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Cord abnormalities, structural lesions, and cord "accidents"

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As the umbilical cord is the lifeline of the fetus, obstruction or disruption of blood flow through the umbilical vessels can lead to severe fetal compromise. Obstruction is usually mechanical in nature and is associated with compression of the umbilical cord and umbilical vessels. Disruption of umbilical or fetal vessels is usually traumatic in origin. These conditions have in common a loss of blood flow to the fetus and an association with adverse perinatal outcome.

Conditions such as abnormal cord length, abnormal coiling, knots, entanglements, constrictions, prolapse and velamentous vessels may lead to cord compression and subsequent diminished blood flow in umbilical vessels. The umbilical vessels may be compressed by fetal parts, against the cervix or by an abnormal configuration of the cord itself. Not surprisingly, these conditions have been associated with fetal demise and adverse perinatal outcome.[1-4] Disruption of the umbilical cord or fetal vessels is usually traumatic in nature and is associated with some degree of fetal hemorrhage. Vascular disruption may occur when excessive traction is used on a short cord[5] or on a placenta with abnormal adherence to the uterus due to a placenta accreta. Disruption may also occur from pathologic processes rendering the cord more friable such as necrotizing funisitis,[6] meconium associated damage,[7] aneurysms[8] and hemangiomas[8] or from direct trauma due to fetal blood sampling or amniocentesis.[4] If large vessels are disrupted, severe fetal hemorrhage can develop relatively quickly, with the potential for acute hypovolemia and circulatory collapse, while hemorrhage from small vessels tends to be more chronic but still may be quite significant. These conditions have also been associated with demise and significant neurologic damage.[4-9]

Cord compression: General considerations

Mechanical obstruction of blood flow through the umbilical cord may occur secondary to any type of force that compresses umbilical vessels.[1-9] The fetus itself may compress the cord when there are cord entanglements, membranous vessels, and cord prolapse. Compression may arise from an abnormal configuration of the cord such as knots, abnormal coiling, abnormal length, or constrictions. Often these structural abnormalities are linked; for example, entanglements and knots are frequently seen in long cords and excessive coiling is often seen with constrictions. These conditions are generally present for many weeks or months and so may cause chronic obstruction of blood flow. However, acute obstruction is also possible when there is an acute progression at or near the time of delivery. This is the case when a true knot or entanglement tightens as the infant descends down the birth canal or when membranous vessels become compressed after membranes rupture and there is loss of the cushioning effect of the amniotic fluid. If obstruction is

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complete, fetal death will be the eventual result, whereas lesser degrees of obstruction can lead to severe neurologic injury.[2,4,9-13] This is consistent with animal studies in which fetal lambs subjected to intermittent partial cord occlusion develop cerebral necrosis and serious fetal neurologic damage.[14] Chronic partial obstruction can also lead to fetal growth restriction. Abnormally coiled cords, abnormally short or long cords, velamentous cord insertions, constrictions, true knots, cord entanglement, and cord prolapse have all been associated with an increased risk of fetal demise, neurologic injury, or abnormal developmental outcome.[1,2,5,6,8-11,13] With the exception of cord entanglement and prolapse, these lesions are easily diagnosed by gross examination of the placenta.

In acute compression, the umbilical vein, being more distensible, will be compressed initially and to a greater degree than the arteries. This leads to vascular congestion in the placenta, and if severe, to hypovolemia and anemia in the fetus. Doppler studies have confirmed that cord obstruction and compression cause impeded venous return.[15] Decreased venous return of oxygenated blood from the placenta will result in distension of the umbilical vessels, particularly the vein, tributaries of the umbilical vein in the chorionic plate, and the villous capillaries. Direct compression of the cord by fetal parts or the cervix may also cause nonspecific damage and degenerative change of Wharton's jelly and the umbilical vessels. Rarely, severe acute compression results in hemorrhage, thrombosis, and even rupture of the cord. Although the above findings are visible on microscopic examination, they are nonspecific and therefore do not enable the definitive diagnosis of cord compression, per se, or indicate the underlying cause of the compression.

Chronic cord compression develops from the same mechanical forces that lead to acute compression; however, the pathologic changes are more clearly delineated. Chronic obstruction of blood flow through the venous circulation initially leads to venous stasis and may ultimately lead to endothelial damage and subsequent fetal vascular thrombosis, further embarrassing blood supply to the fetus. Thrombi in umbilical vessels and fetal circulation can occur secondary to any process associated with cord compression or decreased venous return. The above described cord abnormalities have not only been associated with poor outcome, but with fetal thrombosis, and fetal thrombosis has itself been associated with poor outcome and pathologic lesions in the brain.[9,10,16,17]

Thrombosis in umbilical vessels can be identified by careful gross examination of the placenta. Recent thrombi appear as dark, clotted blood within the vessel, whereas older thrombi may be visible as white streaks in the vessels, which is due to the presence of calcification. Although calcification of the umbilical cord has classically been associated with acute necrotizing funisitis and syphilis infection, it is more commonly due to thrombosis. Venous thrombosis is more common than arterial thrombosis, but the latter is more often lethal.[17] The appearance of thrombi in the cord is often subtle and difficult to identify by external examination. Therefore, serial sectioning is recommended in all areas of cord discoloration. Areas of previous cord clamping should be avoided, as these are associated with artifactual hemorrhage and disruption. Often, only the vague impression of a clamp is visible, so careful gross inspection is necessary to rule this out. Thrombi in umbilical vessels are much less common than those in the large vessels of the chorionic plate and its tributaries. On microscopic examination, recent thrombi can be occlusive or nonocclusive and contain primarily fibrin and clotted blood (Figure 1A). More remote thrombi may show organization in the form of intramural calcification, a finding indicative of duration of many weeks (Figure 1B).

Umbilical cord coiling and constriction

The umbilical cord is usually twisted or coiled counterclockwise, to the left, with a left to right ratio of about 7:1.[4]

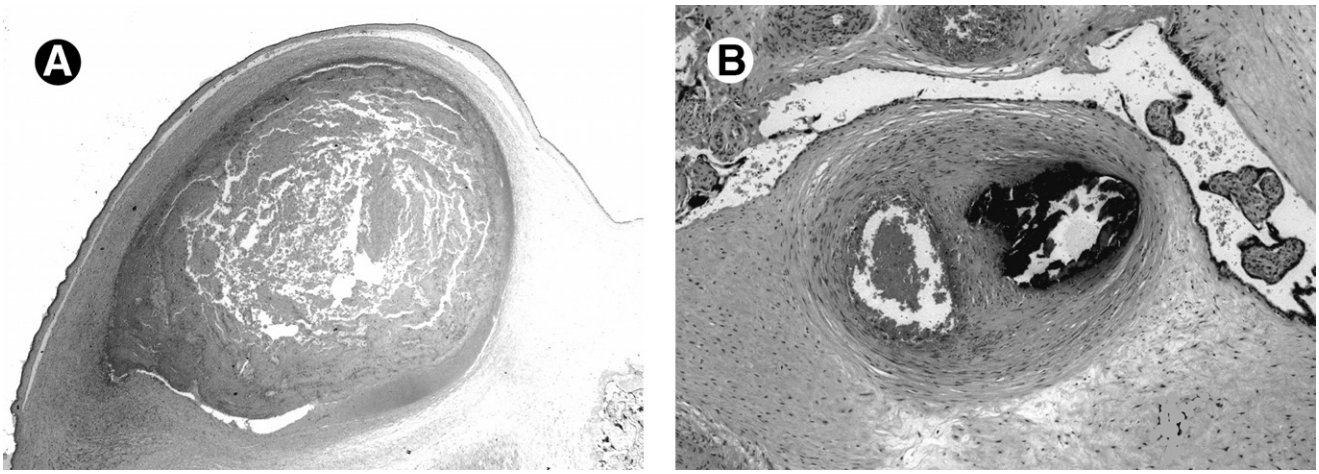

Figure 1 Fetal thrombotic vasculopathy. (A) Histologic section of a large tributary of an umbilical vessel with a recent occlusive thrombus consisting mostly of fibrin. H&E; original magnification 4×. (B) Small fetal stem vessel with calcified mural thrombus indicative of a longstanding event. H&E; original magnification 40×.