

1. DACH ANCA VASKULITIS FORUM 2023
12. & 13. MAI 2023

Was können Vaskulitis-Register leisten? - GeVas – Gemeinsames Vaskulitis-Register im deutschsprachigen Raum

Prof. Dr. Peter Lamprecht
 Lübeck



1. DACH ANCA VASKULITIS FORUM 2023
12. & 13. MAI 2023 | WIEN, HOTEL SAVOYEN

P. Lamprecht



GeVas – Gemeinsames Vaskulitis Register im deutschsprachigen Raum

Wien, 13.05.2023

1ST DACH ANCA VASKULITIS FORUM 2023

12. & 13. MAI 2023 | WIEN, HOTEL SAVOYEN



UNIVERSITÄT ZU LÜBECK
KLINIK FÜR RHEUMATOLOGIE
UND KLINISCHE IMMUNOLOGIE

Peter Lamprecht

Eine kurze Geschichte... – GeVas

- 1. Kick-off, Hamburg, 28.11.2014
Holle / Moosig
Deutsches Vaskulitis Register (D)
German Vasculitis Registry
- “Relaunch”, Frankfurt, 17.03.2017 
Iking-Konert / Lamprecht
Gemeinsames Vaskulitis Register im deutschsprachigen
Raum (DACH)
Joint Vasculitis Registry in German-speaking Countries
- 2017ff. Beobachtungsplan, Zentrenrekrutierung & -verträge,
Ethikvoten, eCRF 2.0 Design, Validierung & Launch, FPI
05.06.2019, *2020 – 2022 Corona, 2022 BfArM Intervention* →
 - Protokollamendment, 2023ff. Zentrenerweiterung,...

GeVas – Steering Board, Arbeitsgruppen & ZKS

➤ Steering Board

- de Groot, Hellmich, Hoyer, Iking-Konert (Co-PI), Lamprecht (PI), Magnus, Müller-Ladner, Rech, Schulze-Koops, Venhoff, Villiger

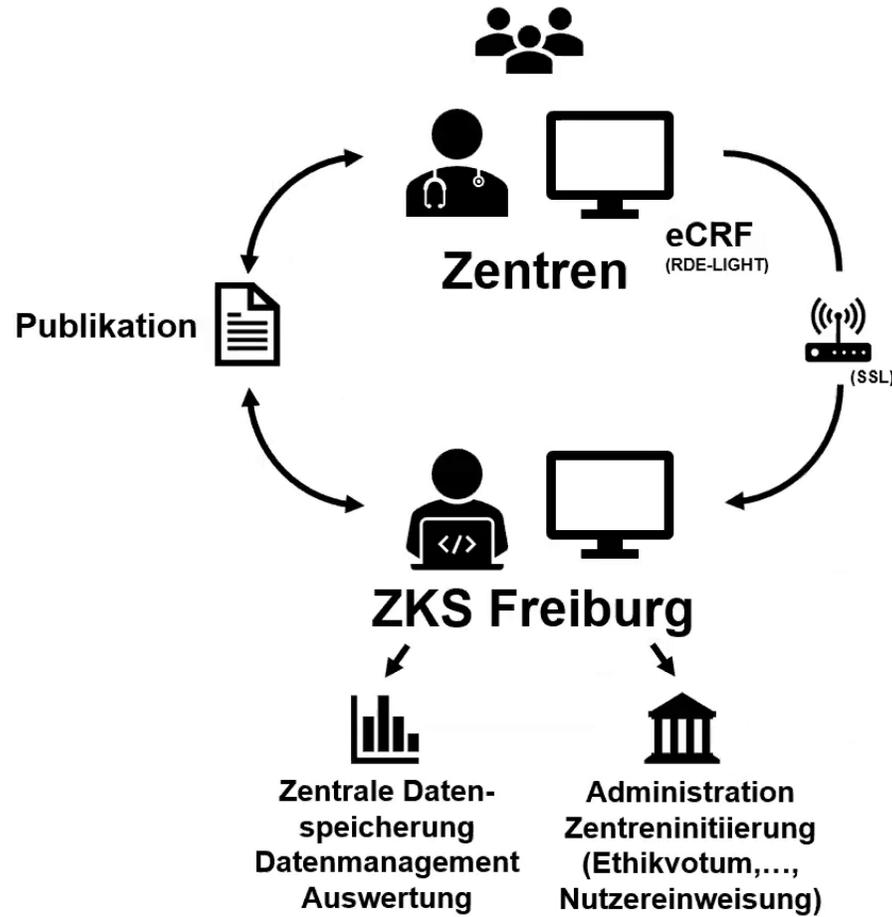
➤ Arbeitsgruppen

- AAV, Behçet & Cogan, Nephrologie, Pathologie, RZA & TAK, ZNS-Vaskulitis

➤ Zentrum Klinische Studien (ZKS) Freiburg

- Tais (Studienkoordinatorin), Bradbury (Projektassistenz), Kaufmann (Programmierung), Ihorst (Auswertung), Jäger (FAIRVASC)

GeVas – Datenerhebung & Auswertung



GeVas – Studienprotokoll

Gemeinsames Vaskulitis Register im deutschsprachigen Raum

(GeVas)



DRKS-Nr.

DRKS00011866

Protokollversion

V5 vom 04.08.2022

Studienleiter

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Stellvertretender

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GeVas
Beobachtungsplan
V5 / 04.08.2022

GeVas – Ziele

- **Systematische Erfassung und Langzeitdokumentation von Vaskulitiden**
- **„Das Ziel des Vaskulitisregisters ist es, alle Patienten mit einer neu diagnostizierten Vaskulitis oder einer Therapieumstellung bei einem Rezidiv in einem prospektiven, webbasierten, nationalen Register multizentrisch, standardisiert, systematisch und pseudonymisiert zu erfassen und so den Langzeitverlauf von Vaskulitiden zu verfolgen“**
- ***Inzeptionskohorte***
- **Datenauswertung & Publikationen**

GeVas – Registerdeskription

The Joint Vasculitis Registry in German-speaking countries (GeVas) – a prospective, multicenter registry for the follow-up of long-term outcomes in vasculitis

Christof Iking-Konert^{1*} , Pia Wallmeier¹, Sabrina Arnold², Sabine Adler³, Kirsten de Groot⁴, Bernhard Hellmich⁵, Bimba F. Hoyer⁶, Konstanze Holl-Ulrich⁷, Gabriele Ihorst⁸, Margit Kaufmann⁸, Ina Kötter⁹, Ulf Müller-Ladner¹⁰, T. Magnus¹¹, Jürgen Rech¹², Fabian Schubach⁸, Hendrik Schulze-Koops¹³, Nils Venhoff¹⁴, Thorsten Wiech¹⁵, Peter Villiger¹⁶ and Peter Lamprecht²

Iking-Konert *et al.* *BMC Rheumatology* (2021) 5:40
<https://doi.org/10.1186/s41927-021-00206-2>

GeVas – Registerdeskription

Abstract

Background: Vasculitides comprise a group of rare diseases which affect less than 5 in 10.000 individuals. Most types of vasculitis can become organ- and life-threatening and are characterized by chronicity, high morbidity and relapses, altogether resulting in significant morbidity and mortality. Previous studies have been either monocentric or mainly retrospective – studies with a prospective design mostly consisted of rather small cohorts of 100 to 200 patients.

The aim of the Joint Vasculitis Registry in German-speaking countries (GeVas) is to record all patients who have been recently diagnosed with vasculitis or who have changed their treatment due to a relapse (inception cohort). In GeVas, data are collected prospectively in a multicenter design in Germany, Austria and Switzerland. By this approach, courses of vasculitis and their outcomes can be monitored over an extended period.

Methods: GeVas is a prospective, web-based, multicenter, clinician-driven registry for the documentation of organ manifestations, damage, long-term progress and other outcomes of various types of vasculitis. The registry started recruiting in June 2019. As of October 2020, 14 centers have been initiated and started recruiting patients in Germany. Involvement of sites in Austria and the German-speaking counties of Switzerland is scheduled in the near future.

Discussion: In June 2019, we successfully established a prospective multicenter vasculitis registry being the first of its kind in German-speaking countries. The participating centers are currently recruiting, and systematic analysis of long-term vasculitis outcomes is expected in the ensuing period.

Trial registration: German Clinical Trials Register (Deutsches Register Klinischer Studien): [DRKS00011866](#). Registered 10 May 2019.

Keywords: Vasculitis, GeVas, ANCA, Prospective, Registry, Giant cell arteritis, Long term, Outcome, Therapy

GeVas – BfArM Intervention 2022

- **Überprüfung durch BfArM auf Veranlassung einer Ethikkommission**

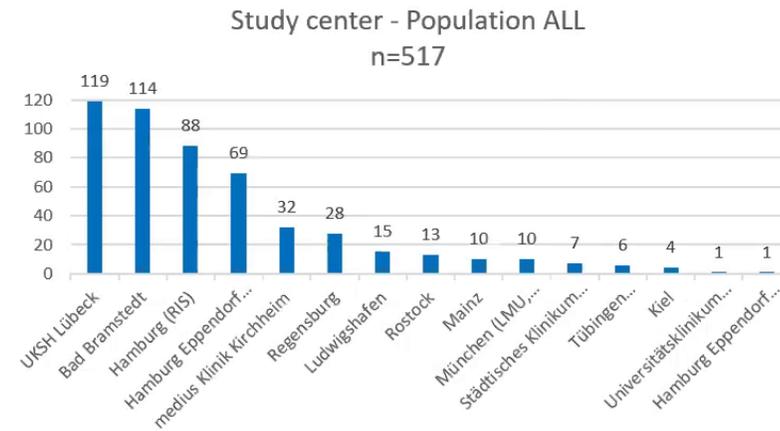
GeVas – BfArM Intervention 2022

- **Überprüfung durch BfArM auf Veranlassung einer Ethikkommission**
- **BfArM: Abgrenzung Register ↔ Nicht-interventionelle Studie (NIS), GeVas fällt nicht unter Arzneimittelgesetz, aber Amendment (V5) zum Protokoll bezgl. AE: kein Bezug zu spezifischen Arzneimitteln**

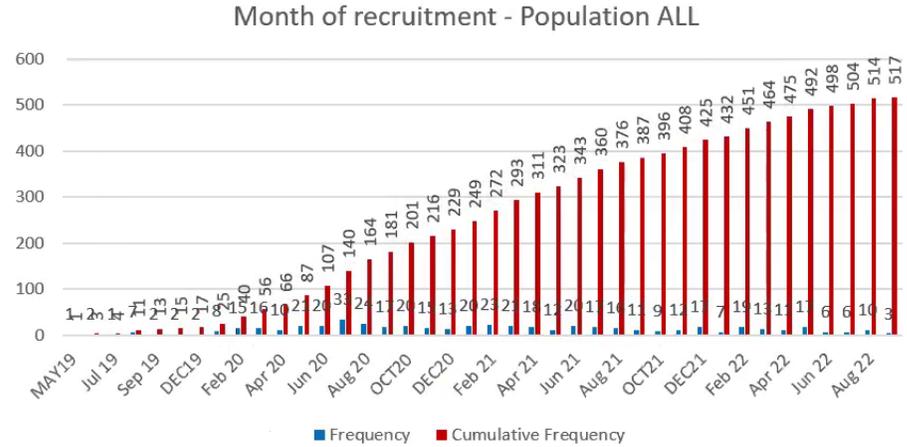
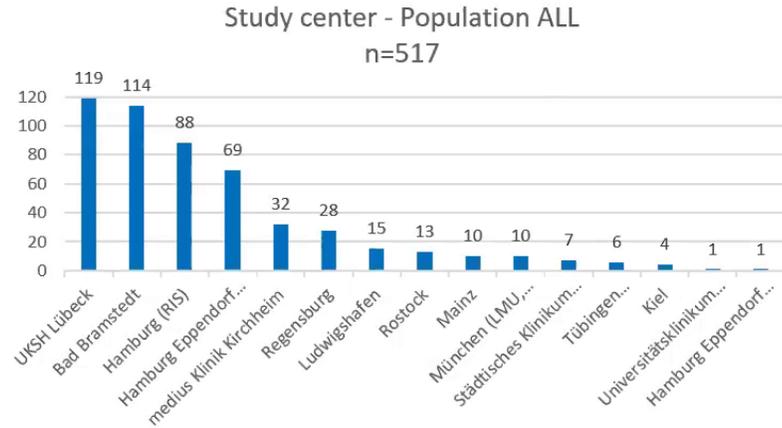
GeVas – BfArM Intervention 2022

- **Überprüfung durch BfArM auf Veranlassung einer Ethikkommission**
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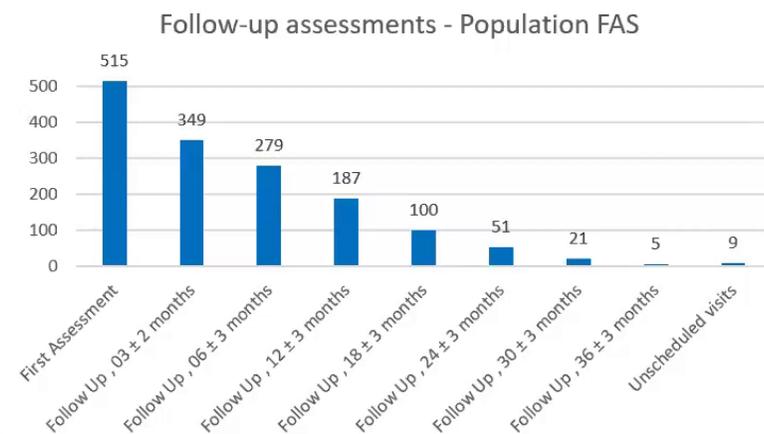
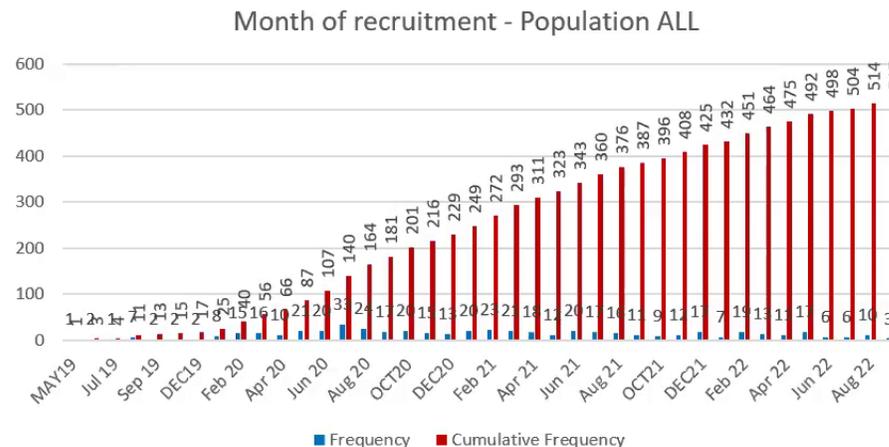
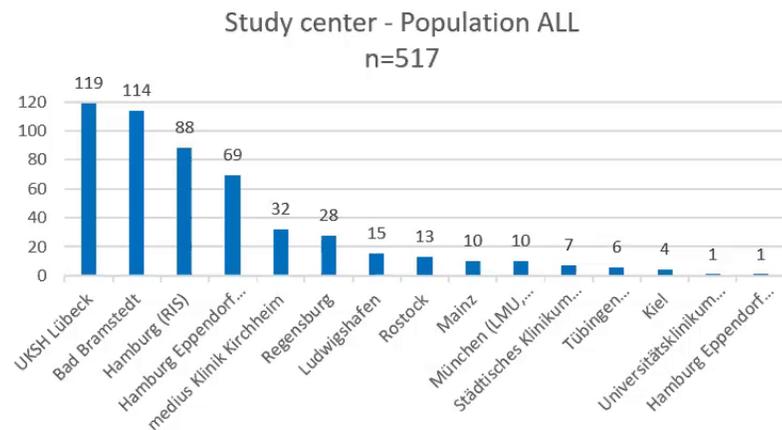
GeVas – Rekrutierung: Stand 10/22



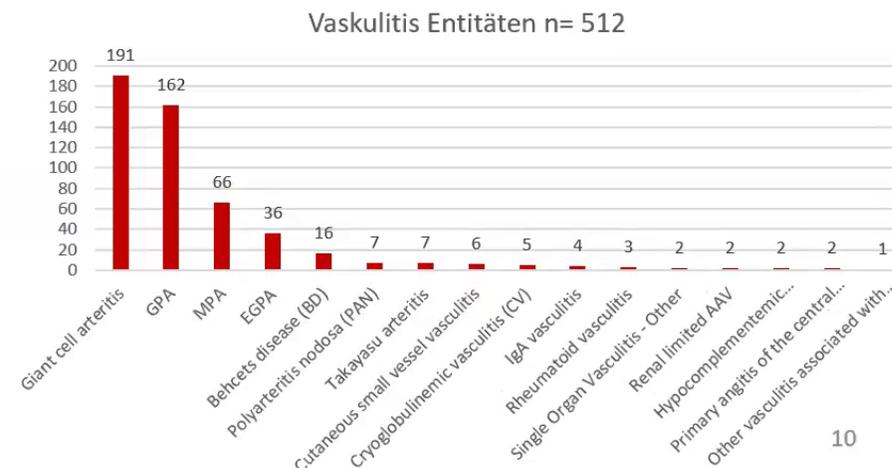
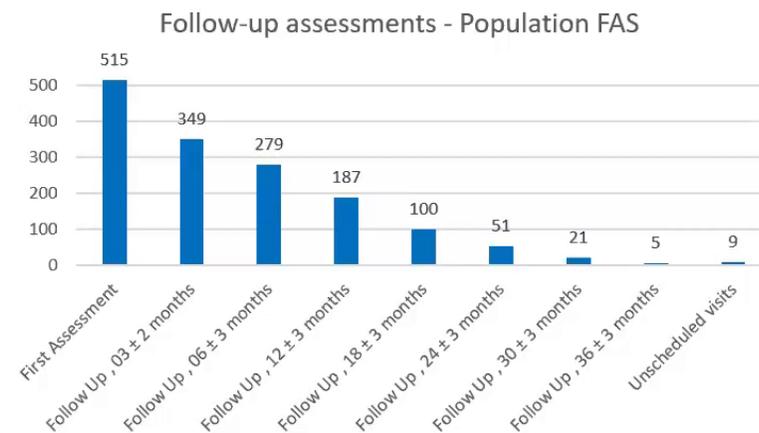
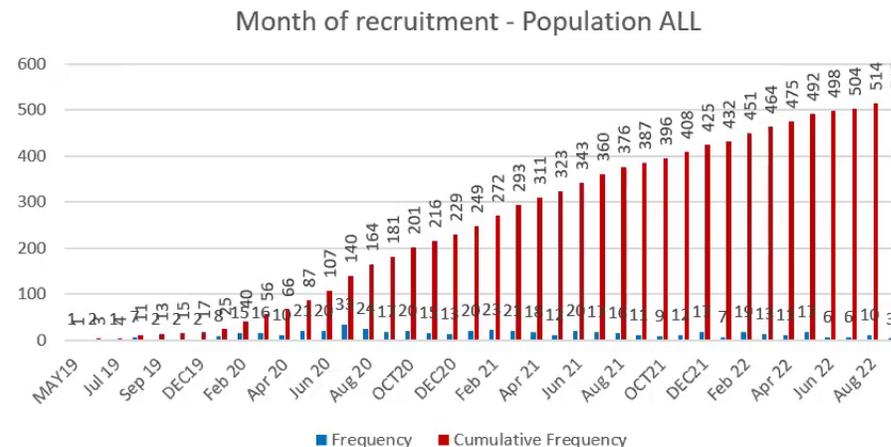
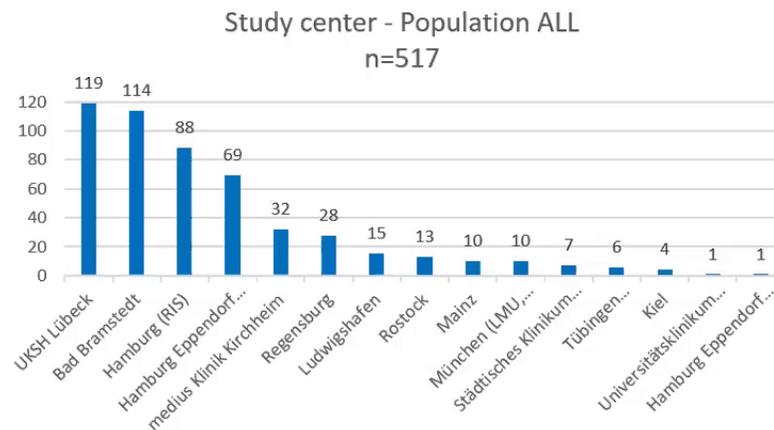
GeVas – Rekrutierung: Stand 10/22



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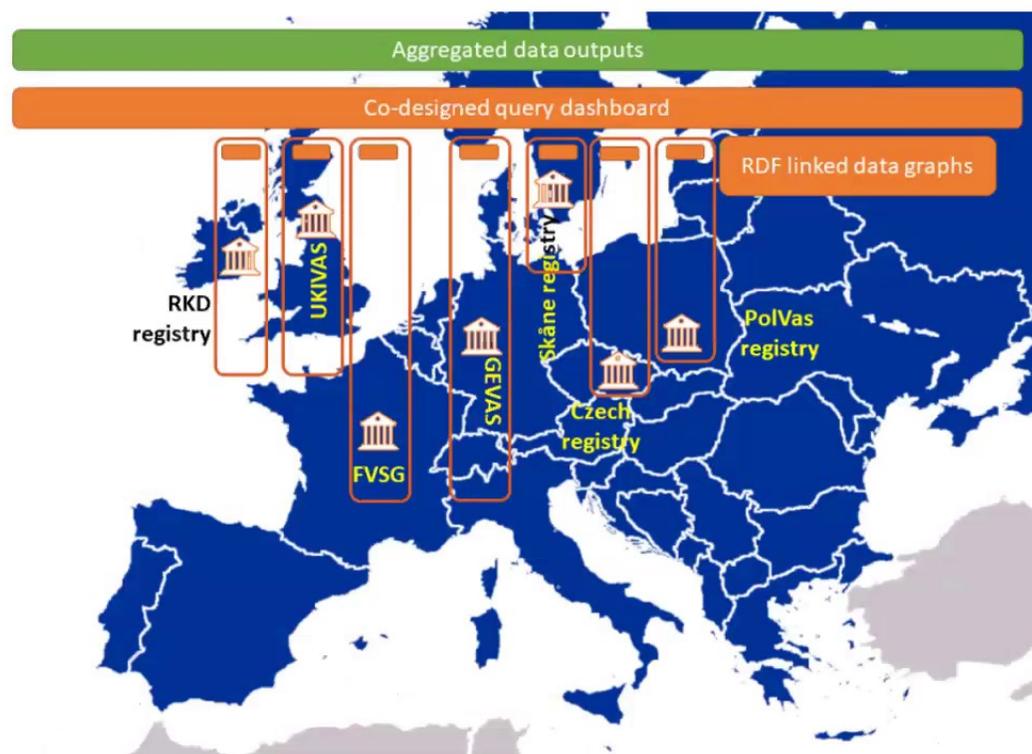


GeVas – Rekrutierung: Stand 10/22



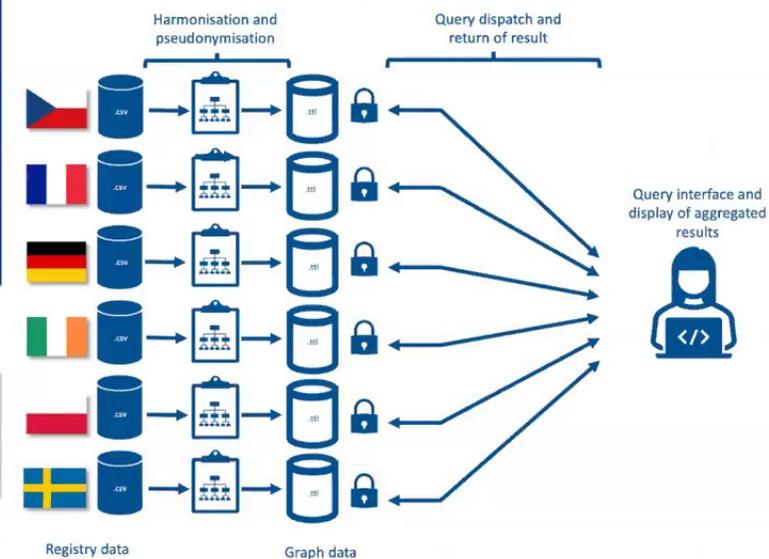
FAIRVASC

➤ EJP RD 2020 – 2023; PI Mark Little, Dublin



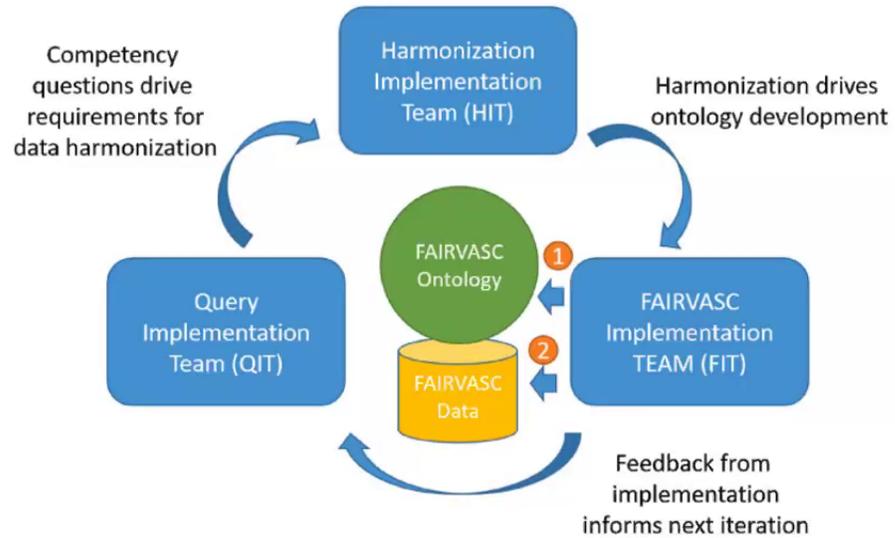
FAIRVASC – building registry interoperability to inform clinical care

FAIRification of national registries

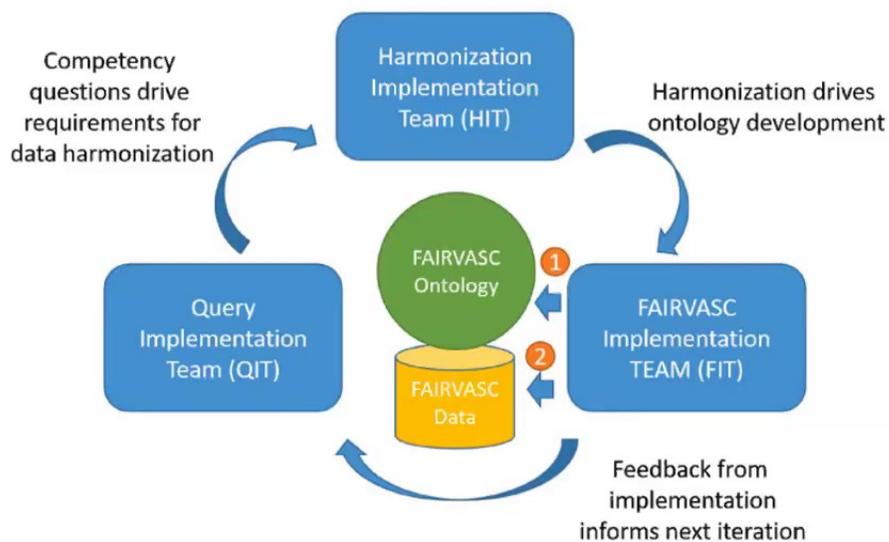


<https://fairvasc.eu/>

FAIRVASC



FAIRVASC



Normalization of terms across registries for complications which are infections. Green cells are those which are present in a registry.

Generic Normalised Term	No. of registries	Specific Normalised Term	No. of RKD registries	GeVas	Czech	Skane	PolVas	UKiVas	GVEF
Respiratory tract infection 275498002	5	Respiratory tract infection 275498002	1		Respiratory system and lungs				
Respiratory tract infection 275498002		Pneumonia 233604007	3	Pneumonia (233604007)		Pneumonia	NA		NA
Respiratory tract infection 275498002		Lower respiratory tract infection 50417007	1					AE Lower-RespInfect	
Respiratory tract infection 275498002	5	Upper respiratory infection 54150009	2	Other	Upper respiratory tract infection	Other	Other >type of inf other	NA	AE Upper-RespInfect
Respiratory tract infection 275498002		Pneumocystosis pneumonia 415125002	1	Other	NA	Other	Pneumocystis jirovecii infection	NA	AE Infection
Urinary tract infectious disease 68566005	5	Urinary tract infectious disease 68566005	2	Other	Urinary tract infection	Kidney and urinary tract	Other >type of inf other	NA	AE Infection
Urinary tract infectious disease 68566005		Pyelonephritis 45816000	3	Pyelonephritis (4581600)	Pyelonephritis		Pyelonephritis	NA	AE Infection

GeVas – Wie geht es weiter... & unmet needs

- **In Vorbereitung: Publikationen zu AAV- und RZA-Basisdaten**
- **eCRF V2.0 Update (ACR/EULAR 2022 Klassifikationskriterien,...)**
- **Datenmanagement & -qualität (→ Queries):**
 - Zentren: MFA**
 - ZKS Freiburg: Query-Mangement**
- **Finanzierung (Maintenance, Erweiterung Datenmanagement,...)**



GeVas – Zusammenfassung

- **Prospektives, webbasiertes, arztbasiertes, überregionales Register zur multizentrischen, standardisierten, systematischen und pseudonymisierten Erfassung des Langzeitverlaufs von Vaskulitiden**
- **FPI: 15.06.2021**
- **15.03.2023: n = 551**
- **Publikationen: Registerdeskription (Iking-Konert et al. BMC Rheumatology 2021), in Vorbereitung: AAV- und RZA-Basisdaten**
- **Internationale Beteiligung: FAIRVASC**

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Die Fachpersonen können bei Vifor Pharma Switzerland AG eine vollständige Kopie des zitierten Prüfungsberichts anfordern.

Gekürzte Verschreibungsinformationen

Deutschland

▼ Dieses Arzneimittel unterliegt einer zusätzlichen Überwachung. Dies ermöglicht eine schnelle Identifizierung neuer Sicherheitsdaten. Angehörige der Gesundheitsberufe werden gebeten, alle Verdachtsfälle von unerwünschten Wirkungen zu melden.

TAVNEOS® ▼ 10 mg Hartkapseln: **Wirkstoff:** Avacopan. **Zusammensetzung:** Jede Hartkapsel enthält 10 mg Avacopan. Sonstige Bestandteile mit bekannter Wirkung: 245 mg Macroglyglycerolhydroxystearat (Ph.Eur.). **Anwendungsgebiete:** Tavneos ist in Kombination mit einem Rituximab- oder Cyclophosphamid-Dosierungsschema indiziert zur Behandlung erwachsener Patienten mit schwerer aktiver Granulomatose mit Polyangiitis (GPA) oder mikroskopischer Polyangiitis (MPA). **Gegenanzeigen:** Überempfindlichkeit gegen den Wirkstoff oder einen der sonstigen Bestandteile. **Nebenwirkungen:** Sehr häufig: Übelkeit, Kopfschmerzen, erniedrigte Leukozytenzahl, Infektion der oberen Atemwege, Diarrhö, Erbrechen, Nasopharyngitis, erhöhte Werte in Leberfunktionstests. Häufig: Pneumonie, Rhinitis, Harnwegsinfektion, Sinusitis, Bronchitis, Gastroenteritis, Infektion der unteren Atemwege, Zellulitis, Herpes zoster, Influenza, Orale Candidose, Orale Herpes, Otitis media, Neutropenie, Schmerzen im Oberbauch, erhöhte Kreatinphosphokinase im Blut. Gelegentlich: Angioödem. **VERSCHREIBUNGSPFLICHTIG. Fachinformation beachten. Pharmazeutischer Unternehmer:** Vifor Fresenius Medical Care Renal Pharma France, 100-101 Terrasse Boieldieu, Tour Franklin La Défense 8, 92042 Paris La Défense Cedex, Frankreich. **Stand der Information:** Februar 2022

Österreich

▼ Dieses Arzneimittel unterliegt einer zusätzlichen Überwachung. Dies ermöglicht eine schnelle Identifizierung neuer Sicherheitsdaten. Angehörige der Gesundheitsberufe werden gebeten, alle Verdachtsfälle von unerwünschten Wirkungen zu melden.

Tavneos® Fachkurzinformation: Tavneos®10mg Hartkapsel. **Zusammensetzung:** Jede Hartkapsel enthält 10 mg Avacopan. Sonstige Bestandteile mit bekannter Wirkung: 245 mg Macroglyglycerolhydroxystearat (Ph.Eur.). **Anwendungsgebiete:** Tavneos® ist in Kombination mit einem Rituximab- oder Cyclophosphamid-Dosierungsschema indiziert zur Behandlung erwachsener Patienten mit schwerer aktiver Granulomatose mit Polyangiitis (GPA) oder mikroskopischer Polyangiitis (MPA). **Gegenanzeigen:** Überempfindlichkeit gegen den Wirkstoff oder einen der sonstigen Bestandteile. Pharmakotherapeutische Gruppe: L04AJ05 Complement Inhibitors **ATC-Code:** L04AJ05. **Inhaber der Zulassung:** Vifor France, 100-101 Terrasse Boieldieu Tour Franklin La Defense 8 92042 Paris La Defense Cedex, Frankreich. Rezept- und apothekenpflichtig. Weitere Angaben zu Warnhinweisen und Vorsichtsmaßnahmen für die Anwendung, Wechselwirkungen mit anderen Arzneimitteln oder sonstigen Wechselwirkungen, Schwangerschaft und Stillzeit und Nebenwirkungen sowie Gewöhnungseffekten sind der veröffentlichten Fachinformation zu entnehmen. Stand der Information: Letzter Stand Fachinformation

Schweiz

▼ Dieses Arzneimittel unterliegt einer zusätzlichen Überwachung. Für weitere Informationen, siehe Fachinformation TAVNEOS® auf www.swissmedicinfo.ch.

Tavneos®: **Z:** Avacopan. **I:** Tavneos, als ergänzende Therapie zu einer immunsuppressiven Standardbehandlung auf Basis von Rituximab oder Cyclophosphamid mit Glukokortikoiden, ist für die Behandlung erwachsener Patienten mit schwerer aktiver ANCA Vaskulitis (GPA/MPA) indiziert. **D:** Orale Einnahme morgens und abends 2x täglich 30 mg (3 Kapseln zu je 10 mg) mit Nahrung. **KI:** Überempfindlichkeit gegen den Wirkstoff oder einen der Hilfsstoffe. **VM:** Hepatotoxizität; Angioödem; Überwachung des Blutbildes (weisse Blutkörperchen); Schwere Infektionen; Reaktivierung des Hepatitis-B-Virus; Herzbeschwerden; Bösartige Tumore; Macroglycerinhydroxystearat. **S/S:** Eine Anwendung während der Schwangerschaft und bei Frauen im gebärfähigen Alter, die keine Verhütungsmethode anwenden, ist nicht empfohlen. Es ist nicht bekannt, ob Avacopan in die Muttermilch ausgeschieden wird. Der Nutzen des Stillens für das Kind sollte gegen den Nutzen der Behandlung für die Patientin abgewogen werden. **UW:** Sehr häufig: Infektion der oberen Atemwege, Nasopharyngitis; Kopfschmerzen; Erbrechen, Durchfall, Übelkeit; erhöhter Lebertest; verminderte Anzahl weisser Blutkörperchen. Häufig: Lungenentzündung, Infektion der unteren Atemwege, Influenza, Bronchitis, Zellulitis, Infektion der Harnwege, Herpes zoster, Sinusitis, orale Candidose, Herpes im Mundbereich, Otitis media, Rhinitis, Gastroenteritis; Neutropenie; Oberbauchschmerzen; Anstieg der Kreatinphosphokinase im Blut. Gelegentlich: Angioödeme. **IA:** Avacopan ist ein Substrat von CYP3A4. Die gleichzeitige Verabreichung von Induktoren oder Inhibitoren dieses Enzyms kann die Pharmakokinetik von Avacopan beeinflussen. Siehe Fachinformation. **P:** Tavneos 10 mg: 30 und 180 Hartkapseln. **Liste B.** Detaillierte Informationen: www.swissmedicinfo.ch. Stand der Information: September 2022. **Zulassungsinhaber:** Vifor Fresenius Medical Care Renal Pharma Ltd., St. Gallen. **Vertrieb:** Vifor Pharma Switzerland AG, CH-1752 Villars-sur-Glâne | CH-AVA-2300011