**Journal of Neurology and Psychology Research**

**RESEARCH** **Open Access**

# Representative Neurological Malformations in Pregnancy

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**Received date:** Feb 14, 2019, **Accepted date:** July 16, 2019, **Published date:** July 18, 2019



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## Abstract

The biggest fear of future parents is related to the possible existence of some anomaly in pregnancy. Congenital malformations are a more frequent cause of perinatal morbidity and mortality.

The most important goals and tasks of perinatal medicine are early diagnosis of fetal anomalies and chromosomal disorders, which means that their early detection, possibly treatment during pregnancy or after delivery, or an termination of pregnancy i fit is life-incompatible anomaly. This concept is called "active gestation of pregnancy" which includes: ultrasonic diagnosis, cytogenetic fetal tissue analysis, fetoscopy, amniography and screening test for neural tube defect and genetic abnormalities.

All neurological malformations in pregnancy are related with Central nervous system. When they happen, key issues are treatment and health care.

Key words: Pregnancy; Malformation; Health

## Introduction

Central nervous system (CNS) infections refer to the involvement of CNS tissues and their surroundings by infectious agents including viruses, bacteria, and parasites [1]. Of these, bacterial meningitis poses the most serious threat to neonates and children, both in terms of numbers of cases affected and mortality rates.

CNS infections especially meningitis are serious life-threatening diseases in neonates and children. Their incidence and type differ according to maturity and also differ in various geographic regions. In neonatal bacterial meningitis, Group B streptococci (GBS; Streptococcus agalactiae), Eschericia coli, Gram-negative bacilli other than E. coli, Haemophilus influenzae, and Listeria monocytogenes are the most important causative organisms in most of the world; in Africa and Asia, other bacteria, including

Streptococcus pneumoniae and salmonella species are also important. As globalization increases, infectious agents are less restricted geographically. Poverty and malnutrition increase susceptibility to infection. Morbidity and mortality vary, indicating differences in health care provision. Multiresistant organisms also contribute to morbidity. In infants and older children, Neisseria meningitidis, H.influenzae (type b) and Streptococcus pneumoniae are the more common responsible organisms.

Immunodeficiency is a major factor in childhood CNS infections. HIV infection, chemotherapy and bone marrow transplant in childhood malignancies and immune suppression in organtran splantation bring the risk of fulminant bacterial sepsis, opportunistic viral, parasitic, and fungal infections.

The skull and spine, meninges, blood–brain and blood–cerebrospinal fluid barriers protect the CNS from entry of organisms. Once these barriers are breached, infection can spread rapidly through the cerebrospinal fluid because the CNS is largely remote from immune protection. The anatomical planes also localize infection (e.g., epidural abscess, subdural empyema, meningitis, encephalitis, brain abscess). Overlap occurs; for example, many viruses cause a meningoencephalomyelitis (inflammation of meninges, brain, and spinal cord), and listeriosis causes meningitis, brainstem encephalitis, andabscesses.

A wide variety of morphologic and functional abnormalities have been identified in the central nervous systems of patients with chromosomal defects [2]. These are reviewed for the more commonly encountered karyotypes, with emphasis given to those aberrations in chromosome number (e.g., trisomy, monosomy) or chromosome morphology (i.e., large deletions andduplications) that affect the CNS. Disorders associated with mosaicism, lesser chromosomal changes (including translocations), or single gene mutations are not included.

Chromosomal changes are encountered in early pregnancy loss, but their true incidence is hard to determine. Acommonly used estimate is 50%. Alterations like trisomy 16 (estimated to occur in 1% of all conceptuses) are unlikely to come to the attention of neuropathologists because of early fetal demise.

Affected individuals and/or their families desire to understand both present and future issues surrounding their condition. Families are concerned about implications for present and future care, as well as prevention, recurrence risk, and family planning. Neuropathologists should be integral members of the team that provides information to patients and their families. Specialists will serve their patients well by providing information that is of direct use to the genetic counselor. In addition to written reports, the pathologist should provide photographs, especially of external phenotypic features (face, head, hands, and so forth), that have diagnostic value.

If there were no symptoms at all suggestive of neurological disease, it is usual to perform a quick examination of the nervous system, and if this examination is normal, then the nervous system is not examined further [3]. There will have been an opportunity to note the patient's posture and gait in the consulting room (or as the patient moves around the bed on the ward). If the patient's face looks normal and moves normally during speech, then there is unlikely to be a cranial nerve abnormality. The patient is then asked to hold both arms out to assess posture, to perform a 'finger-nose' test, to tap each hand on the other in turn, to 'unscrew door knobs', to tap each foot on the floor (or the examiner's hand if in bed), and then to do a 'heel-shin' test with each leg. Finally, reflexes are tested in the arms and legs. If all these are normal (and as emphasized already, there are no symptoms of neurological disorder), then the nervous system is not examined further. If there is a symptom or sign of neurological disorder, then the nervous system has to be examined carefully perhaps beginning with the territory under suspicion.

In adults, the normal resting intracranial pressure (ICP) is 0–10 mm Hg [4]. ICP may rise to 50 mm Hg or so during straining or sneezing, with no impairment in function. It is not, therefore, ICP alone that is important but rather the interpretation of the measurement in pathological conditions. Many of the clinicopathological changes associated with brain injury are the result of pressure differences between the intracranial compartments, with consequent shift of brain structures, rather than the absolute level of ICP.

The deterioration in conscious level accompanying elevation in ICP is probably caused by downward displacement of the diencephalon and midbrain structures. Herniation of the temporal lobe between the brainstem and the tentorial edge into the posterior fossa (tentorial or uncal herniation) causes pupillary dilatation, ptosis, limitation of upgaze and extensor posturing. Tonsillar herniation occurs when the tonsils of the cerebellum herniate through the foramen magnum into the spinal canal. This causes compression of the midbrain with changes in blood pressure, pulse rate and respiratory pattern. Cushing's response, the combination of hypertension and bradycardia, is seen in roughly 1/3 of cases of tonsillar herniation.

## Antenatal Malformations

Encephaloclastic lesions of the second and third trimesters disrupt normal development and are to be distinguished from primary malformations, which result from an intrinsically abnormal developmental process [5]. There are two categories. Smooth-walled lesions originating in mid-gestationand associated with surrounding cortical disorganization include hydranencephaly (synonym, bubble brain), where a thin, translucent, membrane replaces much of the cerebral mantle; porencephaly, exhibiting one or more circumscribed defects in the cerebral wall, variably communicating with the ventricle; and an intermediate lesion of extensive bilateral defects, but with residual intact parasagittal cortex, imaginatively labelled basket brain. By contrast, multicystic encephalopathy refers to ragged defects of hemispheric gray and white matter unaccompanied by cortical malformation, with its origin in the third trimester or, rarely, postnatally.

In porencephaly, although survival to adulthood is possible, clinical manifestations are more usually severe mental restriction and blindness, seizures (less often), tetraplegia, and decerebrate rigidity. Generally, the involvement of the basal ganglia and hypothalamus in hydranencephaly results in impaired thermoregulation, disordered sleep, sucking and swallowing, and therefore early demise. If the basal ganglia are preserved, survival may be prolonged for a few years, but spasticity, epilepsy, and minimal psychomotor development are the rule. The head circumference, usually normal at birth, may increase over the first few months, the hydrocephalus thought to result from obstruction at the aqueduct or foramen of Monro.

Ultrasound and magnetic resonance imaging are the most useful investigations, and are regularly employed for intrauterine evaluation. When these lesions are suspected, possible antecedents should be sought in the clinical history.

## Hormones

Hormones act to trigger transformations in the mother's and baby's body: in pregnancy, during birth, during breastfeeding [6]. The main task for the carer is to create an environment in which hormones that support the labour process can flow freely and abundantly. Hormones work in dyads, with each pair generating opposing effects. When they are in balance, allostasis occurs. They are the messengers in the body, and, with hormone receptors, they develop an intelligent language of the body. Hormones are triggered by, and trigger, emotional states, and transform them into physiological reactions. So, instead of concentrating on the effect of a specific hormone, it is important to be aware of the whole suite of active neurophysiological agents that orchestrate labour progress if they are allowed to work in harmony. The problem with synthetic hormones, such as artificial oxytocin, is that they play alone.

Labour hormones, such as endogenous oxytocin, endorphins and prolactin have an additional function in protecting the baby from danger and helping the fetal–neonatal adaptation process during and after birth. The expression of prolactin, in particular, seems to vary by mode of birth for term infants. These hormones are instrumental in that they prepare the process of bonding, love, happiness and wellbeing that form the ground on which the child can grow. Moreover, they activate a protecting behaviour in the parents towards the child, and damp down aggression. It has been demonstrated in rats that under the influence of high levels of endorphins and oxytocin, circuits of empathy and social capacities become activated in the maternal and neonatalbrain. Recent research has identified new neurological entities, called 'mirror neurons' that seem to allow the reflection and development of socialising behaviour in children, and between adults. The mirror-neuron system seems to develop fully over the first 12 months of a child's life. It is possible that, at the time when the awake and present mother meets her awake and present child for the first time, countless mirror neurons start to build up and work in the baby's brain. This may be part of the process that enables the child to feel empathy later in life, and in the development of social intelligence.

## Stroke

Stroke, also known as cerebrovascular accident (CVA), is an injury to the central nervous system that occurs due to problems with the vasculature (blood vessels) [7]. Stroke can occur anywhere in the central nervous system, including the brain, spinal cord, and retina.

About 87 % of strokes are ischemic, and about 13 % of strokes are hemorrhagic.

Stroke is a cerebrovascular disorder resulting from impairment of cerebral blood supply by occlusion (eg, by thrombi or emboli) or hemorrhage [8]. It is characterized by the abrupt onset of focal neurologic deficits. The clinical manifestation depends on the area of the brain served by the involved blood vessel. Stroke is the most common serious neurologic disorder in adults and occurs most frequently after age 60 years. The mortality rate is 40% within the first month, and 50% of patients who survive will require long-term special care.

Ischemic strokes, comprising thrombotic, embolic, and lacunar occlusions, account for over 80% of all strokes and result in cerebral ischemia or infarction. A variety of disorders of blood, blood vessels, and heart can cause occlusive strokes, but the most common by far are atherosclerotic disease (especially of the carotid and vertebrobasilar arteries) and cardiac abnormalities.

Hypertension, diabetes mellitus, TIAs, hyperlipidemia, smoking, family history, and use of oral contraceptives predispose to atherosclerotic disease. Cardiac disorders such as changing cardiac rhythms (especially atrial fibrillation), dyskinetic myocardium, and valvular heart disease are associated with increased risk for embolic strokes. Bleeding dyscrasias, hypercoagulable states, blood disorders (especially sickle cell disease), and vascular disorders are also associated with a risk for stroke. Carotid artery bruits in patients with TIAs or stroke suggest the possibility of emboli derived from atheromatous plaques.

## Fetal Distress

Fetal distress is a very broad term, which can be used in many clinical situations [9]. Although it is difficult to give a precise clinical definition, obstetricians usually use this term to indicate that the fetus is becoming hypoxic. Immediate delivery has to be considered, because neurological damage may occur when the fetal brain is deprived of oxygen.

The diagnosis of fetal distress based upon heart rate is imprecise because heart rate patterns are only a reflection of the efficiency of physiological mechanisms which depend on blood flow and oxygenation. Furthermore, activity of this control mechanism is influenced by the pre-existing state of fetal oxygenation as with chronic placental insufficiency. Therefore, heart rate patterns are now described as "reassuring" or "non reassuring": in the case of a "non reassuring" heart rate pattern, there is a risk of fetal distress.

The recognition of risk and the knowledge of appropriate measures to treat fetal distress are of the utmost importance. Antepartum fetal testing is used to assess hypoxia in highrisk pregnancies, and monitoring during labor supplies information on the status of the fetus prior to birth.

## Health Care

Critical and intensive care medicine is an integrated discipline that requires the clinician to examine a number of important basic interactions [10]. These include the interactions among organ systems, between the patient and his or her environment, and between the patient and life-support equipment. Gas exchange within the lung, for example, is dependent on the matching of ventilation and perfusion—in quantity, space, and time. Thus, neither the lungs nor the heart are solely responsible; rather, it is the cardiopulmonary interaction that determines the adequacy of gas exchange.

Critical care often entails providing advanced life support through the application of technology. Mechanical ventilation is a common example. Why is it that positive pressure ventilation and positive end-expiratory pressure (PEEP) can result in oliguria or reduction of cardiac output? Many times clinical assessments and your therapeutic plans will be directed at the interaction between the patient and technology; this represents a unique "physiology" in itself.

## Conclusion

Searching for anomalies is the most important task of a gynecologist in the first half of pregnancy. Contemporary perinatology increasingly moves the limit of discovery irregularities to the first trimester of pregnancy, when the

medically indicated abortion is far less traumatizing than in the late pregnancy.

The most common malformations are on the defects of the urogenital system and the central nervous system. Because of this, the kidneys and urinary bladder must be recognized at the end of the first and the beginning of the second trimester. It is important to show the proper brain and brain structures and the correct spinal column in which the spinal cord is located. It is also important to display parts of the digestive tract: the liver, the stomach and the small intestine. To visualize the blood vessels, color-and-power doppler is used.

Regardless of any possible malformations that may occur during pregnancy, each parent expects a healthy child to come to the world. A healthy child is a child capable of life outside from the uterus.

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