0376

DEPARTMENT OF THE AIR FORCE
USAF MEDICAL CENTER, WRIGHT-PATTERSON (AFLC)
WRIGHT-PATTERSON AIR FORCE BASE OHIO 45433

REPLY TO ATTN OF: SGHS\

11 January 1980

SUBJECT: Report of Findings and Conclusions: Project #SGO 79-32

AIR1.941130 052r

TO: AFMSC/SGPA
Brooks AFB, TX 78235

Introduction

This study of Wilson's Disease was done to test the hypothesis that there is a basic underlying enzyme defect in the liver, which accounts for the problem of the body's inability to handle copper and its subsequent deposit in the liver, ganglia of the brain and other areas of the body.

Subject

One subject, a 21 year old female, was entered in the study. She was first seen at this Medical Center in September 1977 and a suggestion of demyelinating disease was made at that time. She was followed and ultimately returned in January 1978 when a diagnosis of Wilson's Disease was made based upon kaiser-fleischer rings in the cornea, decreased serum ceruloplasmin, increased copper stores in the liver on liver biopsy and her general neurologic picture. She was begun on penicillamine at that time and showed a progressive improvement in her neurologic picture.

In August 1978 the patient was evaluated by Dr. I. Herbert Scheinberg at the Albert Einstein College of Medicine in New York. Dr. Scheinberg, a professor of Medicine and Head of the Division of Genetic Medicine at Albert Einstein College of Medicine, Yeshiva University, Bronx, New York, has been studying Wilson's Disease well over 25 years and is considered an expert on this extremely rare condition. In addition to penicillamine, he started her on BAL and maintained an interest in the patient.

The patient had her first episode of upper GI hemorrhage in August 1978. She had a recurrent bleed in October 1978 and at that time was found to have upper GI hemorrhage from bleeding esophageal varicies. She also had an episode of hepatic coma and required nine units of whole blood transfusion. In February 1978 she had an upper GI hemorrhage from her varicies which required six units of whole blood transfusion. Because of the recurrent variceal hemorrhage, she was ultimately referred back to this medical facility for consideration for a porto-systemic shunt procedure. At the time of hospitalization rather massive esaphageal varicies were again noted and she had two episodes of minor upper GI hemorrhage while under observation. Each required approximately three units of blood

transfusion. Diagnosis: 1) Wilson's Disease; 2) Esophageal varices secondary to Wilson's Disease; 3) Recurrent hemorrhage secondary to esophageal varices. Because of recurrent episodes of upper GI hemorrhage, esophageal varices and a massive enlargement of the spleen, it was recommended that the patient undergo a: 1) spleno-renal shunt; 2) splenectomy.

Apparatus

All approvals and licenses were obtained and the Medical Isotopes Committee granted approval for the use of the radiopharmaceutical, Copper 64 Cupric Acetate on a one time basis and authorized Dr. I. Herbert Scheinberg to administer the radioisotope at this Medical Center.

The pre-operative preparation, operating room environment, and post-operative environment were carefully monitored by the Radiation Safety Officer in accordance with the National Committee on Radiation Protection Report No. 37.

Approximately 2 hours pre-surgery, the patient was given orally radioactive Copper 64 in an amount of approximately 1-2 mg copper and 1 millicurie of Copper 64 in approximately 4 oz of skim milk.

Procedure

At the time of surgery, the patient underwent a liver biopsy, in addition to the spleno-renal shunt and splenectomy. The liver biopsy (approximately 5 mg) was taken for radio-assay. The specimen was given to Dr. I. Herbert Scheinberg for quick freezing and return to his laboratory for radio-assay and evaluation of the enzymatic defect of Wilson's Disease. Findings at surgery were those of macro nodular cirrhosis of Wilson's Disease, varices and massive splenomegaly. It was found that a large amount of the isotope which had been administered in a skim milk carrier lay in the stomach in curds and was not absorbed entirely. An additional procedure resulted from the curdled milk which was suspected to be a gastric polyp. The surgery was uncomplicated and the patient had an uneventful post-operative course.

Results

The results of the chemical copper analysis of the biopsy liver sample showed 137 mcg of copper per g dry liver. White clotted milk removed from the stomach was also analyzed and proved to contain over 95% of the radio-copper administered.

Summary

Approximately 2 hours pre-surgery a 21 year old female with a diagnosis of Wilson's Disease was administered approximately 1 mg of Copper 64 in 4 oz of skim milk carrier. During surgery, approximately 5 mg of liver was removed and quick frozen for radio-assay and evaluation of the enzymatic defect of Wilson's Disease. Chemical copper analysis showed 137 mcg of copper per g of dry liver.

Conclusion

The measurable radiation exposure to the patient was considerably less than had been anticipated because of the skim milk curdling. The radiation exposure to the operating team and operating room personnel was negligible. This unexpected finding is extremely important, because much smaller doses of radioactive copper can be used in a different (non-protein - containing) carrier in future work.

GUNTER R. MENG
Colonel, USAF, MC
Vice Commander

Cy to: SGH
SGE
HQ AFLC/SG
AMD/RD