

# Creation of a childhood glaucoma registry database

## Criação de um banco de dados para glaucoma infantil

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**ABSTRACT | Purpose:** The aim is creation of a database to register patients diagnosed with childhood glaucoma. Information collection and storage will utilize a digital platform that permits multiple centers to register patients. This registry will be an important tool to evaluate and group patient profiles, thus allowing identification of risk factors, estimating prevalence, and recruiting participants in future studies. **Methods:** An online form was designed at the Department of Ophthalmology and Visual Sciences of the Federal University of São Paulo to create a registry of patients diagnosed with congenital glaucoma. In addition, a pilot study was conducted in the same institution to validate the Google Forms platform that comprises the registry questionnaire. **Results:** Data can be entered online and stored in the cloud. The database allows the acquisition and storage of epidemiological and clinical data, detection of patterns, and evaluation of the current treatment of children with childhood glaucoma. The data from multiple centers can be combined as collection in online and storage is in the cloud. The form is comprehensive and includes social data, gestational history, family history, clinical data on entering the center, and the treatment received. **Conclusions:** Google Forms is a useful tool for collecting and analyzing statistical data, facilitating the process of registering patients, and analyzing information. Using the online questionnaire, it will be possible to create a multi-center online database that allows identification of risk factors and evaluation of the outcome of interventions and treatment.

**Keywords:** Glaucoma/congenital; Database; Glaucoma/epidemiology; Child

**RESUMO | Objetivo:** Este estudo visa a criação de um programa para a formação de uma base de dados dos pacientes com diagnóstico de glaucoma pediátrico do Departamento de

oftalmologia e ciências visuais da Universidade Federal de São Paulo. O armazenamento das informações se dará por meio de uma plataforma digital, possibilitando que outros centros também possam cadastrar pacientes. Este registro será uma ferramenta importante para avaliar o perfil dos pacientes e agrupá-los, permitindo identificar fatores de risco, estimar prevalência e recrutar indivíduos para futuras pesquisas. **Métodos:** Um formulário on-line foi projetado para criar um registro de pacientes com diagnóstico de glaucoma congênito do Departamento de Oftalmologia e Ciências Visuais da Universidade Federal de São Paulo. Além disso, um estudo piloto foi conduzido para validar a base de dados criada. **Resultados:** Foi criado um questionário usando a plataforma Google Forms, de preenchimento online e armazenamento na nuvem, para registro dos pacientes com diagnóstico de glaucoma infantil do Departamento de Oftalmologia e Ciências Visuais da Universidade Federal de São Paulo. Tal registro permite armazenar dados epidemiológicos e clínicos, detectar padrões e avaliar o tratamento atual de crianças com glaucoma da infância, com a possibilidade de intercâmbio e uso de informações com outros centros, já que a coleta e o armazenamento dos dados se dá de forma online. O formulário é extenso e engloba dados sociais, histórico gestacional, antecedentes familiares, dados clínicos de entrada no nosso centro e o tratamento recebido. **Conclusão:** Este estudo demonstrou que o Google Forms é uma ferramenta útil para coleta e análise de dados estatísticos, facilitando o processo de registro de pacientes e análise de informações. A criação do questionário online e a consequente análise das informações permitirá o maior conhecimento do perfil dos pacientes acompanhados em nosso centro e, com a centralização dos dados num ambiente único - nuvem digital - tornar-se-á possível criar um banco de dados online multicêntrico que permitirá a identificação de fatores de risco e avaliar o resultado das intervenções e tratamentos, assim como recrutar as crianças para pesquisas futuras.

**Descritores:** Glaucoma; Glaucoma congênito; Banco de dados; Epidemiologia; Criança

### INTRODUCTION

Childhood glaucoma is a rare eye disease with changes of the anterior segment and iridocorneal angle that result in increased intraocular pressure (IOP) and optic

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nerve damage<sup>(1,2)</sup>. Epidemiological studies show that the incidence differs regionally, estimated at approximately one case in 10,000 live births<sup>(1,3)</sup>. Because of its rarity, childhood glaucoma is often misdiagnosed or underdiagnosed, resulting in irreversible damage to eyesight from optic nerve involvement, corneal opacities, and amblyopia. Consequently, this rare condition is responsible for more than 18% of childhood blindness worldwide<sup>(4,5)</sup>. In primary pediatric glaucoma, there is isolated anomalous development of the trabecular meshwork and iridocorneal angle, resulting in obstructed flow of the aqueous humor, increased IOP, and optic nerve injury<sup>(6)</sup>. In secondary pediatric glaucoma, the flow of aqueous humor flow is obstructed because of a congenital or acquired ocular alteration or systemic disease<sup>(6)</sup>.

A revised classification has been proposed by the Childhood Glaucoma Research Network (CGRN), an International consortium of glaucoma specialists<sup>(6)</sup> based on clinical findings, timing, and the context in which the glaucoma diagnosis is made. The CGRN includes primary congenital glaucoma (PCG) and juvenile open-angle glaucoma with primary childhood glaucoma. Glaucoma associated with nonacquired ocular anomalies, nonacquired systemic diseases or syndromes, acquired condition, or following cataract surgery is considered as secondary childhood glaucoma<sup>(6,7)</sup>. Because childhood glaucoma is rare, it is difficult sufficient data to pool and analyze. The available epidemiological and therapeutic data are inconsistent<sup>(8)</sup>.

Collecting data acquired at multiple centers in a single patient registry or database is useful as a reference for the study of rare diseases<sup>(3)</sup>, making it easier to study the natural history of the disease, identify risk factors, and evaluate interventions and treatments in patient populations much larger than those available at a single study center. The study aim was to create a database of childhood glaucoma patients that will be usable by multiple treatment and study centers.

## METHODS

An online questionnaire was used to collect epidemiological and clinical data of patients diagnosed with childhood glaucoma at the Department of Ophthalmology and Visual Sciences, Federal University of São Paulo. The patient information was stored in a database designed as a registry to help investigators detect patterns of disease and evaluate current treatment of childhood glaucoma. The objective is to facilitate the exchange

of information with partner centers, functioning as a shared, online database. The registry includes a dataset compiled from children born between 2006 and March 2017 and diagnosed and treated at this center. A second dataset includes patients treated from March 2017 onward. Data are collected during a medical consultation, which allows including children who have already been treated as well as those who will begin follow-up. The registry is managed following the ethical principles of the Declaration of Helsinki. All patients are informed of the nature of the study, its safety, and its benefits before they accept the terms of consent. A copy of the signed consent form is given to the patient, and a copy is filed in the patient's chart. Participation is voluntary, and patients are aware that they can withdraw at any time. To assure confidentiality, patients are identified by a random code that only the investigators can access. All data obtained before a patient's withdrawal are included in the study except in the case of suspension of consent. Previous data in the patient record are monitored at each new consultation.

The study form was created on the Google Forms platform, in which data are entered online and stored in the cloud. Once completed, the form is accessed via an e-mail address, and the answers appear immediately to the respondent who completed the Google Forms page. Visualization of the collected data is a primary advantage of using this platform. The forms are accessed free of charge to the computer without need of registration or a password. The responses are shown as a table that can be exported in various formats including as an Excel spreadsheet. The Google Forms platform was validated in a pilot study that was conducted in the Department of Ophthalmology and Visual Sciences, Federal University of São Paulo.

## RESULTS

The Google Forms include 35 questions that collect demographic data, the patient's gestational history, family history, clinical data from the study center, and the treatment received (Figure 1). Epidemiological data include the patient's name, the center where the patient is being treated, patient registration number, birth date, age at diagnosis, sex, ethnicity, parent consanguinity, father's and mother's education, professions and incomes, history of glaucoma in the father's and mother's families, patient's gestational history. The ophthalmological evaluation data are also recorded on the Google

### Registro Glaucoma pediátrico

**Nome**  
Sua resposta \_\_\_\_\_

**Centro**  
Escolher \_\_\_\_\_

**RH**  
Sua resposta \_\_\_\_\_

**Data de Nascimento**  
Data  
dd/mm/aaaa \_\_\_\_\_

**Idade ao diagnóstico (meses)**  
Sua resposta \_\_\_\_\_

**Sexo**  
 feminino  
 masculino

**Etnia**  
 branco  
 negro  
 pardo  
 amarelo

**Cosanguinidade pais**  
 Sim  
 Não

**Escolaridade mãe**  
 analfabeta  
 1 grau incompleto  
 1 grau completo  
 2 grau incompleto  
 2 grau completo  
 superior incompleto  
 superior completo

**Escolaridade pai**  
 analfabeta  
 1 grau incompleto  
 1 grau completo  
 2 grau incompleto  
 2 grau completo  
 superior incompleto  
 superior completo

**Renda familiar média(salário mínimo/per capita)**  
 sem rendimento  
 até 1/2  
 mais de 1/2 até  
 mais de 1 até 2  
 mais de 2 até 3  
 mais de 3 a 5  
 mais de 5

**número de irmãos (mesmo pai e mãe)**  
Sua resposta \_\_\_\_\_

**Profissão mãe**  
Sua resposta \_\_\_\_\_

**Profissão pai**  
Sua resposta \_\_\_\_\_

**História familiar glaucoma na infância**  
 Ausente  
 Presente

**História familiar glaucoma do adulto**  
 Ausente  
 Presente

**História gestacional**  
 infecção  
 uso de drogas  
 tabagismo  
 ingestão de álcool  
 outro  
 Outro: \_\_\_\_\_

**História gestacional**  
 parto normal  
 cesariana

**História gestacional**  
 a termo  
 prematuro  
 pós-datismo

**Olho acometido**  
 OD  
 OE  
 AMBOS

**Diagnóstico**  
 Glaucoma congênito primário  
 Glaucoma secundário a cirurgia de catarata  
 Glaucoma secundário associado a condições adquiridas  
 Glaucoma secundário associado a condições oculares não adquiridas  
 Glaucoma secundário associado a condições sistêmicas não adquiridas  
 Glaucoma Juvenil de ângulo aberto  
 Outro: \_\_\_\_\_

**Achados biomicroscopia (ao diagnóstico)**  
 estrias de haab  
 edema de córnea  
 megalocórnea  
 alterações irianas  
 embriotoxon posterior  
 catarata  
 Outro: \_\_\_\_\_

**PIO mmHg ( diagnóstico)**  
Sua resposta \_\_\_\_\_

**Relação escavação/disco ( diagnóstico)**  
Sua resposta \_\_\_\_\_

**Perda localizada debrum ( diagnóstico)**  
 sim  
 não

**Defeito camada fibra nervosa ( diagnóstico)**  
 Sim  
 Não

**Diâmetro corneano (mm - diagnóstico)**  
Sua resposta \_\_\_\_\_

**Gonioscopia ( diagnóstico)**  
 ligamentos pectíneos  
 displasia trabecular  
 inserção alta da íris  
 sinéquias

**Comprimento axial( diagnóstico)**  
Sua resposta \_\_\_\_\_

**Quantidade de cirurgias na entrada**  
Sua resposta \_\_\_\_\_

**Cirurgias**

	od	oe	ao
Lensectomia	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Vitreotomia	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Trec	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Trabeculectomia	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Goniotomia	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Implante de tubo	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Transplante de córnea	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

**Uso de colírios (diagnóstico)**  
 Nenhum  
 Latanoprost  
 C. dorzolamida 2,0%  
 T. Brimonidina 0,2%  
 M. Timolol 0,5%  
 Travoprost  
 Bimatoprost

**Intervenção cirúrgica - Glaucoma**

	OD	OE	AO
TREC	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
TRO	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
GONIOTOMIA	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
IMPLANTE DE TUBO	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

**PIO - Após 1 ano - mmHg**  
Sua resposta \_\_\_\_\_

Option 1

**Uso de colírios (após 1 ano)**  
 Nenhum  
 Latanoprost  
 C. dorzolamida 2,0%  
 T. Brimonidina 0,2%  
 M. Timolol 0,5%  
 Travoprost  
 Bimatoprost

**ENVIAR**

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Google Formulários

Figure 1. Google forms pediatric glaucoma registry questionnaire.

Forms questionnaire. Although the patients had a pre-specified diagnosis in their charts, the data collected by the questionnaire forms were used to reclassify the diagnosis according to the CGRN childhood glaucoma classification<sup>(6)</sup>.

A pilot study was conducted to assess the feasibility of using the Google Forms platform. It consisted of an active search of patient electronic health records to identify all patients born between 2006 and 2016 and diagnosed with childhood glaucoma who were registered in the service. Subsequent patients were registered and included in the Google platform when they attended routine evaluations and provided informed consent. The previous diagnosis was confirmed and reclassified using the updated CGRN criteria<sup>(6)</sup>.

The records were of 72 children with childhood glaucoma, 42 (61.5%) with PCG, and 28 (38.5%) with secondary congenital glaucoma (SCG). Most SCG was secondary to congenital cataracts (61.0%), followed by iridocorneal dysgenesis (20.0%) and Sturge-Weber syndrome (12.0%). Following the CGRN classification<sup>(6)</sup>, 56.5% of the patients had PCG. Glaucoma was associated with nonacquired systemic disease in 5.0%, related to cataract surgery in 30.5%, associated with nonacquired ocular anomalies in 4%, and associated with acquired conditions in 4%. Most patients (80.0%) presented at the service with an advanced stage of disease with cloudy cornea, axial length above the 95th percentile, and a vertical cup-to-disc ratio of  $>0.8$ . No problems were found using the Google Forms platform; all data were successfully collected securely, and privacy was assured by creation of an ID code that only the investigators could access.

## DISCUSSION

Access to clinical records allows for identification of patients eligible for retrospective and prospective studies and patients eligible for trials and facilitates research, evaluation, and planning of health services. The patient data can be used to document the real-world effectiveness of generating hypotheses about risk factors for adverse outcomes and provide an overview of the natural history of a disease. Registries also allow evaluation of treatment benefits in patient subgroups, particularly in resource-poor environments that are not well represented in randomized clinical studies<sup>(9)</sup>. Prospective observational records that monitor natural disease evolution, treatments, and clinical outcomes are important

in understanding such diseases. As data collection is difficult in studies of rare diseases, multicenter involvement of centers that specialize in treatment is helpful<sup>(10)</sup>.

Standard treatment protocols, disease markers, and follow-up evaluations are often not available for rare diseases. Registries of heterogeneous treatments and the increased possibility of following up with patients may facilitate evaluation and add to the understanding of the disease. To the best of our knowledge, this is the first online form to collect childhood glaucoma data. The need for these data prompted the creation of a childhood glaucoma registry for São Paulo state that initially included patients from the Department of Ophthalmology and Visual Sciences at the Federal University of São Paulo and later extended to other referral centers in the state. This registry includes socio-demographic characteristics, such as ethnicity, family income, family size, and parents' education. These data are necessary to determine whether disease prevalence is greater in a population segment and whether these factors limit patient monitoring and prognosis. The questionnaire also collects family histories of adult and childhood glaucoma, the patient's gestational history including pregnancy complications, type of delivery, and gestational age. These data may help to identify factors that increased the risk of childhood glaucoma in these patients. The second part of the questionnaire collects clinical characteristics at diagnosis. The diagnostic criteria of childhood glaucoma according to the CGRN classification are recorded and include IOP, axial length, corneal diameter, findings on biomicroscopy (e.g., Haab's striae), optic nerve signs (e.g., cupping size, scarring, retinal nerve fiber layer), and findings on gonioscopy. Recording these factors is important to determine which signs are the most prevalent and to characterize the correct diagnosis according to the Consensus 9 classification of the World Glaucoma Association<sup>(6)</sup>. Finally, the registry also includes information on any treatment received before referral to the center, the pathway of the referral service, and the 1-year follow-up including IOP and the use of eye drops.

The registry was created using Google Forms, which are accessible from any location and at any time. Other advantages include economy of hard-disk space; zero cost; ease of use; and a user-friendly interface with no programming knowledge required. Data collection can be easily implemented by making the electronic form available using an e-mail address. When completed by respondents, the answers appear immediately on the Google Forms page of the form's creator. The form even displays statistical tools to analyze the data collected.

The limitations of the registry include collecting information during an interview with the patient's parents and by a review of the medical records. The answers are primarily parental self-reports and not directly obtained from the patient. Some patients or family members may forget or not remember important information from the past. Finally, the questionnaire depends on accurate reporting by other doctors who have treated the patients.

This online questionnaire and the analysis of the information that it collects will allow a better understanding of the profiles of patients followed in our center. Centralizing the data in a single environment-the digital cloud-it will create a multicenter online database to assist in identifying risk factors and evaluating the outcomes of interventions and treatments. This pilot study verified that Google Forms was a useful tool for collection and statistical analysis of patient and family data, and it facilitated registration.

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