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Date: 29 Sep 2017

Sources Searched: Medline, Embase, DynaMed Plus.

Neurofibromatosis in Pregnancy

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Evidence Summary:

- Neurofibromatosis type 1 (NF1) is an autosomal dominant genetic disorder with highly variable manifestations and accounts for between 90-95% cases of NF.
- Neurofibromatosis type 2 (NF2) is characterized by bilateral vestibular schwannomas (previously termed acoustic neuromas) and meningiomas, as well as other brain and nerve tumors.
- Neurofibromas may grow in size and number during pregnancy which may lead to cord compression due to the enlargement of spinal neurofibromas or impeded delivery as a result of pelvic neurifibromas.
- A recent retrospective [register-based total population study](#) of 176 women (totalling 375 deliveries between 1987 and 2013) reported an increased risk for cesarean delivery and pregnancy complications, including hypertension, preeclampsia, poor fetal growth, placental abruption, maternal care for disproportion, and oligohydramnios among mothers with NF1.
- A [population-based retrospective cohort study](#) conducted in the USA, identified 1553 pregnancies associated with NF1 and reported that pregnancies of patients with NF1 were associated with gestational hypertension, preeclampsia, intrauterine growth restriction, cerebrovascular disease, preterm labour, and caesarean delivery.

Sources:

DynaMed Plus [Internet]. Ipswich (MA): EBSCO Information Services. 1995 - . Record No. 116711, Neurofibromatosis type 1; [updated 2015 Dec 21, cited **29/09/2017**]; [about 16 screens]. Available from <http://www.dynamed.com/login.aspx?direct=true&site=DynaMed&id=116711> . Registration and login required.

DynaMed Plus [Internet]. Ipswich (MA): EBSCO Information Services. 1995 - . Record No. 901367, Neurofibromatosis type 2; [updated 2016 Feb 08, cited: **29/09/2017**]; [about 14 screens]. Available from <http://www.dynamed.com/login.aspx?direct=true&site=DynaMed&id=901367> . Registration and login required.

1. The pregnancy in neurofibromatosis 1: A retrospective register-based total population study.

Author(s): Leppävirta, Jussi; Kallionpää, Roope A; Uusitalo, Elina; Vahlberg, Tero; Pöyhönen, Minna; Timonen, Susanna; Peltonen, Juha; Peltonen, Sirkku

Source: American journal of medical genetics. Part A; Oct 2017; vol. 173 (no. 10); p. 2641-2648

Publication Date: Oct 2017

Publication Type(s): Journal Article

PubMedID: 28815922

Full Text Link: <http://dx.doi.org/10.1002/ajmg.a.38372>

Abstract: The objective of this retrospective total population study was to form a view of the pregnancies of the patients with neurofibromatosis type 1 (NF1). A cohort of 1,410 Finnish patients with NF1 was acquired by searching NF1-related inpatient and outpatient hospital visits and confirming the diagnoses by reviewing the medical records. Ten matched control persons per patient with NF1 were collected from Population Register Centre. Study persons were linked to data from Medical Birth Register and Care Register for Health Care through the personal identity code. Cesarean deliveries, hypertension/preeclampsia, and placental abruptions were more common among mothers with NF1 with adjusted odds ratios of 2.24 (95%CI 1.63-3.07), 1.96 (95%CI 1.18-3.24), and 13.40 (95%CI 4.26-42.13), respectively. The adjusted mean pregnancy duration was 0.65 (95%CI 0.42-0.88) weeks shorter among the mothers with NF1 than in the control group consisting of non-NF1 mothers giving birth to a non-NF1 child. The pregnancies of non-NF1 mothers giving birth to a NF1 child were 0.43 (95%CI 0.24-0.62) weeks shorter than in the control group. In summary, NF1 of the mother was associated with a shortened pregnancy and increased pregnancy complications. Also, the NF1 of the fetus slightly shortened pregnancy. Since mothers with NF1 are at increased risk for pregnancy complications, careful evaluation of their pregnancies is warranted.

Database: Medline

2. Pheochromocytoma in neurofibromatosis type 1 during pregnancy.

Author(s): Remón-Ruiz, Pablo; Aliaga-Verdugo, Alberto; Guerrero-Vázquez, Raquel

Source: Gynecological endocrinology : the official journal of the International Society of Gynecological Endocrinology; Feb 2017; vol. 33 (no. 2); p. 93-95

Publication Date: Feb 2017

Publication Type(s): Case Reports Journal Article

PubMedID: 27908211

Abstract: Pregnant women with neurofibromatosis type 1 (NF-1) have increased complications during gestation, including hypertensive disorders that are sometimes caused by pheochromocytoma. Pheochromocytoma is an extremely rare condition during pregnancy, and the main clinical manifestation is hypertension. If not properly treated, pheochromocytoma has high maternal and fetal mortality rates. Early recognition and adequate clinical management before delivery have led to better outcomes in the last few decades. Despite the association of NF-1 and pheochromocytoma, there are few clinical reports of these two conditions in pregnant patients. We present a rare case of pheochromocytoma diagnosed during pregnancy in a patient with NF-1, and we describe the treatment and the obstetric and fetal outcomes. We also review other medical conditions related to NF-1 that complicated this patient's pregnancy.

Database: Medline

3. One case of severe preeclampsia who died from postpartum complications ten days after caesarian delivery

Author(s): Pacarada M.; Gashi A.M.; Beha A.; Obertinca B.

Source: Italian Journal of Gynaecology and Obstetrics; Sep 2016; vol. 28 (no. 4); p. 31-33

Publication Date: Sep 2016

Publication Type(s): Article

Abstract:Preeclampsia is clinically defined by hypertension and proteinuria, with or without pathologic edema that can happen after 20 week's gestation, but can happen well 4-6 weeks post partum. Worldwide, incidence of preeclampsia is 5-14 percent of all pregnancies, while severe preeclampsia can develop to about 25 percent of all cases of preeclampsia. Severe preeclampsia is a pathology that can often be complicated. This pathology may lead to liver and renal failure, disseminated intravascular coagulopathy (DIC), and central nervous system (CNS) abnormalities. In world, preeclampsia and eclampsia is responsible for about 14 percent of maternal deaths per year. We present a case, from our clinic, which has had serious complications after birth and that ended with the death of the patient. Despite the adequate management with the timely diagnosis and therapy, patient died ten days after Caesarian delivery. Copyright © 2015, Partner-Graf srl, Prato.

Database: EMBASE

4. Pre-eclampsia in neurofibromatosis: The anaesthetist's ordeal

Author(s): Balla S.; Osborn N.

Source: Anaesthesia; Sep 2016; vol. 71 ; p. 24

Publication Date: Sep 2016

Publication Type(s): Conference Abstract

Available at [Anaesthesia](#) - from Wiley Online Library Medicine and Nursing Collection 2017 - NHS

Abstract:Neurofibromatosis is a complex hereditary disease involving many organs and systems. A survey of the relevant literature suggests that management of the pregnant patient with neurofibromatosis is an obstetric challenge [1]. Here, we present a case report of a patient who had pre-eclampsia superimposed on a background of neurofibromatosis. A 27-year-old primigravida presented to the labour ward with a history of neurofibromatosis type 1 diagnosed in childhood. Clinical features identified were coffee coloured skin patches, extensive neurofibromatosis to skin over the back, chest and abdomen and decreased visual acuity with optic nerve head drusen. Pregnancy was uneventful till 36 weeks when symptoms of abdominal pain and high blood pressure prompted an urgent referral to the hospital. Features of severe pre-eclampsia with hepatic and renal involvement were confirmed, i. e. proteinuria, high urate levels, high bilirubin and alanine aminotransferase, low platelet count, hypertension and brisk reflexes. Treatment was commenced immediately, however a poor urine output and no signs of labour with continuing features of severe pre-eclampsia prompted an emergency lower segment caesarean section under a general anaesthetic. The intra-operative period was uneventful. Discussion Type 1 neurofibromatosis is the most common cancer predisposition syndrome affecting the nervous system with an incidence of 1 in 3000 worldwide [2]. Recent research has shown that 75% of neurofibromas carry progesterone receptors. During pregnancy, neurofibromas may grow in size and number and clinicians need to be aware of the risk of cord compression if spinal plexiform neurofibromas expand [3]. Anaesthetic implications relevant to our case were difficult intravenous and intra-arterial access, altered sensitivity to muscle relaxants, difficult airway and hypertension [4]. The patient also had not been seen by an anaesthetist during her pregnancy. Our patient had a Cormack Lehane grade 3 view on intubation and remained hypertensive despite treatment. Indeed, during pregnancy,

phaeochromocytoma may mimic pregnancy-induced hypertension and therefore the correct diagnosis is often made in the operating room or during post-mortem examination [5]. The first case report describing the successful use of epidural analgesia for labour in a parturient with neurofibromatosis was in 1995 [5]. Factors, which proved a deterrent to regional anaesthesia for us, were extensive neurofibromatosis, no previous scans to rule out intradural neurofibromas, absence of active labour and the urgency of blood pressure management. Hence, the management of a parturient with neurofibromatosis needs a multidisciplinary approach with careful planning starting from before pregnancy.

Database: EMBASE

5. Neurofibromatosis type 1-associated hypertension secondary to coarctation of the thoracic aorta.

Author(s): Mavani, Gaurang; Kesar, Vivek; Devita, Maria V; Rosenstock, Jordan L; Michelis, Michael F; Schwimmer, Joshua A

Source: Clinical kidney journal; Aug 2014; vol. 7 (no. 4); p. 394-395

Publication Date: Aug 2014

Publication Type(s): Journal Article

PubMedID: 25852916

Available at [Clinical kidney journal](#) - from Europe PubMed Central - Open Access

Abstract:Neurofibromatosis type 1 (NF-1), also known as von Recklinghausen's disease, is an autosomal dominant genetic disorder. NF-I vasculopathy has been used to describe various vascular malformations associated with NF-1. Secondary hypertension related to NF-1 vasculopathy has been reported because of renal artery stenosis, coarctation of the abdominal aorta and other vascular lesions; however, coarctation of the thoracic aorta has seldom been reported. We report the first case, to our knowledge, of isolated coarctation of thoracic aorta in a pregnant female with NF-1. Healthcare providers caring for patients with NF-1 should be aware of associated vascular complications.

Database: Medline

6. A Rare Case of Sudden Death Due to Hypotension during Cesarean Section in a Woman Suffering from Pheochromocytoma and Neurofibromatosis

Author(s): Cecchi R.; Frati P.; Cipolloni L.; Capri O.

Source: Journal of Forensic Sciences; Nov 2013; vol. 58 (no. 6); p. 1636-1639

Publication Date: Nov 2013

Publication Type(s): Article

PubMedID: 24117722

Available at [Journal of Forensic Sciences](#) - from Wiley Online Library Medicine and Nursing Collection 2017 - NHS

Abstract:Sudden death following acute hypotension due to an undiagnosed pheochromocytoma (PHEO) is a rare event. Moreover, histopathology of the myocardium in such cases is rarely reported. We present a case of a woman who died during delivery. A 37-year-old parturient, who was 38 weeks pregnant, suffering from neurofibromatosis underwent a cesarean section following peridural anesthesia. Acute hypotension, acute intra-operative pulmonary edema and supraventricular paroxysmal tachyarrhythmia occurred during delivery, followed by death. The autopsy revealed the presence of a PHEO, confirmed immunohistochemically with chromogranin-A (CgA), CD20 antibody (L26), anti-Keratocan antibody (KER-1) and neuron-specific enolase (NSE), and a PHEO-induced cardiomyopathy. The physiopathology of both stress-induced cardiomyopathy and PHEO-induced cardiomyopathy, as well as the role of anesthesia in provoking the death, are discussed. The association of an undiagnosed PHEO with neurofibromatosis as the cause of sudden death in pregnancy is an obstetric urgency that raises forensic pathology issues. © 2013 American Academy of Forensic Sciences.

Database: EMBASE

7. Neurofibromatosis type 1 and pregnancy complications: a population-based study.

Author(s): Terry, Anna R; Barker, Fred G; Leffert, Lisa; Bateman, Brian T; Souter, Irene; Plotkin, Scott R

Source: American journal of obstetrics and gynecology; Jul 2013; vol. 209 (no. 1); p. 46

Publication Date: Jul 2013

Publication Type(s): Research Support, Non-u.s. Gov't Journal Article

PubMedID: 23535241

Abstract:OBJECTIVEThe objective of the study was to determine whether vascular and other complications are more common in pregnant women with neurofibromatosis type 1 (NF1).STUDY DESIGNWe performed a population-based retrospective cohort study using the US Nationwide Inpatient Sample, 1988-2009, defining a cohort of pregnancy-related hospitalizations with an associated diagnosis of NF1 and comparing it with the control group not associated with NF1. Multivariable logistic regression was used to adjust for suspected confounders.RESULTSAmong 19 million pregnancy-related admissions between 1988 and 2009, we identified 1553 associated with NF1 (prevalence 0.008%). A diagnosis of NF1 in delivering mothers was associated with gestational hypertension (adjusted odds ratio [AOR], 1.6, 95% confidence interval [CI], 1.2-2.0), preeclampsia (AOR, 2.8, 95% CI, 2.3-3.4), intrauterine growth restriction (AOR, 4.6, 95% CI, 3.7-5.6), cerebrovascular disease (OR, 8.1, 95% CI, 2.6-25.4), preterm labor (AOR, 1.6, 95% CI, 1.4-1.9), and cesarean delivery (AOR, 2.0, 95% CI, 1.8-2.3). Women with NF1 were not significantly more likely to have deep venous thrombosis/pulmonary embolism, acute cardiac events, or stillbirth or to die during their hospitalizations compared with the general obstetric population.CONCLUSION NF1 was associated with increased maternal morbidity in pregnancy (including hypertensive and

cerebrovascular complications) but not increased maternal mortality. Obstetricians should be aware of the potential for increased antenatal and peripartum complications among women with NF1.

Database: Medline

8. Neurofibromatosis type1 and pregnancy

Author(s): Islam S.

Source: Archives of Disease in Childhood: Fetal and Neonatal Edition; Apr 2012; vol. 97

Publication Date: Apr 2012

Publication Type(s): Conference Abstract

Available at [Archives of Disease in Childhood: Fetal and Neonatal Edition](#) - from BMJ Journals - NHS

Abstract:Introduction and background Neurofibromatosis (NF) is a genetic disease that affects multiple organ systems. NF Type I (von Recklinghausen disease) is an autosomal dominant condition caused by mutation in NF1 tumour suppressor gene on chromosome17. There are different opinions about the implications of neurofibromatosis on pregnancy. Some authors have suggested an increased frequency of obstetric complications such as spontaneous miscarriage, preterm delivery, preeclampsia, intrauterine growth restriction, stillbirths and increased rate of caesarean section. However, others suggested that NF1 may not be associated with significant obstetric complications and may have normal pregnancy outcome. Pregnancy sometimes corresponds period of disease progression when cutaneous lesions might increase in size and number. Case summary We managed a 33 years old woman with type1 neurofibromatosis in her first pregnancy. She had cafe au lait patches and extensive cutaneous neurofibromata. An MRI showed multiple pelvic lesions and lesions within the spinal canal and foramina of all lower thoracic and lumbar vertebrae. She was reviewed by anaesthetists. It was suggested that if exit foramina are avoided regional analgesia should be safe for her. Due to aggravation of skin lesions the patient had induction of labour at 40weeks of gestation. She was seen by surgeons to assess risk of bleeding from perineal lesions it case of a perineal tear/episiotomy. She delivered vaginally a healthy male baby weighing 3.2kg with no problems. Conclusion This case shows that careful planning and multidisciplinary management can ensure normal obstetric outcome despite the aggravation of skin and pelvic lesions during pregnancy.

Database: EMBASE

9. Management of pregnancy in women with genetic disorders: Part 2: Inborn errors of metabolism, cystic fibrosis, neurofibromatosis type 1, and Turner syndrome in pregnancy.

Author(s): Chetty, Shilpa P; Shaffer, Brian L; Norton, Mary E

Source: Obstetrical & gynecological survey; Dec 2011; vol. 66 (no. 12); p. 765-776

Publication Date: Dec 2011

Publication Type(s): Journal Article Review

PubMedID: 22192461

Available at [Obstetrical & gynecological survey](#) - from Ovid (LWW Total Access Collection 2015 - Q1 with Neurology)

Abstract:UNLABELLEDWith early diagnosis and increasingly effective medical care, more women with genetic syndromes are undergoing pregnancy, often presenting challenges for providers. Each year more women with genetic disease reach childbearing age. Advances in assisted reproductive technology have enabled pregnancy in a cohort of woman who experience impaired fertility because of their underlying diagnosis. Management of these women requires health care providers from multiple specialties to provide coordinated care to optimize outcomes. Potentially, serious medical issues specific to each diagnosis may exist in the preconception, antepartum, intrapartum, and postpartum periods, all of which must be understood to allow timely diagnosis and treatment. The fetus may also face issues, both related to risk for inheritance of the genetic disorder observed in the mother as well as risks related to her chronic disease status. In this article, the second of a 2-part series, we will review the key issues for managing women with various inborn errors of metabolism during pregnancy. Additionally, we will discuss the care of women with Turner syndrome, neurofibromatosis type 1, and cystic fibrosis.TARGET AUDIENCEObstetricians & Gynecologists and Family Physicians.LEARNING OBJECTIVESAfter the completing the CME activity, physicians should be better able to classify the pulmonary and nutritional issues facing women with cystic fibrosis in pregnancy, assess the baseline evaluation that should take place in women with Turner syndrome, NF1 and cystic fibrosis before attempting pregnancy and evaluate the fetal risks that can be observed in women with untreated inborn errors of metabolism.

Database: Medline

10. Caring for the pregnant woman with neurofibromatosis.

Author(s): Gresham, Deborah D; Braunlin, Jeri Lynn; Vuckovich, Susan K

Source: MCN. The American journal of maternal child nursing; 2010; vol. 35 (no. 1); p. 18

Publication Date: 2010

Publication Type(s): Journal Article

PubMedID: 20032755

Available at [MCN. The American journal of maternal child nursing](#) - from Ovid (LWW Total Access Collection 2015 - Q1 with Neurology)

Abstract:Neurofibromatosis (NF) is one of the most common inherited single-gene disorders in humans, and is expressed as two distinct types: Type I (von Recklinghausen disease) and Type II, which occurs much less frequently. When it was first reported in the obstetric literature in 1906 by Brickner, neurofibromatosis was called "fibroma moluscum contagiosum." This article presents a case of a 20-year-old woman with NF Type I who became pregnant. The clinical challenges of NF and pregnancy are explored through the collaborative relationships between the patient and two clinical nurse specialists from obstetric and neuroscience nursing. The case study provides valuable information for nurses who want to provide evidence-based care.

Database: Medline

11. Neurofibromatosis and pregnancy: a report of maternal cardiopulmonary compromise.

Author(s): Nelson, David B; Greer, Laura; Wendel, George

Source: Obstetrics and gynecology; Aug 2010; vol. 116

Publication Date: Aug 2010

Publication Type(s): Case Reports Journal Article

PubMedID: 20664435

Available at [Obstetrics and gynecology](#) - from Ovid (LWW Total Access Collection 2015 - Q1 with Neurology)

Abstract:BACKGROUNDNeurofibromatosis is a rare pregnancy complication that can present with rapidly enlarging masses that have malignant potential.CASEA 19-year-old primagravid woman with a known history of neurofibromatosis presented in her first trimester with pulmonary complaints. A malignant mediastinal mass was diagnosed and resected, with additional treatment options declined. In less than 3 months, the patient presented with a recurrent mass that resulted in fatal airway compromise during the same gestation.CONCLUSIONNeurofibromatosis type 1 with malignant peripheral nerve sheath tumors complicating pregnancy requires an experienced, multidisciplinary team of care offering an aggressive evaluation to rule out malignant transformation or recurrence when there is any change in clinical status of a patient, as this may signal a potentially fatal change in the lesion.

Database: Medline

12. Sudden postpartum death: An unexpected complication of neurofibromatosis

Author(s): Boyum R.D.; Elliot J.N.; Sexton M.J.

Source: Laboratory Investigation; Jan 2009; vol. 89

Publication Date: Jan 2009

Publication Type(s): Conference Abstract

Available at [Laboratory Investigation](#) - from ProQuest (Hospital Premium Collection) - NHS Version

Abstract:Background: Neurofibromatosis type 1 (NF1) is associated with vascular abnormalities including stenosis, aneurysms, and vessel wall dysplasia, sometimes resulting in vessel rupture. NF1 has also been associated with spontaneous hemothorax and sudden death, and spontaneous hemothorax has been reported in pregnant patients with NF1. Furthermore, neurofibromas in patients with NF1 have been noted to increase in size during pregnancy. We present a case of NF1 resulting in maternal mortality secondary to mediastinal vessel rupture and subsequent massive hemothorax in the early postpartum period. Design: Autopsy was performed with internal gross and microscopic examination of the chest. Results: A 26-year-old with a known history NF1 delivered a healthy male infant following an uneventful pregnancy and labor course remarkable only for mild persistent elevation of the systolic blood pressure. At 27 hours postpartum, the patient complained of left sided pleuritic chest pain, exhibited signs of shock, and died despite extensive resuscitative efforts. Autopsy revealed left hemothorax, causing marked compression of the left lung. The middle and posterior mediastinum contained hemorrhage, and a ruptured arterial aneurysm was located in the mediastinum. The wall of the affected artery was focally necrotic. Adjacent to and completely surrounding the ruptured artery and nearby vessels and nerves was a diffuse neurofibroma (vascular neurofibromatosis) as confirmed by immunohistochemical studies. Granulation tissue was noted at the site of the rupture. Conclusions: The patient in this report died from a sudden massive hemothorax and associated respiratory compromise and hemorrhagic shock. The cause of the hemorrhage was rupture of an arterial aneurysm in the mediastinum. The aneurysm apparently formed due to ischemic necrosis of the arterial wall, secondary to encasement of the artery by a

diffuse neurofibroma. Granulation tissue at the site of rupture indicates that the initial hemorrhage occurred between 1 and 7 days prior to death, and the patient's rapid demise occurred after the initial, relatively slow, mediastinal hemorrhage extended into the pleural cavity and rapidly accelerated.

Database: EMBASE

13. Normal obstetric outcome in neurofibromatosis-1 complicating pregnancy

Author(s): Sangwan N.; Duhan N.

Source: JK Science; 2008; vol. 10 (no. 4); p. 197-198

Publication Date: 2008

Publication Type(s): Article

Abstract:Neurofibromatosis (NF), a genetic disorder, has increased risk of obstetric complications as well as aggravation of maternal disease. However, here is a case of Neurofibromatosis associated with normal obstetric outcome despite the aggravation of dermatological lesions.

Database: EMBASE

14. Guidelines for the diagnosis and management of individuals with neurofibromatosis

Author(s): Ferner R.E.; Huson S.M.; Evans D.G.; Thomas N.; Moss C.; Willshaw H.; Upadhyaya M.; Towers R.; Gleeson M.; Steiger C.; Kirby A.

Source: Journal of Medical Genetics; Feb 2007; vol. 44 (no. 2); p. 81-88

Publication Date: Feb 2007

Publication Type(s): Review

PubMedID: 17105749

Available at [Journal of Medical Genetics](#) - from Europe PubMed Central - Open Access

Abstract:Neurofibromatosis 1 (NF1) is a common neurocutaneous condition with an autosomal dominant pattern of inheritance. The complications are diverse and disease expression varies, even within families. Progress in molecular biology and neuroimaging and the development of mouse models have helped to elucidate the aetiology of NF1 and its clinical manifestations. Furthermore, these advances have raised the prospect of therapeutic intervention for this complex and distressing disease. Members of the United Kingdom Neurofibromatosis Association Clinical Advisory Board collaborated to produce a consensus statement on the current guidelines for diagnosis and management of NF1. The proposals are based on published clinical studies and on the pooled knowledge of experts in neurofibromatosis with experience of providing multidisciplinary clinical and molecular services for NF1 patients. The consensus statement discusses the diagnostic criteria, major differential diagnoses, clinical manifestations and the present strategies for monitoring and management of NF1 complications.

Database: EMBASE

15. Recent onset neurofibromatosis complicating eclampsia with maternal death: a case report.

Author(s): Agarwal, Umber; Dahiya, Pushpa; Sangwan, Krishna

Source: Archives of gynecology and obstetrics; Aug 2003; vol. 268 (no. 3); p. 241-242

Publication Date: Aug 2003

Publication Type(s): Case Reports Journal Article

PubMedID: 14653252

Available at [Archives of gynecology and obstetrics](#) - from SpringerLink

Abstract:Neurofibromatosis in pregnancy has been associated with poor perinatal outcome. A 30-year-old multigravida developed peripheral lesions of neurofibromatosis during the third month of pregnancy. She had eclampsia complicated with fatal left-sided massive intracerebral haemorrhage detected on computed tomography at 8.5 months gestation. Investigations were suggestive of HELLP syndrome. A still-born male baby was delivered. In spite of all supportive measures the patient died on the third postpartum day. We conclude that recent onset neurofibromatosis in current pregnancy should be considered as a predictor of potentially adverse maternal and fetal outcome and such pregnancies be managed in tertiary level referral centre.

Database: Medline

16. Neurofibromatosis in pregnancy. Maternal and perinatal outcome.

Author(s): Segal, D; Holcberg, G; Sapir, O; Sheiner, E; Mazor, M; Katz, M

Source: European journal of obstetrics, gynecology, and reproductive biology; May 1999; vol. 84 (no. 1); p. 59-61

Publication Date: May 1999

Publication Type(s): Journal Article

PubMedID: 10413228

Abstract:OBJECTIVEThe aim of this study was to assess the maternal and perinatal outcome in pregnant patients with neurofibromatosis (NF).STUDY DESIGNDuring the period between January 1994 and December 1996 eight women with NF were delivered at the Soroka University Medical Center. Maternal age, parity, gravidy and ethnic origin were matched with a control group that included 65 healthy parturients out of a total of 31,642 deliveries that occurred in our institution during this period. Maternal outcome and perinatal complications were compared between the two groups.RESULTSThe prevalence of NF during the study period was 1:2434 deliveries. The mean gestational age at delivery was significantly lower in the study group as compared to the control group, 36.8+/-3.3 vs. 39.2+/-1.5 weeks, respectively (P=0.029). The rate of intrauterine growth restriction was significantly higher in the study group, (46.2% vs. 8.95%, respectively, P=0.0005), as well as stillbirth rate (23% vs. 1.5%, respectively, P=0.011) and cesarean section rate (38.5% vs. 7.7%, respectively, P=0.01).CONCLUSIONPatients with NF have an increased risk of perinatal complications. Thus, close antenatal observation at high risk tertiary centers is required.

Database: Medline

17. Neurofibromatosis type 1 with pregnancy-associated renovascular hypertension and the syndrome of hemolysis, elevated liver enzymes, and low platelets.

Author(s): Hagymásy, L; Tóth, M; Szücs, N; Rigó, J

Source: American journal of obstetrics and gynecology; Jul 1998; vol. 179 (no. 1); p. 272-274

Publication Date: Jul 1998

Publication Type(s): Case Reports Journal Article

PubMedID: 9704804

Abstract:The authors detected a case of neurofibromatosis type 1 complicated with preeclampsia and the syndrome of hemolysis, elevated liver enzymes, and low platelets, which developed during pregnancy. When the cause of the patient's hypertension was investigated after birth, renal stenosis of the right artery was detected, which was then successfully treated with percutaneous transluminal angioplasty.

Database: Medline

18. Neurofibromatosis and renovascular hypertension presenting in early pregnancy.

Author(s): Pilmore, H L; Na Nagara, M P; Walker, R J

Source: Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association; Jan 1997; vol. 12 (no. 1); p. 187-189

Publication Date: Jan 1997

Publication Type(s): Case Reports Journal Article

PubMedID: 9027797

Available at [Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association](#) - from Oxford Journals - Medicine

Database: Medline

19. Neurofibromatosis type 1 and pregnancy.

Author(s): Dugoff, L; Sujansky, E

Source: American journal of medical genetics; Dec 1996; vol. 66 (no. 1); p. 7-10

Publication Date: Dec 1996

Publication Type(s): Journal Article

PubMedID: 8957502

Abstract:Neurofibromatosis Type 1 (NF-1) is an autosomal dominant condition which has markedly variable clinical expression, with manifestations ranging from mild cutaneous lesions to severe orthopedic complications and functional impairment. The current obstetrical literature indicates that women with NF-1 have increased complications associated with pregnancy. However, the majority of publications are case reports involving no more than 11 patients each, and are likely biased toward reporting on cases in which complications occurred. This study presents data on pregnancy outcome in 105 women with NF-1. The data were obtained from questionnaires completed by the study participants, and by review of their pregnancy and peripartum medical records. The 105 women had a total of 247 pregnancies, resulting in 182 live births, 44 first trimester spontaneous abortions, 21 elective terminations, and 2 ectopic pregnancies. There were two sets of twins. The cesarean section rate in our series (36%) was greater than the general population rate (9.1-23.5%). In 7 of these patients, the cesarean section was required because of maternal NF-1 complications. The study did not show the previously reported increased incidence of preeclampsia, preterm

delivery, intrauterine growth restriction, pregnancy-induced hypertension, stillbirth, spontaneous abortion, or perinatal mortality. Sixty-four (60%) of the one hundred five women reported growth of new neurofibromas during pregnancy and fifty-five (52%) noted enlargement of existing neurofibromas. Nineteen women observed no changes in the size of their neurofibromas and no growth of new neurofibromas during pregnancy.

Database: Medline

20. Clinical significance of neurofibromatosis in pregnancy.

Author(s): Hadi, H A

Source: American journal of perinatology; Nov 1995; vol. 12 (no. 6); p. 459-461

Publication Date: Nov 1995

Publication Type(s): Case Reports Journal Article Review

PubMedID: 8579663

Abstract:Neurofibromatosis (NF) is a common genetic disease that affects multiple organ systems. We studied eight women with NF and the outcome of their 14 pregnancies. One patient and her fetus died of a massive intracranial hemorrhage as a result of ruptured glioblastoma of the basal ganglia. The incidence of live birth and preterm labor were 50% and 28.6%, respectively. One of 14 pregnancies (7.1%) resulted in spontaneous abortion. There was also one growth-retarded fetus in this series (7.1%). Perinatal implications of this disease and management guidelines are discussed.

Database: Medline

21. Neurofibromatosis and pregnancy. An update.

Author(s): Weissman, A; Jakobi, P; Zaidise, I; Drugan, A

Source: The Journal of reproductive medicine; Nov 1993; vol. 38 (no. 11); p. 890-896

Publication Date: Nov 1993

Publication Type(s): Case Reports Journal Article Review

PubMedID: 8277488

Abstract:Neurofibromatosis is one of the most frequent genetic diseases in humans. Pregnancy in neurofibromatosis patients is, however, less common. Most current information on pregnancy and neurofibromatosis is derived from case reports, which may not reflect the true situation. In the past 15 years only two series of pregnant neurofibromatosis patients were reported in the English-language literature. We present our experience with 34 pregnancies in nine neurofibromatosis patients who delivered at our medical center. While fertility does not seem to be impaired in neurofibromatosis, these patients experience a higher-than-expected rate of first-trimester spontaneous abortions (20.7%), stillbirths (8.7%) and intrauterine growth retardation (13.0%). A high rate of cesarean section (26%) was also observed in our series. We conclude that pregnant neurofibromatosis patients constitute a high-risk group, in danger of developing life-threatening complications. However, with proper antenatal care, most pregnant neurofibromatosis patients can deliver safely if the pregnancy continues beyond the first trimester.

Database: Medline

22. Fetal hydrocephalus associated with maternal neurofibromatosis.

Author(s): Neuman, M; Beller, U; Boldes, R; Schreyer, P; Bukovsky, I; Caspi, E

Source: Journal of perinatal medicine; 1992; vol. 20 (no. 5); p. 397-399

Publication Date: 1992

Publication Type(s): Journal Article

PubMedID: 1479523

Abstract:Neurofibromatosis (NF), an autosomal dominant inherited disorder affecting multiple organ systems, is rare among pregnant women. NF in pregnancy has been reported to be complicated by maternal hypertension, fetal intrauterine growth retardation and fetal wastage. A case of pregnancy associated with neurofibromatosis, complicated by pregnancy aggravated chronic hypertension, asymmetric intrauterine growth retardation and fetal hydrocephalus is described. Possible interrelations between those pathologies are discussed.

Database: Medline

23. Eclampsia complicating a pregnancy with neurofibromatosis. A case report.

Author(s): Sherman, S J; Schwartz, D B

Source: The Journal of reproductive medicine; May 1992; vol. 37 (no. 5); p. 469-472

Publication Date: May 1992

Publication Type(s): Case Reports Journal Article Review

PubMedID: 1507195

Abstract:Neurofibromatosis occurs in approximately 1 in 3,000 births and is known to be associated with the development and/or worsening of hypertension in pregnancy. A woman was treated for eclampsia with neurofibromatosis and had a complicated course.

Database: Medline

24. Maternal and perinatal complications in neurofibromatosis during pregnancy.

Author(s): Sharma, J B; Gulati, N; Malik, S

Source: International journal of gynaecology and obstetrics: the official organ of the International Federation of Gynaecology and Obstetrics; Mar 1991; vol. 34 (no. 3); p. 221-227

Publication Date: Mar 1991

Publication Type(s): Journal Article

PubMedID: 1673938

Abstract:A total of ten patients with lesions of neurofibromatosis during pregnancy were followed up for pregnancy complications. Seven cases (70%) had hypertensive disorders of pregnancy; four had severe PET (pre-eclamptic toxemia) including one case of eclampsia, one had mild PET and the other two had only mild gestational hypertension. A total of 60% had preterm labor and in none of these did the baby survive; thus perinatal mortality was 600/1000. Mean gestation was 33.0 weeks and mean birthweight was only 1.924 kg. Thus, neurofibromatosis during pregnancy is associated with poor obstetrical outcome and requires greater care.

Database: Medline

25. Fetal growth retardation as a complication of pregnancy in patients with neurofibromatosis.

Author(s): Blickstein, I; Lancet, M

Source: American journal of obstetrics and gynecology; Aug 1987; vol. 157 (no. 2); p. 343

Publication Date: Aug 1987

Publication Type(s): Journal Article

PubMedID: 3113250

Abstract:Neurofibromatosis adversely affects reproductive performance. Fetal growth retardation as a complication of pregnancy in patients with neurofibromatosis had not been widely documented. The present cases suggest that pregnant patients with neurofibromatosis may have an increased risk for growth-retarded babies.

Database: Medline

26. Neurofibromatosis and pregnancy: Report of a case complicated by intrauterine growth retardation and oligohydramnios

Author(s): Belton S.R.; Ferguson II J.E.; Catanzarite V.A.

Source: American Journal of Obstetrics and Gynecology; 1984; vol. 149 (no. 4); p. 468-469

Publication Date: 1984

Publication Type(s): Article

PubMedID: 6428234

Abstract:The case of a pregnant patient with neurofibromatosis, complicated by mild chronic hypertension, asymmetric intrauterine growth retardation, and oligohydramnios, is described.

Database: EMBASE

27. Neurofibromatosis and severe hypertension in pregnancy

Author(s): Edwards J.N.T.; Fooks M.; Davey D.A.

Source: British Journal of Obstetrics and Gynaecology; 1983; vol. 90 (no. 6); p. 528-531

Publication Date: 1983

Publication Type(s): Article

PubMedID: 6407515

Abstract:Four pregnant patients with neurofibromatosis (Von Recklinghausen's disease) either developed hypertension during pregnancy or had an exacerbation of a pre-existing chronic hypertension. Two patients required early termination of pregnancy for severe hypertension; a third had severe intrauterine growth retardation resulting in intrauterine fetal death; and one had an otherwise uneventful pregnancy and a live healthy infant. Because of the association between neurofibromatosis and hypertension, patients with neurofibromatosis require special antenatal care and management.

Database: EMBASE

28. Possible maternal and hormonal factors in neurofibromatosis

Author(s): Hall J.G.

Source: Advances in neurology; 1981; vol. 29 ; p. 125-131

Publication Date: 1981

Publication Type(s): Article

PubMedID: 6798833

Database: EMBASE

29. Neurofibromatosis and pregnancy

Author(s): Jarvis G.J.; Crompton A.C.

Source: British Journal of Obstetrics and Gynaecology; 1978; vol. 85 (no. 11); p. 844-846

Publication Date: 1978

Publication Type(s): Article

PubMedID: 102334

Abstract:10 patients with neurofibromatosis were studied in 27 pregnancies. 2 of the patients had evidence of pre-eclampsia in their first pregnancy but not in subsequent ones. In all other pregnancies, no adverse features were noted. It would seem that neurofibromatosis is not specifically associated with any obstetric complications. The need for further reporting of cases is stressed.

Database: EMBASE

Strategy 281205

#	Database	Search term	Results
1	Medline	(neurofibromatosis).ti,ab	12616
2	Medline	exp NEUROFIBROMATOSES/	11162
3	Medline	(1 OR 2)	15547
4	Medline	(pregn*).ti,ab	403238
5	Medline	exp PREGNANCY/	816236
6	Medline	exp "PREGNANCY COMPLICATIONS"/	389149
7	Medline	(4 OR 5 OR 6)	928483
8	Medline	(preeclamps*).ti,ab	20624
9	Medline	("pre eclamps").ti,ab	8941
10	Medline	exp "PRE-ECLAMPSIA"/	27193
11	Medline	exp "FETAL GROWTH RETARDATION"/	14561
12	Medline	(IUGR OR "Intrauterine Growth Retardation").ti,ab	8510
13	Medline	(8 OR 9 OR 10 OR 11 OR 12)	51842
14	Medline	(3 AND 7)	292
15	Medline	(3 AND 13)	27
16	EMBASE	(neurofibromatosis).ti,ab	15412
17	EMBASE	exp NEUROFIBROMATOSES/	18618
18	EMBASE	(16 OR 17)	21169
19	EMBASE	(preeclamps*).ti,ab	26811

20	EMBASE	("pre eclamps*").ti,ab	12580
21	EMBASE	exp "PRE-ECLAMPSIA"/	48068
22	EMBASE	exp "FETAL GROWTH RETARDATION"/	35941
23	EMBASE	(IUGR OR "Intrauterine Growth Retardation").ti,ab	12791
24	EMBASE	(19 OR 20 OR 21 OR 22 OR 23)	85439
25	EMBASE	(pregn*).ti,ab	567923
26	EMBASE	exp PREGNANCY/	679754
27	EMBASE	exp "PREGNANCY COMPLICATIONS"/	123727
28	EMBASE	(25 OR 26 OR 27)	900527
29	EMBASE	(18 AND 24)	94
30	EMBASE	(18 AND 28)	339