high at the end of therapy. Suppressor cells normalized faster, whereas helper cell levels remained abnormally low for a long period. Most children became nonimmune to polio, whereas the majority had antibodies to measles, mumps, and rubella. Despite depressed immunity, serious infections were not documented.

SUPPRESSION OF THE OLIVOCOCHLEAR REFLEX: A NEUROTOXIC ADVERSE EFFECT OF VINCRISTINE

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OBJECTIVE: The purpose of this study was to examine the effects of a known neurotoxic regimen, such as the acute lymphocytic leukemia (ALL) Berlin-Frankfurt-Münster 95 (ALL-BFM-95) protocol, on the function of the medial olivocochlear bundle, which was assessed by recording suppression of the amplitudes of distortion-product otoacoustic emissions (DPOAEs) when white noise was applied simultaneously to the contralateral ear.
METHODS: Our population consisted of 3 groups of children with ALL. A baseline examination was performed before the beginning of therapy. DPOAE-suppression measurements were repeated after 4 weekly doses of vincristine in the first group ($n = 12$), after 8 weekly doses in the second group ($n = 12$), and 3 years after completion of the protocol in the third group ($n = 23$). In the third group, a subgroup of 12 children who were exposed to low-dose gentamicin (<13 days) and another 11 children who were exposed to high gentamicin doses (>23 days) were evaluated.
RESULTS: At baseline examination, all groups presented significant suppression at all frequencies. Efferent mediated DPOAE suppression was still present after 4 sessions of vincristine. However, after 8 vincristine sessions, instead of suppression, an increase of amplitudes was noted at 5 (of 12) frequencies. In the subgroups examined 3 years after ALL-BFM-95, the olivocochlear reflex had recovered.
CONCLUSIONS: Enhancement or no significant suppression of OAEs by contralateral noise indicates a probable vincristine-induced insult to the efferent cochlear innervation. This adverse effect seems to take place early in the course of chemotherapy and is slowly reversed a few years after chemotherapy. The clinical implications of these findings may need additional investigation.

SCREENING NEWBORNS FOR SICKLE CELL DISEASE IN GHANA

Submitted by Kwaku Ohene-Frempong
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INTRODUCTION: Screening of newborns for sickle cell disease (SCD) allows early initiation of prophylactic therapy, parental education, and comprehensive management, which results in reduced mortality. Since April 1993, a demonstration project to develop and implement a program of newborn screening for SCD has been conducted in Kumasi, Ghana, by the Comprehensive Sickle Cell Center at the Children's Hospital of Philadelphia in collaboration with the Ministry of Health and other institutions in Ghana.
OBJECTIVE: Our goal was to assess the program of screening and follow-up of children with SCD in Ghana.
METHODS: Infants are screened at birth or at well-infant visits within days or a few weeks after birth. Mothers are asked to come for results within 4 weeks, and failing that, an extensive tracking system is used to deliver results to the homes of families with infants with possible SCD. Tracking relies solely on information obtained from mothers at the time of screening. The goal is to enroll infants with possible SCD into the sickle cell clinic by 8 weeks of age. Pregnant women, parents with children, and the general public are educated regularly about the screening program. Children with SCD receive comprehensive care through the Sickle Cell Clinic at Komfo Anokye Teaching Hospital (Kumasi, Ghana).
RESULTS: From February 13, 1995 (when newborn testing was started), to December 31, 2005, a total of 202 244 infants were screened through 8 public health institutions and 14 private clinics in Kumasi and 1 private maternity center and 1 public health Center in Tikrom, a nearby, rural community. A total of 3745 (1.9%) infants were identified as having possible SCD with the following hemoglobin phenotypes according to isoelectric focusing: 2047 (1.04%) fetal sickle cell hemoglobin; 1684 (0.83%) fetal SC hemoglobin; and, 14 (0.003%) fetal SA hemoglobin (Table 1).

TABLE 1. Screening and Tracking Results: February 1995 to December 2005

	No.	%
Total No. of infants screened	202 244	100.0
Infants with possible SCD	3745	1.9 (of infants screened)
Under active tracking (newly diagnosed)	(125)	3.3 (of infants with possible SCD; excluded)
Total possible SCD accounted for	3620	96.7 (of infants with possible SCD; reported)
Lost to follow-up/never found	(494)	13.6 (of infants with possible SCD; reported)
Dead before contact made	(34)	0.9 (of infants with possible SCD; reported)
No. contacted with results (eligible for enrollment)	3092	85.4 (of infants with possible SCD; reported)
Came for results	651	21.0 (of those contacted)
Through home visiting	2441	79.0 (of those contacted)
Eligible but not enrolled after contact	477	15.4 (of those eligible)
Enrolled in clinic	2615	84.6 (of those eligible)
Known deaths after enrollment	109	4.2 (of those enrolled)

CONCLUSIONS: Screening and follow-up of newborns for SCD is feasible in a developing country in Africa. Extra effort in tracking is necessary to ensure that infants with disease are found early and referred for medical management.

PROSPECTIVE FOLLOW-UP OF PATIENTS WITH EWING SARCOMA WITHIN THE LATE EFFECTS SURVEILLANCE SYSTEM

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INTRODUCTION: It is known that antineoplastic treatment may induce early and late organ toxicities depending on treatment modalities and intensity.
OBJECTIVE: The aim of this study was to determine the cumulative incidence of sequelae within our cohort of patients treated within the EICESS-92 (European Intergroup Cooperative Ewing's Sarcoma Study, 1992) treatment trial.
METHODS: Since 1998, the Late Effects Surveillance System (LESS) of the German Society for Pediatric Oncology and Hematology has prospectively registered late effects in patients of all ages with relapse-free bone and soft tissue sarcoma in Austria, Germany, and Switzerland. The follow-up is conducted locally in accordance with LESS guidelines. Data are reported to the LESS center for collation and analysis.
RESULTS: There were 67 patients available for analysis (42 male, 25 female) with a median age at diagnosis of 13 years and a median follow-up of 3.5 years. Registration had to be terminated for 17 patients as a result of relapse. In total, 43.3% (29 of 67) of the patients were reported to have at least 1 sequelae of treatment. Sixteen patients suffered toxicity in 1 organ system, 9 patients developed toxicity in 2 organ systems, and there were 3 organ systems affected in 4 patients. Nephrotoxicity was reported in 10.4% (7 of 67), cardiotoxicity in 8.9% (6 of 67), peripheral polyneuropathy in 5.9% (4 of 67), and other toxicities in 34.3% (23 of 67) of the patients.
CONCLUSIONS: Sequelae of treatment for Ewing sarcoma within this cohort of the EICESS-92 study were not more frequent than reported previously. Patients are at risk for the development of several toxicities after treatment for Ewing sarcoma, and they should receive adequate medical follow-up.

HEMATOPOIETIC STEM CELL TRANSPLANTATION FOR THE TREATMENT OF THALASSEMIA: THE GREEK EXPERIENCE

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INTRODUCTION: Although prevention remains the cornerstone for the management of thalassemia, hematopoietic stem cell transplantation (HSCT) is the only curative approach.
OBJECTIVE: Our goal was to assess our experience with HSCTs for the treatment of patients with thalassemia.
METHODS: From 1994 to 2006, 96 HSCTs have been performed in 84 thalassemic children from Greece, 3 with sickle cell/thalassemia and 1 with sickle cell disease. According to Pesaro classification, of these 84 children, 20 were in class I, 35 were in class II, and 29 were in class III. Donors were 84 histocompatible siblings and 4 unrelated volunteers. The graft was of bone marrow in 85, cord blood in 3, bone marrow and cord blood in 4, and peripheral blood stem cells in 4. The conditioning regimen consisted of busulfan, cyclophosphamide, and antilymphocyte globulin.
RESULTS: All except 1 patient received engraftments. Ten patients rejected the graft. Eight received another transplant from the same donor, 7 of which were successful. Four patients died; causes of death were graft-versus-host disease (GVHD) (2), disseminated toxoplasmosis (1), and brain hemorrhage (1). At a median follow-up time of 6.5 years, 84 of 88 children survived, 81 were cured and free from transfusions, and 3 remained transfusion-dependent. Severe acute GVHD developed in 18 children, and chronic GVHD developed in 8 patients. The overall survival rate, event-free survival rate, rejection rate, and transplant-related mortality rate were 95%, 94%, 11%, and 5%, respectively. Event-free survival was 100% for class I, 95% for class II, and 87% for class III. Eleven children had mixed chimera (residual recipient hematopoiesis) with normal levels of hemoglobin.
CONCLUSIONS: HSCT is a highly effective treatment for thalassemic patients who have a fully matched donor (related or unrelated). Younger age at transplant secures excellent results with reduced morbidity and mortality rates.

TWO-DECADE EXPERIENCE AND LONG-TERM SURVIVAL IN PEDIATRIC NON-HODGKIN'S LYMPHOMA

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