Objective: To design a relational database integrating clinical and basic science data needed for multidisciplinary treatment and research in the field of vascular anomalies. Based on data points agreed on by the American Society of Pediatric Otolaryngology (ASPO) Vascular Anomalies Task Force. The database design enables sharing of data subsets in a Health Insurance Portability and Accountability Act (HIPAA)-compliant manner for multisite collaborative trials. Vascular anomalies pose diagnostic and therapeutical challenges. Our understanding of these lesions and treatment improvement is limited by nonstandard terminology, severity assessment, and measures of treatment efficacy. The rarity of these lesions places a premium on coordinated studies among multiple participant sites.

Setting: Vascular anomaly programs in the United States.

Results: A relational database correlating clinical findings and photographic, radiologic, histologic, and treatment data for vascular anomalies was created for standalone and multiuser networked systems. Proof of concept for independent site data gathering and HIPAA-compliant sharing of data subsets was demonstrated.

Conclusions: The collaborative effort by the ASPO Vascular Anomalies Task Force to create the database helped define a common vascular anomaly data set. The resulting relational database software is a powerful tool to further the study of vascular anomalies and the development of evidence-based treatment innovation.


Congenital vascular lesions (ie, hemangioma and lymphatic malformation), or vascular anomalies, have commonly been grouped into generic categories because they are difficult to describe. Lymphatic malformations illustrate the difficulties surrounding separation of vascular lesions into categories. The pathologic description of lymphatic malformations does not reflect their clinical appearance or behavior. Consequently, it is difficult to develop new treatments based on biological principles. More recently, biological and epidemiological data from these lesions are allowing some clinically relevant lesion differentiation. "Infantile" hemangiomas, tumors of proliferating endothelium first appearing in infancy, were initially differentiated from other vascular malformations by their natural history. This distinction is supported by detection of a biomarker unique to hemangioma endothelium, not present in other types of vascular lesions.

Combining careful clinical observation of congenital vascular lesions and cellular biological discoveries has resulted in a classification scheme for vascular anomalies. In this scheme, vascular lesions demonstrating neoplastic characteristics are "vascular tumors," whereas lesions consisting of abnormal vessel groups are "vascular malformations" (Table). Classifying vascular anomalies with this scheme has furthered our understanding of many types of vascular lesions. Some vascular lesions look like hemangiomas histologically but do not have the same clinical behavior and immunohistochemical staining pattern. These are called "congenital" hemangiomas and are distinct from "infantile" hemangiomas. Radiographic observations have characterized some lymphatic malformations as pre-
dominantly “macrocystic,” as opposed to “microcystic” lesions. Physiologically, certain infantile hemangio-
mas demonstrate abnormal transient arteriovenous shunt-
ing of blood flow, which is associated with high-output cardiac failure. These findings and others open the po-
sibility of further clinically relevant differences between vascular anomalies.

While discoveries in histologic, radiologic, and clinical categorization have changed our understanding of vascular anomalies, the subtle distinctions between these lesions are not widely recognized and have not been corre-
lated with treatment outcomes. Consequently, many vascular anomalies are still incorrectly called “hemangio-
mas,” when they are more appropriately called “vascular malformations.” Given the rarity of and nonstandardized nomenclature for congenital vascular anomalies, our abil-
ity to clearly define a lesion and develop effective treatment standards is hampered. As information about vascu-
lar anomalies has accumulated, changes have occurred in treatment and treatment philosophy. To improve vascu-
lar anomaly treatment, these ideas need refinement and stan-
dardization through objective measures.

Four years ago, a group of interested otolaryngolo-
gists (the Vascular Anomalies Task Force [VATF]) be-
gan meeting to discuss vascular anomalies and how changes in their classification and treatment can be ap-
plied to otolaryngology–head and neck surgery. One of the initial decisions was to use the vascular anomaly class-
sification scheme to standardize nomenclature.8 Building off this decision was the recognition that multidis-
ciplinary management and multi-institutional research collaboration would be necessary to accomplish the goal of developing evidence-based treatment approaches to head and neck vascular anomalies. Central to this en-
deavor was the development of a vascular anomaly re-
lation database capable of collecting data unique to vas-
cular anomaly treatment.15

Storage of data and database design have changed over the past several decades. In this evolution, relational da-
tabases have become important in prospectively connect-
ing clinical observational data with radiographic, sur-
gical, pathologic, and genomic data. The well-
designed relational database has features that eliminate redundancy of stored data and allow for the comparison of differing data types (ie, pictorial, categorical, and ordi-
 nal) while maintaining data integrity. In otolaryngol-
ogy, widely disseminated relational databases have been developed for use in quality-of-life assessments in head and neck cancer and staging of recurrent respiratory pap-
illomatosis. Other disciplines have adopted similar da-
tabases to study all aspects of disease care.

Designing relational databases is a labor-intensive pro-
cess, which requires the input of experts from multiple medical disciplines as well as programming expertise. Se-
lecting data points for inclusion in the database forces groups such as the VATF to further refine vascular anomaly nomenclature as well as standardize evaluation,
management, and follow-up of patients with vascu-
lar anomalies. The design process becomes a new way for experts to understand the disease processes they are studying. Properly performed, the design process builds a foundation that can facilitate multi-institutional re-

<table>
<thead>
<tr>
<th>Table. Vascular Anomaly Classificationa</th>
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<tr>
<td><strong>Vascular Malformation</strong></td>
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<tr>
<td>Single vessel type</td>
</tr>
<tr>
<td>Capillary</td>
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<tr>
<td>Venous</td>
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<tr>
<td>Lymphatic</td>
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<tr>
<td>Arteriovenous</td>
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<tr>
<td>Combined/complex malformations</td>
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<tr>
<td>Lymphaticovenous</td>
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<td>Capillary-venous</td>
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<td>Capillary-arteriovenous</td>
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search networks and clinical trials. This report de-
scribes the methods used to create a relational database to study vascular anomalies, as well as the capabilities of this software to further our understanding of vascular anomalies.

**METHODS**

Through a group discussion among the VATF, the purpose, data collection, and structure priorities of the database were identified. To aid this process, several hierarchical databases were created to identify key data fields and tables for collecting data specific for vascular anomalies. Additional input was obtained from related medical, surgical, and radiologic disciplines to capture data relevant to vascular anomaly assessment and/or treatment. This input was obtained through direct interview and medical literature review.

The relational database design process was focused on inter-
disciplinary identification of essential information specific to all vascular anomalies. This information was grouped in tables that have “one-to-many” and “many-to-many” relationships (Figure 1). This eliminates commonly duplicated data, such as lesion location, and minimizes data redundancy, often seen in pa-
tients being tracked with multiple lesions. Each of the tables con-
tains data on a specific aspect of vascular anomaly assessment, diagnosis, or treatment. Data tables include information pertaining to demographics, quality-of-life data, medical therapy, pro-
cedural data, imaging data, histologic and laboratory data, special-
ity unique data, and treatment outcome measures.

Design of the data entry user interface is a critical issue. Ease of use, speed of data entry, and accuracy of entered informa-
tion are equally important. Because there is a diversity of im-
portant data surrounding vascular anomalies, data are entered in fields using a variety of formats. These include real num-
bers (ie, patient weight of 13.2 kg), integers (ie, 1=yes and 2=no), text (ie, pathologic description), pictorial data (ie, cli-
nical or radiographic images), and binary data forms (ie, binary large object) (Figures 2, 3, and 4).

Boolean fields (ie, false/true and no/yes) are not typically used in well-designed databases. Proper database design re-
quires that the default value for all fields in a new record be “N.D.” for not done (Figure 2). The entry of “No” (or a zero in a numeric field) should never be the default because it is a
finding that requires active evaluation and entry by an observer. What if a clinician never evaluated the vascular anomaly completely? Can it really be said that the anomaly did not involve an adjacent area? When searching the database in the future, one may choose to include anomalies for which complete data are not available, but the potential limitations on the data will be known. “No” can be the answer to many of the questions on the form. Data entry design can create logical subgroups of data fields that can be modified en masse, streamlining the entry process and improving the overall quality of the collected data.

Complete descriptions of vascular anomalies can be complex. They are necessary for comparison of serial examinations to evaluate natural progression and response to therapies. In addition to descriptors such as size measurements, appearance, and symptoms, graphical entry can be used to map lesions and their treatments (Figure 3 and Figure 4). Mapping vascular lesions allows a clinician to quickly compare involved areas during serial examinations (Figure 3). Mapping can also be placed in the context of anatomic overlays, which may lead to insights in lesion development or treatment (Figure 3). Mapping the actual area being treated (ie, using a test laser patch at varied energy levels or spot sizes or treating a lesion with differing energy settings) allows for separation evaluation of the treated and untreated areas (Figure 4).

The intent throughout the project was to create an end product that would be a commercial-grade software package for Macintosh and Windows computers. To be acceptable to a wide audience of users, it needed to be compliant with the Health Insurance Portability and Accountability Act (HIPAA) and to have been vetted for this. To achieve the design requirements, the Otobase database kernel was used as the basis for the customized vascular anomalies database. Designed at the University of Washington, Seattle, the Otobase kernel has been used to create databases for a number of different medically related groups. Otobase kernel–based databases have been used to study head and neck cancers, recurrent respiratory papillomatosis, sarcomas, pediatric urologic anomalies, communication disorders, and behavioral problems, among others. The Otobase kernel has been accepted for use by organizations such as the American Head and Neck Society (AHNS), the American Society for Pediatric Otolaryngology (ASPO), and the National Institutes of Health (NIH). Otobase kernel–based databases include a robust set of search and data export tools, which eases the process of sharing data in a HIPAA-compliant manner for multi-institutional studies. In addition, databases created using the Otobase kernel can be used as stand-alone programs or in multiclient server environments.

RESULTS

After the VATF meetings, priorities and data structure for the database were identified. For the database to have clinical and research functionality, the following features were essential: serial data collection by each episode of care, incorporation of demographic data on all patients, ability to share data in a HIPAA-compliant manner, and ability to create clinical treatment trial documentation. Otolaryngologic-specific priorities were focused on areas of controversy and/or ineffective management, such as standardized collection of data on the evaluation and management of vascular anomalies involving the airway and head and neck lymphatic malformations failing standard treatment or causing macroGLOSSIA. The flexibility to add new areas for analysis could also be added to the database. To gain clinically relevant information from this data, it was believed that correlation of imaging and clinical measures of treatment outcomes was necessary, especially for lesions associated with complications (ie, high-output cardiac failure or death). This would be assisted through real-time manual mapping of superficial and deeper (soft tissue and bone) components of vascular lesions, using clinical and lesion-specific radiographic data (ie, tumor volume and measures of blood flow). The ability to maintain data on other factors that currently or potentially affect treatment planning (ie, psychosocial, basic science, and concomitant medical conditions) was thought to be essential.

The beta version of the database, which can be used on Macintosh or Windows systems, was created and released for evaluation. Based on feedback, further design changes will be made and the database released for use by any individual interested in studying these rare lesions.

COMMENT

We report the process by which a relational database for the study of a rare condition, vascular anomalies,
has been created. This database was designed through
the input of multiple health care providers and reflects
current and future vascular anomaly treatment and
assessment patterns. It has capabilities that will refine
and characterize vascular anomalies in a manner that
captures the multiple facets of these complex lesions
and diseases. It will enable multi-institutional research
in vascular anomalies by standardized data collection,
HIPAA-compliant data sharing, and built-in data integ-
ritv checks. This tool is usable at an individual provider
or departmental level. Since our understanding of vas-
cular anomaly physiology is expanding rapidly, a rela-
tional database is essential in allowing comparison of
standardized clinical and demographic data with cellu-
lar and/or genetic data.4,30 In this manner, laboratory-
derived knowledge of vascular anomalies can be more
rapidly translated into clinically relevant information.
Through careful collection of defined data in this da-
base, at multiple clinical sites, a cohesive body of
knowledge will be created that will have significance for
multiple medical and surgical disciplines.

In addition to clinical and basic science data, vas-
cular anomalies are also assessed through sophisticated ra-
diographic and photographic images.31 While these im-
ages accurately describe vascular anomalies, how these
images correlate with vascular anomaly type and treat-
ment outcome is not well understood. This relational da-
tabase has the capacity to store image data and allow map-
ing of lesion size, location, and characteristic, which can
be used to compare treatment type and outcome for a va-
riety of vascular lesions, something missing from exist-
ing studies of vascular anomalies.
Decisions to treat various diseases are based on many variables. Improving or maintaining normal function is always a high priority in vascular anomaly treatment. In the process of accomplishing this, the anomaly can defy curative therapy, and long-term management is necessary. Relational databases have been essential in measuring how treatment affects quality of life. Built into this database are the capabilities to measure caregiver and patient assessments of quality of life.

Any database has limitations. In this case, most of the data in this product will initially need manual input. In the future, with institutional cooperation, automated data entry is possible. In addition, database use will require the user to input data in a standardized fashion, understood through training manuals.

This relational database is available to any interested party and is a powerful tool to further understand vascular anomalies and their treatment, through the comparison of existing clinical, radiographic, photographic, pathologic, and basic science data. Future use of this tool will be instrumental in studying current treatment outcomes and developing evidence-based treatment innovation for vascular anomalies that involve the head and neck. The process of creating this database illustrates a model that could be applied to any rare disease encountered in the field of otorhinolaryngology–head and neck surgery.

Submitted for Publication: May 17, 2007; final revision received September 4, 2007; accepted September 12, 2007.

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Author Contributions: Dr Coltrera had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Study concept and design: Perkins and Coltrera. Acquisition of data: Perkins. Drafting of the manuscript: Perkins and Coltrera. Critical revision of the manuscript for important intellectual content: Perkins and Coltrera. Administrative, technical, and material support: Perkins and Coltrera.

Financial Disclosure: None reported.

Previous Presentation: This article was presented at The American Society of Pediatric Otolaryngology 2007 Annual Meeting; April 28, 2007; San Diego, California.

Additional Contributions: ASPO Vascular Anomalies Task Force Members provided specific data points for the database as well as editorial comments on the database design and development. Prior to development, these individuals provided a conceptual framework for the database to answer specific questions germane to the treatment of head and neck vascular malformations.

REFERENCES