The Swedish National Paediatric Cataract Register (PECARE)

Annual Report 2021

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Swedish paediatric cataract surgery

2021 Annual Report, based on data from the Swedish National Paediatric Cataract Register (PECARE).

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Background

The Swedish Paediatric Cataract Register (PECARE) is a subdivision of the adult cataract register. Data reporting began in autumn 2006. PECARE is a web-based surgical register that covers children aged 0-8. Around 40 children are born with cataracts in Sweden each year. Additionally, surgery resulting from trauma, uveitis (inflammation of the iris) and lens subluxation are also registered. Globally, paediatric cataracts are among the most common curable causes of blindness in children. Incidence is lower in Sweden as a result of our national vaccination programme, as infection during pregnancy is a common cause of cataracts.

Congenital cataracts are associated with higher degrees and a differing complex of problems than the age-related form. In adults, the goal is to restore previous visual function, while in children, the aim of treatment is to achieve visual function. The development of visual pathways is highly dependent on early visual stimuli. The treatment of dense congenital cataracts involves early surgery, i.e., during the first months of life, and requires early detection in the maternity ward or paediatric health centre. Surgery may indeed be necessary as early as 4-8 weeks of age in the most severe cases. If left untreated or detected too late, the disease can lead to severe visual impairment or blindness. Infants have a significantly higher tendency to have surgical complications that require frequent healthcare visits over many years. One explanation for this is a more aggressive healing process due to growth factors in infants' eyes. Complications may result in permanent damage, affecting both their choice of profession and quality of life, and can emerge later during any time in the child's life.

The most common complications are visual axis opacification (VAO) and secondary glaucoma. In studies, glaucoma has been reported to occur in up to 30% of all surgical cases, and VAO occurs in even higher rates. These complications effectively inhibit normal visual development. In the worst cases, one disease is replaced with another lifelong disease.

Childhood cataracts are a curable condition, provided that they are detected and treated early. In the case of dense congenital cataracts, the earlier surgery is performed, the greater the chance for normal visual development, but paradoxically, this also carries a greater risk of surgical complications. This is a dilemma. The exact timing for surgery and the best possible surgical method are therefore constant topics of discussion, especially considering secondary glaucoma, which in severe cases leads to enucleation (surgical removal of the eye). Conflicting explanatory models and results can be found in scientific literature. One of the reasons is likely that the cohorts are too small and the groups of children are heterogeneous.

The US and UK have national regulations for eye screening of infants. The current applicable recommendations for cataract screening in Sweden are described in the National Handbook for Child Health Services, but are suggested guidelines rather than absolute rules. This guidance has been interpreted in different ways in Sweden: in the vast majority of regions, screening is carried out in maternity wards; it is only in exceptional cases that no routine screenings are conducted at all. There is need for a joint position as to whether eye examinations in maternity wards should be included as a mandatory part of the routine exams of newborns, and how to create the resources for these exams.

Unilateral cataracts generally have worse visual outcomes than bilateral cataracts, and the percentage who become visually impaired in that eye despite treatment is not insignificant. For children with bilateral cataracts, the prognosis is better for visual aquity. During childhood visual development, the brain prioritises the healthiest eye and “disconnects from” the unhealthy one. A patch is then applied over the stronger eye in order to achieve balance and visual development of the weaker eye. This applies to both bilateral and unilateral cases. Patching treatment is handled by parents and is crucial to resulting vision, but it is highly demanding. Some children become completely passive and apathetic, while others become frustrated and angry. It takes strongly motivated parents with an understanding of the maturation and development of the visual system during childhood to achieve good results for vision. Engaging with parents, communication, and information are crucial to treatment outcomes.

Purpose

The register aims to optimise screening strategies and the efficacy of paediatric cataract treatment. The goal is to be a comprehensive register for all paediatric cataract surgery in Sweden, to constitute a national foundation for quality assurance and to define and analyse unexpected treatment outcomes.

Development of the register

All eye clinics in Sweden that perform surgery on infants have submitted data since autumn 2006. In 2007, a clinic in Denmark began submitting data, and in 2011, all

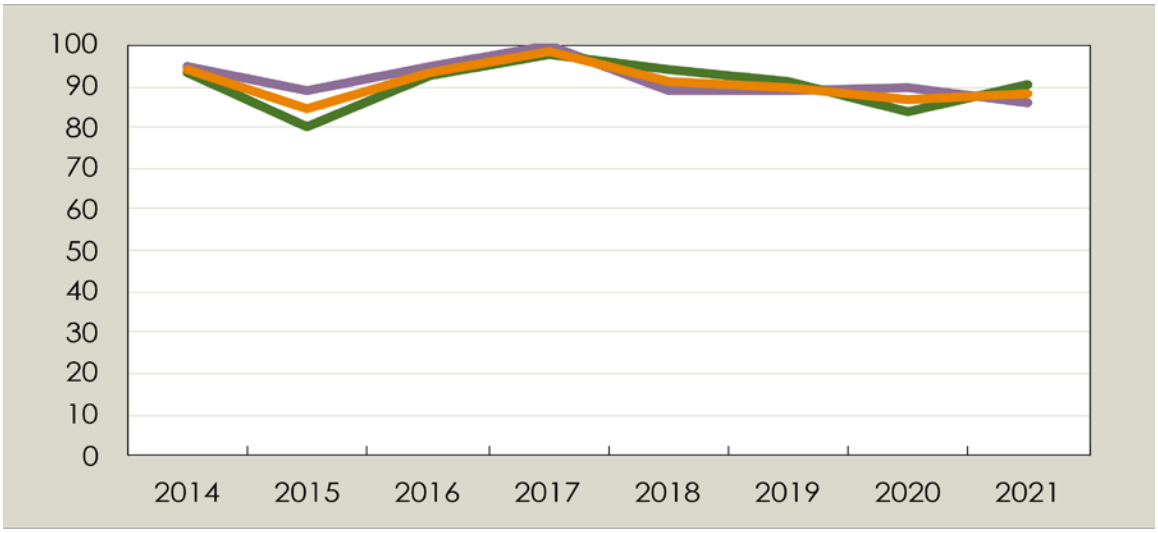
surgical clinics in Denmark decided to participate. From 2016 onwards, registration of Danish data was paused in conjunction with the negotiation of new contracts. Denmark then chose to start their own register. Efforts to improve report retrieval have progressed, as have corrections of follow-up questionnaires and help files with respect to selection and correction variables. The website is in use and is continuously being updated and developed. On the website, patients can receive easily accessible information and personal contact with a patient representative. The steering committee is interprofessional, consisting of ophthalmic nurses, paediatric ophthalmologists, ophthalmic surgeons and patient representatives.

In 2021, PECARE also collaborated with the European paediatric cataract register EuReCCA (European Registry for Childhood Cataract Surgery), which recently became fully operational.

Coverage rate

The paediatric cataract register rate of coverage has been high over the years, as can be seen in the figure below representing the years 2014-2021. The rate of coverage was 88% in 2021. Comparisons are made with the patient administrative register (PAR). As can be seen in the figure, both genders are represented to essentially the same degree.

Coverage rate (%), overview for PECARE



Source: PECARE and the patient administrative register (PAR).

Surgery registration

Data pertaining to the detection process (referring physician, date of diagnosis, clinical findings) and surgical parameters (cataract type, surgical technique, lens type, planned refraction, axial length, corneal dimensions, prior treatment, postoperative treatment) are registered at each surgery, as well as information on heredity and visual acuity, and any systemic disorders or eye anomalies.

Follow-up registration

Treatment outcomes regarding visual development and occurrence of complications are registered via follow-up questionnaires at specific intervals: at the ages of 1, 2, 5 and 10. These check-ups are conducted by 30 paediatric ophthalmology clinics around Sweden who have the designated contact persons for the register. Data are collected regarding strabismus, nystagmus, visual acuity, glaucoma and VAO, including number and type of treatments, as well as types of correction.

Reporting data

Data are reported via the Swedish National Cataract Register website (http://www.kataraktreg.se) using a specific questionnaire. Care is centralised to a small number of clinics, and in Sweden, Stockholm and Gothenburg hold licences for providing what was previously known as *National Medical Care*, and is now and National Specialized Medical Care of children with paediatric cataract under the age of three years. The remaining reporting clinics are Umeå, Malmö-Lund, Linköping, Örebro and Eskilstuna.

Retrieving data

All clinics may take out a report on data, both at the individual level and the eye level, for their own clinic at any time.

Results

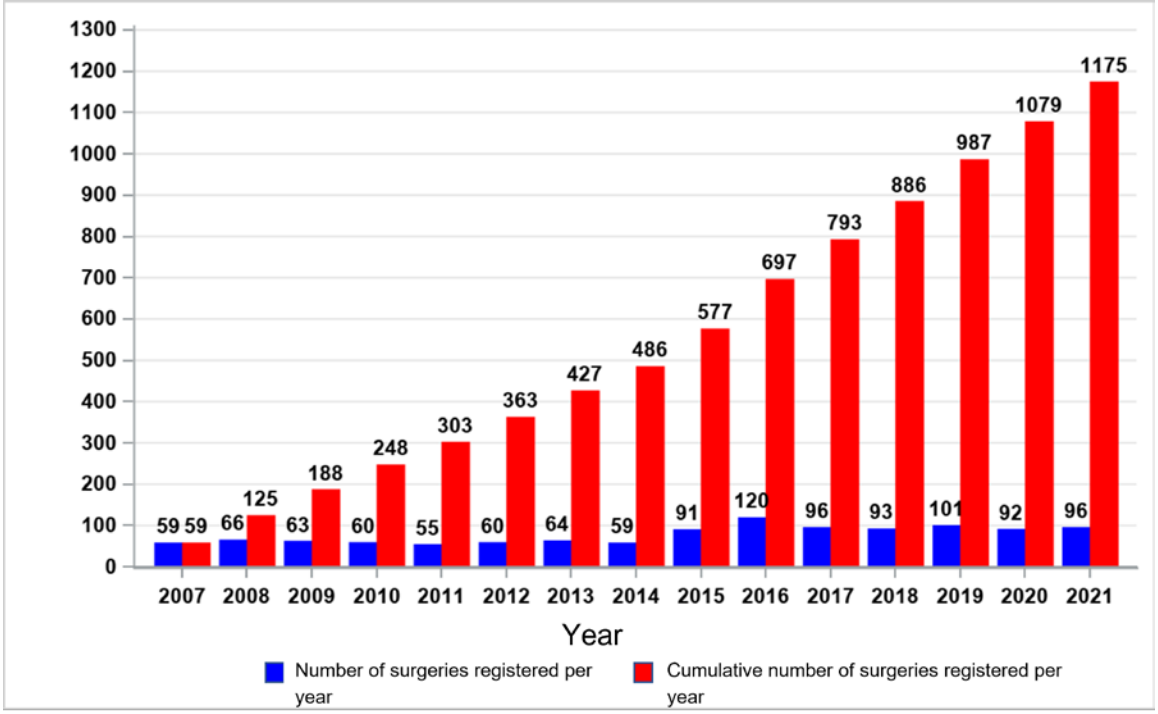


Figure 1a. Number of registered surgeries in PECARE per year and accumulated in 2007-2021.

Information	Number
Operated patients in PECARE	846
Operated eyes in PECARE	1175
Deceased patients in PECARE	13
Unavailable patients	37
Number of individual girls	399
Number of individual boys	447

Figure 1b. Number of registered children and eye surgeries, number of deceased children, as well as total number of children who were unavailable for follow-up during 2007-2021.

Data for a total of 1,175 eye surgeries were registered as of 31 December 2021. Accumulated number of registered surgeries per year can be seen in Figure 1a and 1b.

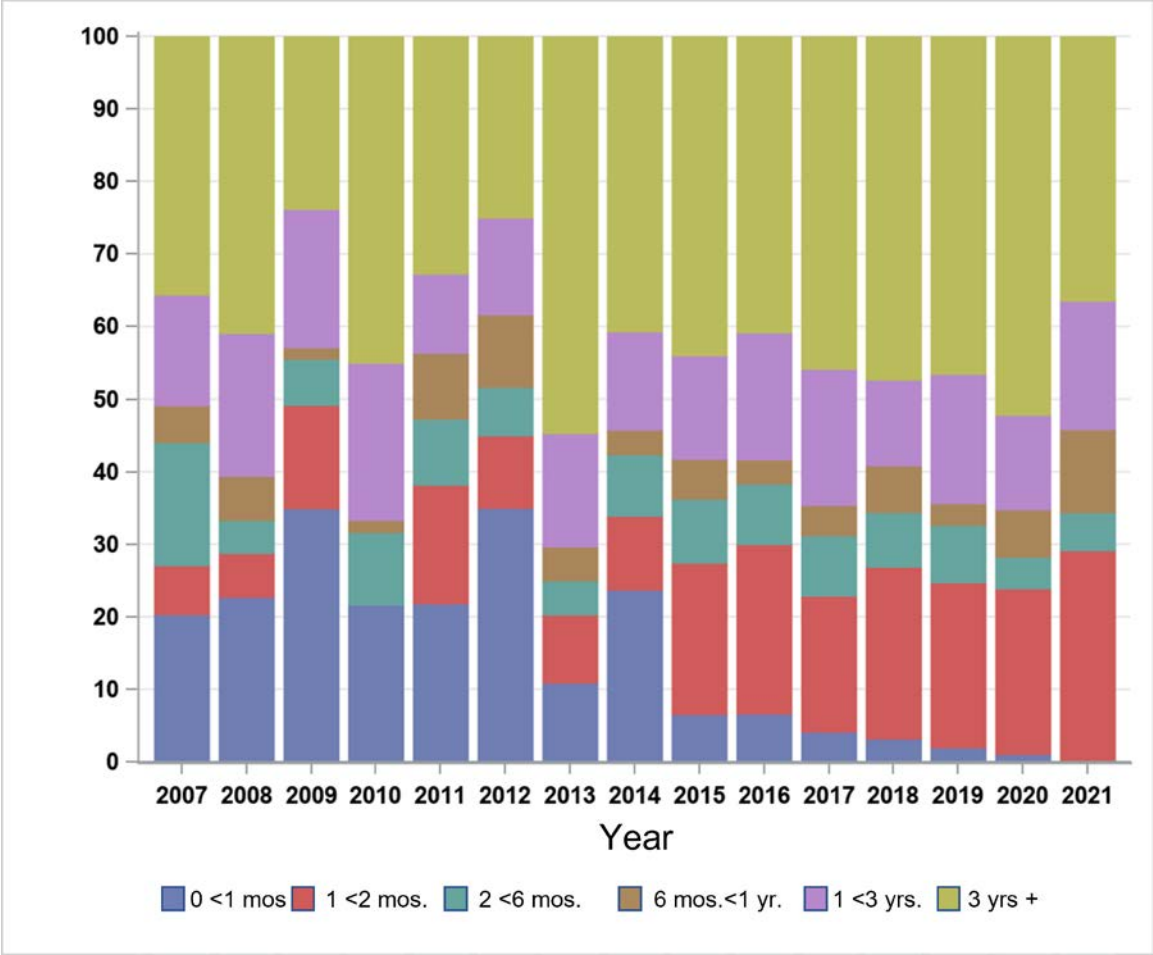


Figure 2. Age distribution at time of surgery in Sweden expressed in percentage and divided by year, 2007-2021.

Age at the time of surgery for the entire register in Sweden, 2007-2021, is shown by Figure 2. In 2021, barely any surgeries were performed during first month of life (dark blue) compared with previous years, whereas a larger proportion were operated on at 1-2 months of age (red). This is a conscious strategy, as analysis of surgical complications in the register revealed that surgery in the first four weeks of life increases the risk of complications.

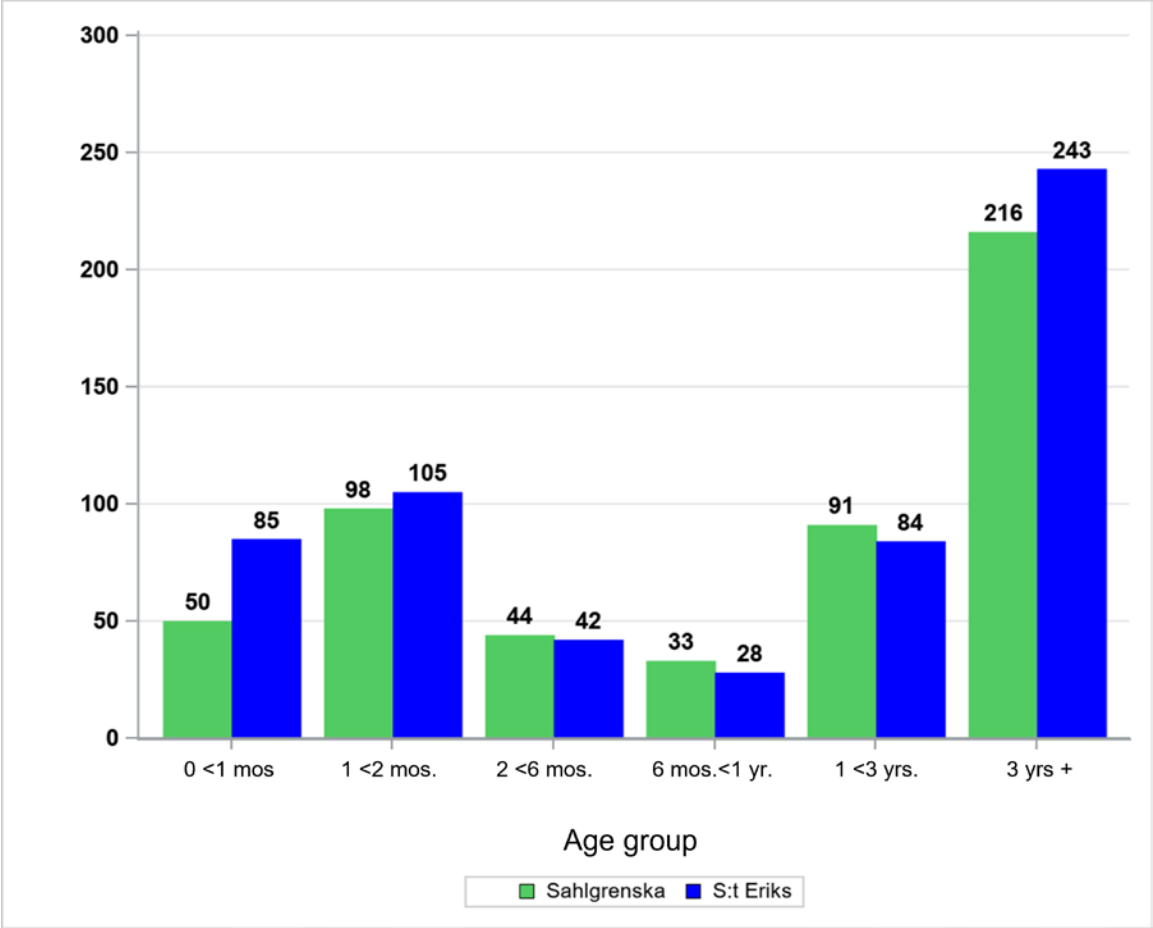


Figure 3. Number of surgeries by age group at St. Erik Eye Hospital, Stockholm, and Sahlgrenska University Hospital, Gothenburg, in 2007-2021.

Figure 3. Total number of eyes operated on in 2007-2021 conducted at Sweden's two largest eye clinics. Surgeries on patients younger than the age of 3 are only performed at two centres in Sweden, as they fall under National Specialised Medical Care and the youngest children are the most severely ill. These centres are located in Stockholm and Gothenburg. This figure shows a comparison of ages at the time of surgery between St. Erik Eye Hospital, Stockholm (blue column) and Sahlgrenska University Hospital, Gothenburg (green column).

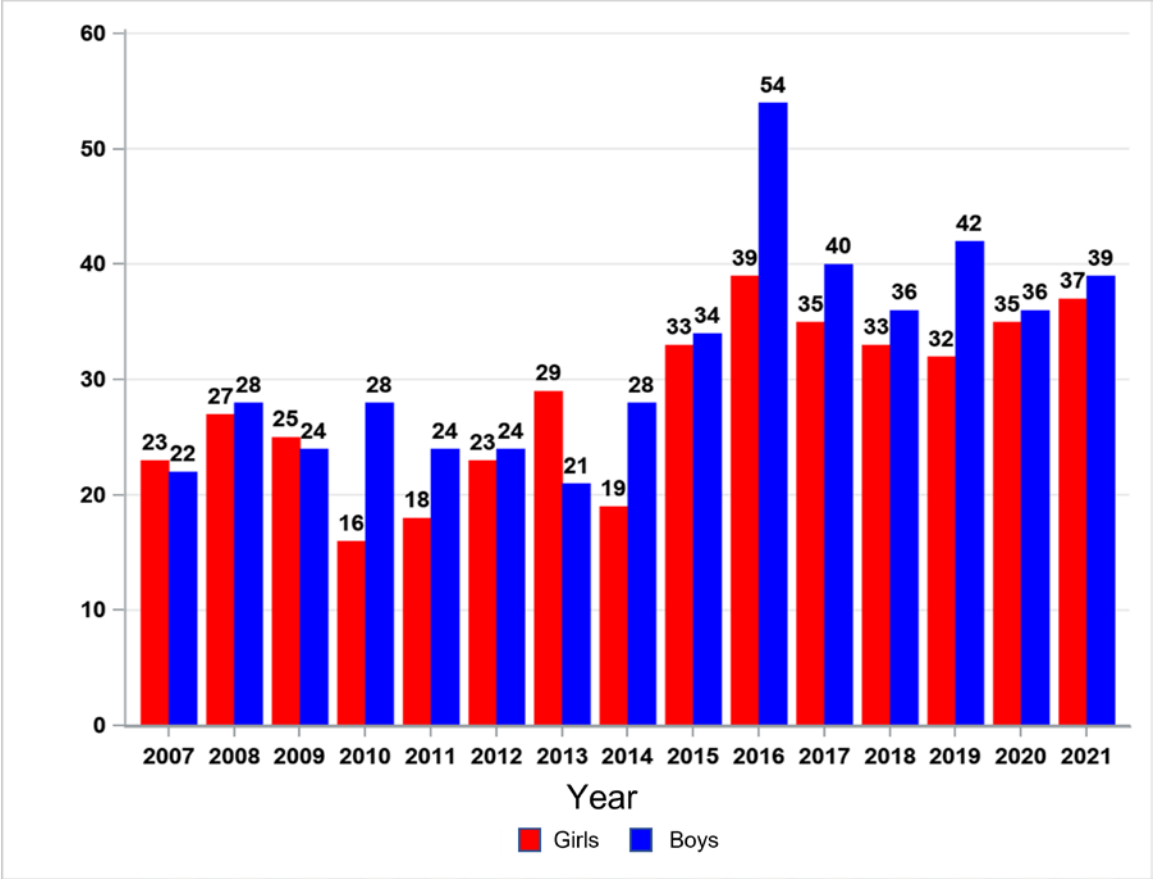


Figure 4. Distribution by gender per year, 2007-2021.

Figure 4 shows the distribution by gender of Swedish paediatric patients over time. In total, there were 401 girls and 458 boys in the register at the end of 2021, corresponding to 859 children.

Incidence of VAO and secondary glaucoma are shown in Figure 5 and 6, respectively.

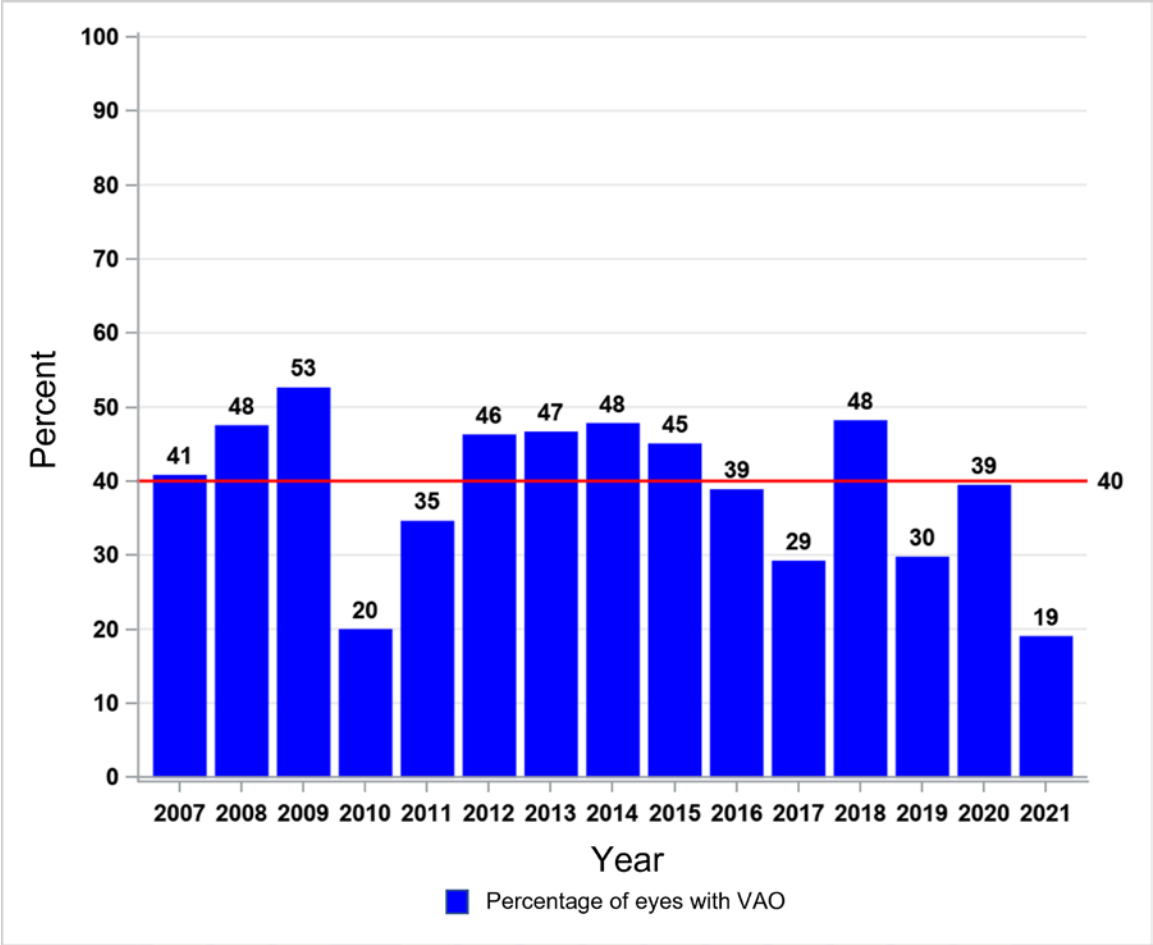


Figure 5. Percentage of eyes with VAO (visual axis opacification) occurrence at some point during follow-up reporting, divided by year of surgery 2007-2021. The reference line indicates the median incidence of VAO for all years.

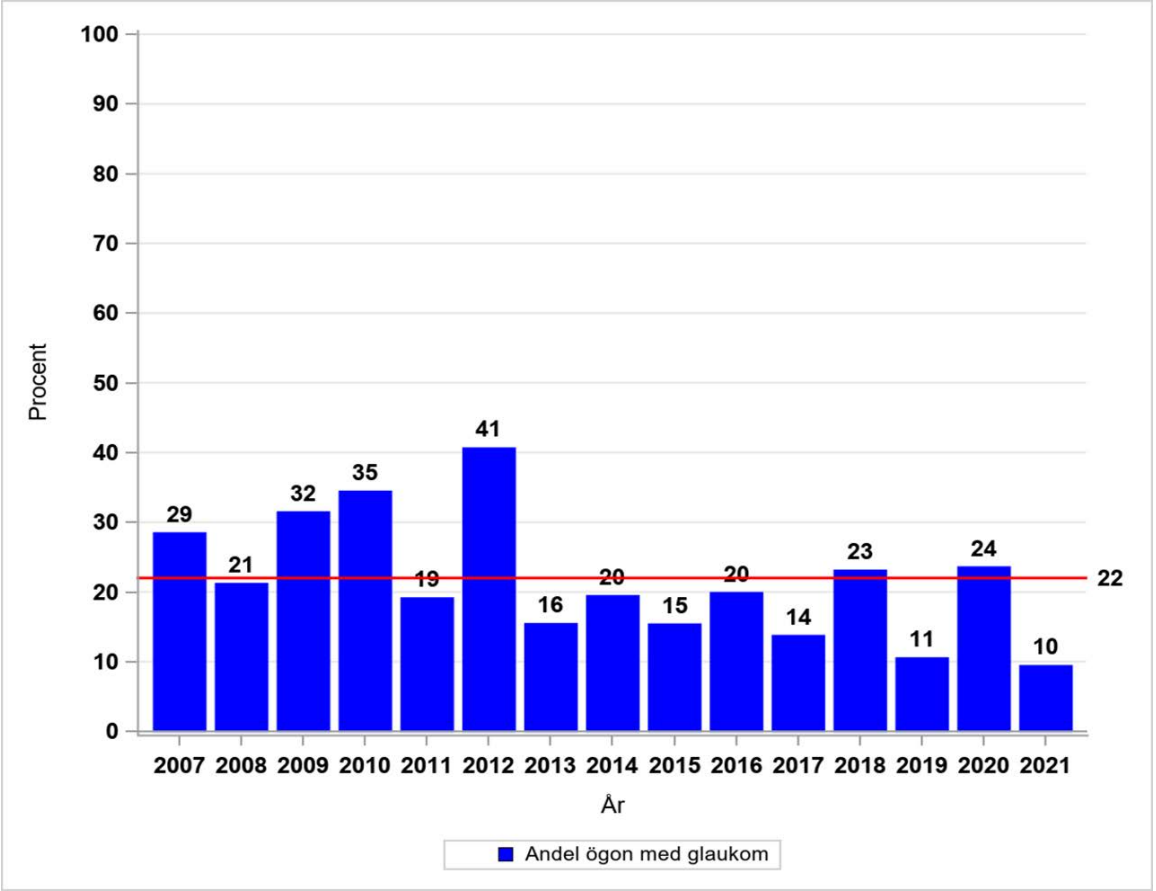


Figure 6. Percentage of eyes with postoperative secondary glaucoma in Sweden 2007-2021, where indicated years refer to year of surgery. The data are based solely on follow-up reporting. The reference line indicates the median incidence of eyes with glaucoma for all years.

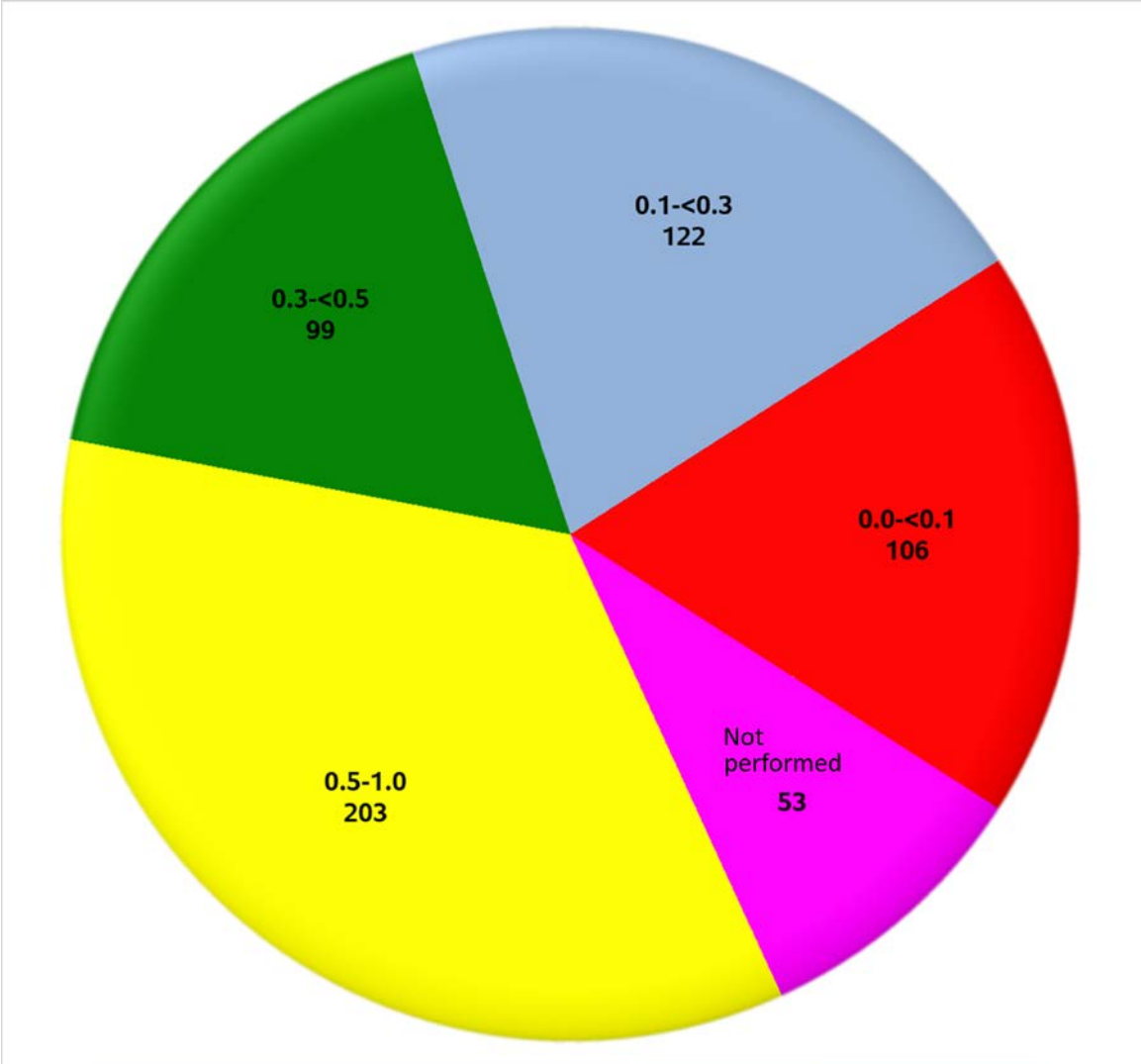


Figure 7a. Postoperative decimal visual acuity per eye at 5 years of age following both unilateral and bilateral cataract surgery in 2007-2021, n= 583.

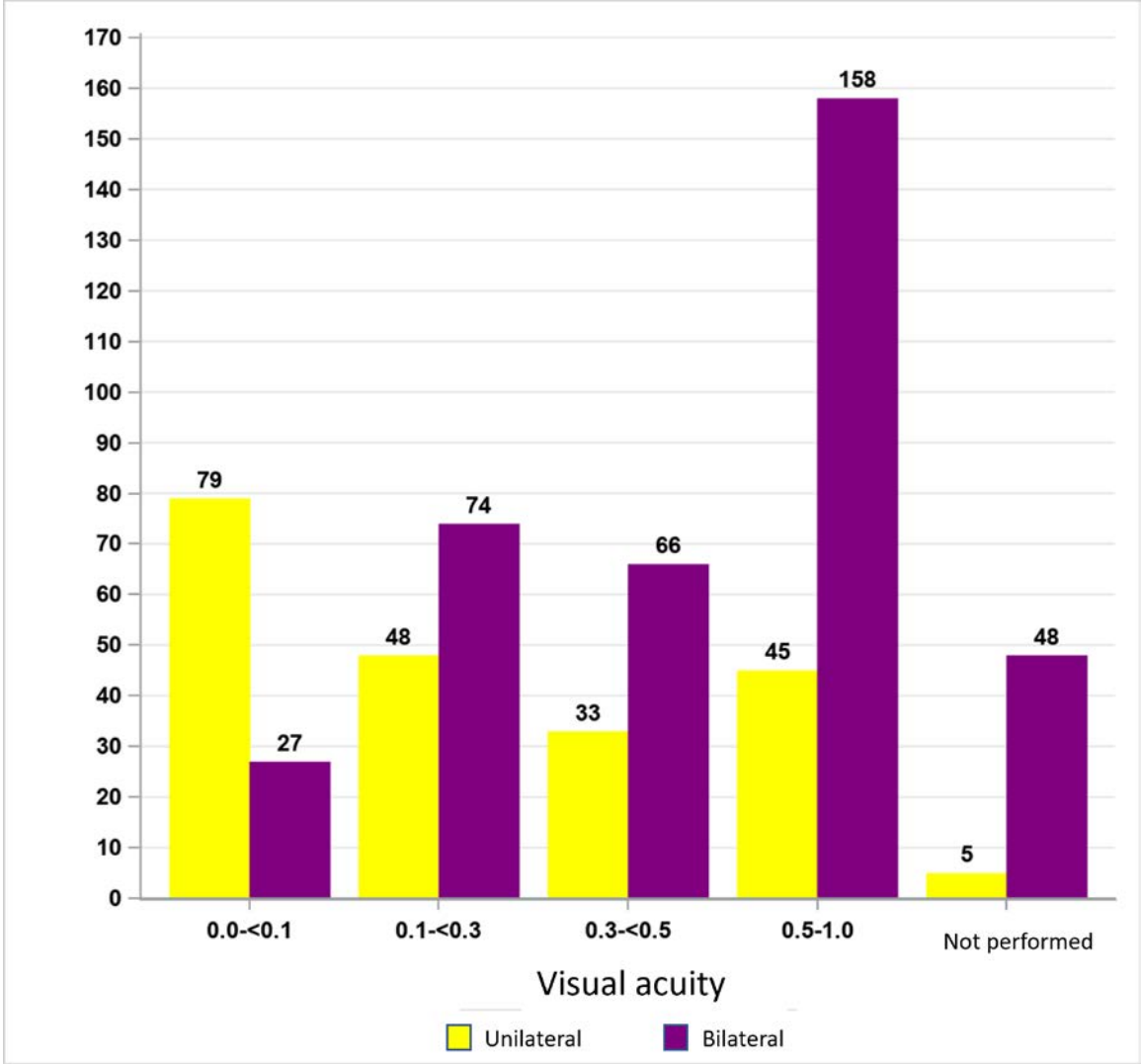


Figure 7b. Postoperative decimal visual acuity per eye at 5 years of age categorised by unilateral and bilateral cataract surgery performed during 2007-2021, n= 583.

The visual outcomes in figures 7a and 8a show that visual acuity increases significantly between the ages of 5 and 10. Approximately 50% of eyes attain visual acuity of 0.5 or higher at the age of 10, which is equivalent to the necessary level of visual acuity to be permitted to drive a car.

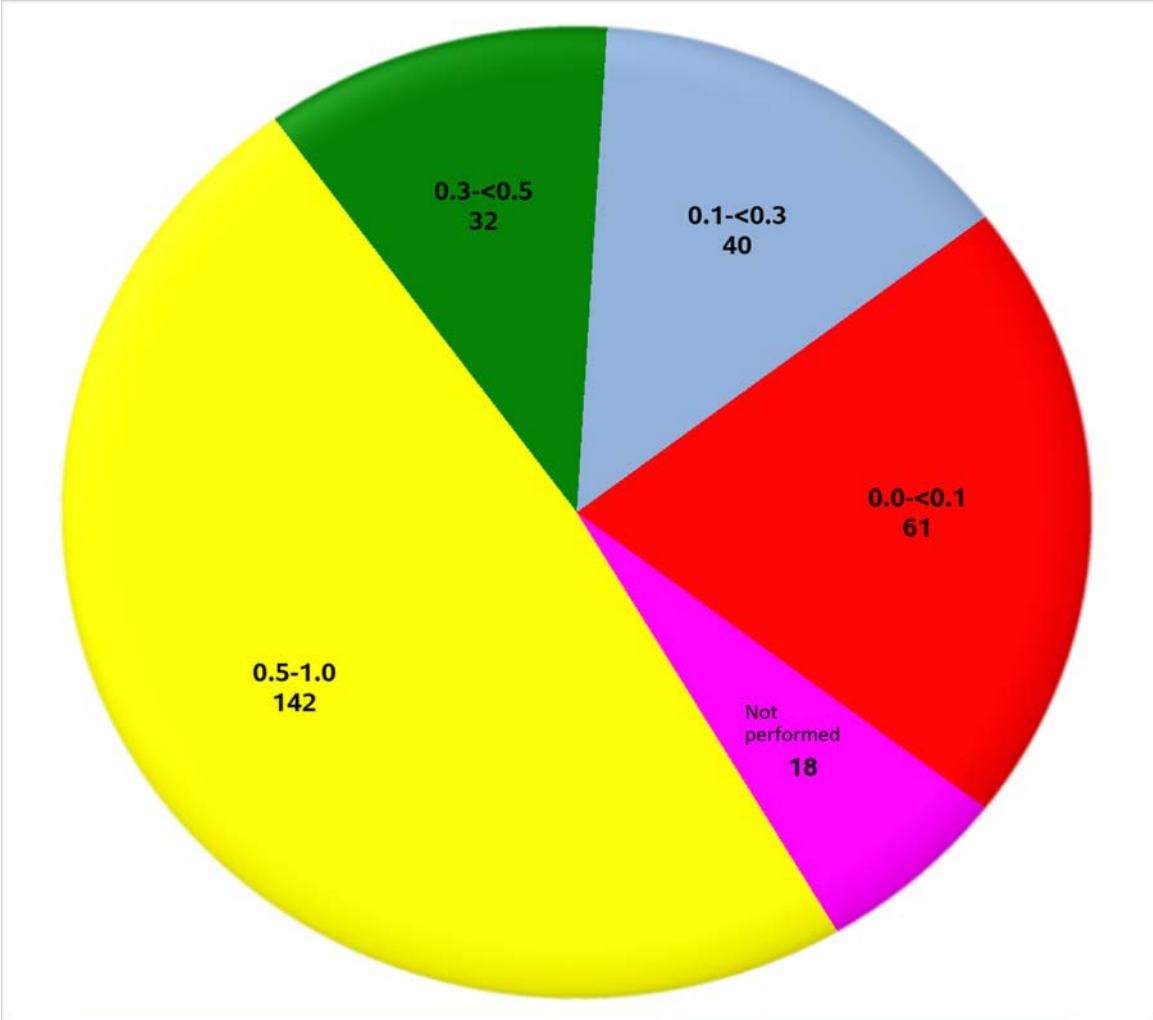


Figure 8a. Postoperative decimal visual acuity per eye at 10 years of age following both unilateral and bilateral cataract surgery during 2007-2021, n=293.

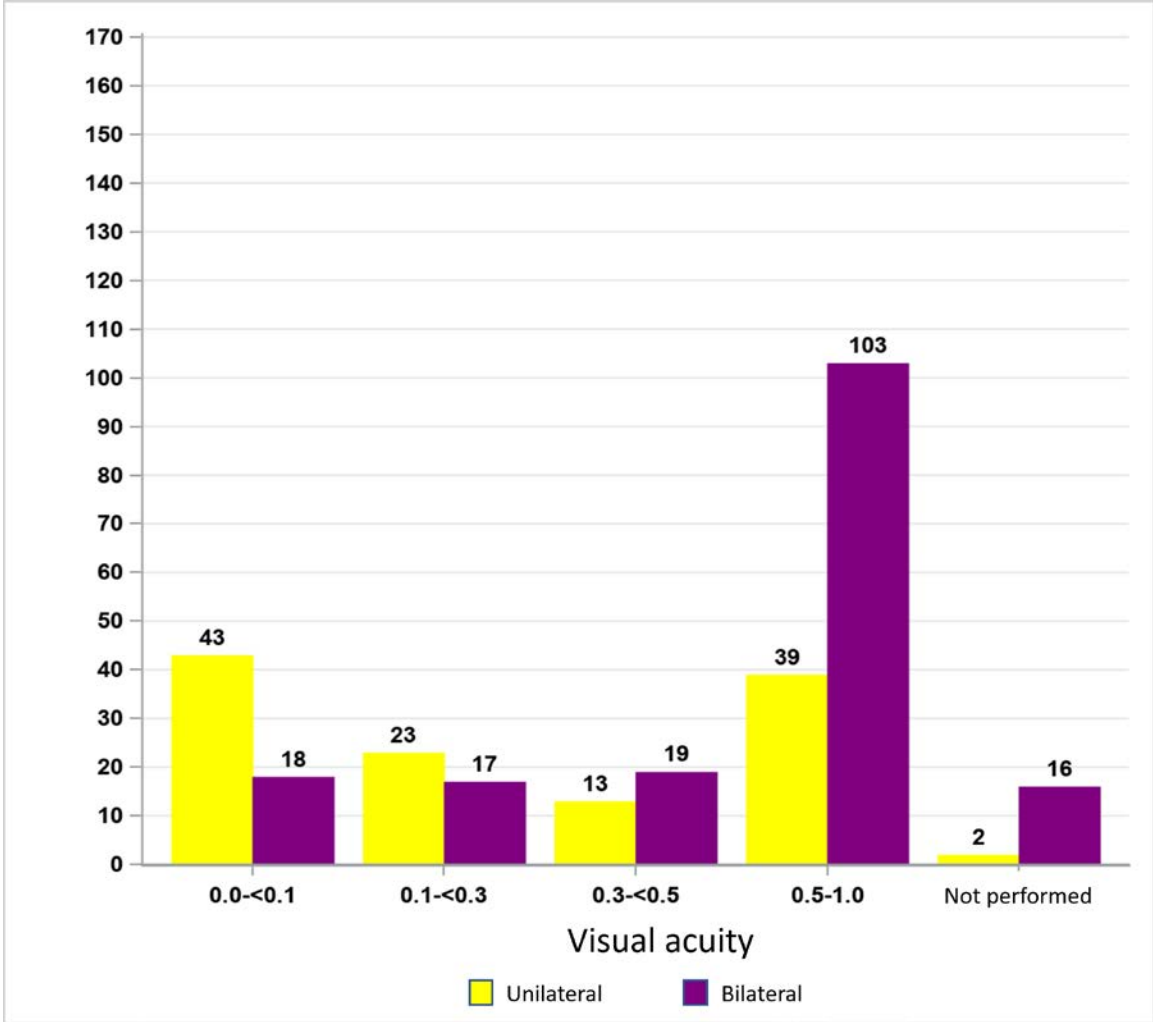


Figure 8b. Postoperative decimal visual acuity per eye at 10 years of age categorised by unilateral and bilateral cataract surgery performed during 2007-2021, n= 293.

Table 1 shows the proportion of registered quality indicators given in percentages for the years 2014-2021.

Variable	2014	2015	2016	2017	2018	2019	2020	2021
Surgical method	**100**	**100**	**100**	**100**	**100**	**100**	**100**	**100**
Initiator of referral to ophthalmologist	**100**	**100**	**100**	**100**	**100**	**100**	**100**	**100**
Time interval between detection and diagnosis	**100**	**100**	**100**	**100**	**100**	**100**	**100**	**100**
Other eye anomalies	**100**	**100**	**100**	**100**	**100**	**100**	**100**	**100**
Other systemic disorders	**100**	**100**	**100**	**100**	**100**	**100**	**100**	**100**

Table 1. Proportion of registered quality indicators for PECARE given in % for the years 2014-2021.

IMPACT OF THE REGISTER'S CONTRIBUTIONS IN HEALTHCARE

1. Early detection and treatment

The diagnosis and treatment of cataracts early in a child's life is crucial to visual development. Sweden is unique in the world as we have established an efficient eye screening in maternity wards, and through the design of the register we have been able to ensure since early 2006 that this screening continues to function well, which it has done. Early detection is the basis for early treatment. The register contains data on children's eyes that were operated on very early, and there is no appreciable difference between the country's two highly specialised centres, as can be seen in Figure 3.

2. Strategies for reducing complication rates

The difficulty lies in that early surgery results in better vision, but also a higher rate of postoperative complications. Complications inhibit visual development.

Incidence of secondary glaucoma has been reviewed and, after scientific analysis and discussion within the framework of the register, children are now no longer operated on before the age of four weeks, with the common policy being to delay the operation in Sweden until the age of five to six weeks for the most serious cases, which is shown in Figure 2.

With regard to secondary VAO, the register has enabled it to be established that there is a dramatic reduction of occurrence when using the new type of intraocular lens known as the bag-in-the-lens implant, which is technically more difficult to insert but reduces the rate of VAO to approximately 6%. This surgical technique is now used throughout the country and the results are promising.

3. Optimised communication with patients and families

The Swedish Patient Safety Act emphasises the importance of strengthening the position of the patient through accessibility, participation and information from the healthcare team. Collaborative efforts in the register have enabled the mapping of the well-being and needs of parents across the country. Fundamentally, there is a great need for close, informative communication. A mobile app has been developed to be a practical tool and it is currently in use. The advantage of this app in comparison to our usual channels for accessing healthcare (phone/email/1177.se) is that it allows parents to contact healthcare providers around the clock, send pictures of their child for quick digital medical assessments without delays and that the design is easily adaptable in response to changes in the target group's needs over time. The mobile phone as a means of communication is here to stay. In our experience, parents feel more secure as they always have their mobile phone with them.

The register website is important for patients and their practical care. The steering committee's patient representative, has a separate tab on the website where parents have access to various topics, such as being able to read each other's stories, look into tips and tricks for how to go about with contact lenses, templates with information for day-care centre staff about conditions, and many other useful elements. Kim connects with parents in a Facebook group called "Barn med medfödd grå starr" [Children with congenital cataracts].

4. Equality of care

When comparing care in Stockholm and Gothenburg, there are no differences regarding access to care, competence of care, age at surgery or gender.

Clinical improvement initiatives

Assessment of screening effectiveness

Knowledge concerning the effectiveness of eye screening is generally limited. When recommending a screening routine for all infants in Sweden, it is sensible to follow-up on and assess outcomes. With this in mind, parameters were constructed when the register was created to enable answering questions about who initiated a referral during an examination, how the condition was detected in a child and which symptoms prompted the referral. This data was analysed in 2011 and published in an article in *Läkartidningen*, a Swedish medical journal, in 2012 as well as in *Acta Paediatrica*. The coverage rate of neonatal eye screening is 90% in Sweden; the majority of all children who need early surgery are first detected on the maternity ward.

Assessment has been carried out of the Danish procedure for neonatal eye screening and its relationship to early detection and the data show that without routine maternity ward eye screening, children are diagnosed and operated on statistically significantly later than in Sweden. The results were published in *Acta Ophthalmologica* in 2014.

Training in early detection screening techniques

Being able to make early diagnoses requires knowledge and training in examination techniques for doctors at maternity wards and paediatric health centres. An instructional film on screening techniques has been posted online in collaboration with Linköping University, which has also been translated into English This was carried out in conjunction with the publication of the article, "Ögonscreening på BB är effektiv" [Eye screening on maternity wards is effective] in *Läkartidningen*, which was based on data from PECARE.

Improved communication with families

Optimal treatment requires the insight and participation of parents considering the complex of issues specific to the condition and how visual development functions in children. A project regarding improving information for families with childhood cataracts has been implemented. A survey of relevant parents from the register and ophthalmologists has been analysed with regard to how families received information and how they would like to receive information. Essentially the same questions were asked of follow-up ophthalmologists – how they provide information and how they would prefer it to be. On the basis of this survey, a new training and information leaflet has been prepared, a scientific article on the subject was published in 2015 and a mobile app called Kind has come into use as an aid to communication.

The well-being of patients' parents: in-depth interviews and questionnaire studies

The concept of self-management involves an individual's ability to cope with symptoms, treatment and the psychosocial consequences and lifestyle changes associated with living with a chronic illness. In-depth interviews of 26 parents were carried out on a scientific basis with open-ended questions and verbatim transcriptions. These have been analysed and form the basis of two scientific articles, including models of parents' strategies as well as concrete proposals for improvement measures for eye clinic teams. The goal is to create a foundation for intervention and a programme of supportive strategies for parents who have children with cataracts. This work has been carried out by Jenny Gyllén, a member of the paediatric eye team at Queen Silvia's Hospital for Children and Youth, a part of Sahlgrenska University Hospital, where she works as an ophthalmic nurse. A total of four scientific articles have been published on the subject. Her thesis, *On parental self-efficacy in families with paediatric cataract and its clinical implications,* was defended in the fall of 2020. The resulting findings are being implemented in clinical practice, as well as in the afore mentioned mobile app. Forty percent of parents suffer from fatigue, tiredness that is not ameliorated by sleep. An action plan to help parents support their children is being prepared.

Further development

Information leaflet

A detailed information leaflet has been written based on the preferences of parents and ophthalmologists as described above under "Improved communication with families." It was posted online in 2014. The information is also available on the PECARE register's section on the NCR website.

Analysis of complications

A detailed, easy to use report on secondary glaucoma has been written. Two scientific analyses of the incidence of glaucoma in the register and associated factors have been conducted. One of these has been published and the second text is nearly finished.

A text for scientific publication on secondary VAO is in progress and is expected to be published in the autumn 2022 semester. A specialist trainee has conducted scientific analyses of the incidence of VAO within the framework of a specialised medicine project.

Study of comorbidity in children with bilateral cataracts

Bilateral cataracts may be a symptom and/or an element of a syndrome or other disease, such as a metabolic disease. Scientific evidence of this has been published as genetic analysis has become more accessible and cost-effective. An interprofessional collaboration to establish a recommendation for more extensive study of paediatric bilateral cataracts is in progress. Discussions have been held with representatives of the Swedish Registry for Inherited Metabolic Diseases (RMMS). A decision on genetic screening of all non-hereditary congenital cataracts has been discussed and partially implemented and will be reviewed and assessed.

Goal attainment and discussion

Systematic data registration on a national basis facilitates a unique statistical basis for evaluation and improvement of treatment methods and thus healthcare.

The goal involving optimisation of screening strategy was largely attained in 2021. This is particularly important for dense unilateral cataracts that require early surgical intervention before the age of 6 weeks. Data in the register shows that visual acuity for unilateral cataracts is higher than that published earlier, about two decades ago, which shows through the register that we are moving in the right direction.

The level of participation in the register is very high and all regions in the country report follow-up data to the register. The goal for PECARE is to be a full-coverage national register and continuous efforts are being made towards that goal. The degree of coverage in 2021 remains high at 88% when statisticians with the Swedish National Board of Health and Welfare compare against the patient register and the goal of being a full-coverage national register is assessed as having been met. The two centres that perform the majority of surgeries in Stockholm and Gothenburg are engaged in frequent dialogue by means of collaboration related to the register, including discussions of treatment outcomes and strategies for improvement. New

strategies have been implemented over the years and will be continuously evaluated.

Care provision is assessed as equitable and of equally high quality nationwide as regards accessibility, age at detection and diagnosis, age at surgery, gender and treatment outcomes.

PECARE's relevance from a global perspective

The register's research findings have had significant impact in connection with international reviews and analyses by a WHO Guideline Group for ophthalmological conditions composed of internationally recognised researchers who systematically reviewed the scientific literature aimed at providing advice based on the latest research findings to healthcare systems worldwide.

In the most recent "WHO recommendations on maternal and newborn care for a positive postnatal experience" (Geneva: World Health Organization Review), PECARE's results of newborn eye screening in the maternity ward played the biggest role in the recommendations for eye screening of all newborns in the world during the first weeks of life, as no other similar studies exist.

PECARE's steering committee is actively engaged in national and international partnerships. Two members of the steering committee are members of the steering committee for a parallel project, the EuReCCA (European Registry for Childhood Cataract Surgery).

VAO is a complication that affects 40% of operated eyes and causes vision to worsen again. Bag-in-the-lens IOL has been used in Sweden since 2009 and this is an artificial lens that virtually eliminates secondary VAO, which requires extensive care and thus also the high frequency of sedation of young children, which is a controversial subject. Only a few eye clinics in Europe currently have surgeons who have been specially trained in this surgical technique. The reduced frequency of complications is highly interesting from a global perspective and it is therefore important to also spread the positive outcomes of paediatric cataract surgery performed in Sweden outside national borders.

The impact of Covid

Paediatric cataract surgeries were prioritised during the pandemic and were performed as usual. Register data were reported and most steering committee meetings were held via Skype instead of face-to-face. Only one meeting was cancelled: the Nordic training meeting that was to have been held in 2021, "PECARE: 1,000 eyes later – what we have learnt", has been postponed for the second time.

Publications

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The Annual Report has also been published on the website:

http://rcsyd.se/pecare/