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CONGENITAL ANOMALY EVALUATION PROGRAM IN ZEKAİ TAHİR BURAK WOMEN'S HOSPITAL: PRELIMINARY RESULTS

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Aim: We are presenting the first results of our new and computer based evaluation program of congenital abnormalities in our hospital. This program is started to record of cases with congenital abnormalities more systematically. These findings can help us to discuss the screening programs we need in our hospital and in Turkey. In this study we are only presenting prenatally diagnosed cases in 2001.
Material and Methods: 148 congenital abnormality cases were evaluated. Cases diagnosed by ultrasound had been evaluated not only prenatally but also after pregnancy termination. Fetal examination, anthropometrical measurements, X-ray studies had done in all cases and families invited to Genetics outpatient clinic for genetic counseling. Autopsy was performed to 35 cases who approved. Both prenatal and postnatal findings of the cases enrolled to a computer program. All of the cases classified as in "International Classification of Diseases".
Results: 55 central nervous system defects (44 neural tube defects, 8 hydrocephaly, 2 holoprosencephaly and 1 microcephaly), 21 had chromosomal defect, 17 had musculoskeletal system disorder, [6 skeletal displasias (1 achondroplasia, 1 thanatophoric dysplasia, 1 Langer type mesomelic dysplasia, 1 hypophosphatasia, two campomelic dysplasia), 2 osteogenesis imperfecta (1 of type I, 1 of type III), 3 disostosis (1 spondylothorasic- Jarco Levin, 1 spondylocostal, 1 hemivertebra), 2 artrogryphosis multiplex congenital, 1 bilateral 1 unilateral club foot], 14 had congenital malformation syndromes, 17 had non-immune hidrops, 11 had genitourinary system disorder, 7 had cystic hygroma, 3 had congenital infection, 2 had gastrointestinal disorder, 2 had cardiovascular system disorder, 1 had respiratory system disorder and 5 had other disorders.
Conclusions: Multidisciplinary and computer registration based studies are necessary for systematic screening programs for congenital malformations. Multidisciplinary studies are more effective than the studies preformed by only one department. The biggest problem is the classification of cases. "International Classification of Diseases" is one of the most useful classifications to use in a computer program for screening studies.

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RESULTS OF POSTMORTEM EVALUATION OF 602 FOETUSES

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Aim: We are presenting the results of routine postmortem evaluations of foetuses in our hospital to stress the importance of this evaluation on prenatal diagnosis in further pregnancies and genetic counselling.
Material and method: 602 cases were evaluated between 1998-2001 in this study. The sources of the cases were 1) prenatally diagnosed and terminated pregnancies in our hospital 2) Foetuses detected during daily routine visit to morgue, 3) Cases referred from other hospitals. Postmortem examination of all cases and X- ray studies of 416 cases and autopsy (when family approved-60 cases) were done. Records of dysmorphic findings, antropometric measurements and photographs of the foetuses were archived. Cases were classified by using "International Classification of Disorders".
Results: 245 central nervous system disorder, 57 chromosomal disorder, 62 musculoskeletal system disorder, 115 multiple malformation syndrome, 35 hydrops fetalis, 26 genitourinary system disorder, 17 congenital infections, 11 cardiovascular defects, 10 gastrointestinal defects, 1 respiratory system anomaly, 5 unclassified cases (2 anhydroamnios sequence due to premature rupture of membranes, 2 conjoined twins, 1 type II sacrococsigeal teratoma) were detected.
Discussion: Postnatal evaluation of foetuses is very important diagnostic study for adequate and accurate genetic counselling and planning prenatal diagnostic method for further pregnancies. Exact diagnosis of

a case presents aetiology and inheritance pattern of disorder. Risk assessment, determination of laboratory methods and detection of cases have risk in the same family are possible after exact diagnosis of case. On the other hand, prenatal diagnostic studies are getting cheaper and easier in case of effective postmortem evaluation.
The biggest group of anomalies was neural tube defect (181 cases) both in study group and in cases with CNS anomalies. This finding presents the importance of folic acid replacement. This is a very cheap and effective way of prevention.
Skeletal dysplasias were the biggest group in musculoskeletal system disorders. These disorders can be classified easily by X ray studies. On the other hand, all class of cases can have X-ray findings, because of that, X-ray studies necessary in all cases.
Chromosome analysis is also very important during this evaluation. Postnatal foetal blood samples and skin biopsy are the best choice. Foetal urine sample can sometimes helpful for chromosomal studies.
Conclusion: All foetuses must be evaluated by an experienced person. If this is not possible, foetal blood and skin samples must be send to a genetic centre and foetal photographs and X rays can be archived for consultation with an experienced centre. TORCH studies must be done from both foetal and maternal samples in perinatal period. These records are very important for the evaluation of these cases.

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KONJENİTAL KİSTİK ADENOMATOİD MALFORMASYON: İKİ OLGU

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Özet: Bu çalışmamızda nadir görülen akciğerin konjenital kistik adenomatoid malformasyonlu iki olguyu inceledik.
Anahtar Kelime: Konjenital kistik adenomatoid malformasyon
Giriş: Konjenital kistik adenoid malformasyon (KKAM), değişen klinik bulgu ve belirtilerle karşımıza çıkabilen, akciğerlere ait gelişimsel bir anomalidir. KKAM 1:25000-1:35000 gebelik oranında gözlemlenmektedir. KKAM, intralobar sekestrasyon, ekstralobar sekestrasyon, bronkopulmoner foregut malformasyon, bronşial atrezi, lobar amfizem kombinasyonlarını içine alabilecek bir yelpazeye sahiptir. KKAM prenatal tanısında kullanılacak yöntemler içerisinde en iyisi ultrasonografi, postnatal araştırmalarda ise X-ray ve bilgisayarlı yöntemlerdir.
Bu çalışmamızda ikinci trimestrede ultrasonografik olarak gözlemlediğimiz Tip III KKAM'lu iki olguyu irdeledik.
Gereç ve Yöntem: Bu çalışma Haziran-Ağustos 2002 tarihleri arasında SSK Ege Doğumevi ve Kadın Hastalıkları Eğitim Hastanesi Perinatoloji departmanına başvuran hastalar arasından tesbit edildi. Fetusların otopsi ve mikroskobi incelemeleri hastanemizin patolojik anatomi ünitesinde gerçekleştirilmiştir.
Olgu İncelemeleri: Olgu I: Olgu 36 yaşında, G3P2Y2, anamnezinde patolojik bir söylem saptanmadı. Son adet tarihine göre fetus 19 haftalıktı. Yapılan ultrasonografik incelemede fetus 20 haftalık, fetal hidrops hali, mediastinal kayma, akciğerler yoğun sıvıya bağlı olarak sıkışmış, Tip II KKAM görünümü izlendi. Yapılan fetal karyotip incelemesinde üreme sağlanamadı.
Olgu II: Olgu 30 yaşında G2P1Y1 anamnezinde patolojik bir söylem saptanmadı. Son adet tarihine göre fetus 18 haftalıktı. Yapılan ultrasonografik incelemede fetus 19 haftalık, fetal hidrops hali, mediastinal kayma, akciğerler Tip III KKAM görünümündeydi. Yapılan fetal karyotip incelemesi 46 XY idi. Her iki gebelik de aile onayı ve hastanemiz Perinatoloji Konseyi'nde tartışılarak sonlandırıldı. Otopsi incelemelerinde unilateral Tip II KKAM tespit edildi. Fetuslardan biri kız diğeri erkek fenotipindeydi.
Tartışma: KKAM nadir görülen akciğer hastalıklarından biridir. Kötü prognostik faktörler olarak geçen çift taraflılık, hidrops fetalis, mediastinal kayma, sağlam akciğerin ileri derecede hipoplazisi veya amfizematöz değişikliği, akut gelişen polihidramnios varlığı durumlarında postnatal sonuçlar yüz güldürücü değildir. Monni G'nin çalışmasında; KKAM'lu olgularda, tek taraf tutulumu ve küçük lezyon olması, akut polihidramnios ve hidrops hali olmaması iyi prognoz göstergesi olarak saptanmıştır. Kendi çalışmamızda hidrops gelişimi, mediastinal kayma, polihidroamnios izlenmesi kötü prognostik gösterge olarak bulundu. Hastanemiz Perinatoloji Konseyi'nde yapılan tartışma ve hasta ile yapılan görüşmede gebelik sonlan-