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Chapter 26

The human umbilical cord

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Disruption of the umbilical cord may be a source of harm to the developing fetus.[1] An estimated 30% of pregnancies carried to term have some type of umbilical–placental abnormality. How these findings affect the fetus and to what degree is unknown.

The most obvious unwanted effect of umbilical cord disruption is stillbirth. A review of stillbirth literature from 1930–98 suggests that 15% of all stillbirths with an identifiable cause were associated with umbilical cord complications.[2] Although difficult to prove in humans, umbilical cord complications are not uncommon in mammals, especially horses.[3] Umbilical cord complications occur throughout pregnancy. Javert and Barton reviewed 1000 cases of spontaneous abortion and reported that 56% had umbilical cord complications while controls had only 6%.[4] These authors noted that 'most of the cord complications were such as to compromise the fetal circulation so as to cause death *in utero*'. Frkovic *et al.* reviewed stillbirth between 20 and 36 weeks of gestation, of which 5.4% were associated with umbilical cord complications.[5] Defining the cause of death with supporting evidence varies greatly from country to country. Wang *et al.* describe a case of stillbirth where the diagnosis of umbilical cord complications is made based on careful pathologic observations.[6] Although the stillbirth did not have delivery evidence of cord entanglement, autopsy findings suggested entanglement and cord compression as the cause of death. Part of the difficulty in determining the cause of death is that the event of delivery itself can undo the puzzle of umbilical cord complications. Delivery can also confuse the statistics of umbilical cord complications by creating false impressions of cause and effect. True knots may not always be present prior to delivery. Conversely, a body loop may be passed down the body of the infant by vaginal delivery, creating the false impression of a knot which was not problematic or dangerous. Nuchal loops and shoulder loops may be misinterpreted and miscounted as nuchal cords.[7]

The most dramatic examples of umbilical cord complications include prolapsed umbilical cord, and rupture of the cord or a velamentous vessel. The chances of having a prolapsed cord during labour range from 1/239 cases to 2.4/1000 cases. The reported chances of fetal loss from a prolapsed cord are 8.6–49% and it appears that the ability to respond in a timely fashion may not improve outcomes.[8] The possibility of avoiding umbilical cord complications needs to be

investigated.[9] In monoamniotic twins, stillbirths often occur in one twin or both because of knots and cord encirclement, a risk of 75%.

Many variations of umbilical cord complications exist in which factors such as the time and degree of compression are important. Fetal gestation may also play a role. Compression of the umbilical cord obstructs blood flow and oxygen and releases stress factors from endocrine organs which contribute to the immediate danger to the fetus.[10,11] On the basis of this knowledge, examination and anticipation of other cord complications is important and not difficult. Table 26.1 illustrates the current awareness of umbilical cord complications.

Between 50% and 70.8% of stillbirths are unexplained.[12] It is possible that the criteria used to assess causality could be missing umbilical cord complications related to stillbirth.[13] A review of 40 unexplained stillbirths found a significant incidence of tight nuchal cords among intrapartum fetal deaths. Another study reported on perinatal deaths which may have been averted.[14] A dominant observation was failure to respond to abnormal heart-rate patterns and the authors suggested that the term fetuses were well equipped to survive until the terminal event and that such deaths could have been avoided.

The Perinatal Umbilical Cord Project

The possible morbidity associated with umbilical cord complications ranges from neurological damage to isolated organ injury, such as the bowel and kidney.[15,16] Indirect morbidity may be associated with meconium, premature labour and unrecognised fetal heart failure secondary to cord anomalies. In spite of numerous publications since the 1900s, no prospective well-defined study has been conducted to investigate umbilical cord complications. To initiate such a large study, a pilot observational review was undertaken: the Perinatal Umbilical Cord Project (PUCP), for which local ethics committee approval was obtained.[17–19]

In a community of low-risk suburban mothers, 4948 deliveries were retrospectively reviewed for umbilical cord complications; 14 were documented at delivery, with the possibility of more being unrecognised. All cases were stillborn and suggested a mortality rate of 2/1000 live births associated with such

Table 26.1. Estimated incidence of umbilical cord complications (UCC) and stillbirth

UCC	Incidence	Mortality
Nuchal loops	14–30%?	Unknown
Nuchal cords	14–30%	Unknown
Torsion	6–10%	20%
True knots	1%	6%
Body loops	1%	10%
Single umbilical artery	0.2–3.6%	7%
Velamentous insertion	0.54–2.2%	30%
Short cord	Unknown	Unknown

complications. Using this rate as a reference point, a goal of 1000 deliveries was established for the project.

Objective

The objective of the study was to conduct a prospective observational study, in a low-risk population (cohort), to determine the effect of umbilical cord complications on pregnancy.

Method

All patients accepted for management were informed of the PUCP. Preliminary observations suggested that ultrasound was accurate in identifying umbilical cord complications antenatally and later publications have confirmed this view.[20–24] Fetal monitoring is known to identify accurately fetal heart rate decelerations due to umbilical cord compression.[25,26]

In our study, 1250 pregnancies were accepted for management and 1064 deliveries occurred between 1989 and 1999. Routine screening tests were performed.

Antenatal ultrasound screening consisted of a 10–12-week vaginal ultrasound to confirm intrauterine pregnancy and pregnancy number. Abdominal scans at 20 weeks, 24 weeks, 28 weeks and 30 weeks documented the presence or absence of

Table 26.2. Umbilical cord complications (UCC) observed antenatally and postnatally

UCC	Observed antenatally	Observed postnatally	Expected
Nuchal loop	>220	120	Unknown
Nuchal cord	184	137	250
True knot	4	16	10
Torsion	10	18	0
Body loops	1	4	10
Ankle loops	1	5	0
Single umbilical artery	4	4	1
Velamentous	9	9	2
Marginal	23	23	10
Total UCC	456	336	319

Table 26.3. Umbilical cord complications (UCC) observed with poor outcome

UCC outcome	Expected	Observed
Cerebral palsy	2	none
Autism	none	2×nuchal cord
Meconium	10%	none
Stillbirth >28weeks	2–4	1×32 weeks; 3×nuchal cord
Persistent pulmonary hypertension of the newborn	none	2×nuchal cord
Hydrops	none	1×torsion

umbilical cord complications and continued as indicated with subsequent antenatal visits. Placental lie and placental umbilical cord insertion were noted.

Fetal heart rate monitoring was undertaken for five minutes at each antenatal visit, starting at 20 weeks. Prolonged non-stress tests were used when appropriate. Labour occurred spontaneously or by induction at term. Repeat caesarean section was performed one week prior to the due date. Patients were managed according to traditional obstetrical principles. Patients with identified umbilical cord complications were managed routinely and interventions were undertaken for obstetric indications and not according to ultrasound findings. All patients were informed of umbilical cord complications when these were documented. Fetal activity awareness was emphasised and follow-up of any complaints was immediate.

A total of 1064 deliveries occurred with an overall caesarean section rate of approximately 25%. This rate was average for our community standard of practice. No litigation was experienced and patients appreciated knowing the detail of their ultrasound examinations. Patients were helpful in monitoring identified cases of umbilical cord complications.

One case of Down syndrome, one duodenal atresia, one cleft lip, two polycystic kidneys and one heart defect (tetralogy of Fallot) were identified. Table 26.2 reviews the umbilical cord complications encountered antenatally and at delivery and Table 26.3 reviews the outcomes observed after delivery that may be associated with umbilical cord complications.

Duration and unravelling of nuchal cords has been discussed elsewhere.[27] Of the 184 fetuses identified with entangled nuchal cords, 47 became unravelled and were not present prior to delivery. This observation raises the question of antenatal injury without witnessing an aetiology. The difference in rate between nuchal loops and nuchal cords was approximately 50%. This observation may explain the difficulty of previous studies to delineate cause and effect. Also, a trend was noted in a few patients with repeat pregnancies over ten years with a tendency to repeat entanglement. Our experience summarised here is that one can expect to encounter a potentially harmful umbilical cord complication in at least 1/100 pregnancies managed to delivery. Ultrasound is helpful in identifying these umbilical cord complications antenatally. What remains to be determined by a larger study is the potential of identifying umbilical cord complications as an obstetric risk factor, for example at 36 weeks of gestation, and thus improve perinatal outcome.

Discussion

Early published studies would suggest that no significant harm results from umbilical cord complications.[28–32] More recently, McLennan *et al.* noted, in a study of 1115 vaginal deliveries, that no clinical indicators warned of stillbirth risk from umbilical cord complications.[33] Our impression suggests that fetal hiccups may be a clinical indicator of umbilical cord compression.[34] Also noticed was a consistent report by the umbilical cord complication group of decreased fetal movement and fetal heart rate umbilical cord compression patterns.[35] A retrospective review of 706 consecutive deliveries in a community hospital reported no poor outcomes in the group with umbilical cord complications.[36]

Table 26.4. Singleton term livebirths with true knots and associated placental findings

GP	Weight (g)	Gestational age (weeks)	Apgar scores	Umbilical length (cm)	Umbilical weight (g)	Placental weight (g)	Placental dimensions (cm)	Histology	UCC
32002	2948	37	8/8	38.1	24	493	16.1×2.7	Normal	K
10000	3260	39	4/6	64.0	54	600	22.0×1.6	Chorioangiosis	K
32002	3969	42	8/8	87.0	80	744	22.0×1.3	Immature	K/NC
54004	4167	43	8/9	62.3	51	638	22.0×2.7	Immature	K
21001	3402	41	8/9	65.5	94	648	20.0×3.6	Normal	K/BL
43003	3884	37	8/8	84.1	98	607	20.7×4.2	Normal	K/NC
54004	3317	40	8/9	81.7	80	645	21.4×3.4	Immature	K
21001	2742	40	8/9	70.5	42	602	20.0×2.4	Chorioangiosis	K/PR
21001	????	40	8/9	66.5	70	470	19.5×1.4	Immature	K
41111	2807	38	8/9	54.5	45	401	17.0×2.0	Normal	K
10000	3147	38	8/8	47.9	34	492	20.0×2.6	Normal	K2/NC2
53103	3856	38	8/9	47.3	59	532	21.7×3.7	Immature	K/NC
20010	3147	39	8/9	48.7	56	390	19.7×2.4	Normal	K2

BL=body loop, GP=gravity parity; Immature=immature placenta with enlarged hypervascular villi, K=true knot, NC=nuchal cord, PR=prolapse cord, UCC =umbilical cord complication

Table 26.5. Umbilical cord accident: time of death

Patient	Gestation (weeks)[a]	Fetal movement 7 p.m. to 12 midnight	Fetal movement after 7 a.m.	Umbilical cord accident
1	38	+	–	NC×2
2	36	+	–	NC×2; TN
3	36	+	–	TN
4	39	+	–	NC×2
5	38	+	–	NC×2
6	39	+	–	TN×2
7	39	+	–	NC×1; TN
8	38	+	–	NC×2
9	37	+	–	Torsion
10	38	+	–	NC×1
11	32	+	–	NC×1; BL×2
12	36	+	–	AL×1; 183 cm
13	38	+	–	TN
14	36	+	–	NC×1
15	40	+	–	NC×2; AL×1
16	39	+	–	BL? (meconium)
17	39	+	–	NC×3; TN
18	32	+	–	NC×3
19	40	+	–	TN
20	32	+	–	Torsion
21	39	+	–	Cord compression
22	40	+	Nap 3 p.m.	FM–; TN
23	38	+	–	NC×1; TN×1; BL
24	39	+	–	NC×5
25	38	+	–	NC×2
26	38	+	–	NC×4
27	39	+	–	NC; BL; SB×2
28	39	+	Nap 1 p.m.	NC×2; TN
29	36	+	–	NC×1
30	35	+	–	Abnormal insertion
31	40	+	–	NC×1; BL×3
32	38	+	–	Torsion
33	39	+	–	NC×4; AL×1 ;LL
34	39	+	–	NC×1; pain 3 a.m.
35	40	+	–	LL×3; constriction
36	39	+	–	TN×1
37	39	+	–	AL×2 (meconium)
38	39	+	–	Cord rupture
39	40	+	–	NC×1; AL×1
40	40	+	–	NC×5
41	37	+	–	Rupture; FI
42	39	+	–	Haematoma; pain 5 a.m.
43	38	+	–	NC×1
44	38	+	–	NC×1
45	39	+	–	Marginal insertion
46	33	+	–	Cord haematoma
47	33	+	–	NC ×3; TN×1
48	36	+	–	NC×1
49	39	+	–	Cord haematoma
50	38	+	–	Torsion, constriction
51	37	+	–	Cord compression
52	35	+	–	AL
53	40	+	–	TN
54	38	+	–	NC×1; AL×1
55	39	+	–	NC×2 (previous NC)
56	30	+	–	NC×1; AL×1

Table 26.5. *Continued*

Patient	Gestation (weeks)[a]	Fetal movement 7 p.m. to 12 midnight	Fetal movement after 7 a.m.	Umbilical cord accident
57	40	+	–	BL×1; AL×1
58	38	+	–	NC×2
59	39	+	–	NC×1
60	37	+	–	NC×3
61	38	+	–	NC×2 (fetal heart rate +)
62	39	+	–	Anomaly
63	41	+	–	Cord compression
64	39	+	–	TN×1 (repeat entanglement)
65	37	+	–	TN×1; AL×1
66	39	+	–	NC×2; BL×1
67	37?	+	Nap 1 p.m.	TN×2; NC×2
68	38	+	–	Cord prolapse
69	41	+	–	NC×1; AL×1
70	28?	+	–	NC×3; BL×2
71	37	+	–	NC×1
72	39	+	–	Two vessel
73	36	+	?	Two vessel
74	38	+	–	NC×1BL×1; LL
75	39	+	–	NC×3
76	29	+	–	Constriction
77	36	+	–	NC×2; AL×1
78	38	+	–	Haemorrhage
79	39	+	–	TN×1
80	26	+	–	TN×1
81	38	+	–	TN×2
82	37	+	–	NC×2; AL×1
83	40	+	–	TN×1
84	37	+	–	Cord prolapse
85	40	+	?	NC×3; BL×1
86	38	+	–	AL; constriction
87	38	+	–	Cord compression
88	26	+	–	Torsion
89	35	+	–	NC×1; constriction
90	35	+	–	NC×1
91	28	+	Nap 3 p.m.	Torsion; BL
92	32	+	–	BL×1
93	39	+	–	NC×2;BL×1; AL
94	40	+	–	NC×1; <40 cm
95	39	+	–	NC×2
96	40	+	–	NC×3; TN×1
97	40	+	–	Haemorrhage
98	40	+	?	NC×2; BL×1; TN
99	26	+	–	Torsion
100	38	+	–	TN×1

[a] Average gestational age approximately 38 weeks; AL=ankle loop; BL=body loop; FI=furcate insertion; FM–=fetal movement negative; LL=leg loop; NC=nuchal cord; SB=stillbirth; TN=true knot

Larson *et al.* retrospectively reviewed singleton, vertex term cases with multiple nuchal loops and noticed increased meconium and abnormal fetal heart rate patterns.[37] Our experience was negative for meconium and may relate to delivery by LMP due date.

Torsion was not recognised in these studies.[38,39] The concept of torsion is an integral part of how nuchal cords and true knots are formed. Another discrepancy in previous studies is our observation of type A and type B nuchal cords.[40] These two groups must be studied separately, just as nuchal loops must be distinguished from nuchal cords. Crawford first recognised the need for a definition in 1962; he described a nuchal cord as 'the condition in which the umbilical cord is wound at least once around the neck of the fetus'.[41] This initial description was applied to a study which led to the following conclusion: '[the] nuchal cord is well recognised as being commonly associated with fetal distress and neonatal depression. It is all the more remarkable, therefore, that little work has been published to demonstrate the incidence of the condition, and to analyse its effects during labour and delivery'.

There are several consequences that can occur in fetuses with umbilical cord complications antenatally which resolve and in fetuses experiencing labour with umbilical cord complications. Other than stillbirth, recent studies have noted effects from umbilical cord complications which may include admission to a neonatal intensive care unit (but no longer hospital stays) when compared to controls.[42,43]

Neurological damage is the worst morbidity outcome. Manning *et al.* reviewed 84 947 live births.[18] In a low-risk untested group, 4.74/1000 cases of cerebral palsy were noted compared to the tested high-risk group rate of 2/1000. Antenatal asphyxia was considered a potentially avoidable cause of cerebral palsy, especially when biophysical profile testing is considered. The PUCP observed no case of cerebral palsy in either the group with umbilical cord complications or the group without such complications. However, two cases of autism were suspicious for antenatal cord compression as noted by ultrasound and antenatal nonstress tests. Nelson and Grether retrospectively reviewed cases for cerebral palsy and noted an association with tight nuchal cords,[44] although antenatal ultrasound confirmation of tightness was not available. The cerebral palsy rate was 1/1500. In addition, 39% of children with unexplained cerebral palsy, 47% of children with quadriplegic cerebral palsy and 19% of control cases had umbilical cord encirclement.

Clapp *et al.* reviewed nuchal cord cases and their neurodevelopmental performance over a period of one year. They suggested a significant association with subclinical neurological deficits.[45] The Physicians' Insurance Association of America reviewed neurological damage claims and found a significant association with nuchal cords.[46] O'Callaghan *et al.* suggested that obstetric complications may play a role in nonfamilial schizophrenia.[47] Another study suggested that, in nonfamilial schizophrenia, umbilical cord knots and encirclement was related to development in 23.7% of cases.[48]

As the problem of umbilical cord complication is studied it may become possible to identify the fetus at risk and to manage the fetus successfully to avoid injury or stillbirth. Various authors have described the ability to identify umbilical cord complications with ultrasound as an initial step in its management.[49–54] Sherer and Manning, in a recent editorial, suggested that ultrasound should be considered at 36 weeks to scan for umbilical cord complications.[55]

Once born, the fetus, umbilical cord and placenta should be carefully examined for signs of subtle compromise. Chorioangiosis (a placental change noted by hypervascularity) is one placental finding which may imply cord compression leading to ischaemia.[56] Table 26.4 shows a review of cases of true knots at the Pregnancy Institute, which suggests that this may be a helpful indicator.

Table 26.5 lists the stillbirths associated with umbilical cord complications (>26 weeks) in 100 consecutive interviews from parents with stillbirth loss from umbilical cord complications. This unique list, which exceeds any known published data on stillbirth attributed to umbilical cord complications, suggests that the events may be occurring during maternal sleep.[57,58] These data are, of course, not part of a controlled study but the importance of presenting the information is the possibility that these patient insights may lead to clues to the aetiology of how these normal fetuses die. What is needed is a careful interview process with the family when an umbilical cord complication is encountered. It may be worthwhile exploring the possible link with stillbirth during maternal sleep. Good controlled research studies are required to determine whether umbilical cord complications are indeed an important contributor to perinatal morbidity and mortality.

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