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The Epidemiology of Conjoined Twins in Latin America

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Abstract. Twenty-three cases of symmetrical conjoined twins were registered by the Latin-American Collaborative Study of Congenital Malformations (ECLAMC) in 1,714,952 births, which were observed during the 1967-1986 period in 95 maternity hospitals distributed in eleven Latin-American countries. This results in a birth prevalence rate of about 1/75,000 births. The secular and geographic distribution of this material do not depart from random in spite of one hospital with three cases, and two hospitals with two cases each, within a short time period. These 23 cases include one diprosopus, 3 dicephalus, one ischiopagus, 5 pygopagus, none dipygus, 3 syncephalus, none craniopagus, 9 thoracopagus, one omphalopagus, and none rachipagus. Sex distribution is even, with 12 male and 11 female cases.

Key words: Conjoined twins, Collaborative study, Congenital malformations

INTRODUCTION

Conjoined twinning is a very rare event, the epidemiology of which is poorly understood since most publications deal with the anatomical and physiological description of isolated cases.

Its prevalence rate at birth has been estimated around one in 75,000 births [5], that is, as infrequent as sirenomelia, cyclopia, or true phocomelia. Therefore, just a couple of cases occurring within a given discrete geographic area or time interval may easily be taken as an epidemic even when difficult to prove because the rareness of the event demands enormous birth sample sizes.

Table 1 - Observed number of births, and observed/expect-ed numbers of conjoined twins by year and country of bir

Year	Argentina	Bolivia	Brazil	Chile	Colombia	C. Rica	Ecuador	Paraguay	Peru	Uruguay	Venezuela	Total
1967	12,430	...	...	...	...	...	...	...	...	...	...	12,430
	0											0/0.2
1968	25,923	...	...	...	...	...	...	...	...	7,951	...	33,874
	0									1		1/0.4
1969	33,504	...	...	4,103	...	...	...	...	...	5,267	...	42,874
	0			0						0		0/0.6
1970	36,342	...	...	9,317	...	...	...	...	...	5,876	...	51,535
	0			0						0		0/0.7
1971	32,733	...	...	10,709	...	...	...	...	...	3,714	...	47,156
	0			0						0		0/0.6
1972	31,991	...	4,433	11,295	...	...	...	...	...	3,067	...	50,786
	0		0	0						0		0/0.7
1973	30,800	...	10,600	10,983	...	...	197	...	182	6,117	6,130	65,009
	0		0	0			0		0	0	0	0/0.9
1974	27,137	...	6,842	10,128	...	...	11,813	...	382	6,908	21,751	84,961
	2		0	0			0		0	0	0	2/1.1
1975	19,247	...	6,403	6,803	...	...	5,211	...	449	2,899	24,202	65,214
	0		0	0			0		0	0	0	0/0.9
1976	24,770	...	10,970	6,976	...	...	13,431	...	419	2,836	18,590	77,992
	1		0	0			0		0	0	0	1/1.1

(Continued)

Table 1 - *Continued*

Year	Argentina	Bolivia	Brazil	Chile	Colombia	C. Rica	Ecuador	Paraguay	Peru	Uruguay	Venezuela	Total
1977	24,795	...	8,737	5,673	...	...	15,389	...	525	2,822	9,491	67,432
	0		0	0			0		0	0	0	0/
1978	15,272	...	8,207	12,525	...	...	15,135	...	2,134	3,434	15,524	72,231
	2		0	0			1		0	0	0	3/
1979	13,798	...	3,752	13,822	...	...	6,723	...	4,091	4,911	21,548	68,645
	0		1	0			0		0	0	0	1/
1980	17,957	...	1,435	9,688	...	...	2,589	...	4,648	813	17,363	54,493
	0		0	0			0		0	0	0	0/
1981	12,831	3,354	8,095	4,198	...	...	...	...	4,491	...	21,780	54,749
	0		0	0			0		0	0	0	0/
1982	42,812	6,364	34,089	11,648	1,624	...	16,864	3,441	6,701	10,427	11,445	145,415
	0	0	2	0	0		0	0	0	0	0	2/
1983	46,014	3,143	53,832	14,132	1,630	...	...	9,154	5,616	9,752	11,324	154,597
	1	0	3	0	0			1	0	0	0	5/
1984	53,905	2,530	56,066	12,945	3,321	...	1,548	8,135	5,867	10,015	12,326	167,258
	1	0	2	0	0		0	1	0	0	0	4/
1985	60,465	1,950	54,082	12,741	9,318	3,239	...	8,193	6,481	12,272	13,330	182,071
	0	0	1	0	0	0		0	1	0	0	2/
1986	76,134	2,718	50,951	15,874	10,217	13,425	2,057	7,378	7,441	12,354	17,681	216,230
	0	0	1	0	0	0	0	0	1	0	0	2/
Total	638,860	20,059	319,094	183,560	26,110	16,624	90,957	36,301	49,427	111,435	222,485	1,714,952
	7/8.6	0/0.3	10/4.3	0/2.5	0/0.3	0/0.2	1/1.2	2/0.5	2/0.7	1/1.5	0/3.0	23

Eight conjoined twins born in Venezuela during 1986 were largely commented by the lay press. This prompted us to analyze the material accumulated into the files of the Latin American Collaborative Study of Congenital Malformations (ECLAMC) [3] in order to study the time and space distribution of conjoined twins. This was done in more than 1.5 million births registered during the last twenty years in selected maternity hospitals from eleven Latin-American countries. These results are reported here.

MATERIAL AND METHODS

The Latin American Collaborative Study of Congenital Malformations (ECLAMC) is a hospital-based, clinical-epidemiological program, aimed to the study of birth defects, including the monitoring of their birth prevalence rates, as well as the systematic surveillance of 50 genetic and environmental risk factors [3].

The ECLAMC registers all major and minor congenital anomalies defined and described according to standards given in a procedures manual. Since even innocent pigmented naevi or partial toe syndactylies are routinely recorded [4], the under-registration of a severe, external, and conspicuous defect as conjoined twins seems highly unlikely.

In the 1967-1986 period, a total of 1,714,952 births were examined and registered in 95 maternity hospitals distributed in 11 Latin-American countries. Since the participating hospitals entered and left the program at different times, the annual number of covered births has been variable. The most important change in this respect occurred between the years of 1981 an 1982 (Table 1), when the annual number of observed births experienced a three-fold increase due to the implementation of a simplified methodology, making the study suitable to a large number of hospitals. At that point, the average number of participating hospitals suddenly increased from 25 to 70.

Twenty-six cases of conjoined twins were registered in the ECLAMC hospitals in the 1967-1986 period. Three of them were discarded from this study being asymmetric cases, two "fetus-in-fetus" and one partial duplication of the face, or incomplete diprosopus. Therefore, the material included here is restricted to the 23 cases presented in Table 3.

RESULTS

The observed birth prevalence rate for symmetrical conjoined twinning was $23/1{,}714{,}952 = 1/74{,}563$. The distribution of cases by year and country of birth is homogeneous, showing no obvious clusters (Table 1). However, in order to analyze the time distribution, yearly, from 1967 to 1986, the interannual variation in the number of examined births was too large: $\bar{x} = 85{,}748$, SD = 55,581 births (Table 1).

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In an attempt to overcome this drawback, the 20-year time series was divided into 26 intervals with similar number of births: $\bar{x} = 65,603$, SD = 3,265 (Table 2). Under this arrangement, the time distribution of the 23 cases of conjoined twinning was studied, and it did fit well to a random expected Poisson distribution. Such fitness was both proven by chi-squared ($\chi^2 = 0.69$; df = 1; P = 0.41) as well as by the Kolmogorov-Smirnov (DM = 0.05, P > 0.05) tests.

Table 2 - Time distribution of births and conjoined twins in 26 intervals of similar sample size of births

Secular interval	Calendar correspondence	No. of months	Births No.	Conjoined twins No.
1	JUL 67–MAY 69	23	61,328	1
2	JUN 69–SEP 70	16	66,530	0
3	OCT 70–JAN 72	16	63,767	0
4	FEB 72–APR 73	15	65,587	0
5	MAY 73–MAR 74	11	66,389	0
6	APR 74–DEC 74	9	65,230	2
7	JAN 75–DEC 75	12	65,209	0
8	JAN 76–OCT 76	10	65,814	1
9	NOV 76–OCT 77	12	67,698	0
10	NOV 77–SEP 78	11	64,473	2
11	OCT 78–AUG 79	11	66,030	2
12	SEP 79–SEP 80	13	65,143	0
13	OCT 80–DEC 81	15	66,384	0
14	JAN 82–JUN 82	6	65,396	0
15	JUL 82–NOV 82	5	66,363	2
16	DEC 82–APR 83	5	66,809	1
17	MAY 83–SEP 83	5	63,720	2
18	OCT 83–FEB 84	5	65,585	3
19	MAR 84–JUL 84	5	71,331	2
20	AUG 84–DEC 84	5	64,232	1
21	JAN 85–APR 85	4	67,916	1
22	MAY 85–AUG 85	4	74,229	0
23	SEP 85–DEC 85	4	67,934	1
24	JAN 86–APR 86	4	56,647	1
25	MAY 86–AUG 86	4	60,504	1
26	SEP 86–DEC 86	4	65,456	0
Total No.	JUL 67–DEC 86	234	1,714,952	23
$\bar{x}$			65,603.8	0.88
SD			3,236.5	0.90

The 10 Brazilian cases fall within the 99% Poisson confidence limits for the expected value of 4.28 cases. They were born in 7 different hospitals located in 6 different cities. Three were born in a single hospital of the city of Fortaleza ($03°19'S$, $41°25'W$), two in a single hospital of the city of Ribeirão Preto ($21°11'S$, $42°48'W$), two, one in each hospital, in the city of São Paulo ($23°32'S$, $46°37'W$), one case in Porto Alegre ($30°04'S$, $51°11'W$), one case in Rio Grande

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(32°02′S, 52°05′W), and one in Rio de Janeiro (22°54′S, 43°14′W). The hospital located in Fortaleza participated in the ECLAMC program since January 1982, having examined 47,660 births. Therefore, the expected number of conjoined twin cases in this hospital is 0.64, while 3 cases were observed, born 20 October 1982, 16 June 1983, and 26 November 1983, that is, within a range of 13 months. Likewise, the two cases born in Riberão Preto occurred in one hospital with 19,232 births examined since 1973, and their birth dates were only 60 days apart: 28 February and 28 April 1984. In a single hospital of Lima (12°03′S, 77°03′W), Peru, with 14,555 births examined between 1982 and 1986, there were two conjoined twins born 8 December 1985 and 19 March 1986, that is, only three months apart. The remaining 16 conjoined twins were all born each in a different hospital.

Table 3 - Descriptive data for the 23 conjoined twins reported here

Case	Country	Birth date	Birth[b]	Sex	Birth weight (g)	Matern. age (yr)	Birth order	Type
1	URU	10 JUL 68	LBD	M	?	30	02	Thoracopagus
2	ARG	20 OCT 74	LBD	M	?	22	01	Pygopagus
3	ARG	29 DEC 74	LBD	F	3,500	25	01	Thoracopagus
4	ARG	22 APR 76	LBA	M	4,940	20	04	Ischiopagus
5	ARG	23 NOV 78	LBA	F	4,070	24	01	Thoracopagus
6	ARG	02 JUN 78	LBD	F	2,000	41	02	Thoracopagus
7	ECU	08 APR 78	LBD	M	2,600	32	04	Diprosopus
8[a]	BRZ	05 MAY 79	LBD	F	4,450	33	04	Thoracopagus
9	BRZ	20 OCT 82	SB	F	4,400	36	10	Dicephalus
10	BRZ	25 AUG 82	LBD	M	3,750	28	02	Thoracopagus
11	ARG	28 NOV 83	SB	M	1,700	25	02	Syncephalus
12	BRZ	16 MAY 83	SB	F	4,000	21	02	Dicephalus
13	BRZ	26 NOV 83	LBD	F	1,010	30	06	Syncephalus
14	BRZ	04 MAR 83	LBD	M	2,750	14	01	Dicephalus
15	PAR	13 AUG 83	LBD	M	2,600	25	01	Syncephalus
16	ARG	09 AUG 84	LBD	M	3,300	23	06	Pygopagus
17	BRZ	24 FEB 84	LBD	M	2,900	24	02	Pygopagus
18	BRZ	28 APR 84	LBD	F	2,570	27	06	Omphalopagus
19	PAR	15 MAY 84	LBD	F	1,920	22	05	Thoracopagus
20	BRZ	10 JAN 85	LBD	F	850	21	01	Thoracopagus
21	PER	08 DEC 85	LBD	M	3,700	35	04	Pygopagus
22	BRZ	04 JUN 86	LBD	F	2,850	25	02	Pygopagus
23	PER	19 MAR 86	LBD	M	3,830	30	09	Thoracopagus

[a] Product of first cousin parents.
[b] LBA: Livebirth, alive at 3rd day; LBD: Livebirth, dead at 3rd day; SB: Stillbirth.

As shown in Table 3, the 23 conjoined twin cases included 3 stillbirths, while only 2 of the 20 liveborn cases survived the first three days of life. Sex, maternal age, and birth order did not differ from expected values for this birth population. There were 12 male and 11 female pairs; mean maternal age was 26.8 years (SD = 6.1; SE = 1.3) while the expected mean value is 25.8 years; and there were 7/23 primigravity products, with an expected value of 6.24 cases.

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Eight cases (Cases 1 to 8 in Table 3), belonging to a case/control subgroup of malformed babies, had further data available on risk factors. None of the 8 cases had reported prenatal exposure to griseofulvin; and one of the 8 cases was a consanguineous product born from first cousin parents.

DISCUSSION

The birth prevalence rate for conjoined twinning observed in this Latin-American series fits well with the 1/75,000 value estimated by Källén and Rybo from corrected nationwide Swedish data [5]. Other published observations usually give a higher value, around 1/50,000 births, which seems to be produced by systematic sample bias [5].

Conjoined twinning has frequently been reported to occur in clusters which are difficult to prove on statistical grounds, probably because of the rareness of the event. So, it is noteworthy that, in spite of the scanty information available concerning the epidemiology of this birth defect, several reports deal with the suspicion of small epidemics. One is that of Milham [6] in the State of New York; another comes from southern Africa [1]; and the third belongs to a single hospital in Skövde, Sweden [5]. The latter is very similar to an observation made in our series. Källén and Rybo [5] reported three cases born in a 14-month range within a single hospital with 2,300 births per year. Our hospital in Fortaleza, Brazil, also recorded three conjoined twin sets within a 13-month lapse, even though this is a large maternity hospital, with 10,000 annual births.

Furthermore, the three cases born in Fortaleza are not the only indication of a nonrandom time/space distribution of conjoined twinning in this Latin-American material. The two cases born in Ribeirão Preto within a 60-day range, and the other two born in Lima 101 days apart, also suggest clustering. However, the distribution of cases by years, or by country of birth, does not depart from random expectation. More precise or punctual distributions, like those by time and hospital of occurrence, could not be tested due to sample size limitations in spite of having at hand the systematic observation of more than 1.5 million births.

The original Venezuelan rumor did not show up in our material. This is not surprising because the ECLAMC did only represent about 3% of all Venezuelan births in the 1973-1985 period. On the other hand, probably there was no epidemic at all in Venezuela. Eight cases in one year do approximately represent 8/500,000 births. Under the expected Latin-American frequency of 1/74,563, 6.70 cases are expected annually, which, for a Poisson distribution, varies from 0 to 14 cases within 99% confidence limits.

Two obervations reported here deviate from common knowledge found in the literature. One concerns the sex of conjoined twins, which is said to be predominantly female [4,7], while an even distribution was observed in our material. The other one has to do with the relative frequencies of the different anatomical types of conjoined twinning. According to the literature [4,7], more than 90% of cases are

either thoracopagus or pygopagus. Only 14 of our 23 cases fell into any of these two categories, and the remaining 9 cases represented all known types, with the only exception of dipygus, craniopagus, and rachipagus, for which we did not record any observation. If differences between this material and the literature concerning frequency of sexes and types in conjoined twins are not simply due to random variation, then probably the reason might be survival. Only 2 of the 23 cases survived more than three days. If published data have been based on reported cases from the literature, instead than from consecutive birth series, chances are that those cases frequently survived longer than just the immediate neonatal period. In that case, less severe forms like thoracopagus are likely to be overrepresented, and, furthermore, the low sex ratio could also be a characteristic of less severe types, rather than of all cases of conjoined twins.

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