

## ORIGINAL PAPER

**LIMB BODY WALL COMPLEX – THE HISTORY OF THE ENTITY AND PRESENTATION OF OUR SERIES OF CASES**

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We present an analysis of two first historically documented limb body wall complex (LBWC) cases and our own contemporary perinatal autopsy series of this rare complex. So far it was supposed that the first case of this complex was reported in 1685 by Paul Portal. Studying the Joachim Oelhaf's autopsy report from 1613 with attached engraving showing the neonate with multiple birth defects led our research team to a conclusion that it was genuinely the first description of LBWC in the medical literature so far. We compared the Oelhaf's case from 1613 and the Portal's autopsy report from 1685 with our series of LBWC cases dissected in the Medical University of Gdansk between 1999 and 2011. Reviewing 1100 autopsy reports performed we encountered 9 cases of this unique complex. The analysis was supported by the literature review.

**Key words:** LBWC, autopsy, teratology, birth-defects, amniotic band syndrome.

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## Introduction

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Human body deformations had been already noted from the very beginning. The oldest images date back about 15,000 years. The ones that attracted the greatest attention were conjoined twins with all variations. They were portrayed quite accurately, and one of the oldest comes from Tlatilco, Mexico and dates from 3000 years ago [1]. Hippocrates, Aristotle, Pliny, and even Galen wrote about monstrous fetuses and newborns and also tried to find the mechanisms of or the reasons for different malformations [2, 3, 4]. Nevertheless, the unusual newborns were considered either as prodigies or portents, divine information or a suggestion for the people. As such, they were taken rather as curiosities, but not to be treated from a scientific point of view as a subject

for medical investigation. Centuries later, descriptions of congenital malformations were not regarded by scholars as credible contributions to knowledge. The first known teratological autopsy reported in literature is a case of conjoined female twins born in 1533 in Hispaniola. The following cases were dissected in Italy (1540 in Zarzara and 1544 in Milan) [4]. In the sixteenth century and later, while dissecting conjoined twins the main reason for the dissection was to prove the existence of single or two souls for baptism, whereas when alive, the behaviour of separate heads were taken more seriously and proved much more that medical examination.

Several classifications of "monsters" were published by anatomists and physicians, including Ambroise Pare's (ca 1510-1590) "Des Monstres", published in 1573 and as a definitive edition as Les

Oevres in 1575. But even his work was dismissed as “fit for amusing little children” with all due respect to his immense achievements as a surgeon. Later, Caspar Bauhin (1560-1624) and other researchers published books, yet at that time fetuses with congenital defects were treated as curiosities of nature [5, 6]. Very few other interesting dissected cases were reported until the nineteenth century. Another rare malformation – holoprosencephaly – though described as an entity much earlier, was reported after autopsy until 1839 [7].

The part of medical science that studies the causes, mechanisms, and different patterns of abnormal development of either part or the whole body is teratology. The term was coined and introduced by father and son – Etienne Geoffroy Saint-Hilaire (1772-1844) and Isidore Geoffroy Saint-Hilaire (1805-1861) in the nineteenth century, but as a separate branch it dates barely from the beginning of the twentieth century [8, 9].

In the following years the increasing knowledge in teratology and dysmorphology was supported by genetics and molecular biology. Birth defects can be classified in many ways. The Polish Registry of Birth Defects classifies them as to pathogenesis: malformations, disruptions, deformations, and dysplasias; epidemiology: multiple and isolated, major defects (impairing the functions of the organism) and minor defects (without such impairment); lethal defects and defects not affecting survival; and sporadic and familial defects. Multiple birth defects are classified as sequences, complexes, syndromes, and associations [10].

In 2013 the 400<sup>th</sup> anniversary of the first public autopsy in Gdansk and North-East Europe was celebrated and commemorated by the conference “Joachim Oelhaf i jego następcy” (Joachim Oelhaf and his followers). It was organised by the Medical University of Gdansk and held at the National Museum, which in the sixteenth century was the original place of Atheneum Gedanense. Joachim Oelhaf (1570-1630), educated in the best European universities, was a major city physician, an anatomist, and an Atheneum lecturer. He performed the dissection and published its results. Studying the scans of this original autopsy report led our team to the conclusion that it was a case of limb body wall complex (LBWC) and most probably the first case of this complex malformation published in history [11].

Limb body wall complex is a rare lethal birth defect with different and variable phenotypes. Limb body wall complex covers several areas of malformations, and when possible the diagnosis is based on established criteria formulated by Van Allen in 1987, who mentioned the first available, in his opinion, description made by Paul Portal (1630-1703). Usually, to diagnose this complex, two out of three of the following anomalies are required: exenceph-

aly or encephalocele with facial clefts and/or thoraco- and/or abdominoschisis and/or limb defects [12]. In 1993 Russo with his team identified two distinct phenotypes. Type I includes craniofacial defects with facial clefts present, amniotic adhesions localised between the placenta and cranial defects and amniotic band syndrome (ABS). Type II is noted when there are no craniofacial defects, but we can spot urogenital anomalies, imperforate anus, lumbosacral myelomeningocele, severe kyphoscoliosis, persistence of the extraembryonic coelom, and some placental anomalies with usually short umbilical cord. Those two phenotypes probably result from two different pathomechanisms [13].

This study presents an analysis of the first two historically documented LBWC cases from 1613 and 1685 and our own contemporary perinatal autopsy series with a literature review.

## Material and methods

We analysed the original source text from the Library of Polish Academy of Sciences Department in Gdansk – Oelhaf Joachim (1613) “Foetus monstruosus in pago Prust territorii Dantiscani editus Anno Domini MDCXIII die 27 Februarii Bene fide delineatus et descriptus. Typis Hünefeldi, Dantisci” (*PAN Biblioteka Gdańska*, sygn. XIX q 79a adl. 28) [14]. The original text was scanned, and further work was performed on a digital version of the manuscript. After transliteration it was translated with participation of a specialist in seventeenth century Latin to Polish and English. Two available engravings attached to the report were analysed in detail, namely the first original dating from 1613 and the second from the copy of Bauhin’s work from 1614 quoting in full the report by Oelhaf.

The second known historical case, reported by Paul Portal from 1685, was analysed based only on the original French text and an available translation to English, found after citation in van Allen’s work. This description was devoid of the foetus depiction [12, 15].

The third part of the study was analysis of our own material of nine perinatal autopsies performed by one of the authors (JG) at the Pathomorphology Department of the Medical University in Gdansk in the years 1999 to 2011 out of a total of over 1100 perinatal dissections. All cases came from the Department of Obstetrics of the Medical University Clinical Hospital in Gdansk. The clinical data and autopsy reports are supported by the photographic documentation.

Finally, we performed a critical review of the available source material and research published in the databases PubMed, Researchgate, and Academia.edu focusing on the LBWC cases and case series, prenatal diagnostics, and morphology types.

## Results

### Historical case 1 (1613) by Oelhaf

This case was of triple pregnancy with one foetus with several severe malformations, which was the subject for consecutive dissection [11]. The mother was 25 years old at the moment of delivery, primipara, with triple pregnancy. Two daughters were live-born but died soon afterwards, the third neonate with malformations survived only for several minutes. The written description was very detailed and divided into two distinct sections: first “QUO AD FORMAM externam” – dealing with external appearance, while the second section – “QUO AD PARTES internas” – referred to the internal parts, starting with a description of the brain and further describing internal organs and their topography.

The final report reveals a complex of malformations including foetal cerebral ventriculomegaly, neck cystic hygroma, lack of left eyeball, cleft lip, thorax deformation with narrow chest and sternoschisis, omphalocoele with viscera of the thorax and abdomen (heart, single lung, liver, spleen, and stomach), ectopic heart with unusual shape, hypoplastic lung (single reported), lack of urogenital organs, anal atresia, single lower extremity, and deformation of up-



Fig. 1. Engraving of the foetus with malformations (1614)

per extremities (including fingers and their number). The whole picture after thoughtful analysis corresponds to the diagnosis of LBWC. The autopsy report included one engraving depicting the neonate with malformations. The 1614 version of the picture is slightly “enhanced” as far as the lines and their thickness are concerned, but we must not state that corrected in any way (Fig. 1).

### Historical case 2 (1685) by Paul Portal

This description of the case is one made by an obstetrician, or as Portal was called “a man-midwife”, not a pathologist. He started describing the procedure of extraction of the stillborn foetus to conclude with the anomalies found. “The membranes chorion and amnion were joined to the right side of the head just behind the temples... On the same side, above and behind the ear, was excrescency like a small horn, of the length of half an inch, or something more. On the other side there was such another, but rising somewhere higher, and betwixt both a musculous flesh, without any appearance of brains”. He also noted “the upper lip split like a hare-lip” and that “left arm was shorter than the other, and crooked at the elbow”,

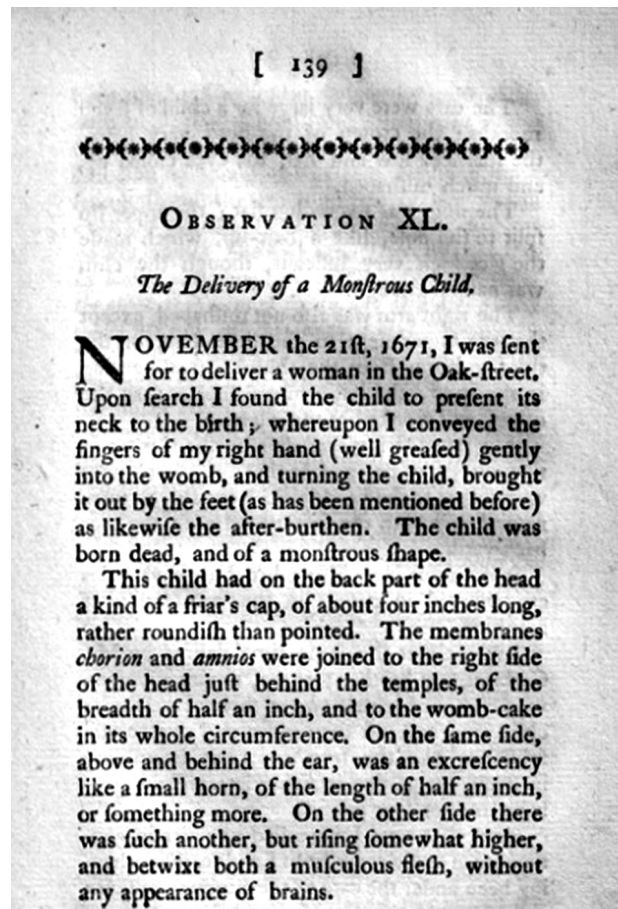


Fig. 2. Page 139 from Portal's manual with the beginning of the case description

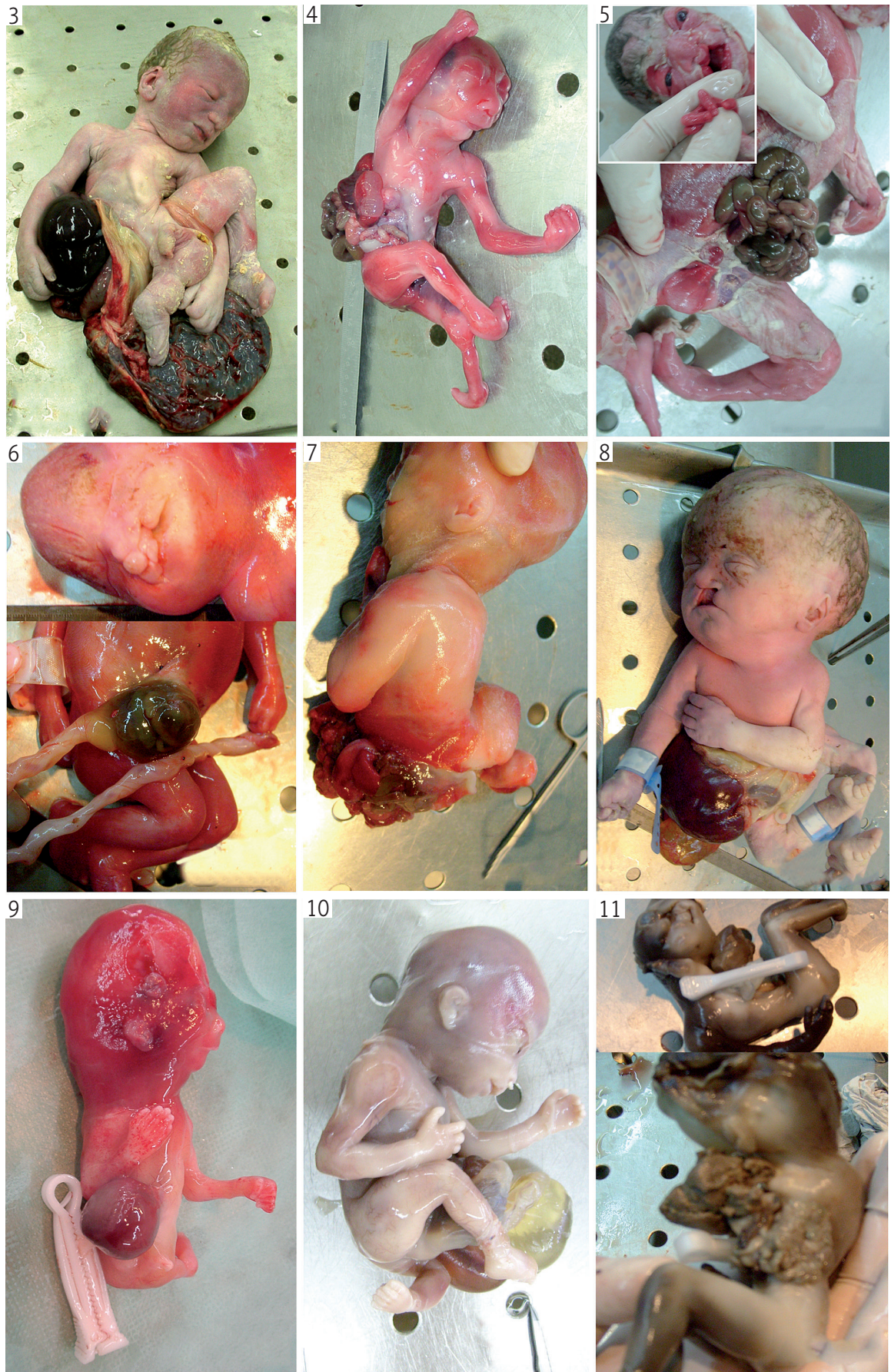


Fig. 3-11. Cases 3-11

“both the thighs were natural, but both the feet turned with the toes backward”. This is where the description ended. Information given there are sufficient to diagnose LBWC. This report does not contain inspection of internal organs [15] (Fig. 2).

The following is presented our own series of contemporary cases.

### Case 3 (GD-01)

Mother, age 28 years at the delivery term, primipara, 37<sup>th</sup> week of gestation. Stillborn male foetus was 40 cm long, weight 1850 g. There was no external alteration within the head, but microgyria was disclosed on the brain inspection. Severe kyphoscoliosis was noted, narrow and deformed chest with hypoplastic lungs. The next finding was ruptured omphalocele containing liver, spleen, stomach, small intestine, and ascending colon with right kidney with suprarenal gland. Deformation of the hips and lower right extremity (complete lack of right femur with only two toes) (Fig. 3).

### Case 4 (GD-02)

Mother, age 26 years at the delivery, second pregnancy (first without known malformations), 20<sup>th</sup> week of gestation. Stillborn male foetus was 20.5 cm long with weight 200 g. Deformation of the cranium was noted, with hypognathia and posterior encephalocele. Hypoplastic sternum and ruptured omphalocele resulted in ectopic heart and eventration of the stomach with the small and part of the large intestine with the liver. The lower extremities rotated inwards and had clubfoot shape (Fig. 4).

### Case 5 (GD-03)

Mother, age 23 years at the delivery, primipara, 31<sup>st</sup> week of gestation. Stillborn male foetus was 27 cm long with weight 900 g and with macera-



Fig. 12. US scan showing cleft lip and cleft palate



Fig. 13. US scan showing abdomen with omphalocele and two vessel umbilical cord

tion of the third degree. The autopsy with opening of the cranium showed autolysis of the brain with traces of intraventricular haemorrhage. Bilateral cleft-lip and unilateral cleft-palate were noted. Both palms had only four fingers with ectrodactyly on the right side (cleft-hand). Both lower extremities were in the shape of clubfoot. Ruptured omphalocele contained part of the small intestine and transverse colon (Fig. 5).

### Case 6 (GD-04)

Mother, age 27 years at the delivery, third pregnancy (no information concerning previous pregnancies), 21<sup>st</sup> week of gestation. Stillborn male foetus was 25 cm long with weight 420 g and maceration of the first degree. Bilateral cleft-lip and cleft-palate were noted, both ears set very low. Upper and lower extremities did not show any abnormalities. Small heart, round in shape, had VSD (ventricle sept defect). Ruptured omphalocele contained part of the small intestine and transverse colon (Fig. 6).

### Case 7 (GD-05)

Mother, age 41 years at the delivery, second pregnancy (first without known malformations), 20<sup>th</sup> week of gestation. Stillborn male foetus was 20 cm long with weight of 325 g. Severe deformation of the vertebral column with hyperlordosis in the lumbar region and rotation was noted. Hypognathia and low set of ears were noted. Upper extremities without abnormalities, absent left lower extremity with deformation of the right one including deformation of the right hip and polydactyly (six toes). The ruptured omphalocele contained part of the small and large intestines with liver. Narrow chest resulted in hypoplastic lungs and the heart with the VSD (Fig. 7).

**Case 8 (GD-06)**

Mother, age 34 years at the delivery, second pregnancy (first without known malformations) – this time twin pregnancy, 32<sup>nd</sup> week of gestation (second twin without defects). Live-born male neonate, survived 90 minutes, was 40 cm long with weight 1900 g. Deformation of the vertebral column with hyperkyphosis in the thoracic region and scoliosis in the lumbar region were noted with rachischisis and myelomeningocele in the sacral area. Internal hydrocephaly with lateral ventricles widened up to 4 cm each. Hypertelorismus with unilateral cleft lip and cleft palate. Upper extremities without abnormalities, both lower extremities clubfoot. The ruptured omphalocele contained the whole liver with stomach, small intestine, and part of the large intestine. Narrow chest rotated a little to the left, with hypoplastic lungs (right bilobar, left - unilobar) and the heart had VSD with coarctation of the aorta. The other autopsy findings confirmed the clinical diagnose of circulatory-respiratory failure as the cause of death (Fig. 8).

**Case 9 (GD-07)**

Mother, age 31 years at delivery, second pregnancy (no data on the previous one), 17<sup>th</sup> week of gestation. Stillborn foetus with autolysis of the second degree – 12 cm long, weight 70 g. Both lungs were small (1 cm long) and unilobar, the heart spherical in shape. The omphalocele contained only the liver and was not ruptured. Neck hygroma was noted, but no other defects. Upper extremities were normal, whereas the lower were both deformed. The left consisted only of a single long bone ending with a foot without knee or ankle joints. The right consisted only of the deformed foot with two toes and no joint connection with the hip region. The sex organs were ambiguous – a penis like structure was noted, without the scrotum. A vaginal entrance was not found despite present uterus and ovaries within the abdominal cavity, hence hermaphroditism was diagnosed (Fig. 9).

**Case 10 (GD-08)**

Mother, age 28 years at delivery, third pregnancy (twin, no data on the previous ones), 16<sup>th</sup> week of gestation. Stillborn female foetus with autolysis of the second degree – 16 cm long, weight 190 g (the second twin presented no malformation). The omphalocele contained part of the liver, four cysts filled with translucent liquid, stomach, and part of the intestines. Upper extremities were normal, whereas left lower was slightly deformed, rotated inwards, and in the shape of a clubfoot. Internal sex organs were stretched over an enlarged urinary bladder. The anus was imperforated, the intestines were blind ending shortly behind the stomach (Fig. 10).

**Case 11 (GD-09)**

Mother, age 26 years at delivery, primipara, 17<sup>th</sup> week of gestation. Stillborn male foetus with autolysis of the second degree (the skull was not examined) – 17 cm long, weight 180 g. The skin on the non-symmetric head ruptured over deformed cerebral tissue with abnormal ventricle system. Lack of left eyeball. Bilateral cleft lip, on the right side cleft palate progressing to maxilla and infraorbital region. The nose very small and deformed. The ruptured omphalocele contained liver, stomach, and part of the intestines. The vertebral column rotated to the right and bent forward. The right lung on gross examination was normal, whereas the left was small (1 cm) and unilobar. Left forearm shortened, lack of fifth finger in the palm, syndactyly between fingers third and fourth. Both feet in the shape of clubfoot, with the left additionally rotated inwards (Fig. 11).

**Summary**

Table I presents comparison of all analysed cases.

The age of mothers in analysed group ranged from 23 to 41 years, with one exception, where there is no data (case 2). The gestational age varied from 16 to 37 years.

The majority of the cases (9/11) can be described as the first phenotypical type, according to Russo (type I), with craniofacial defects. Limb abnormalities were noted in 10 out of 11 cases. Apart from the case reported by Paul Portal (case 2) all foetuses presented abdominal wall defect, in two cases presenting thoracic wall defects as well. The pathology within the central nervous system was present in five out of nine cases (two were sectioned sine capite), in one case hydrocephaly was accompanied by sacral myelomeningocele. The same number (five out of ten) showed the deformation of the vertebral column, but it was not consistent with exencephaly or encephalocele. Two-vessel umbilical cord was reported only in two cases.

What is most worth mentioning is the fact that two cases (1 and 8) were live-born and both came from multiple pregnancies.

**Discussion**

Limb body wall complex is a very rare fatal foetal congenital disorder, usually resulting in intrauterine death of the foetus, or in cases of liveborn the demise comes shortly after delivery. Reported cases so far show no predilection as to sex of the foetus or the age of parents [19], although in our material seven cases were male, with two not given, and one diagnosed with hermaphroditism. It occurs sporadically, with the normal karyotype of the foetus (in most cases), and the incidence is quoted as 1 : 10,000 to 1 : 42,000 depending on the authors [16, 17].

**Table I.** Comparison of all analysed cases

	J. OELHAF CASE 1	P. PORTAL CASE 2	CASE 3	CASE 4	CASE 5	CASE 6	CASE 7	CASE 8	CASE 9	CASE 10	CASE 11
Maternal age	25	ND	28	26	23	27	41	34	31	28	26
Gravidity	1	ND	1	2	1	3	2	2	2	3	1
Single/Multiple	M	S	S	S	S	S	S	M	S	M	S
Gestational age	28-32 (?)	ND	37	20	31	21	20	32	17	16	17
Sex	ND	ND	M	M	M	M	M	M	M/F	F	M
Exencephaly or encephalocele	+	+	-	+	-	-	-	+	ND	ND	+
Amniotic adhesions	ND	+	+	ND	ND	ND	ND	+	ND	ND	ND
Craniofacial defects	+	+	-	+	+	+	+	+	-	+	+
Thoracic wall defect	+	ND	-	+	-	-	-	-	-	-	-
Abdominal body wall defect	+	ND	+	+	+	+	+	+	+	+	+
Limb abnormalities	+	+	+	+	+	-	+	+	+	+	+
Low-set ears	-	ND	-	+	-	+	+	-	-	-	-
Abnormal cardiac shape	+	ND	-	-	-	+	-	+	+	-	-
Two-vessel umbilical cord	-	ND	+	-	-	+	-	-	-	+	+
Hypoplastic lung(s)	+	ND	+	-	-	-	+	+	+	-	+
Deformation of the vertebral column	+	ND	+	-	-	-	+	+	-	-	+
Urogenital anomalies	+	ND	-	-	-	+	-	-	+	+	+
LBWC type according to Russo	I	I	II	I	I	I	I	I	II	I	I

(+) present; (-) absent; ND – no data

The PubMed database quotes 159 papers published up to now concerning the analysed complex, presenting altogether about 250 cases. Apart from several manuscripts (Luehr – 11 cases [18], Russo – eight cases [13], Bhat – three cases [19]), usually only single cases are reported.

Hippocrates was the first to propose that foetal deformities can be the consequence of a small or just insufficient uterine cavity, other authors propose that the short umbilical cord and/or amniotic bands (ABS – amniotic band syndrome) is/are responsible for at least some of the malformations, but the full origin of this complex remains unestablished. There are three mechanisms that have been proposed in LBWC so far, but they do not necessarily have to be taken into consideration separately [20, 21]. The first is the extrinsic theory proposed by Torpin, which considers early amniotic sac rupture followed by the formation of amniotic bands. They entrap and disrupt

limb formation leading to amputations and constriction bands. This model, however, does not explain the malformations of internal organs (liver, heart, kidneys) often found with LBWC [22]. The second proposed mechanism is vascular disruption causing the internal malformations seen in LBWC and the persistence of the extraembryonic coelom as the origin of amniotic bands [12, 23]. The third, intrinsic theory, proposed by Streeter, focuses on an abnormality in the germinal disk at the early development stage, resulting in final structural malformations. We should stress that the main aim for the latest researcher was the phenomenon of limb ring-constrictions [24]. All those theories were later supported by other researchers [13, 25].

It was also suggested that some specific gene mutation may also play a role in developing this complex. Cocaine smoking by mothers in the first trimester was also mentioned in two LBWC cases [3, 26, 27].

Nowadays in most cases LBWC is diagnosed antenatally. Currently there are no prenatal or postnatal possible treatment options for fetuses with severe malformations within this complex. The main features can usually be identified by routine obstetric ultrasound examination. However, performing it during the second trimester can be difficult due to quite often coexisting oligohydramnios, and the diagnosis may not be easy or accurate. In such cases magnetic resonance imaging (MRI) providing more informative images of congenital anomalies might be more helpful. The sonographic hallmarks are thoraco- and/or abdominoschisis, neural-tube abnormalities, severe scoliosis, limb deformations, positional deformities, and abnormalities of foetal membranes and umbilical cord [19, 28] (Figs. 12, 13).

It is very important to diagnose the complex as early as possible, because termination of such a pregnancy is suggested by all authors. In some cases, autopsy is not performed after the precise prenatal diagnosis (ultrasonography – US or MRI).

Basing on Russo's distinction of the LBWC phenotypes and our own material, we must focus on the range and severity of the spotted anomalies. CLPs (cleft lip, cleft palate) are quite common birth defects with various aetiology, and they affect one in 700 live births. And as a non-syndromic phenomenon (isolated), which occurs more often, the aetiology is rather unknown, the outcome in developed countries is rather good [29]. A similar situation may exist with isolated ABS. It carries an incidence ranging from 1/1200 to 1/15,000 live births. The clinical manifestation of these congenital anomalies covers constricting rings, tissue synechiae, and amputation of body parts distal to the constriction bands. The following treatment depends on the severity and localisation of the defect [30, 31].

In CLP and ABS the aetiology might be totally separate and thus not inflicting one another or be conjugated. Another conjunction is any of the above and abdominal and/or thoracic wall defect. Several separate entities constitute this group. Gastroschisis and omphalocele (exomphalos), as congenital wall defects, present unique challenges to clinicians. Gastroschisis occurs in three per 10,000 births, and omphalocele has an incidence of about two per 10,000 births. The aetiology is still under research; nevertheless, gastroschisis is probably less affected by genetics and more dependent on environmental factors (such as ischaemia), hence an association between gastroschisis and intestinal atresia. Omphalocele is probably caused by genetic factors. It is associated with anomalies such as Beckwith-Wiedemann syndrome, pentalogy of Cantrell, bladder/cloacal exstrophy, Down syndrome, and of course in LBWC [32, 33]. But as such, again, they do not have to be fatal.

Van Allen was reported by some authors as the first to describe LBWC as a separate entity. But even he declared in his work from 1987 that it “was the largest collection of fetuses with LBW complex published to date” – and definitely not the first one. Later he added that “the earliest cases were described by Portal [1685] and Ziegler [1896]...”, which now we dare to question [12 van Allen]. However, a few years ago we discovered that Joachim Oelhaf (1579-1639), the anatomist and lecturer from Gdansk Academic Gymnasium, reported such a case released in print in 1613. We have not so far found information of any LBWC case reported before Oelhaf. The information about this interesting, curious, and unusual complex of different defects was shared by Oelhaf in correspondence with Caspar Bauhin (1560-1624). The latter incorporated a full autopsy report sent in the second edition of the largest compendium for those times describing different malformations – *De hermaphroditorum monstrosorumque partuum natura ex theologorum, jureconsultorum, medicorum et rabbinorum sententia: libri duo* (1614). The single original engraving was slightly enhanced, as mentioned before it slightly differs from the original, but rather as far as the line width and thickness are concerned [6, 11].

The author of the second description of LBWC in history – Paul Portal (1630-1703) completed his medical studies in Paris. He changed his area of expertise from being a “companion surgeon” and performing autopsies (among other duties), to helping midwives in difficult and complicated labours. It is also where he helped to give birth to a monstrous foetus, later described in his manual. Paul Portal left us a vivid, gross description of another case, unfortunately without any depiction or engraving and without the description of the internal organs [15].

Ernst Ziegler (1849-1905), mentioned by van Allen, in paragraph 136 of his book “Faulty Closure of Abdominal and Thoracic Cavities, and the Accompanying Malformations”, described the list of possible defects of abdominal or thoracic wall that can be found during the examination or autopsy. Some of them may be applied to make the diagnose of LBWC, but may also be seen in several other malformations. However, the author did not put them in a separate syndrome or syndromes, but just listed the possible autopsy findings. Neither did he present a case-report, as Oelhaf did or Portal before him [15, 34, 35].

We added to the two first 17<sup>th</sup> century cases our own series. The Department of Obstetrics of Medical University of Gdansk is a referential perinatal care centre; hence, we had a chance to analyse so many cases of this rarely occurring complex. The first LBWC case reported by a Gdansk anatomist was hereby supported by a substantial series of contemporary Gdansk cases [11, 36].

Limb body wall complex is a rare birth defect covering several defects that can be spotted during



prenatal sonography examination and/or by a pathologist during autopsy. The range of anomalies is vast, and the exact pathomechanism is still not established, with some theories already reported. The discussion goes on, whether it should be divided into two separate entities, as Russo suggests (according to different probable mechanism), or it should stay under one wide diagnosis of LBWC with subtypes as it is in e.g. osteogenesis imperfecta or thanatophoric dysplasia. Eventually, both in phenotypic types of LBWC or cases presenting at least two out of three criteria proposed by van Allen, the prognosis is fatal. Maybe the name of the complex should be changed into LLBWC (lethal limb body wall complex) in contrast to coincidence (or associations according to the registry of birth defects) of two deformations (CLPs and ABS or others). Therefore, pathoclinical cooperation demands from obstetricians thorough prenatal examination, ultrasound, or MRI, later on confirmed by pathological analysis by obducent skilled in this narrow teratological part of perinatal autopsy, if possible supported by genetic tests to prove or dismiss the latest theories.

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