1.0; $p=0.47$) or JIA (RR=1.0, 95% CrI: 1.0 to 1.0; $p=0.63$) over the study period. There was no evidence that the trend in the prevalence of SIA, duodenal atresia or JIA, was significantly different between registers ($p=0.67$, $p=1.00$ and $p=0.35$ respectively). Considering the UK data only, SIA, duodenal atresia and JIA prevalence did not increase over time (RR=1.0, 95% CrI: 1.0 to 1.0, $p=0.94$; RR=1.0, 95% CrI: 1.0 to 1.1, $p=0.48$; RR=1.0, 95% CrI: 0.9 to 1.0, $p=0.63$).

Maternal age

The percentage of mothers aged less than 20 years varied significantly between registers ($p<0.001$), ranging from 0.9% in Paris (France) to 10.7% in the North of England (table 2). The proportion of births to mothers less than 20 years also increased over time ($p<0.001$), from 3.8% in 1990 to 5.1% in 2006. Maternal age less than 20 years was borderline significantly associated with an increased risk of SIA (RR=1.3, 95% CrI 1.0 to 1.8, $p=0.08$), but was not associated with duodenal atresia (RR=1.2, 95% CrI 0.8 to 1.9, $p=0.35$) or JIA (RR=1.4, 95% CrI 0.9 to 2.2, $p=0.12$) (table 3). Adjusting for register and year of delivery did not change these associations (table 3). Considering the UK register data only, maternal age less than 20 years, adjusted for register and year of delivery, was not associated with SIA, duodenal atresia or JIA prevalence ($p=0.62$, $p=0.57$ and $p=0.19$, respectively).

DISCUSSION

This study is the largest to examine the epidemiology of SIA combined, or by SIA subtype. Using data from 20 population-based European registers over a 17-year period, we found a total prevalence of 1.6 per 10 000 births for SIA, 0.9 per 10 000 for duodenal atresia and 0.7 per 10 000 for JIA. There was no evidence that the prevalence of SIA, or either subtype, had altered over the study period. However, the prevalence of SIA and duodenal atresia, but not JIA, varied by geographical location. Furthermore, there was weak evidence that the prevalence of duodenal atresia was greater in the registers in the UK than in those in the rest of Europe and that SIA prevalence was greater in mothers aged less than 20 years. There was no evidence that SIA prevalence was greater in mothers aged less

Table 3 RR for SIA and SIA subtype by maternal age, adjusted for register and year*

Maternal age (years)	Unadjusted RR (95% CrI)			Adjusted RR (95% CrI)		
	SIA	Duodenal atresia	JIA	SIA	Duodenal atresia	JIA
<20	1.3 (1.0 to 1.8); p=0.08	1.2 (0.8 to 1.9); p=0.35	1.4 (0.9 to 2.2); p=0.12	1.3 (1.0 to 1.8); p=0.09	1.2 (0.8 to 1.8); p=0.39	1.4 (0.9 to 2.1); p=0.12
20–29	1 (reference)	1 (reference)	1 (reference)	1 (reference)	1 (reference)	1 (reference)
≥30	0.9 (0.8 to 1.1); p=0.46	1.0 (0.8 to 1.2); p=0.83	0.9 (0.7 to 1.1); p=0.38	0.9 (0.8 to 1.1); p=0.33	1.0 (0.8 to 1.2); p=0.67	0.9 (0.7 to 1.1); p=0.34

*Cases with chromosomal or genetic syndromes, gastroschisis or multiple (or unknown) births were excluded from this analysis.

CrI, credible interval; JIA, jejunoileal atresia; SIA, small intestinal atresia.

than 20 years. Cases of duodenal atresia were more likely to have associated anomalies than JIA, with a quarter of all duodenal atresia cases being associated with Down syndrome. Pregnancies associated with duodenal atresia were less likely to result in live birth than those associated with JIA.

The primary strength of this study is that it is based on data derived from established, high-quality, population-based congenital anomaly registers. Standard methods of identifying cases across all registers and the use of multiple sources of notifications ensure high case ascertainment.

Multilevel methods provide more accurate standard error estimates for nested data than classical approaches. Furthermore, this method eliminates the potential for confounding due to registers contributing data from different time points.¹⁸

However, the study also has some limitations. The variation between registers could be partially explained by differences in case ascertainment. For example, the register in the North of England is long established (since 1985) which could partially explain its greater prevalence. However, geographical variation in prevalence may reflect a true difference, for example in exposures between regions. Our study did not have access to routine data on potential exposures such as smoking status or ethnicity, and there is some evidence that these are associated with SIA prevalence.^{7 20 21} In addition, this study could not investigate possible geographical differences in the prevalence of infertility and infertility treatment as none of the UK registers are legally allowed to hold this information, and thus a comparison with data from continental Europe, was not possible.

Excluding cases associated with chromosomal anomalies, the total prevalence of SIA was 1.7 per 10 000 births, which is lower than that reported in several studies from the USA where prevalence varied between 2.8 and 3.19 per 10 000 live births.^{5 20 22}

Several studies found no trend in the prevalence of SIA, duodenal atresia or JIA over time^{5 6 20} which is consistent with our findings.

We found a suggestion of an increased occurrence of SIA among mothers aged less than 20 years. Fracannet and Roberts reported an increased risk of SIA in mothers under 20 years, but gastroschisis cases were included.⁹ Husain *et al* identified a significant U-shaped relationship with maternal age and SIA prevalence,²² but they included cases associated with chromosomal anomalies, which are more common in older mothers. Cragan *et al* found no evidence of a maternal age relationship with SIA prevalence,²⁰ but this was a small study investigating only 176 cases.

A quarter of the duodenal atresia cases occurred in children with Down syndrome, confirming this well-known association. Other studies have found associations in 17% to 27.5% of cases.^{5 7 20 23} We also found that those children with duodenal atresia and normal karyotype were at substantially greater risk of having additional structural anomalies compared with those with JIA, concurring with previous reports.^{6 20}

Pregnancies affected by duodenal atresia were significantly less likely to result in a live birth compared with those affected by JIA. This observation was previously reported by Hemming and Rankin in their study from the North of England.⁶ Interestingly, our study also indicates that this difference is not related to the increased proportion of associated anomalies in cases of duodenal atresia. It is possible that cases of duodenal atresia are associated with more serious anomalies than cases of JIA.

Although the exact pathogenesis of gastroschisis is not known,²⁴ gastroschisis and JIA may share a similar aetiology. There is increasing evidence to suggest that gastroschisis prevalence is rising in the UK^{12 13} but not in Europe.²⁵ We did not find an increase in JIA prevalence over time in the UK registers, but this may be due to the smaller number of JIA cases available to the study.

Further research is required to confirm the geographical differences in SIA prevalence, particularly the apparent higher prevalence of duodenal atresia in the UK compared with continental Europe, and to investigate possible reasons for this variation.

CONCLUSION

This large population-based study found no evidence of an increase in the prevalence of SIA, duodenal atresia or JIA prevalence over time. However, SIA and duodenal atresia rates differed according to geographical location, a finding which requires further investigation. A greater proportion of duodenal atresia cases were associated with other congenital anomalies and fewer resulted in a live birth compared with cases with JIA.

Correction notice This article has been corrected since it was published Online First. The last 4 authors were not in the correct order and this has now been rectified.

Contributors KEB undertook the analysis and interpretation of the data, and drafted the manuscript. JR conceived the project and, with PWGT, participated in the analysis and interpretation of data and critically reviewed the manuscript. MCA, FB, PB, EC, CD, BD, ED, EG, MG, RG, MH, BK, BM, CM, VN, HR, AR, JS, DT and DW, were involved in the data collection and the critical review of the manuscript. All authors read and approved the final version of the report before submission.

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