



**Figure 1.** Chest x-ray showing bilateral fine granular infiltrates.

Dr. Jeffrey Whitsett, Cincinnati Children's Hospital). DNA analysis revealed the homozygous 122delT mutation, while both parents were heterozygous for the same mutation (Courtesy of Dr. Lawrence Nogee, Johns Hopkins University).

SP-B is a hydrophobic protein involved in the adsorption of surfactant phospholipids to the air-liquid interface. It is coded by a gene of 11 exons on chromosome 2. In 1993, Nogee et al reported SP-B deficiency causing severe respiratory disease, as described in our patient.<sup>1</sup> The patient and a sibling who had died earlier had a frame-shift mutation caused by a 2 base-pair insertion (121ins2) in exon 4 of the SP-B gene.<sup>2</sup> Radiologically, SP-B deficiency presents like hyaline membrane disease. Histopathologically, the distal airspaces appear filled with lipid-rich, periodic acid Schiff-positive, eosinophilic proteinaceous material.<sup>1</sup>

The diagnosis is established by failing to identify SP-B in the tracheal effluent and is confirmed by genetic studies, which show a mutation on the SP-B gene. More than 13 mutations have been described,<sup>3</sup> of which (121ins2) accounts for about 70%. Its frequency in the United States is estimated to be 1 per 1000-3000 individuals.<sup>4</sup> The 1043ins3 mutation was detected in 2 unrelated Pakistani

families.<sup>3</sup> The mutation described in the present report (122delT) was described in a consanguineous kindred of Kurdish descent,<sup>5</sup> and in three unrelated Lebanese families (L. Nogee, personal communication). The recognition of specific mutations in various ethnic groups may allow diagnosis in individual patients and population-wide studies for the determination of gene frequency. This would gain particular importance in our population, where consanguinity is prevalent. SP-B deficiency is usually fatal, unless treated with lung transplantation.<sup>6</sup> Gene transfer therapy may be the treatment modality of the future.

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## Serum immunoglobulin A, G and M in healthy adults in Dhofar, Oman

**To the Editor:** There is little data available on normal levels of serum immunoglobulin in the healthy adult populations of the Gulf countries and the Arab world. In many instances, the normal ranges for immunoglobulin, which are used by many hospitals within the Arab world, are those that are supplied by the manufacturer of the equipment or the reagents, and these values may not reflect the normal values of the local populations. Therefore, it is essential that each population establish its own normal values that can be used locally.

Although Oman has a climate that is generally hot and dry, similar to other Gulf Countries, Dhofar's (the southern region of Oman) climate is relatively cool and rainy, particularly during the summer monsoon. Individuals from this part of Oman may have their own distinct levels of immunoglobulin as this region has a distinct pattern of infections.<sup>1</sup>

Serum samples were collected from 489 (389 males and 100 females) Omani healthy adults from Dhofar recruited from healthy blood bank donors attending Sultan Qaboos Hospital in Salalah. Individuals with a history of acute or chronic illness, present or past allergy, parasitic infestation, chronic drug use, or present immunization were excluded from the study. After informed consent was secured, blood samples were obtained and allowed to clot at room temperature. Sera were separated and stored at -20°C until assayed for immunoglobulin G, M, and A, using a rate nephelometry system (Beckman Image System).

The majority of the participants, 54% (n=262), were 20-29 years old, followed by 23% (n=114) that were 30-39 years old and 8% (n=40) were 40-49 years old. Only 5 individuals (1%) were above the age of 50 years, while 68 individuals (14%) were 20 years or younger. The mean age for the whole cohort was 28.6 years (males and females were 29.9 years and 23.6 years, respectively).

The mean serum levels of IgM, IgG, and IgA for the whole cohort are shown in Table 1. The immunoglobulin M, G, and A normal ranges are shown as the range between the 5th and 95th percentile. When we compared serum immunoglobulin levels in individuals below the age of 20 years (n=68), we observed a significant difference with regard to serum IgA levels, which occurred at lower levels in those young individuals compared to those above the age of 20 years (n=421), ( $P<0.01$ ).

Comparing our results with those obtained from the neighboring Saudi population,<sup>2</sup> similar levels of IgM (1.14 g/L for Saudis versus 1.01 g/L for Omanis) and similar levels for IgA were noted. However, a significantly higher level ( $P<0.05$ ) of IgG (14.63 g/L for the Saudis versus 12.88g/L for

Omanis) was detected. This may be due to environmental factors, since the climate in Saudi Arabia is quite different from the climate in Dhofar. Whereas the climate in Saudi Arabia is hot and dry all year, the climate in Dhofar is cold and rainy most of the year. Therefore, different antigen exposure in the two groups may account for the different levels of IgG. However, data from a cross-sectional study are needed to verify that the Dhofar population are different, per the reviewer comment.

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## N-Butyl-2-Cyanoacrylate (Histoacryl) Complication: A Case Report

**To the Editor:** The tissue adhesive N-butyl-2-cyanoacrylate (*Histoacryl*, Trihawk International, Montreal, Canada) is a well-known and effective modality for treatment of gastric varices secondary to portal hypertension of various causes. It has been used safely in many centers for up to 20 years.<sup>1,2</sup> Nonetheless, in a minority of cases, its use has been associated with adverse effects like portal vein thrombosis. We report on the management of a secondary bleeding complication by placement of a transjugular intrahepatic portosystemic shunt (TIPS).

A 41-year-old woman was referred to our hospital with esophageal and gastric varices secondary to liver disease due to bilharziasis. She had multiple episodes of upper gastrointestinal bleeding. Esophagogastroduodenoscopy identified two esophageal and three gastric varices with evidence

**Table 1.** Serum IgM, IgG and IgA in healthy adult Omanis (ages 18 to 54 years) from Dhofar, Oman.

Variable	IgM			IgG			IgA		
	Both (n=489)	Males (n=389)	Females (n=100)	Both (n=489)	Males (n=389)	Females (n=100)	Both (n=489)	Males (n=389)	Females (n=100)
Mean	1.006	0.886*	1.471*	12.88	12.85	13.00	2.64	2.57*	2.90*
SD	0.511	0.450	0.468	2.76	2.97	1.71	1.12	1.07	1.27
95% CI	0.960, 1.051	0.841, 0.931	1.378, 1.564	12.63, 13.12	12.55, 13.14	12.66, 13.34	2.54, 2.74	2.47, 2.68	2.65, 3.15
5th Percentile	0.374	0.354	0.647	8.52	8.17	10.41	1.23	1.18	1.50
95th Percentile	2.050	1.640	2.469	17.45	18.30	15.60	4.77	4.72	6.51

Serum immunoglobulin levels are in grams per liter

Asterisks (\*) indicate significant difference in values between sexes.