

SOFT TISSUE SARCOMAS OF THE HEAD AND NECK

NORMAN D. BLOOM, MD, FACS

Soft tissue sarcomas of the head and neck are rare. These tumors arise in either the extraskeletal connective tissues of the body that connect or support anatomic structures or within the bony or cartilaginous structures. Approximately 6,000 soft tissue sarcomas occur in the United States with approximately 3,300 deaths each year. These tumors comprise less than 1% of all cancers in adults but, 6.5% of all cancers in children younger than 15 years of age. Approximately 10% of all soft tissue sarcomas, or about six hundred a year, arise in the head and neck area.

The most common histologic types of soft tissue sarcomas in adults are the malignant fibrous histiocytomas, liposarcomas, and fibrosarcomas which occur in the head and neck. Other less common histologic types include the angiosarcomas, leiomyosarcomas, rhabdomyosarcomas and the sarcomas of neural origin. These tumors present most often as asymptomatic soft tissue masses. These tumors tend to grow in the path of least resistance and push surrounding tissues before them. Surrounding these tumors is a pseudocapsule which always contains invasive tumor. Shelling out soft tissue sarcomas is never curative.

An excisional biopsy is appropriate for establishing the diagnosis of small tumors less than 2 cm in size that are found in the head and neck area. Tumors larger than this should have a core or incisional biopsy performed in order to establish the proper diagnosis. Fine needle aspiration biopsies may often be misleading.

The importance of establishing a tissue diagnosis prior to definitive surgery is related to establishing the grade and size of

the tumor as well as the exact tumor type. Tumor grade and size are important prognostic factors for patients with soft tissue sarcomas. Grades are assigned from 1 (well differentiated) to 3 (poorly differentiated) and the stage of the disease is directly related to the grade of the tumor. Histologic type also appears to have prognostic value with poor prognosis being predicted for angiosarcomas, esthesioneuroblastomas, Ewing's sarcoma, synovial sarcoma and neuroblastomas.

Once the diagnosis is established, radiologic studies to evaluate the extent of local disease and distant metastasis are important. An MRI of the neck and a CT scan of the neck and chest evaluate the local regional disease as well as the most probable metastatic site, the lungs. These imaging studies are invaluable in planning any radiotherapy treatment in conjunction with surgery.

Combination surgery and radiation therapy is the primary treatment for those sarcomas that are of an intermediate or high grade or for all tumors where the margins are close or contaminated. With this approach to primary treatment, local control of the disease can be achieved in 95% of all cases with 5 year survival rates for clinical Stage I (G1), Stage II (G2) and Stage III (G3) of 79%, 65% and 45% respectively. Stage IV disease is present when lymph nodes are positive or there is metastatic disease to the lungs. Survival rate for clinical stage IV is less than 10% for five years.

Approximately 5% of all patients with soft tissue sarcomas develop lymph node metastasis during the course of their disease, however, the incidence is higher for patients with epitheloid sarcoma, synovial cell sarcomas and rhabdomyosarcomas (40%, 17%, and 12% respectively). An elective neck dissection is not indicated for the majority of head and neck soft tissue sarcomas.

Surgical excision with negative margins followed by radiation therapy is the mainstay of therapy. Since the head and neck region is crowded with vital structures, obtaining a negative margin may be difficult without sacrificing a vital organ. Obtaining margins on the face, for instance, may cause damage to an esthetic unit such as the mouth, eye or nose. Radiation therapy can be delivered by external beam radiation alone to 6300 cGy or a combination of brachytherapy and external beam therapy can be employed.

Those individuals who die of their disease usually manifest distant metastasis to the lungs. In an attempt to decrease the distant failure rate, numerous randomized adjuvant trials have been conducted. Clinical trials to date that have included trunk or head and neck sarcomas have failed to show any statistically significant benefit from any of the adjuvant regimens utilized.

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While most of the soft tissue sarcomas that occur in the head and neck area arise within the soft tissues of the neck, tumors can appear within the organs of the neck as well, though these are exceedingly rare. Forty cases of liposarcomas have been reported in the literature; twenty-six cases arising in the larynx; seven, in the hypopharynx; six, in the cervical esophagus and one indeterminate site. Thirty-five cases of leiomyosarcomas have been reported in the larynx as well as nine cases of rhabdomyosarcomas. Treatment involves removal of the affected organ as well as adjunctive radiation therapy where indicated.

In addition to the standard head and neck sarcomas some individuals will present with locally aggressive tumors such as dermatofibrosarcoma protuberans and desmoid tumors. While these tumors have low potential to spread to other sites, local control of these tumors can be more difficult than for the sarcomas.

Dermatofibrosarcoma protuberans is a slow growing malignant tumor of the skin. Control of the disease can be achieved by obtaining a wide surgical margin (>3cm) in all directions. This can be difficult to achieve in the head and neck region because of both functional and cosmetic aspects. None the less the end result of not treating these tumors aggressively can lead to a greater than 50% recurrence rate. The effectiveness of radiation therapy for these tumors has not been proven.

The desmoid tumors, also known as aggressive fibromatosis arise primarily from the deep fascial sheaths in the head and neck. They infiltrate extensively into the surrounding tissues of the brachial plexus which is the collection of nerves that provide for all function and sensation to the arm. Encasement of the subclavian artery (part of the main artery of the arm) and vein (part of the main vein of the arm) is also common. Surgical resection is the treatment of choice for these tumors. For those patients with clear margins, or minimally positive margins, close observation is preferred. Local recurrences that can occur in as many as 20% of patients up to six years after resection can be treated with resection and radiation therapy.

Finally there is a category of soft tissue sarcomas that may arise following radiation therapy for squamous cell cancers of the head and neck. These radiation induced sarcomas develop anywhere from 9 to 45 years after the completion of the radiation therapy. These sarcomas are predominantly malignant fibrous histiocytomas and usually high grade. Complete surgical excision offers the only realistic chance for long term survival. The distant failure rate for these tumors exceeds 70%.

Rhabdomyosarcoma is the most frequent soft tissue sarcoma in children. Slightly more than 200 new cases are diagnosed each year in the United States. The head and neck is the most frequent site of occurrence. More than half of the tumors will be diagnosed in children under the age of 5 and the tumor is associated with distinct genetic alterations.

In the head and neck, 60% of the tumors arise from a parameningeal site (orbit, middle ear, nasal cavity, the sinuses, nasopharynx, and infratemporal fossa). The remaining tumors arise in the scalp, parotid gland, oral cavity, larynx, oropharynx, cheek, hypopharynx, thyroid, parathyroid and neck. After determining the site of origin, invasiveness of the primary tumor

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A TIME FOR SHARING I've Taken the Time to Smell the Flowers

It was a routine physical. Morton Silverblatt, a Pittsburgh florist, was having a medical check-up as required for scuba-diving certification. As Mort opened his mouth, the doctor inserted a tongue depressor. Purely routine. But then there was a startled moment of silence culminating in a question by the doctor. "Aren't you having any difficulty swallowing?" he asked. During examination of Mort's mouth, the doctor had discovered a tumor the size of a golf ball at the back of his tongue. Mort had never felt a thing.

Mort had cancer. It was 1973 and he was 46 years of age. At that time the word "cancer" felt like a death sentence to him. But that was 28 years ago and today he is a "cancer survivor" enjoying his retirement in the Florida sunshine.

Prior to his diagnosis of cancer, Mort had been deeply involved in the family flower business and was helping to raise four children. He worked hard and he smoked heavily. Suddenly his world stopped. "Cancer. Think of the sounds of the word: "Malignant." "Metastasize." His priorities changed. "One of the best things to happen to me was cancer, said Mort. "It gave me a whole different outlook on life."

"The worst thing was breaking the news to my father who was quite elderly, and to a couple of aunts." Mort did not want pity. "I told them, 'You can cry now, but I feel sorry enough for me. I don't need your help with that.'"

It was his 47th birthday when he underwent 8 1/2 hours of surgery. The surgeons literally split his face in half, cutting his jaw down the middle and slicing open his tongue to remove the tumor. He is able to laugh about it now as he retells the story. "For a year afterward, I spoke with a forked tongue," he quipped

Following his surgery, Mort resumed his life much as he had before. He even began smoking again. However, about 2 1/2 years later, he found himself back in the doctor's office listening to discomfort-

ing words. "I don't like the way it feels," the physician said as he palpated a knot on Mort's neck. Recurrence! This time the cancer had metastasized to the lymph nodes. Mort was given less than a 50% chance of survival.

The day after Thanksgiving in 1975, Morton Silverblatt underwent a radical neck dissection for control of neck metastases. The radiation therapy that followed consumed most of his strength. Nerves had been clipped during surgery leaving him with a tremor in his right hand and not very much strength in his left arm.

But Mort approached this recurrence of cancer with much the same tenacity as he had the initial diagnosis, and it worked. "And the more I understood about the disease, the more it helped," he said. In ad-

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The only day you can
live is this day....
Every day is so damn
important."*

dition, "I had many people from many different religions and walks of life saying prayers for me and I think that helped a lot."

Mort and his wife, Alice sold the family business and traveled for a couple of years before finally settling in Miami in 1979. Cancer often imposes additional strains on many marriages but in Mort and Alice's case, "If it did anything, it strengthened our marriage." The Silverblatts have been happily married more than 47 years.

In 1983, Mort joined the American Cancer Society's "CanSurmount" program, where he is a volunteer counselor. "I get very emotionally involved, because if I didn't I wouldn't be worth a damn," he said. He became especially close to one 36 year-old patient some years back. "She

lived a "quality life" to the end because she fought back," he said. "There is a time when you have to accept the inevitable." Mort said. "It isn't always easy...I don't like pain. I don't know anybody who does."

Today Mort continues to enjoy his life with Alice in their North Dade County town house where he counts his blessings for his good fortune, his four children and his grandchildren. He continues his many hours of volunteer work with cancer patients and spends numerous hours caring for his dozens of African violets and working on his computer tucked away in an upstairs studio.

"We're all terminal," he says. "The only day you can live is this day...Every day is so damn important. If I die tomorrow, don't feel sorry for me. I've taken the time to smell the flowers, as well as time to grow them."

Editor's note: I was privileged to meet Morton Silverblatt at the National Coalition of Cancer Survivorship Assembly in Charlotte, North Carolina in October of 1992. We became friends and met again in November of 1993 at the Assembly in Seattle, Washington. Mort has spent many years with a pet project of his called "Hope Lodge." "Hope Lodge," sponsored by Winn Dixie and the American Cancer Society, is a residence for cancer patients receiving out-patient treatment (radiation and/or chemotherapy). Here, cancer patients whose homes are a distance away are able to stay with their significant others free of charge. This program is offered to cancer patients living in the greater Miami area. In addition to his volunteer work at "Hope Lodge," Mort is interested in starting a support group for head and neck cancer patients. (1994)

Editor's Comments: The article that appears above originally appeared in the February, 1994 issue of News From SPOHNC. It has been modified to reflect Mort's present status. We are indeed pleased and proud to have SPOHNC-MIAMI, FL, Mort Silverblatt Head and Neck Support Group as part of our network of local chapters of SPOHNC. (2001) ■

CORRECTION

The correct phone number to contact Andrea S. Barber, Esq. is 631-271-4558. Ms. Barber is the author of "Advance Directives: Health Care Proxies" which appeared in the April, 2001 issue of *News From SPOHNC*.

THE SHAKER EXERCISE A TREATMENT FOR DYSPHAGIC PATIENTS

CARYN EASTERLING, M.S., CCC

Many of us take the act of eating and swallowing for granted. However, this seemingly uncomplicated act of swallowing is actually very complex and involves more than 20 groups of muscles that open and close the round sphincter muscle at the top of the esophagus.

Dysphagia or difficulty in swallowing, includes symptoms such as coughing, throat clearing, food sticking and choking while eating, and affects one out of every seventeen Americans. These symptoms of dysphagia may lead to malnutrition, pneumonia and asphyxia. Abnormalities of swallowing are common especially among elderly individuals and may involve one or all four phases of swallowing, that is, the preparatory, oral, pharyngeal or esophageal phases of swallowing. Dysphagia is a common sequela of central nervous system diseases, neuromuscular disorders and therapy for head and neck cancers.

Researchers at the Medical College of Wisconsin led by Reza Shaker, MD, Director of the Digestive Disease Center, have found a simple and effective way to help people with dysphagia and aspiration that occurs after the swallow and is caused by disruption in the opening of the upper esophageal sphincter. The upper esophageal sphincter (UES) is the opening to the esophagus or food tube. If this sphincter is unable to open, there will be residual food in the throat after the swallow is completed and this may result in aspiration.

The Shaker Exercise, developed to increase upper esophageal sphincter (UES) opening, is an example of one possibility for scientific advancement through interdisciplinary team cooperation. The contributing influences for the development of the Shaker Exercise span a broad spectrum of disciplines including radiology, medicine, speech pathology, surgery and biomechanics.

The physiologic basis of the Shaker Exercise is not new. It is based on the fundamental principle that the upper esophageal sphincter (UES) opening is a me-

chanical event, consisting of a series of external forces applied to the UES combined with relaxation of the sphincter musculature. The Shaker Exercise is aimed at making the swallowing muscles, located in front of the neck, stronger; and therefore they are able to work harder.

Thirty-one elderly subjects who were healthy, but had swallowing problems, participated in the Shaker Exercise Study. These individuals were assessed before and after random assignment to the Shaker Exercise or the sham exercise (the control exercise).

Under the supervision of Caryn Easterling, M.S. CCC a licensed Speech Language Pathologist, patients were instructed to do the Shaker Exercise in the following manner:

- Lay flat on your back on the bed or floor.
- Raise your head and look at your toes for one minute.
- Do not raise your shoulders off the bed or floor as you raise your head and look at your toes.
- Lay your head flat and rest for one minute.
- Repeat this sequence a total of three times.
- Then raise your head thirty times consecutively without holding it. Again, do not raise your shoulders while looking at your toes.
- Do the entire exercise three times per day for six weeks.

The results of the study were as predicted; an increase in anterior movement of the larynx, increased opening of the top of the food tube (UES) and a decrease in the pressure exerted on the food as it passed through the throat.

These findings resulted in an ongoing clinical investigation utilizing the Shaker Exercise and focusing on analysis of the exact muscle group(s) that were physiologically affected by the Shaker Exercise and application and effect of the Shaker Exercise on the antero-posterior (A-P) UES opening of dysphagic patients with aspira-

tion after the swallow secondary to UES opening abnormalities and residual food in the pyriform sinuses (crevices in the throat or pharynx that are located on either side of the vocal cords).

As previously stated, multiple factors influence the food tube or UES opening mechanism. However, the specific muscles responsible for "distraction" (pulling open) of the UES opening are of primary interest in swallowing rehabilitation of the dysphagic patient. Using an electromyograph (EMG) an instrument that displays and records the use and fatigue of muscles, researchers were able to determine which muscles had been affected and therefore could be strengthened by the Shaker Exercise.

For the purpose of this clinical research study of dysphagic patients, researchers focused on tube-fed patients with pharyngeal pyriform sinus residue and post deglutitive aspiration due to abnormal upper esophageal sphincter opening. Application of the Shaker Exercise to the rehabilitation of nineteen dysphagic patients was presented to the Eighth Annual Dysphagia Research Society. The aim of this clinical research study was to:

- determine the effect of the Shaker Exercise on swallowing function and biomechanics in tube fed patients with deglutitive failure defined by the presence of post deglutitive pyriform sinus residue and aspiration due to UES dysfunction,
- assess follow-up nutritional and swallow status after completion of the Shaker Exercise.

Seven of the nineteen patients in the study were randomly assigned to the Shaker Exercise or a "sham" Exercise. The "Sham" Group subsequently performed the Shaker Exercise after completion of six weeks of "Sham" Exercise.

Researchers concluded, that by "using the Shaker Exercise, safe oral feeding could be restored in tube fed patients

deemed previously to have an unsafe swallow and aspiration due to abnormal UES opening." The Shaker Exercise appeared to have an enduring effect in maintaining oral nutrition.

Although researchers now had an understanding of the muscle groups that the Shaker Exercise physiologically strengthens, and saw that the Shaker Exercise had a rehabilitating effect on deglutitive function in dysphagic patients, it was unknown whether the efficacy of this exercise was influenced by the etiology and/or the duration of dysphagia.

Therefore, the aim of the most recent study was to determine the effect of the exercise on deglutitive function and biomechanics in dysphagic patients with different etiologies and duration of dysphagia. Twenty-seven tube-fed dysphagic patients were assessed before and after six weeks of the Shaker Exercise Protocol. Prior to the exercise regime, all patients experienced aspiration or entrance of food into the airway, residual food in the throat after the swallow, and decreased opening of the top of the food tube or UES. The radiation patients had not had surgery or chemotherapy. Methods of assessment included videofluoroscopic evaluation of aspiration, biomechanics, and the Functional Outcome Assessment Measurement of Swallowing (FOAMS)

The etiology of the dysphagia was

- Hemispheric CVA - 9 patients (stroke)
- Brainstem CVA - 7 patients (stroke)
- Pharyngeal radiation injury - 5 patients
- various etiologies - 6 patients.

The duration of dysphagia ranged from

- less than two months in 8 patients
- two to four months in 6 patients
- four to six months in 4 patients
- greater than six months in 9 patients.

For every tested etiology and duration of dysphagia, six weeks of exercise resulted in resolution of post deglutitive aspiration and resumption of oral feeding. Analysis of antero-posterior upper esophageal sphincter opening after six weeks of exercise showed significant increases compared to pre-exercise values. Furthermore, the Functional Outcome Assessment Measurement of Swallowing (FOAMS) scores were also significantly higher after completing the exercise regimen compared to pre-ex-

ercise scores.

The Shaker Exercise is a simple and cost effective rehabilitative technique for post deglutitive aspiration. With the guidance of a Speech Language Pathologist specializing in swallowing disorders, this exercise may be helpful to some patients with abnormal upper esophageal sphincter opening.

Editor's note: Caryn Easterling, M.S. CCC is a doctoral student at the University of Wisconsin-Madison. She is President of the Wisconsin Speech Language Pathology and Audiology Association and a researcher and Speech Language Pathologist in the Division of Gastroenterology and Hepatology at the Medical College of Wisconsin, Milwaukee, WI. For additional information, please call 414-805-3870.

Attention: Patients should consult their physicians before using any treatment.

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SARCOMAS continued from page 2

and presence or absence of regional lymph nodes or metastatic disease, surgical excision of the primary site is performed. Less than total excision may be accomplished due to the proximity to vital structures.

Surgical excision is followed by chemotherapy and localized radiation therapy to 4000 cGy for microscopic residual disease with higher doses for gross residual disease. This yields a 10 year survival of greater than 60% in those children who do not have metastatic disease at the time of diagnosis and 20% for those with metastatic disease.

It is clear that for this histologic type of tumor in children, chemotherapy is very effective and a key component of therapy. This type of success is not the case for other histologic types of soft tissue sarcomas in either children or adults.

Adjuvant chemotherapy trials for sarcomas of the head and neck have failed to show any statistically significant benefit. The most active agents used against this disease have been adriamycin and ifosfamide. For metastatic disease, most large randomized cooperative group studies have reported response rates of 17-30% with a median survival of 12 months.

Editor's Note: Norman D. Bloom MD FACS is Chief of Surgical Oncology at the Cabrini Medical Center New York City and Clinical Professor of Surgery at New York Medical College. He is also a Lecturer in Surgery at Mt. Sinai School of Medicine. ■

LOCAL CHAPTERS OF SPOHNC

SPOHNC-ATLANTA

Harmon Grotzky; Coordinator
404-284-8045 <H26C30@aol.com>

Active for more than three years, this chapter of SPOHNC meets on the 2nd Monday of each month at St. Joseph's Hospital in Atlanta, GA

SPOHNC-BOCA RATON

Darci Lypson-McNally, LCSW
Coordinator/Facilitator

561-637-7216: <DMcNally@brch.com>

The SPOHNC-BOCA RATON chapter meets at the Boca Raton Community Hospital on the 1st Tuesday of each month from 3:00PM-4:00 PM.

SPOHNC-BOSTON, MA

Valerie Hope Goldstein:
Coordinator/Facilitator

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SPOHNC-BOSTON meets on a Tuesday at Massachussetts General Hospital. Please call for additional information.

SPOHNC-HOUSTON

William A. Phelan: Coordinator
281-259-4110 <w_phelan@yahoo.com>

This newly formed chapter will be meeting at MD Anderson in Houston, TX, Please contact Bill for more information.

SPOHNC-LONG ISLAND, NY

Nancy Leupold:
Coordinator/Facilitator

1-800-377-0928 <Nleupold@spohnc.org>
This is the original chapter of SPOHNC which was founded in 1991. It presently meets on the second Tuesday evening of the month at the Holistic Center, in Syosset, NY.

SPOHNC-MIAMI, FL

Marsha A. Braunstein:
Coordinator/Facilitator

305-596-6566 <marshab@the-beach.net>
SPOHNC-MIAMI meets on the first Wednesday of each month at 5:30 PM at the Baptist Hospital Radiation Oncology waiting room.

SPOHNC-MIAMI, FL

Mort Silverblatt Head and Neck Support Group
Mort Silverblatt; Coordinador Emmeritus
Martin Mash: Coordinator
Penny Fisher, Faciliator/Coordinator
305-243-4952; <pfisher@med.miami.edu>
This newly formed chapter meets from 1:00 PM-2:00 PM on the first and third Monday of the month at the Sylvester Comprehensive Cancer Center on the University of Miami Medical Campus.

SPOHNC-NJ-PA

Micki Naimoli: Coordinator
856-722-5574

This New Jersey/Pennsylvania chapter meets at the University of Pennsylvania Hospital in Philadelphia. Please contact Micki for additional information.

SPOHNC-OMAHA, NE

Katherine Jones:
Coordinator/Facilitator
402-559-8940 <kjonesj@unmc.edu>

SPOHNC-OMAHA meets on the third Friday of the month at the University Medical Center. Please contact Katherine for time and place.

SPOHNC-SAN DIEGO

Valerie Targie:
Coordinator/Facilitator
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This chapter is presently meeting on the last Saturday of each month. Please contact Valerie for place and time.

SPOHNC-UPMC-PITTSBURGH, PA

Marilyn Hudak, RN:
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SPOHNC-PITTSBURGH, PA, UPMC will hold its first meeting on September 21st at 2:00 PM. The meeting is entitled "Coping with Changes in Body Image and Function." Please contact Marilyn for exact location.

SPOHNC-FAIRFAX, VA, Heads Up!

Pam Black, LCSW:
Coordinator/Facilitator

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This is the newest chapter of SPOHNC. Although the group has been in existence since last fall, the members recently asked to become a chapter of SPOHNC. This chapter meets on the 2nd and 4th Wednesdays of each month from 5:30-7:00 PM at Inova Fairfax Hospital in Falls Church, VA.

SPOHNC-BOUND BROOK, NJ

Bernadette Maszczak
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SPOHNC-BOUND BROOK, NJ is in the development stages and is looking for interested survivors and their families to become involved. For more information, please contact Bernadette.■

BOOK REVIEW

Making Informed Medical Decisions;

Where to Look and

How to Use What You Find

Nancy Oster, Lucy Thomas

Darol Joseff, MD.

O'Reilly Press © 2000

ISBN1-56592-459-2 \$19.95

Web site: www.patientcenters.com

Reviewed by Betsy Wilson

O'Reilly press is "committed to empowering individuals to evolve into informed consumers armed with the latest information and heartfelt support for their journey." In this 350 page paperback they do exactly that.

We are coached by the authors to see how high quality care can result from educating ourselves about our situations. The reasons for our giving attention to research, alternative therapies, statistics, the internet, etc., are all carefully and clearly presented. The resources to do our "searching" are all here in this book.

My favorite part of the book is how it continually teaches us to become equal partners with our medical teams. Life in medical settings may not always encourage partnerships between patient and doctor, but this book gives all of us, who want to become a vital part of our medical care, the skills to participate. This book would be a welcome gift to anyone newly diagnosed.■

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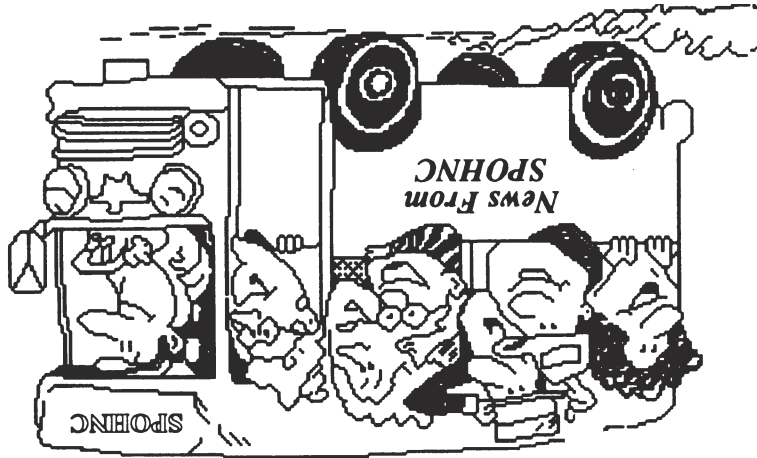
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