Annals of Clinical and Medical Case Reports

DOI: https://doi.org/10.47829/ACMCR.2023.11602

Research Paper ISSN 2639-8109 | Volume 11

Superior Umbilical Ectopias and Ectopia Cordis, A "New" Syndrome with an Unusual Epidemiology

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Received: 03 Aug 2023
Accepted: 25 Sep 2023
Published: 02 Oct 2023
J Short Name: ACMCR

Keywords:

Ectopia Cordis; Epidemiology; Male; Primagravida, Umbilicus.

Citation:

Lubinsky M, Superior Umbilical Ectopias and Ectopia Cordis, A "New" Syndrome with an Unusual Epidemiology. Ann Clin Med Case Rep. 2023; V11(6): 1-6

1. Abstract

1.1. Introduction

Superior/anterior umbilical ectopias with non-disruptive origins are uncommon, and have been minimally investigated.

1.2. Methods: Literature Review.

1.3. Results: Thirty-three of 36 cases were associated with ectopia cordis, with a continuous gap extending from the cardiac defect to a superiorly displaced umbilicus. Three quarters showed non-European ancestry. For patients without other issues, 16 were males and 8 were females. Eight others with additional disturbances, mostly midline, that may represent predispositions, had a sex ratio of 3:5. There was a predominance of primagravida births without a corresponding decreased maternal age effect.

1.4. Conclusions: Findings support a specific dual ectopia syndrome of ectopia cordis associated with a superiorly displaced umbilicus and a distinct epidemiology, with a male preponderance and non-European ancestry, consistent with a deficient ventral closure above the umbilicus. A predominance of primagravida births without a corresponding decreased maternal age effect suggests an etiological role for fertility related issues.

2. Introduction

Typically associated with surgically significant abdominal defects, but with largely cosmetic implications themselves, umbilical ectopias per se have been minimally investigated. Inferior displacements are common with lower wall defects such as bladder exstrophy [1], but the superior ectopias reviewed here are considerably rarer, and may provide important insights into normal and abnormal development. Of 36 primary superior/anterior umbilical ectopias without disruptive origins (e.g., limb-body wall defects) in the English language literature, 33 were associated with ectopia cordis, with a continuous gap typically extending from the cardiac defect to a superiorly displaced umbilicus. Most showed non-European ancestry, and a predominance of primagravida births without an apparent decreased maternal age effect suggests fertility issues. Twenty-five patients without other issues had a 2:1 sex ratio, while 8 others with additional disturbances, mostly midline, that may represent predispositions, had a 3:5 ratio, 3 with atypical features (Table 3) suggesting alternative etiologies. These findings support a syndrome of dual ectopias with ectopia cordis with supra-umbilical displacement, primarily males and non-European ancestry, and a predominance of primagravidas without a decreased maternal age effect.

3. Materials and Methods

English language reports of primary upward midline displacement of the umbilicus, excluding limb-body wall defects, amniotic bands affecting the thorax, notable asymmetry, and aneuploidy were reviewed. A displaced umbilicus was either noted in case reports or subjectively placed by the sternum. Author institutions provided countries of origin. Two Turkish and one Brazilian patient were considered to have non-European ancestry. U.S. patients were considered to have European ancestry. Pubmed had no ectopic umbilicus heading, and an initial ascertainment was through Google Scholar.

Most also had ectopia cordis, leading to a further search for that term plus "umbilicus" or "umbilical, " and reviews of various reports of ectopia cordis descriptions and/or pictures, the latter with an inevitable subjective component. Inferior umbilical ectopia was first found in Google Scholar using low set umbilicus, and was largely associated with exstrophy of the bladder. Syndrome links were primarily through an OMIM (1a) search for "umbilicus," and syndrome cases with high ectopic umbilici but without ectopia cordus were excluded (2, 3 patient BPS3, 4).

4. Results

Thirty-six patients had superiorly displaced umbilici, 33 with a distinct phenotype connected to a thoracic ectopia cordis through a typically continuous open ventral wall defect. When information was available, cardiac lesions were present in most, generally with a full Pentalogy of Cantrell, although 3 patients apparently lacked intracardiac lesions [5, 6, 7]. The most common absent finding documented was the diaphragmatic hernia, while sternal defects were always present save for an atypical case of Bae et al. [8] (Table 1 and 2). Ectopia cordis findings were divided into simplex (N = 25) (Tables 1 and 4), without, and complex (N = 8) with (Tables 2 and 5), other issues. Six of the latter had midline lesions (Table 2): Cleft lip and palate in 2 [9, 10], esophageal atresia plus IUGR and micrencephaly [11], a mediastinal teratoma [12], and 2 syndromes affecting the sternum (PHACES [13] and Forzano (Personal Communication, PN Kantaputra). For non-midline findings, 2 had amniotic bands with mild finger issues, one with a band attached to the vertex as well [14, 15]. Of mothers with available information, 21 of 25 were primagravidas, and, for 17, the average age was 26, the youngest aged 19 and 20 years, and the oldest 33, 35, and 42 years. Only 2 paternal ages were noted, 36 years [16] and 40 years [5]. Of the 25 simplex cases, all but 3, one Dutch, and 2 from the U.S., were of non-European ancestry. There were 16 males and 8 females, plus one unknown (Table 1). Of the 8 complex cases, 5 were non-European, and 3 were males (Table 2).

Table 1: Ectopia Cordis Simplex

Case	Sex	Nation	Maternal Age	Parity	Type of Ectopia
Akhtar et al. (21).	M	Pakistan	na	na	Pentalogy
Apte (22)	M	India	na	na	na
Batur et al. (14).	M	Turkish	33 y.o.	Primagravida	Pentalogy
Çelik et al. (23).	F	Turkey	20 y.o.	Primagravida	na
Chen et al. (24).	M	Formosa	na	na	Pentalogy
Chen et al. (24).	M	Cameroon	23 y.o	G3P1010	Pentalogy
Guanmei et al. (26).	M	India	24 y.o.	Primagravida	na
Ito et al. (27).	M	Japan	na	na	na
Jones et al. (28).	M	Colorado	21 y.o	Primagravida	Pentalogy
Júnior et al. (29).	F	Brazil	42 y.o.	Primigravida na	
Kaouthat et al. (30).	F	Tunisia	na	na	Pentalogy
Kumar, Sharma Sinha (31).	M	India	28 y.o.	Primagravida	Pentalogy
Meena, Jangid (6).	na	Nigeria	na	Primagravida na	
Momin, Sangma, Saha (32).	F	India	28 y.o.	Primagravida na	
Muktan, Singh, Bhatta (33).	M	Nepal	28 y.o.	Primigravida	na
Naburi et al. (5).	F	Tanzania	26 y.o.	na	no dh
Pius et al. (11).	F	Nigeria	na	Para 3	na
Puvabanditsin et al. (34).	M	U. S	19 y.o.	Primigravida	na
Shad, Budhwani Biswas (35).	M	India	na	na	na
Shrestha et al.	M	Nepal	22 y.o.	Primagravida	na
Shwe et al. (36)	F	Nigeria	27 y.o.	Primagravida	Pentalogy
Tan et al. (37).	M	Ireland	na	Primagravida	Pentalogy
Van der Horst et al. (38).	M	S. Africa (black)	na	na	na
Van Hoorn et al. (39).	F	Dutch	26 y.o.	Primagravida.	Pentalogy
Yuko-Jowi, Simiyu, Musoke (40).	M	E. Africa	na	na	na

dh = diaphragmatic hernia na = not available

Table 2: Ectopia Cordis Complex

Case	Sex	Nation	Maternal Age	Parity	Ectopia	Additional
Blaszczyński F et al. (41).	F	Polish	22 y.o.	Primagravida	Pentalogy	Teratoma
Kaplan et al.	F	US	20 y.o.	G4POAb3	no dh	Amniotic (10).
Kantaputra	M	Thai	na	Primagravida	Covered	Abs. Umbilicus (personal com munication & rectus abd.) Forzano syndr. situs inversus
Kragt et al. (42)	F	Dutch	35 y.o.	3 prior abs.	no dh	Esophageal Atr., IUGR
Oumarou et al. (9).	M	Nigeria	24 y.o.	Primagravida	no dh	Amniotic bands
Patil et al. (43).	M	India	24 y.o.	Primagravida	Pentalogy	Left CL/P
Sharma, Bagri	F	India	na	na	Partial	PHACES
Jangid (16).	F	na	na	na	na	Consang.
Jafarian et al.* (44).	F	Iran	39 y.o.	Primagravida	Pentalogy	bilateral CL/P

*Uncertain- clip of umbilical cord seems high, but origin is obscured
dh = diaphragmatic hernia na = not available

Table 3: High Umbilicus without Ectopia Cordis.

1. Black Nigerian male with a grand multiparous mother, with an absent sternum under a membranous diamond shaped defect with a hyperpigmented line extending to a high umbilicus. The heart was normally placed with a secundum ASD. There was also a narrow urachal diverticulum off the superior bladder. (45).
2. Adult white female with a soft tissue gap at the lower sternal border and an absent umbilicus (19).
3. Korean female with an additional cord going from a normally placed umbilicus to the xiphoid and the left ventricle, mesocardia, a focal pericardial defect on mid-inferior cardiac apex, 2-mm perimembranous VSD and 3-mm secundum ASD, thought by the authors to be a “mild” Pentology (8).

ASD = atrial septal defect; VSD = ventricular septal defect

Table 4: Cardiac Findings: Simplex Cases

Akhtar et al. (21).	Restrictive perimembranous VSD and small patent foramen ovale both spontaneously closed in infancy. Left ventricular apex herniated into epigastrium. Ventriculi form a tail looking like a crocodile.
Apte (22).	No pericardium, apex of heart pointing anteriorly.
Batur et al. (14).	Umbilical cord directly joined to right atrium. Single-cavity heart without a septum.
Çelik et al. (23).	Single atrium, hypoplasic right ventricle, ventricular septal defect, pulmonary atresia, no pericardium.
Chen et al. (24).	VSD and intra-abdominal left ventricular diverticulum.
Chishugi. (25).	na
Guanmei et al.	(26). na
Ito et al. (27).	Transposition of great arteries, VSD, left ventricular outflow tract obstruction, hypoplastic right ventricle and bilateral superior vena cava.
Jafarian et al. (44).	ASD, VSD, Patent ductus.
Jones et al. (28).	One great vessel. Strikingly elongated ascending aorta, pulmonary valve atresia, VSD, patent ductus arteriosus approximately ten times diameter of hypoplastic main pulmonary artery. Long tubular ventricles.
Júnior et al. (29).	Small VSD measuring 2.7 mm
Kaouthar et al. (30).	Double-outlet right ventricle with malposition of the great arteries.
Kumar, Sharma and Sinha (31).	Interventricular septal defect.
Meena and Jangid (6).	Without intracardiac defect
Momin, Sangma and Saha (32).	na
Muktan, Singh and Bhatta (33).	na
Naburi et al. (5).	No defect
Pius et al. (11).	na

Puvabanditsin et al. (34).	Double-outlet right ventricle with malposition of great arteries, severe pulmonary valve stenosis, moderate pulmonary artery hypoplasia, aortic and pulmonary valve insufficiency, perimembranous ventricular septal defect
Shad, Budhwani and Biswas (35).	Ventricular septal and pericardial defects.
Shrestha et al. (7).	Normal
Shwe et al (36).	VSD, dilated left ventricle and inferior vena cava.*
Tan, Lim and Sharif (37).	Double outlet right ventricle, subaortic VSD and severe right ventricular outflow obstruction. Dextrocardia.
Van der Horst Mitha, and Chesler (38).	Single ventricle, common atrioventricular valve
Van Hoorn et al. (39).	Tetralogy of Fallot with VSD, pulmonary valve stenosis, aberrant aortic valve, large ASD.

*Also: Spina bifida oculta, suspected twin gestation with empty sac.
ASD = atrial septal defect; VSD = ventricular septal defect. na = not available.

Table 5: Cardiovascular Findings: Complex Cases

Blaszczyński et al. (2015).41.	Membranous VSD with large left to right shunt, ASD type II, tricuspid valve leaflet insufficiency. Teratoma superior cavoatrial junction.
Jafarian et al. (44).	ASD, VSD, Patent ductus.
Kantaputra PN (Personal Communication).	Dextrocardia with situs inversus, ASD and VSD.
Kaplan et al. (10).	Tetralogy of Fallot with severe pulmonary infundibular and valvular stenosis, secundum-type ASD due to valve incompetence, common pulmonary vein inserting into the left atrium, bilateral superior vena cavae.
Kragt et al. (42).	Aberrant right subclavian artery. Left persistent superior vena cava draining into identifiable coronary sinus, transposition of the aorta and marked pulmonary artery hypoplasia with double-outlet right ventricle and perimembranous VSD.
Patil et al. (43).	Truncus arteriosus and VSD.
Oumarou et al. (9).	na
Sharma, Bagri and Jangid (16).	Severe aortic arch coarctation with left ventricular hypertrophy. Left common carotid artery and internal carotid artery severely narrowed with reformation of left terminal internal carotid artery/ anterior cerebral artery- middle cerebral artery through collateral circulation.

ASD = atrial septal defect; VSD = ventricular septal defect. na = not available.

5. Discussion

While there are undoubted biases and missing information, the reviewed reports support a specific syndrome of ectopia cordis and umbilical ectopia with a distinct epidemiology. An isolated superior umbilical ectopia is exceptional (Table 3), and is unlikely to be a forme fruste, suggesting that the ectopia cordis is the initiating lesion. Structurally, a ventral wall defect extends continuously from the heart ectopia to a superiorly displaced umbilicus; 75% of patients were without other findings, and 25% had additional, mostly midline, defects. The non-midline defects were amniotic bands with mild peripheral disruptions. Kaplan et al., who discussed one such case [10], suggested that this was a remnant of a more extensive band disruption that had caused the ectopia cordis. While I excluded patients with visible thoracic bands, the infrequency of bands elsewhere with dual ectopias suggests instead an occasional independent manifestation apart from the causative disturbance. The other additional findings are most likely markers for midline risk factors for occasional additional expressions of some separate cause.

After the 6th week of development, the abdominal muscles expand ventrally and caudally to form the infraumbilical body wall [12], and cardiac lesions (Tables 4 and 5) often involve rotational abnormalities suporting very early developmental events [13]. Batur et al. [14] found a short umbilical cord that directly joined the right atrium, indicating abnormal umbilical positioning from the start, instead of some sort of secondary migration. With this, several sequences are possible. In one, there may be an issue with the straightening of the embryonic body axis, which also occurs at this time [12]. In another, abnormal wall fold deficiencies result in both ectopias. Alternatively, abnormal placement of the umbilicus interferes with folding and closure. Sternal defects in all patients save for the atypical case of Bae et al. [8] go against the latter, but other pathogeneses may be involved [10], and this remains speculative. Still, the distinct epidemiology of the dual ectopias (below) supports a distinct etiology, making this separate from other forms of ectopia cordis.

There were 4 major epidemiological findings. For patients with available information (Tables 1 and 2):

1. Non - European ancestry involved 85% of the simplex and 50% of the complex cases. The reason for this bias is unknown, and both genetic and environmental factors may be involved.

2. Males comprised 16 of 24 of the simplexes and 3 of 8 of the complex cases. Ectopia cordis is etiologically heterogeneous, so good comparison figures are difficult to come by, but a large survey failed to note a skewed sex ratio [10]. A shift to an equal sex-ratio for the complex cases is not unexpected: Disturbances of

differentiation are generally female biased, as with hemangiomas and teratomas, while males are more susceptible to migrational issues [15], so females might show milder umbilical displacements that would have been missed in our survey. One case [16] was associated with the predominantly female PHACE syndrome, which may not be identifiable early on [17], so this disorder with dual ectopias could be undiagnosed in neonates.

3. For parity, 84% were first born, and both simplex and complex cases seemed to be similarly affected. This is particularly impressive since the non-European countries of origin typically have relatively high birth rates, decreasing the population percentages of primagravidas.

4. Primagravida status was unaccompanied by a corresponding decrease in maternal age, which averaged 24 years of age for 15 mothers. Epidemiology can be an important clue to the etiology of congenital disorders. Here, a predominance of primagravida mothers without a decreased maternal age effect, a pattern that has not, to my knowledge, been described before, suggests fertility issues, and may be a helpful clue to the etiology of other disorders. It is of interest here that subfertile couples had an increased incidence of abdominal wall defects and right ventricular outflow obstructive anomalies that included Tetralogy of Fallot [18].

Excluding ectopia cordis, amniotic bands, and limb-body wall defects, only 3 isolated superior umbilical ectopias were found (Table 3). The case of Lee et al. [19] is difficult to explain, with sub-sternal displacement without any other findings. Interestingly, Akman et al. [20] noted an analogous isolated inferior ectopic umbilicus.

The patient of Bae et al. [8] with a normally located umbilicus and an additional umbilicus attached to the xiphoid area and left ventricle is truly unique, and clearly represents another entity. Finally, a membrane covered evisceration with ectopia cordis is sometimes referred to as an omphalocele despite asymmetric relationships to the umbilicus, or even separation from it. There are actually 3 phenotypes of non-disruptive midline defects here: 1. A “true” omphalocele with the umbilicus at the center of the defect, often secondary to mechanical factors physically interfering with closure. 2. With deficient upper closure, either separate from the umbilicus, or asymmetrically related superiorly. Findings here support a distinct embryonic closure superior to the umbilicus. 3. Affecting lower closure, and asymmetrically related to the umbilicus inferiorly, typically associated with bladder exstrophy. With this, vertically asymmetric midline involvement of the umbilical ring represents superior or inferior defects, rather than omphaloceles.

6. Disclosures

There are no conflicts of interest.

7. Funding

There was no financial support.

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