

CASE REPORT

Open Access



# Myelin oligodendrocyte glycoprotein (MOG) associated optic neuritis in a patient with idiopathic intracranial hypertension (IIH) and compressive optic neuropathy case report

Ryan D. Sorensen<sup>1\*</sup>, Ashtyn Vogt<sup>2</sup>, Noor Laylani<sup>3</sup>, Mohammad Pakravan<sup>3</sup> and Andrew G. Lee<sup>3,4,5,6,7,8</sup>

## Abstract

**Background** Myelin oligodendrocyte glycoprotein-associated disease (MOGAD) has a wide phenotypic expression and should be considered in a differential diagnosis of patients with optic disc edema and increased intracranial pressure because MOGAD can mimic IIH and compressive optic neuropathy.

**Case presentation** A 53-year-old woman with a history of presumed idiopathic intracranial hypertension (“IIH”) presented with new headache and visual loss. She had a BMI of 35.44 kg/m<sup>2</sup> and a past medical history significant for depression, hepatitis C, hyperlipidemia, and uterine cancer post-hysterectomy. She had undergone multiple lumboperitoneal shunts for presumed IIH and had a prior pituitary adenoma resection. Her visual acuity was no light perception OD and counting fingers OS. After neuro-ophthalmic consultation, a repeat cranial MRI showed symmetric thin peripheral optic nerve sheath enhancement of the intra-orbital optic nerves OU. Serum MOG antibody was positive at 1:100 and she was treated with intravenous steroids followed by plasma exchange and rituximab.

**Conclusions** This case highlights the importance of considering MOGAD in the differential diagnosis of optic neuropathy. Although likely multifactorial, we believe that the lack of improvement in our case from presumed IIH and despite adequate neurosurgical decompression of a pituitary adenoma with compression of the optic apparatus reflected underlying unrecognized MOGAD. Clinicians should consider repeat imaging of the orbit (in addition to the head) in cases of atypical IIH or compressive optic neuropathy especially when the clinical course or response to therapy is poor or progressive.

**Keywords** Idiopathic intracranial hypertension, Myelin oligodendrocyte glycoprotein antibody, Pituitary adenoma

\*Correspondence:

Ryan D. Sorensen

ryan.sorensen@bcm.edu

<sup>1</sup>Baylor College of Medicine, 2535 Shakespeare St Unit 2, 77030 Houston, TX, USA

<sup>2</sup>Texas A and M College of Medicine, Dallas, TX, USA

<sup>3</sup>Department of Ophthalmology, Blanton Eye Institute, Houston Methodist Hospital, Houston, TX, USA

<sup>4</sup>Departments of Ophthalmology, Neurology, and Neurosurgery, Weill Cornell Medicine, New York, USA

<sup>5</sup>Department of Ophthalmology, University of Texas Medical Branch, Galveston, TX, USA

<sup>6</sup>University of Texas MD Anderson Cancer Center, Houston, TX, USA

<sup>7</sup>Texas A and M College of Medicine, Bryan, TX, USA

<sup>8</sup>Department of Ophthalmology, The University of Iowa Hospitals and Clinics, Iowa City, IA, USA



© The Author(s) 2024. **Open Access** This article is licensed under a Creative Commons Attribution 4.0 International License, which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if changes were made. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit <http://creativecommons.org/licenses/by/4.0/>. The Creative Commons Public Domain Dedication waiver (<http://creativecommons.org/publicdomain/zero/1.0/>) applies to the data made available in this article, unless otherwise stated in a credit line to the data.

## Background

Myelin oligodendrocyte glycoprotein-associated disease (MOGAD) has a wide phenotypic expression that includes optic neuritis, transverse myelitis, acute demyelinating encephalomyelitis (ADEM), as well as other neurologic syndromes. MOG antibody (MOG-Ab) is an autoantibody that has been associated with neuromyelitis optica spectrum disorder (NMOSD), optic neuritis (ON), and serous retinal detachment [1–4]. We report a case of MOGAD optic neuritis in a patient with presumed idiopathic intracranial hypertension (IIH) and compressive optic neuropathy who did not recover vision despite aggressive surgical treatment for both IIH and compressive optic neuropathy. We hypothesize that MOGAD mimicked these conditions in our case.

## Case presentation

A 53-year-old woman presented with bilateral visual loss. She had a prior history of “IIH” by modified Dandy criteria 19 years prior but aggressive surgical treatment with lumboperitoneal shunt placement and two revisions failed to improve the vision. She had a pituitary adenoma that was resected but the vision did not improve and she presented with 3 weeks later with new-onset headache and visual loss. Her BMI was 35.44 kg/m<sup>2</sup>. Past medical history included depression, hepatitis C, hyperlipidemia, and uterine cancer post-hysterectomy. The patient was taking trazodone for anxiety and buprenorphine for chronic pain. She had allergies to penicillin, tolterodine, ondansetron, metoclopramide, and prochlorperazine. She never smoked and had no history of alcohol or drug use. The family history was non-contributory.

At presentation she reported new headaches, transient visual obscurations, and a history of “papilledema” and “IIH” in 2003, at the age of 35. In 2003, cranial magnetic resonance imaging (MRI) was normal and lumbar puncture (LP) revealed normal CSF contents but an elevated opening pressure, which was compatible with the modified Dandy criteria for IIH. The patient had progressive visual loss that required LP shunt and had a subsequent shunt revision.

From 2015 to 2018, she began experiencing painless peripheral vision loss, which she and her physicians attributed to “old damage from the pseudotumor cerebri”.

The patient presented again in 2018 with progressive peripheral vision loss in both eyes. A repeat cranial MRI showed a prior stable mild Chiari I malformation but a new suprasellar pituitary adenoma (Fig. 1). A diagnosis of compression optic neuropathy was made. Visual acuity was hand motion in the right eye (OD) and 20/60 in the left eye (OS). Automated perimetry (Humphrey visual field (HVF) 24–2) revealed a temporal hemianopic impairment with a superimposed inferior altitudinal field loss OS and diffuse depression OD. Repeat LP results

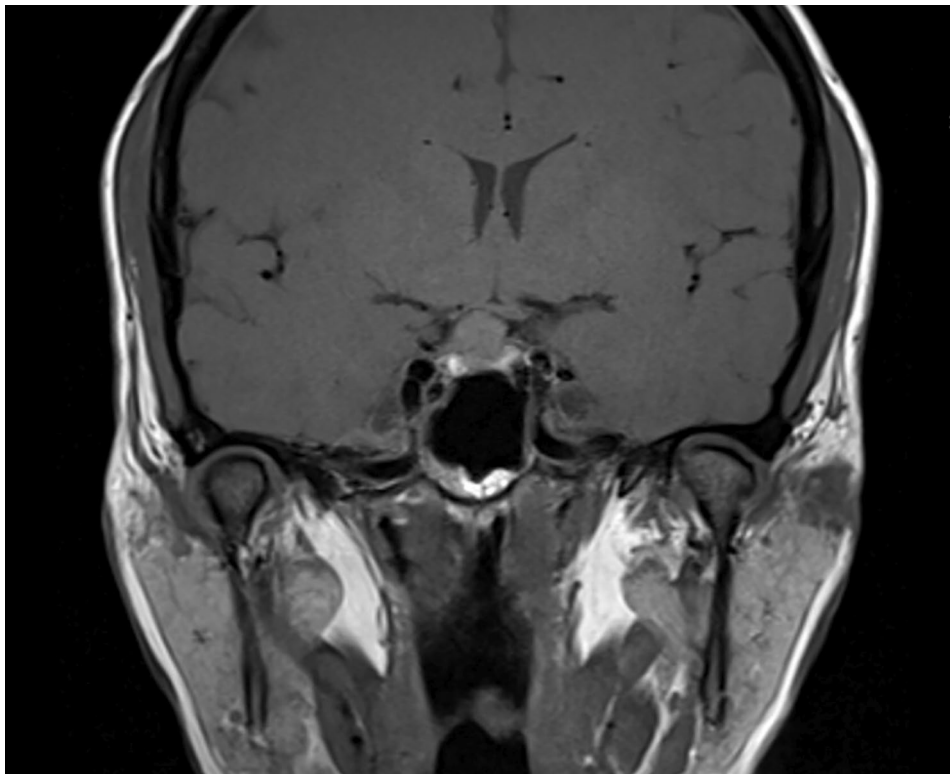
revealed normal CSF and opening pressure. The patient had a Grade 1 pituitary adenoma removed completely via a transnasal technique, but her vision did not improve, likely due to the prior optic atrophy that was present for years prior to resection.

In February 2020 the patient presented to the neuro-ophthalmology clinic at Houston Methodist Hospital with one-month worsening headaches and vision loss for which she was admitted. On neuro-ophthalmic examination, the visual acuity was no light perception OD and counting fingers OS. The right pupil was amaurotic. Optical coherence tomography (OCT) of the retinal nerve fiber layer and the macular ganglion cell layer showed diffuse loss OU. New subretinal and intraretinal edema was seen on OCT (Fig. 2) and ophthalmoscopy showed diffuse optic atrophy OU.

Repeat cranial MRI showed gross total resection of the pituitary adenoma with no recurrent or residual disease. LP showed normal opening pressure and CSF content. Repeat orbital MRI with contrast however showed symmetric thin peripheral optic nerve sheath enhancement of the intra-orbital optic nerves OU (Fig. 3). Serum MOG-Ab testing was positive at a 1:100 titer. She was treated with intravenous steroids followed by plasma exchange and rituximab. There was an improvement of subretinal fluid collection following the plasma exchange, and she continued rituximab every 6 months. In September of 2020 her VA was NLP OD and 20/400 OS but the incomplete improvement was attributed to prior optic atrophy. In August of 2021 OCT was done (Fig. 2) and her VA was stable at NLP OD and 20/400 OS. On May 3, 2022, she presented with 3 weeks of worsening vision and headaches, her VA was NLP OD and CF OS, with a right RAPD, on OCT global was 75 OD and 67 OS after being 99 OD and 84 OS and showed bilateral optic atrophy despite treatment for MOGAD likely due to the delayed treatment of MOGAD.

## Discussion and conclusions

Both IIH and MOGAD-related optic neuritis can present with visual loss, headache, and optic disc and macular edema. There have been previous cases of MOGAD patients with typical symptoms who were initially diagnosed with IIH but then developed visual deficits suggestive of optic neuritis, in certain clinical contexts (e.g., fulminant IIH) it may be difficult to initially differentiate visual loss from papilledema due to IIH and MOGAD optic neuritis [1, 2]. Table 1 lists the prior cases of MOGAD associated with increased intracranial pressure mimicking IIH. The proposed mechanism for MOGAD-related increased intracranial pressure has been CSF inflammation and secondary decreased CSF reabsorption [2]. We believe that this patient had MOGAD with secondary increased intracranial pressure in the setting

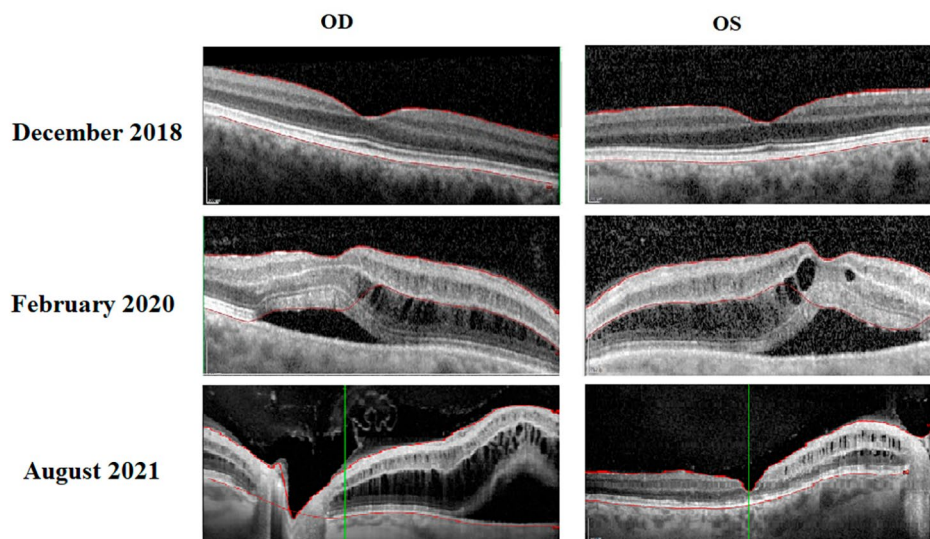


**Fig. 1** Coronal T1 preoperation. Small possibly necrotic slow-enhancing component in the inferior aspect of the pituitary gland

of a concomitant pituitary adenoma. Although the optic neuropathy is likely multifactorial, we hypothesize that MOGAD was the primary cause for the visual loss and that the presentation mimicked the visual loss from prior “IIH” and then later from pituitary adenoma.

MOGAD can present bilaterally and with optic disc edema, in contrast to typical optic neuritis from multiple

sclerosis which is usually unilateral and associated with a normal fundus or mild if any optic disc edema. In addition, optic nerve sheath enhancement has been shown to be a differentiating and distinctive finding for MOGAD (see table) that is not typically seen in IIH or MS-related optic neuritis [5]. Finally, although macular edema can

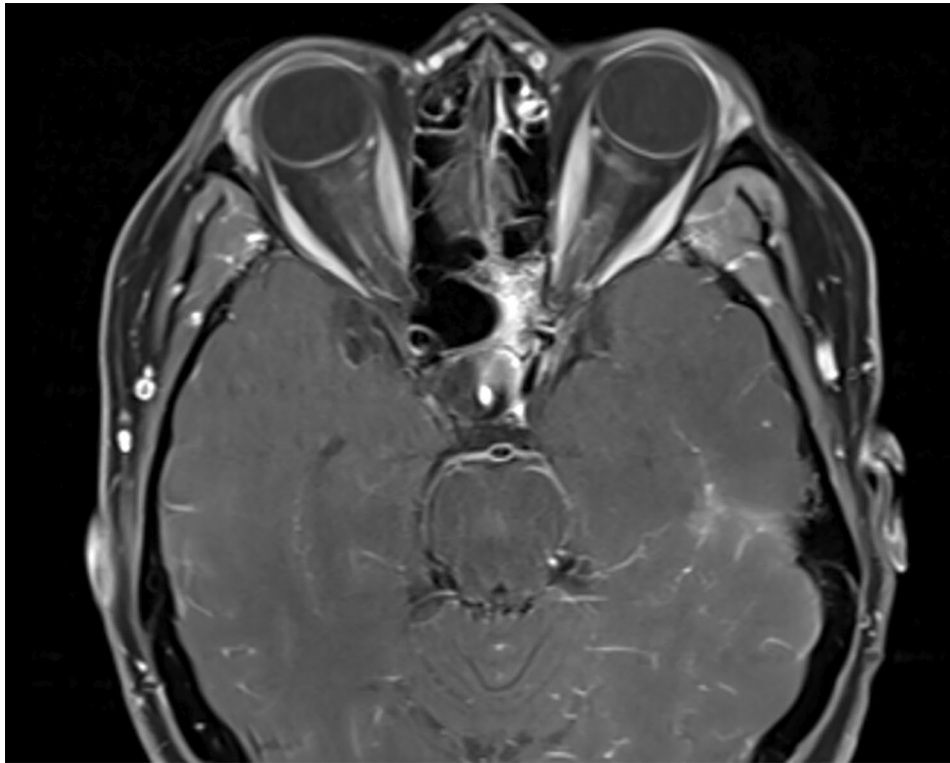


**Fig. 2** OCT scans from 2018–2021

**Table 1** Prior cases of MOGAD associated with increased intracranial pressure mimicking IIH compared to the case presented

Presenting Case	Patient(1) from 'Case of atypical Anti-MOG Syndrome' [1]	Patient(2) from 'Case of atypical Anti-MOG Syndrome' [1]	'Atypical MOG Presenting with isolated elevate ICP' [2]	'Case of MOG associated with serious retinal detachment' [3]	Patient(1) from 'MOG masquerading as IIH' [4]	Patient(2) from 'MOG masquerading as IIH' [4]	Patient(3) from 'MOG masquerading as IIH' [4]	Patient(4) from 'MOG masquerading as IIH' [4]	Patient(5) from 'MOG masquerading as IIH' [4]	Case/Averages from MOG Clinical Characteristics [5]
<b>Gender</b>	Female.	Male.	Male.	Female.	Female.	Male.	Female.	Male.	Female.	57% Female.
<b>BMI</b>	35.44 kg/m <sup>2</sup>	NA	Obese	NA	38 kg/m <sup>2</sup>	32 kg/m <sup>2</sup>	41 kg/m <sup>2</sup>	32 kg/m <sup>2</sup>	23 kg/m <sup>2</sup>	NA
<b>Visual Acuity</b>	HA OD and 20/60 in OS.	HA OD. RAPD OD.	Normal.	20/50 OD 20/40 OS.	Mildly decreased.	Mildly decreased.	20/60 bilaterally.	Unable to obtain.	20/60 OD. RAPD OD.	20/30 Bilateral.
<b>Visual Fields</b>	Diffuse depressed OD and a temporal hemianopia superimposed on inferior altitudinal field defect OS.	NA	Normal.	Mild enlargement of the physiological cal blind spot OS.	Bilaterally enlarged blind spots.	Bilaterally enlarged blind spots.	Nasal step defect in one eye and a normal field in the other.	NA	NA	NA
<b>OCT</b>	OCT 37 OD.40 OS with diffuse optic atrophy. Edema in macula.	NA	NA	OCT revealed subretinal fluid in macula bilaterally.	Thickened peripapillary retinal nerve fiber layer (RNFL) bilaterally. 172 μm.	Thickened RNFL bilaterally. 140 μm, LE bilaterally. 172 μm.	Thickened RNFL bilaterally (RE 140 μm, LE 172 μm).	Thickened RNFL bilaterally.	NA	NA
<b>Fundoscopic Findings</b>	Intra-retinal cystic change in the inner retina corresponding with optic atrophy.	Disk edema OD.	Bilateral papilloedema.	Bilateral disk edema.	Bilateral disc edema.	Bilateral disc edema.	Bilateral disc edema.	Bilateral disc edema.	Bilateral disc edema.	36/42. Present in 86% of cases.
<b>Symptoms</b>	Painless progressive peripheral vision loss.	Headache, neck pain, neck stiffness, photophobia, nausea, vomiting.	Headache, blurry vision, nausea, and vomiting.	1-week history of blurred vision.	Headaches, blurred vision.	Headaches, blurred vision.	Headaches, blurred vision.	Headaches, blurred vision.	Headaches, blurred vision.	Pain (86%) Recurrent optic neuritis (10%) Monophasic optic neuritis (10%) ADEM (18%) NMSOD-like phenotype (22%)
<b>MRI findings</b>	ONSE.	Inflammation of the right optic nerve.	Signs of elevated ICP, dilation of nerve sheaths, flattening of posterior globes, and a partially empty sella.	ONSE.	Hyperintense signal in both optic nerves with gadolinium enhancement of the right optic nerve.	ONSE.	ONSE.	ONSE.	Enhancement of the right optic nerve.	Perineural enhancement: 50% Length involved more than half: 80% Orbital portion involved: 92% Intracranial portion involved: 72% Chiasm involved: 12%
<b>CSF Opening Pressure and Analysis</b>	Markedly elevated. Normal constituents	24 cmH <sub>2</sub> O Lymphocytic Pleocytosis	52 cmH <sub>2</sub> O Lymphocytic Pleocytosis	NA	26 cmH <sub>2</sub> O Normal constituents	33 cmH <sub>2</sub> O Normal constituents	23 cmH <sub>2</sub> O Normal constituents	24 cmH <sub>2</sub> O Normal constituents	35 cmH <sub>2</sub> O Normal	44% had pleocytosis
<b>MOG Titer</b>	1:100	Positive.	1:10,000	1:512	Positive.	Positive.	Positive.	Positive.	Positive.	100*

NA – not available, HA – Hand motion, CF – count fingers, ONSE – Optic Nerve Sheath Enhancement, \*Criteria for study was MOG positive titer.



**Fig. 3** Axial T1 fat-suppressed magnetic resonance imaging shows optic nerve sheath enhancement bilaterally

rarely be seen in optic neuritis this finding has been reported in MOGAD.

This patient has a very unusual case of MOG-related optic neuritis which was complicated by the presence of a pituitary adenoma 15 years after the initial presenting symptoms. Either MOG-related optic neuritis manifested as IIH from the beginning of this patient's history or it developed in the setting of a long-standing IIH. Clinicians should be aware that IIH symptoms (e.g., fulminant IIH) can mirror MOGAD-related optic disc edema. Orbital and cranial MRI should be performed to evaluate for optic nerve sheath enhancement which can be more suggestive of MOGAD than IIH. In addition to having a positive titer, LP may also exhibit inflammatory CSF indices, which could be the cause of the elevated ICP caused by MOGAD, however, there are many cases reported of MOGAD masquerading as IIH which had no inflammatory markers noted in the CSF as shown in Table 1 similar to this patient's first CSF study [4, 5].

We believe that our case highlights the importance of considering MOGAD in the differential diagnosis of any acute bilateral optic neuropathy. Acute fulminant presentations of IIH can produce a similar clinical picture to MOGAD. Likewise lack of visual improvement despite adequate neurosurgical decompression of a pituitary adenoma should also prompt consideration for testing for MOGAD. In addition, clinicians should consider imaging of the orbit and brain with contrast in cases of fulminant,

progressive, or atypical IIH and in cases of compressive optic neuropathy which do not recover following surgical decompression. The presence of optic nerve sheath enhancement on orbital MRI should prompt consideration for MOGAD.

#### Abbreviations

MOGAD	Myelin oligodendrocyte glycoprotein-associated disease
ADEM	Acute demyelinating encephalomyelitis
MOG-Ab	Myelin oligodendrocyte glycoprotein antibody
NMOSD	Neuromyelitis optica spectrum disorder
ON	Optic Neuritis
IIH	Idiopathic intracranial hypertension

#### Author contributions

R.S., A.L. and N.L. wrote the main manuscript text and prepared Table 1. M.P. and A.V. gathered the relevant patient information and prepared Figs. 1, 2 and 3. All authors reviewed the manuscript.

#### Funding

None.

#### Data availability

The images in the figures are from the patient's medical record and are used with the written consent of the patient. Additional data is available upon request.

#### Declarations

##### Ethics approval and consent to participate

The patient consented to participate.

##### Consent for publication

Written informed consent was obtained from the participant for publication of identifiable information or images.



**Competing interests**

The authors declare that they have no competing interests

Received: 20 September 2023 / Accepted: 27 December 2023

Published online: 13 February 2024

**References**

1. Narayan RN, Wang C, Sguigna P, Husari K, Greenberg B. Atypical Anti-MOG syndrome with aseptic meningoencephalitis and pseudotumor cerebri-like presentations. *Multiple Scler Relat Disorders*. 2019;27:30–3.
2. Valdrighi A, Russ J, Wauban E, Rasool N, Francisco C. Atypical myelin oligodendrocyte glycoprotein antibody Disease presenting with isolated elevated intracranial pressure. *Neuroimmunol Rep*. 2021;1:100028.
3. Kon T, Hikichi H, Ueno T, Suzuki C, Nunomura J, Kaneko K, Takahashi T, Nakashima I, Tomiyama M. Myelin oligodendrocyte Glycoprotein-IgG-positive recurrent bilateral Optic Papillitis with Serous Retinal detachment. *Intern Med*. 2018;57:3307–12.
4. Lotan I, Brody J, Hellmann MA, Bialer O, Ganelin-Cohen E, Michaeli N, Marignier R, Stiebel-Kalish H. Myelin oligodendrocyte glycoprotein-positive optic neuritis masquerading as pseudotumor cerebri at presentation. *J Neurol*. 2018;265:1985–8.
5. Chen JJ, Flanagan EP, Jitrapaikulsan J, López-Chiriboga ASS, Fryer JP, Leavitt JA, Weinshenker BG, McKeon A, Tillemma JM, Lennon VA, Tobin WO, Keegan BM, Lucchinetti CF, Kantarci OH, McClelland CM, Lee MS, Bennett JL, Pelak VS, Chen Y, VanStavern G, Adesina OO, Eggenberger ER, Acierno MD, Wingerchuk DM, Brazis PW, Sagen J, Pittock SJ. Myelin oligodendrocyte glycoprotein antibody-positive Optic neuritis: clinical characteristics, radiologic clues, and Outcome. *Am J Ophthalmol*. 2018;195:8–15.

**Publisher's Note**

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.