



Malignant Orbital Tumors Diagnosis, Treatment and Rehabilitation

DANIEL G. DESCHLER, MD, FACS

Introduction

The term “tumor” refers to an abnormal growth, which can occur anywhere in the body. Tumors may be *benign*, meaning that although they can continue to grow, they do not spread to other sites in the body or aggressively invade structures near them. Tumors may also be *malignant* indicating that the cells which make up the tumor can invade and destroy nearby tissues as well as spread to other parts of the body – metastases. If a benign tumor occurs in a delicate place, which does not allow room for it to expand, such as within the brain, it can be an extremely serious matter even though the tumor cells are not malignant.

The treatment of orbital tumors provides a significant challenge within the realm of head and neck surgery. Because of the strategic location of the orbit, numerous processes can affect the orbit from externally as well as begin within the structure itself. A basic knowledge of orbital anatomy is helpful to understand the diagnosis, treatment and rehabilitation of tumors within this region.

The orbit consists of a bony cup, which contains many critical elements. The bony framework of the orbit consists of portions of seven bones within the skull: the ethmoid, sphenoid, palatine, frontal, lacrimal, maxillary and zygomatic bones. Located within this bony skeleton are numerous structures, which may provide the origin of cancers within the orbit as well as be affected by cancers that occur in proximity to it. The contents of the orbit are largely contained within a tough membrane called the periorbita.

The periorbita is made up of the lining, which provides blood and nutrients to the bony skeleton. The eyeball, or globe, is the critical item within the orbit. The remaining structures within the orbit provide supportive roles to the globe. These include the optic nerve, which provides vision; the first and second branches of the trigeminal nerve, which provide sensation to the upper and middle face; as well as the nerves which allow movement of the muscles to the eyeball, the extraocular muscles. In addition to the six muscles, which move the eyeball, is another muscle, the levator palpebre, which allows opening of the upper lid. These structures are all surrounded by orbital fat, which provides the remaining volume of the orbit. Finally, within the orbits are the lacrimal glands and draining ducts, which provide lubrication to the eye and maintain hygiene. The eyelids with their skin, hair and glands may also be a source of tumors involving the orbit.

The orbit has three major entry points through the facial skeleton. These include the optic canal – through which passes the optic nerve providing vision; the superior orbital fissure – through which run the nerves for eye movement as well as sensation to the upper face; and the inferior orbital fissure – which contains the nerve for the middle face. Tumors from outside the orbit may enter the orbit through these openings or tumors originating in the orbit may pass through these holes to involve areas outside of the orbit.

As important as the structures within the orbit itself are those which surround it. Directly superior to the orbits is the floor of the cranial vault, the bony case that contains the brain. Above this bony floor is the front portion of the brain. Between and below the orbits are the paranasal sinuses, which are separated by thin bone. This thin bone can often be easily broken down by tumors occurring within the sinuses. Behind the orbit is a region filled with critical nerves and blood vessels called the cavernous sinus.

As with other areas of the head and neck, there are specific regions of lymph nodes that receive drainage from the orbital areas. These include lymph nodes within the parotid gland, the facial lymph nodes at the undersurface of the jaw, the submandibular gland and its lymph nodes as well as remaining lymph node regions within the neck.

To accurately and successfully manage tumors occurring within the orbit attention to all the anatomy discussed above is essential.

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For Head & Neck Cancers
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Malignancies of the Orbit

Malignancies involving the orbits may occur within the orbit itself and are referred to as primary lesions. Tumors may also arise in areas around the orbit and involve it secondarily. These are referred to as secondary malignancies. Finally, cancers may travel from other parts of the body far removed from the orbits and begin to grow there. These are referred to as metastatic lesions or metastases.

The treatment of malignancies of the orbit often involves a multidisciplinary approach. This brings to bear the latest advances in multiple medical specialties such as chemotherapy, radiation therapy, rehabilitation as well as surgical techniques to provide the most thorough and potentially successful approach to these challenging tumors. Surgeons may be involved from various specialties including otolaryngology, ophthalmology, neurosurgery and plastic and reconstructive surgery. The various methods of therapy may be used in any combination based upon clinical experience, which has shown which combinations to be the most successful. When the orbits are addressed surgically, the approach can vary. Extensive removal of all contents within the orbits referred to as an orbital exenteration, may be performed. Surgery may involve only the removal of the eyeball itself, which is known as enucleation. Other surgical variations involve the partial resection of those portions of the orbit involved by the tumor with maximum efforts used to safely preserve remaining structures.

Reconstructive efforts are planned to maximize the function of remaining orbital structures as well as provide a stable wound which can be cosmetically acceptable with the use of appropriate prostheses.

Primary Orbital Tumors – Tumors from Within the Orbit

The most common tumor of the globe in children is the retinoblastoma. This usually occurs before the age of 6 and may be invasive beyond the globe itself and be prone to distant metastases. Sixty-five percent of retinoblastomas are bilateral and thirty-five percent are unilateral. The majority occur sporadically with no specific cause, but there are a certain number which are genetic and passed down in families. The most common presentations are a white pupil on examination or abnormality in eye movement. Treatment involves chemotherapy, radiation therapy and possible enucleation – removal of the eyeball. The survival of patients with retinoblastoma treated with aggressive combined therapy is between 90 - 95 percent. Numerous other rare tumors may occur within the very specialize structures of the globe. Melanomas, although rare, are particularly aggressive and require similar therapy.

The lacrimal gland is a common site of orbital tumors. As the lacrimal gland is similar in structure to the salivary glands, lacrimal gland tumors share similar types to salivary gland tumors. The adenoid cystic carcinoma is a particularly insidious tumor of the

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lacrimal gland and orbit. It has a high tendency to travel along nerves and therefore even with aggressive therapy, 40% of these tumors can come back within the orbit. Another one-third of the tumors will have distant metastases to lungs, bone, liver, or brain. Aggressive surgical therapy combined with radiation therapy is the standard treatment for these lesions. An even more aggressive lacrimal tumor is the pleomorphic ex tumor. This is a malignant version of the benign pleomorphic adenoma, which occurs in the lacrimal gland and likewise requires aggressive combined therapy.

The rhabdomyosarcoma is a soft tissue tumor, which may arise within the orbit. There are numerous subtypes and each carries a different prognosis and behavior pattern. This tumor can present with the rapid bulging of the eye, called proptosis. The treatment of this lesion is based primarily upon chemotherapy and radiation therapy with surgery reserved for those tumors that do not respond. Early lesions have shown 83% survival rate while more advanced lesions have a survival rate of 20%. Sarcomas are another type of tumor, which can occur from the bony tissues making up the orbit. The treatment of these lesions is likewise based in surgery with the use of possible radiation therapy and chemotherapy. Recent efforts have been placed on using specialized proton radiation for the treatment of these tumors. Orbital tumors may also occur which are related to the nerves contained within the orbit. These include schwannomas and meningiomas. Although benign by strict definitions, these tumors can have significant impact because of their growth within a confined space.

Secondary Tumors – Tumors from Around the Orbit

As stated before, tumors may grow from areas surrounding the orbit and significantly involve this region. Basal cell carcinomas and squamous cell carcinomas, as well as melanomas, of the skin surrounding it may grow to involve the orbit. Treat-

ment of these lesions is based upon how extensively the tumors involve the orbit. Treatment can involve surgeries as simple as local excision to full orbital exenteration.

Tumors that may commonly involve the orbits secondarily are those that originate in the sinuses surrounding them. There are numerous types of sinus tumors including squamous cell carcinomas, adenocarcinomas, salivary gland tumors and olfactory neuroblastomas (tumors arising from the nerves which provide the sense of smell). When sinus tumors are large and advanced, surgery may not only require extensive removal of the tumor within the sinuses, but also structures within the orbit. This may involve only the bone between the sinuses and the orbits. Surgery may also involve removing the lining of the orbit, called the periorbita, as well as portions of the lacrimal system. If these tumors invade through the periorbita, and grow into the fat and muscle surrounding the eyeball, orbital exenteration is usually required. Additional therapy with chemotherapy and radiation therapy is then added depending on the specific type of tumor. Tumors originating from the lining of the brain may also grow to invade the orbit. These are called meningiomas and are usually benign, but because of their growth in this critical location, can have an aggressive and malignant behavior. These are usually approached with surgery and radiation therapy in conjunction with the efforts of the neurosurgical team.

Metastases to the Orbits – Tumors from Elsewhere in the Body

The majority of metastatic tumors from other parts of the body which occur within the orbital region, occur within the globe. They may also involve the additional structures of the orbit. Roughly 50% of metastatic tumors within the orbit come from breast cancers. The next highest incidence is from lung and prostate cancers with gastrointestinal and kidney tumors providing the remainder. Metastatic tumors within the orbit itself are usually addressed surgi-

cally if possible, with chemotherapy being added to provide some control for the spreading nature of the original tumor.

Reconstruction and Rehabilitation

As noted earlier, surgery in the orbit can take various forms from limited removal of specific structures to removal of all the orbital contents. When limited resection is undertaken, the reconstructive efforts are aimed at replacing these tissues while maximizing the function of the remaining tissues within the orbit. If the critical portion of orbital bone is removed, this can be replaced with bone grafts from the skull or elsewhere in the body. Likewise specialized mesh made from titanium or other prostheses can be created and secured to reapproximate the lost structural support. Fascia, the thin, strong tissue which surrounds muscles, can be used to reconstruct not only the missing lining known as periorbita, but also to provide structure and support of the orbital contents.

The goals of limited and focused reconstructions are to maintain vision, movement, volume and appearance, and tear drainage from the orbit as close to normal as possible. If enucleation must be undertaken, then an eyeball prosthesis may be placed which is cosmetically acceptable. In cases of orbital exenteration, where all the orbital contents are removed, there are often significant portions of nearby bone from the sinuses and skull missing. This may include bone which separates the brain from the orbit. The goal of reconstruction in this setting is initially aimed at covering these areas with healthy and reliable tissue from elsewhere in the body. When tissue is moved from one location in the body to another, it is referred to as a flap. Flaps may be from right near the orbit such as the forehead or further away, such as the chest, back, arm or abdomen. Tissue flaps can include skin, muscle and bone as needed to rebuild an area. One form of flap reconstruction is called free flap reconstruction. With this method, a piece of tissue is obtained from elsewhere in the

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Psychosocial and Nutritional Aspects of Head and Neck Cancer

Jayne Wiprovnick, MS, RD, CDN

Over time, people tend to develop strong relationships with food that are difficult to change. Dietary habits are life time acquired, and when a person is faced with head and neck cancer, the most fundamental functions of eating may be completely compromised.

Although head and neck cancer accounts for only 5% of all cancers in the United States, it can be one of the most challenging. Since this type of cancer often affects the oral cavity, a person's ability to consume a normal diet and to maintain adequate nutrition may be at risk. Alcohol abuse, which is often a predisposing factor for developing head and neck cancer, also leads to poor dietary habits, poor dentition, as well as vitamin and mineral deficiencies.

Treatment options for head and neck cancer including surgery, radiation therapy and chemotherapy, may further compromise nutritional status and lifestyle. Five common side effects of treatments are : impaired mastication function (difficulty chewing), dysphagia (difficulty swallowing), xerostomia (dry mouth) and thick saliva, mucositis (mouth sores) and dysgeusia (taste changes such as intolerance to sweet, bitter and salty flavors). One or more of these side effects can ultimately affect the consistency of one's diet and lead to poor nutritional health. Therefore, it is crucial to implement a plan for good nutrition that will help to provide the optimum success of cancer treatments while considering the patient's quality of life.

The goals of nutritional therapy for patients with oral and head and neck cancer should provide sources for sufficient calories, protein, fluid, vitamins and minerals to meet nutritional needs; guidance for maintaining weight and hydration; ways for aiding in the regeneration of healthy tissue; and encouragement to the survivor to follow nutritional guidelines to help improve quality of life.

Common Side Effects of Treatments and Dietary Interventions

Mastication and Dysphagia Problems

- Difficulty chewing – try mechanical soft diet (foods that require little or no chewing and break apart easily), chopped, ground or soft meats and poultry, eggs, fish and pastas.
- Difficulty swallowing – try pureed or blenderized diet (foods that require no chewing, are blenderized or strained to reach desired texture and form a cohesive bolus), pureed or blenderized meats, poultry and fish, baby food (stage 1 and 2), souffles, puddings, and smooth cottage cheese.
- Eat small frequent meals; include high calorie liquids, com-

mercial liquid supplements and snacks between meals (see tips for boosting calories).

- When eating, sit upright, chew and swallow slowly.
- Follow special eating or swallowing techniques taught by your Speech Pathologist..

Xerostomia and Thick Saliva

- Add gravies and sauces, applesauce or other liquids to foods.
- Have a spoonful of warm liquid such as soup between mouthfuls of food.
- Ask your physician about artificial saliva.
- Use pureed or mechanical soft and moist foods such as casseroles, macaroni and cheese, yogurt (if you have a heart condition or high blood cholesterol you may wish to try low fat varieties of these foods).
- Drink 8 to 10 cups of fluid per day; always carry a bottle of water with you (or a small spray bottle filled with water and spray your mouth continuously throughout the day).
- If mucositis is not present, try sucking on sugar-free sour candy, lemon drops, sugar-free gum; use citrus beverages, lemonade, or citrus flavored soft drinks to stimulate saliva.

Mucositis

- Eat moist, bland foods such as cream soups, jello, mashed potatoes, ice cream and pureed foods.
- Serve foods at room temperature.
- Drink through a straw to bypass mouth sores.
- Avoid alcohol and caffeine; avoid rough, salty, raw course foods such as crackers.
- Avoid spicy foods, irritating spices and seasoning such as chili powder, cloves, nutmeg, pepper and horseradish.
- Avoid acidic foods such as tomato sauce, citrus fruits and juices. Use fruit nectars such as apricot, mango, papaya and guava.
- Ask your physician for a topical anesthetic or prescription pain medication and use 15 minutes before eating.
- Rinse mouth often with baking soda and water (one quart water to one Tablespoon baking soda).

Dysgeusia*

- To reduce metallic taste, use plastic utensils instead of silverware.
- Try tart foods such as lemon custard, orange sorbet, or citrus juices.

- Try herbs and spices such as basil, oregano, rosemary and mint, onion and garlic.
- Use pineapple to neutralize taste.
- Chew on sugar-free gum, fresh parsley or mint.
- Marinate foods in fruit juices, nectars, duck sauce, or sweet and sour sauce.
- Select fresh vegetables and frozen fruits such as cantaloupe, grapes, and tomatoes.

* *These suggestions are meant for patients with taste changes alone and not for patients who have other side effects such as mouth sores (mucositis).*

Emotional Awareness

From the moment we are born we begin to form relationships with people and things. How a person relates to food is determined by many factors. Our relationship with food begins in infancy, continues through childhood and adolescence, and carries into adulthood. Food has profound effects on our daily lives and health. Our experiences surrounding food are closely intertwined with our emotions and feelings. Family, friends, peers, social habits, as well as the media, individually and in combination, influence our acquired dietary habits. Food is often accompanied by emotional ties and feelings; hence, eating disorders such as anorexia and over-eating have become prevalent in our society.

Maslow's hierarchy of needs states that the act of "taking nourishment" is a basic physiological need that must be met before any other need. Next in order of importance is the need for safety and security, followed by affection and social activity and finally, self-realization. Food can satisfy many of these needs. For example having ample food can make us feel safe and secure.

It is therefore natural for a mother or mother figure to "nurture" her baby. Her instincts tell her to love and provide nourishment for her offspring. An infant who is held, loved and fed appropriately becomes a healthy secure child and adult.

Food has many uses. When used as a comfort measure, we tend to eat more particularly when we are stressed. It may be used as a reward for something achieved; or as a punishment for some wrong-doing. If varied and combined properly food can provide the nutrients necessary for good health. More importantly, food is an integral part of our social lives and eating is a social function. The celebration of any major life event such as a wedding, birthday, or a graduation, are all planned around a meal. In most any culture, the offering of food or drink is a friendly gesture and many of our cultural and religious rituals involve food and drink.

Food Transitions

Head and neck cancer patients are faced with the realization that they may never be the same. Their overall appearance may change and they may communicate differently. In addition, their ability to chew and swallow may be altered as well as their sense of taste and smell. They may no longer be able to eat the foods they enjoy and to which they are accustomed. They may need to adapt to an entirely new way of eating and drinking. When eating becomes a challenge and no longer a pleasurable event, people tend to become socially isolated, compromising their quality of life.

Obtaining nourishment is a basic life requirement; we need food and fluids in order to survive. Unfortunately, the physical changes associated with head and neck cancer make it even more difficult for a person to take in adequate nutrients. These physical changes undoubtedly lead to emotional responses such as shock, fear, anger, and withdrawal in response to this overwhelming major life transition.

The act of eating is second nature to us; it is as natural as waking up in the morning and washing your face. It provides us with a sense of consistency. Very early on, we learn that food is our link to life; an infant cries when it is hungry. When an illness strikes that effects the way we eat and *what* we eat, food takes on a whole new meaning.

This is when the Dietitian may be of help. The Dietitian assists the individual in adjusting to and compensating for this major lifestyle change. He or she becomes a facilitator in helping the patient to modify the consistency of his diet, while maintaining nutritious, palatable meals. The Dietitian, along with the Social Worker can help lead the patient to see this change as a new and exciting challenge and assist in coping with psychosocial stages of illness. The patient must learn to become independent in his/her food choices and preparation, taking into consideration all the limiting factors. The individual may need to rely on memory and imagination to help taste and smell certain foods.

It has become apparent that living with head and neck cancer is a life-changing event that can be managed appropriately with the assistance of health care practitioners.■

Editor's Note: Jayne Wiprovnick, MS, RD, CDN works as a Clinical Dietician/Nutritionist in the Department of Food and Nutrition Services at Memorial Sloan-Kettering Cancer Center in New York City. For the past nine years, Ms. Wiprovnick has worked with the head and neck cancer population and has presented lectures on the topic of "Head and Neck Cancer and Nutrition."

Part II, "Nutritional Growth for Head and Neck Cancer Survivors" will appear in the Winter issue of *News From SPOHNC*.

A TIME FOR SHARING

I want to describe my experience with a new procedure that delivers a highly concentrated dose of Cisplatin to the tumor site. It is my understanding that this treatment is generally effective when used in combination with radiation and thus is generally not being used when cancer reappears in an area that has already been radiated.

I am a 55 year-old resident of Washington, D.C. I was diagnosed in August 1998 as having a primary tumor at the base of my tongue and secondary tumors in three lymph nodes on the upper left side of my neck. Despite being a non-smoker and non-drinker, I found myself with stage IV cancer. Initially, I was stunned, feeling that I would die in three months. That pity party lasted one week. Then I got down to fighting this thing.

After much consultation and reading, I opted for the protocol at George Washington University Hospital. The procedure seems to be primarily used for killing tumors in areas (e.g., base of tongue) where surgery would greatly impair bodily functions. It combines standard treatment (i.e., radiation, neck dissection, and systemic chemotherapy) with a new procedure called Intra-Arterial Infusion (IAI). Generally, IAI is used as the very first step in the treatment process. It either shrinks the tumor to a very small size or eliminates it.

In my case, the tongue tumor was reduced from the size of a half-dollar to the size of a pea after four infusions--and was undetectable after six infusions. (Because I began radiation after the 4th infusion, the subsequent disappearance of the tumor cannot be entirely attributed to the IAI alone.)

My surgeon tells me that some patients have started out with swollen tongues hanging out their mouths and, by the time they start radiation, find that their tongue tumors have been reduced to nearly nothing. GWU doctors report having good results over the past five years with approximately one hundred patients. Five-year data are not yet available but GWU doctors are planning to release their results, probably next year.

A team of 5 health care professionals (2 radiologists and 3 assistants) performed the IAI. They first inserted a catheter in one of my leg arteries at the groin area. Using

X-rays and video monitors to observe the catheter, they worked this tube up various arterial pathways until it was at a branching point that supplied blood to both the tongue base and the lymph node area. The catheter was positioned carefully so that it could bathe all the tumors with Cisplatin.

The radiologist chose a position that would release 75% of the Cisplatin to the tongue base and 25% to the lymph node area. To do so, he first did a trial run with dye. Because the dye shoots rapidly into the bloodstream, the dye release was played back in slow motion on video monitors to ensure that placement was correct. In this way, the radiologist was able to deliver a dose of Cisplatin to my tongue base that is a thousand times what could be delivered to that area using systemic release (i.e., infusion into the arm).

I could tolerate the lethal dose of Cisplatin only because an antidote was released at the same time. It was released through a second catheter, which had been snaked up my veins to a position near the first catheter. The antidote neutralized most of the poison. The remaining poison, however, was sufficient to keep me in bed for five days (only one night in the hospital).

Because the IAI released a mega-dose of poison only a few inches from my brain, success requires an expert radiologist who is well seasoned in positioning such catheters. It therefore is not surprising that only a few cancer centers are performing these infusions.

I had six of these IAIs spread over two months. (Most patients, however, need only four IAIs.) I also had two months of radiation, followed by the neck dissection and then systemic infusion of Taxol, administered once every 3 weeks over a period of 3 months.

Fortunately, my insurance covered my treatments. Generally IAI is not considered experimental for insurance purposes because it is a new way of delivering an approved and well established drug, Cisplatin.

I would like to share with you the following abstract of a paper presented at the 5th International Conference on Head and Neck Cancer, San Francisco, CA, July 30, 2000. (The George Washington University

Medical Center, Washington DC 20037 USA) entitled, "Intra-Arterial Cisplatin followed by Radiation Therapy for Advanced Squamous Cell Carcinoma of the Head and Neck" Wilson WR, Siegel RS, Harisiadis L, Davis DO, Nguyenm HH, Bank WO,

"Improving survival and simultaneously preserving organ integrity remain challenges in the management of advanced (stage III and IV) squamous cell carcinoma of the head and neck. Since August 1994 we have utilized a treatment regimen consisting of four weekly intra-arterial infusions of Cisplatin (150 mg/m²) targeting the tumor bed followed by 6 weeks of radiation therapy.

We have treated approximately 100 patients with this regimen; the 58 patients (44 men and 14 women) who have been followed at least 2 years (median follow up of 25 months) form the basis of this report. There were 42 previously untreated patients (4 Stage III and 38 Stage IV) and 16 previously treated patients with recurrent disease in this cohort. Response to treatment was determined by magnetic resonance imaging (MRI) and clinical examination.

Of the 42 previously untreated patients, 27 are alive and disease free, corresponding to a sustained complete response rate of 64.3% with a median follow up of 30 months. Among 16 patients with recurrent, previously treated carcinoma, there are 4 survivors, corresponding to a sustained complete response rate of 25% with a median follow up of 15.5 months.

There have been no deaths or serious complications related to the treatment in either group. Only one patient required resection of the tumor site (laryngectomy).

In conclusion, the combination of high-dose, intra-arterial cisplatin and radiation therapy is effective in improving survival and organ preservation rates in previously untreated, advanced squamous cell carcinoma of the head and neck."

Chuck Needy
Washington, DC

Editor's Note: Additional information about Intra-Arterial Infusion can be found in the May, 2000 issue of News From SPOHNC.

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body, such as a back muscle, and placed in the orbital defect. The small blood vessels – an artery and a vein – are then reconnected in the neck using a microscope. This way, the transferred tissue has a good blood supply to allow successful healing at the reconstruction site. When successful healing has occurred, prostheses can be made to provide a more acceptable cosmetic result.

Summary

As orbital tumors involve an intricate part of human facial anatomy, their treatment is likewise challenging and intricate. A multidisciplinary approach involving chemotherapy, radiation therapy and surgical techniques from multiple specialties, has served to provide improved success of treatment. These efforts, in conjunction with newer reconstructive methods, provide hope for the successful treatment and rehabilitation of malignancies affecting the orbit. ■

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from PAT'S PANTRY
PROVENÇAL

CHICKEN BRAISED IN CIDER

- 3 boneless chicken breasts, sliced rather thinly
- 3 tablesppons olive oil
- 2 onions (chopped)
- 2 1/4 cups apple cider or juice
- 2 cups button mushrooms (fresh or canned)
- 1/2 cup plain yogurt or sour cream
- Salt to taste
- Milk as needed

Fry chopped onion in the olive oil until soft . Salt the chicken pieces and add to onion. Cook chicken gently five minutes on each side. Add mushrooms and cider and simmer for 20 minutes or until the chicken is cooked through. Put mixture into blender and add yogurt and milk as needed.

A good dessert for this dish would be baked apple (core before baking; remove peeling after baking) with cinammon, sugar, a little milk or cream, Blend.

November's Tip: You can now find delicious free range chickens in many supermarkets. You can always tell they're good, because the meat is darker in color than the industrial chickens, and the flavor is wonderful.

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The State-of-the-Art

A replay of this teleconference will
be available on or about
November 1, 2000

Please visit SPOHNC's Web site at
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