Short communication

Lenticonus diagnosis in Alport’s syndrome: Anterior capsule apical angle calculation using Scheimpflug imagery

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ABSTRACT

Objective: We describe a simplified method to detect anterior lenticonus. Three eyes of 2 patients with anterior lenticonus, plus 16 eyes from 16 healthy controls underwent Scheimpflug imaging of their anterior segment with Pentacam. The anterior capsule apex angle was manually identified and automatically measured by AutoCAD.

Results: The mean angle was 173.06° (SD: 1.91) in healthy subjects, and 158.33° (SD: 3.05) in anterior lenticonus eyes. The angle obtained from patients was more than 3 SD steeper than those from healthy subjects.

Conclusions: The apical angle calculation method seems to discriminate well between normal eyes and eyes suspected of having anterior lenticonus.

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Diagnóstico del lenticono en el síndrome de Alport: cálculo del ángulo apical de la cápsula anterior del cristalino mediante imágenes Scheimpflug

RESUMEN

Objetivo: Se describe un método simplificado para detectar lenteconico anterior. Realizamos imágenes Scheimpflug del segmento anterior con Pentacam a 3 ojos de 2 pacientes con lenteconico anterior y 16 ojos de controles sanos. Se identificó el ángulo del ápex de la cápsula anterior y se midió con AutoCAD.
Introduction

Anterior lenticonus is an infrequent condition in which the anterior portion of the anterior lens and the underlying cor-
tex protrude anteriorly due to a genetic defect in the synthesis
of type IV collagen. It is frequently associated with Alport’s
syndrome, a hereditary nephritis accompanied by ocular
anomalies and neurosensory deafness.

The development of the protrusion gives rise to irregular
astigmatism, myopia and high order aberrations which
sometimes cannot be corrected with spectacles or contact
lenses.

As changes in the environment are generally subtle, ante-
rior lenticonus may not be detected in ophthalmological
examination. Scheimpflug imaging technology is very useful
to analyze anterior segment morphology. This paper presents
2 patients with lenticonus to describe a diagnostic by means
of quantitative Scheimpflug analysis.

Case reports

Case 1

Male, 25, diagnosed with Alport’s syndrome due to kidney dis-
ease who visited due to progressive bilateral loss of vision.
Best corrected visual acuity (BCVA) was 0.49 logMAR in right
eye (RE) and 1 logMAR in left eye (LE). Refraction: −5 −1.5 to
168° RE and −10 −4 to 176° LE. Biomicroscopy suggested
the presence of bilateral anterior lenticones.

Case 2

Male, 52, diagnosed with Alport’s syndrome, had 3 kidney
transplants in addition to LE cataract operation several years
ago, visited to request treatment for RE. BCVA in RE: 1.30 log-
MAR and in LE: 0.20 logMAR. Examination revealed anterior
lenticon with right posterior capsular opacity and left pseu-
dophakia.

Scheimpflug anterior segment images were taken (Pen-
tacam, Oculus, USA) of the 3 eyes (cases 1 and 2) and of
16 eyes of 16 healthy controls between 20 and 60 years old after
pupil dilatation (2 drops of cycloplegic Coliricusi eyedrops at
1% by Alcon Cusi Laboratorios). All the subjects accepted the
examination and signed informed consents. The study was
reviewed and approved by the Ethics Committee of the Ramón
y Cajal Hospital (Madrid) and carried out in accordance with
the criteria established in the Helsinki declaration. A standard
Scheimpflug image was selected (image 183-3). The images
were exported and analyzed with the AutoCAD graphic design
application (Audodesk, Inc., California, USA). The apex of the
anterior capsule was identified and a tangent line projected
manually from the capsule at each side. These lines made up
an angle that was automatically measured by the AutoCAD
application (Fig. 1).

The mean angle measurements were 173.06° (standard
device [SD]: 1.91) in checkups and 158.33° (SD: 3.05) in ante-
rior lenticons. The mean apical angle of the lenticons was
more acute, over 3 SD, then that of controls.

Discussion

The above cases illustrate the usefulness of the Scheimpflug
technology to detect anterior lenticons with a simple method
that calculates the anterior capsule apex angle. Early diagno-
istic would enable rehabilitation of these patients’ vision
and prevent and treat the amblyopia generated by irregular
astigmatism in developing eyes.

Routine ophthalmological exploration was the only tool
for detecting lenticon but does not reveal subcutaneous
anomalies. With biomicroscopy it is possible to observe ante-
rior chamber protrusion, which is sometimes opacified. Retin-
scopy shows an “oil drop” image.

Wavefront analysis is of help in screening lenticon. Analy-
ing the difference between high order aberrations derived
from the wavefront analysis of the high order aberrations map
of corneal topography, it is possible to differentiate the corneal
or lens origin. A markedly negative spherical aberration
indicates anterior surface protrusion, although it requires
morphometric measurements.

Lens anterior pole Scheimpflug images can show ante-
rior lenticon, although qualitative analysis on its own is
subjective and not very precise. Quantitative analysis of the
protrusion provides more objective and earlier data. In a sin-
gle case, Xu measures apex protrusion from a 4 mm long line
that is manually drawn. This measure is over 3 SD larger in
lenticon than in the normal population.

A different and simpler approach is proposed: the angle
formed by the protrusion is measured, finding that it is over
3 SD more acute in lenticon than in healthy controls. This
method seems less subjective and is based on the measures
of 3 eyes of 2 patients, although it requires validation with
additional cases.

However, this process has the drawback that it cannot be
performed with Pentacam software, which means that the
image has to be exported. Even so, after validation it would
be easy to include said process.
It would be interesting to assess whether this method can detect pre-clinic anterior lenticone in patients diagnosed with Alport syndrome as kidney alterations and diagnostic usually appear before visual impairment.

New imaging technologies allow us to perform qualitative and quantitative analysis of the crystalline lens in order to detect and monitor its disorders. The present method would facilitate discriminating between normal lenses and anterior lenticone suspects before the anomaly became evident. In addition, it would allow us to assess the evolution thereof with successive examinations.

Conflict of interest

No conflict of interest has been declared by the authors.

REFERENCES


Fig. 1 – Left: Scheimpflug images showing crystalline lens outline and anterior angle curvature in patients with lenticone. Right: Scheimpflug images showing lens outline and anterior angle curvature in normal eyes (pupils in pharmacological midriasis in all cases).