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# DEVELOPMENT OF A HEAD SUPPORT DEVICE FOR PEOPLE WITH HYPERMOBILE-TYPE EHLER-DANLOS SYNDROME

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## INTRODUCTION

Ehler-Danlos Syndrome (EDS) is a group of heritable disorders that are characterized by abnormal collagen synthesis, leading to multisystemic connective tissue disorder. Six major types of EDS are recognized, the most common of which is the hypermobile type. EDS hypermobile-type (EDS-HT) is characterized by connective tissue laxity resulting in excessive joint mobility due to weak and less elastic ligaments and tendons. Patients with EDS-HT suffer from frequent joint dislocations and tears in their connective tissue that are slow to heal. Cumulative damage to joint connective tissue often results in frequent, intense pain in several different joints. This pain, combined with joint fragility, has the effect of severely limiting the activity levels of many EDS-HT patients, with a corresponding decrease their quality of life. [1]

A particularly common symptom amongst EDS-HT patients is pain and instability in the neck. Laxity of the ligaments of the neck may result in multilevel cervical instability, including craniocervical (CCI) and atlanto-axial instability (AAI). CCI and AAI may cause repetitive brainstem and spinal cord compression and stretch (medullary kyphosis), producing neurologic symptoms and longstanding damage. The vertebral arteries pass through the vertebrae in the cervical region, so instability at these levels may also result in repetitive decrease in blood flow through the vertebral arteries. In EDS-HT, the extensive complex of cervical ligaments may fail to provide the normal support and endpoints for range of motion, thus the muscles of the neck and upper back must fire almost continuously to stabilize the head and neck. This leads to muscle fatigue and pain from muscle spasm. EDS-HT patients describe the feeling of balancing a bowling ball on top of their necks, constantly working to support and stabilize their heads and unable to rest the muscles in their necks and shoulders.

In cases in which there is CCI and/or AAI, many EDS_HT patients will choose to undergo surgical fusion of occiput to C1, C1-C2, or more extensive multilevel fusion. These procedures have been shown to significantly decrease head instability, however the procedures carry a high risk, are extremely expensive, are somewhat controversial in the neurosurgical community with regard to the EDS population, and there are few neurosurgeons willing to do them. Furthermore, the long-term effects of this surgery on the spines of EDS-HT patients is uncertain. The procedures may transfer loads to vertebrae located below the fusion site, raising the possibility of need for subsequent fusions.

An alternative to spinal fusion surgery for EDS-HT patients is long-term use of a cervical collar in order to provide support for the head. It is common for EDS-HT patients to own one or more different types of cervical collar and to use these collars when they need to unload their neck. However, commercially-available collars were not designed with the needs of EDS-HT patients in mind. Many cervical collars are designed to be used after surgery or after an injury. They are stiff and completely immobilize the head, making it impossible to participate in many common activities. Furthermore, by completely immobilizing the head the post-surgical type of collars may contribute to atrophy and weakening of the muscles in the neck that are needed to support the head. An alternative to the post-surgical collars is the soft collar, usually made from foam wrapped in a cloth cover. These collars allow some movement of the head, however they are bulky and hot and they completely enclose the neck. This makes them very uncomfortable for many EDS-HT patients who are intolerant of pressure surrounding their necks.

In this abstract we describe work that has been performed to design a head support specifically for EDS-HT patients. We applied the methods of "human-centered design" (HCD), a method that has its origins in the field of information technology and was developed in

response to the need for better human-machine interfaces [2]. A key to the method is to develop an understanding of the physical and psychological needs of the human user. As it is applied to product development, HCD treats the end-user of a product as a member of the design team. The designer works to build an understanding of the user community at a human level. Product design becomes a highly iterative process in which customer interviews are quickly followed by a round of brainstorming, design, and prototyping. In this manner, designers and customers work together to converge on a design solution.

## METHODS

HCD dictates that the designer become familiar with the community of intended users. Towards this end, a survey of websites that provide support for people with EDS was performed. This provided insight into the type of head support devices that people with EDS-HT currently find useful. Next, we contacted the leaders of the Asheville area EDS Support Group who invited us to attend a group meeting. At this meeting we met several EDS patients who described their experiences with neck pain and confirmed the need for an improved head support device.

Seven people with EDS-HT (six female and one male) were recruited to participate in interviews. The goal of these interviews was to uncover desirable characteristics of a head support. The interview script and process were approved by the Institutional Review Board. Transcripts of these interviews were analyzed to determine the frequency with which interviewees gave particular responses. Table 1 gives a summary of the most frequent interview responses.

Using Table 1 as a starting point, a "voice of the customer" statement was formulated: "I want a device that will support and stabilize the weight of my head while allowing limited motion. It should fit well, be well-ventilated, and be unobtrusive."

| Interview Question | Most frequent responses |
|---|---|
| What causes pain? | Sitting upright with head unsupported<br>Pressure on occipital region of head<br>Looking up or down<br>Physical activity<br>Staying in any one position for long |
| How is the pain manifested? | Sharp pain in back of neck<br>Fatigue and tightness in neck muscles<br>Headache centered on occipital region<br>Sensation of upper vertebrae rubbing together<br>Looseness and pain in the jaw |
| Thoughts on existing "hard" cervical collar designs | Too confining<br>Fear that immobilizing head will lead to atrophy of neck muscles<br>Fit is often poor<br>Draws unwanted attention in public |
| Thoughts on existing "soft" cervical collar designs | Some allowed motion is a plus<br>Contact around entire neck is confining<br>Doesn't provide enough support of head weight<br>Draws unwanted attention in public |
| Desirable characteristics of a new head support design | Allows some motion<br>Stabilizes the head on the neck<br>Supports the weight of the head<br>Well ventilated<br>Adjustable or custom fit<br>Unobtrusive |

**Table 1: Most frequent responses given by EDS-HT patients during interviews**

Starting with this statement, a functional analysis of the head support was performed [3]. Primary functions were identified and broken down into sub-functions in order to construct a function tree. Several solution methods were then formulated for each sub-function. Sub-function solutions were then combined in order to look for promising solution variants. Two of these variants were selected for detailed design and prototyping.

## RESULTS

Figure 1 shows solid models of the two approaches that were selected for the first round of design. The "headband" design, shown in Figure 1a is intended to stabilize the head while allowing limited motion in all three rotation axes. The "space-frame" design is intended to support the weight of the head while also resisting motion. Both designs are constructed from molded ribs that can be engineered to flex in a prescribed manner. This allows limited motion of the head, while providing support and preventing excessive motion. The range of motion in different axes can be controlled by changing the stiffness and geometry of the ribs. Both designs contact the user's body at points that are not painful for people with EDS-HT. The designs are open, well-ventilated and unobtrusive. They leave the user's face and the front of the neck open and they follow the contours of the user's body so that they can be worn under clothing.

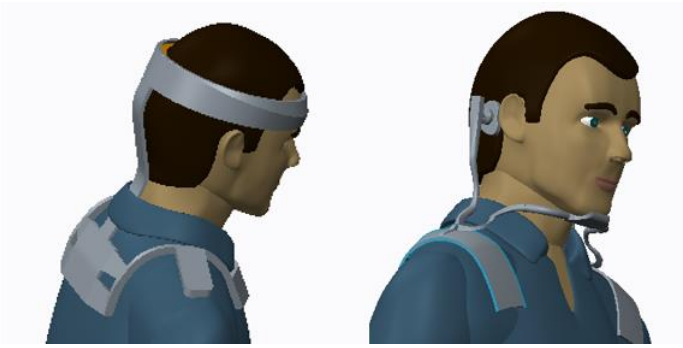

**Figure 1: Two head support designs. a) Headband design, b) Space frame design.**

## DISCUSSION

Using the methods of HCD, two head supports have been designed to meet the needs and desires of people with EDS-HT. The supports stabilize and support the head while allowing limited, prescribed motion. They contact the body in areas that people with EDS-HT do not find painful. They are open, well-ventilated, and can be worn under clothes, increasing the probability that they will be used in public.

This work is the first iteration in a highly-iterative process. Prototypes of the two designs are currently under construction. These prototypes will be taken to EDS-HT support group meetings in for feedback that will be used in the next design iteration. Over time, we hope to converge on designs that provide highly-usable, affordable devices for head stabilization and pain relief for EDS-HT patients.


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