ERKNet REG Registry for Rare Kidney Diseases (1.01.2018-31.12.2020)

UMOWA GRANTOWA 777304

Grant z Trzeciego Programu Zdrowia Unii Europejskiej na lata 2014-2020

Aleksandra Żurowska Katedra i Klinika Pediatrii, Nefrologii i Nadciśnienia , Gdański Uniwersytet Medyczny

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Czym są Europejskie Sieci referencyjne?

Rzadkie choroby
to choroby, które dotykają
niewielką liczbę osób w
porównaniu do całej populacji.
W Europie choroba uznawana
jest za rzadką, jeśli dotyka
1 osobę na 2000.

Europejskie Sieci Referencyjne służą gromadzeniu oraz rozpowszechnianiu informacji ***o rzadkich chorobach***
aby zapewnić możliwie najlepszą opiekę dla chorych w całej Unii Europejskiej.
Najlepsze ośrodki europejskie są wyłaniane w drodze konkursu do prowadzenia wspólnej działalności.

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ERKNET: EUROPEJSKA SIEC REFERENCYJNA DLA RZADKICH CHORÓB NEREK

- ERKNet jest jednym z 24 Europejskich Sieci referencyjnych (tzwn. ERNów) ERNy sa współwinansowane przez Komisję Europejską.

http://ec.europa.eu/health/ern

ERKNet jest Europejską Siecią Referencyjna dla rzadkich chorób nerek . Łączy 38 ośrodków eksperckich, zajmujących się ponad 40 000 dorosłymi oraz dziećmi z rzadkimi chorobami nerek.

W pierwszym konkursie kwalifikującym w 2018r dla ośrodków nefrologicznych do sieci europejskiej

z Polski zakwalifikował się jedynie ośrodek nefrologii dziecięcej (Prof. Aleksandra Żurowska) oraz nefrologii dla dorosłych (Prof. Alicja Dębska- Ślizień).

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ERKNet

The European Rare Kidney Disease Reference Network

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Co-funded by the European Union

About us | Our Experts | Disease Information | Virtual Consultation | Guidelines & Pathways | Databases | Education & Training | Research | News | Events | Patient website | The ERNs

ERKReg

The European Rare Kidney Disease Registry

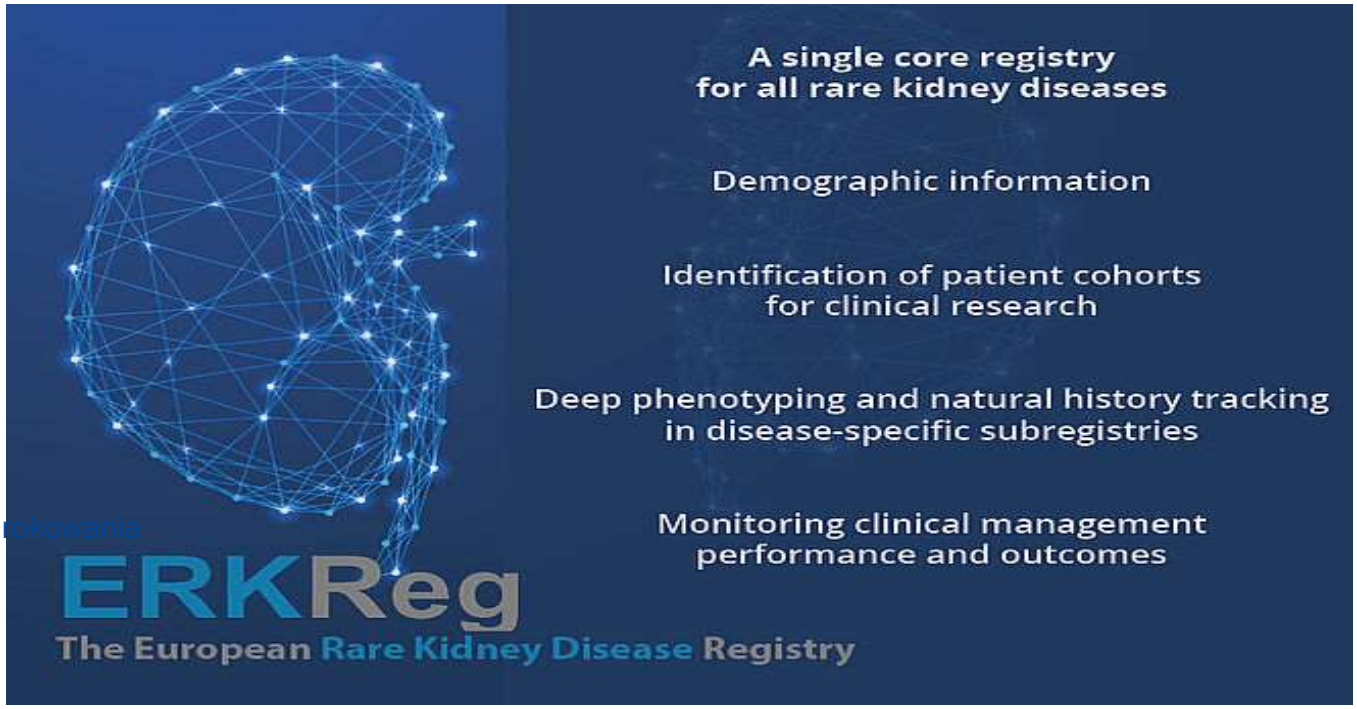
Jeden wspólny europejski rejestr dla rzadkich chorób nerek

Informacje demograficzne

Identyfikacja grup chorych do nowych terapii

Opisywanie historii naturalnej

Monitorowanie jakości prowadzenia chorych i ich odległego rokowania



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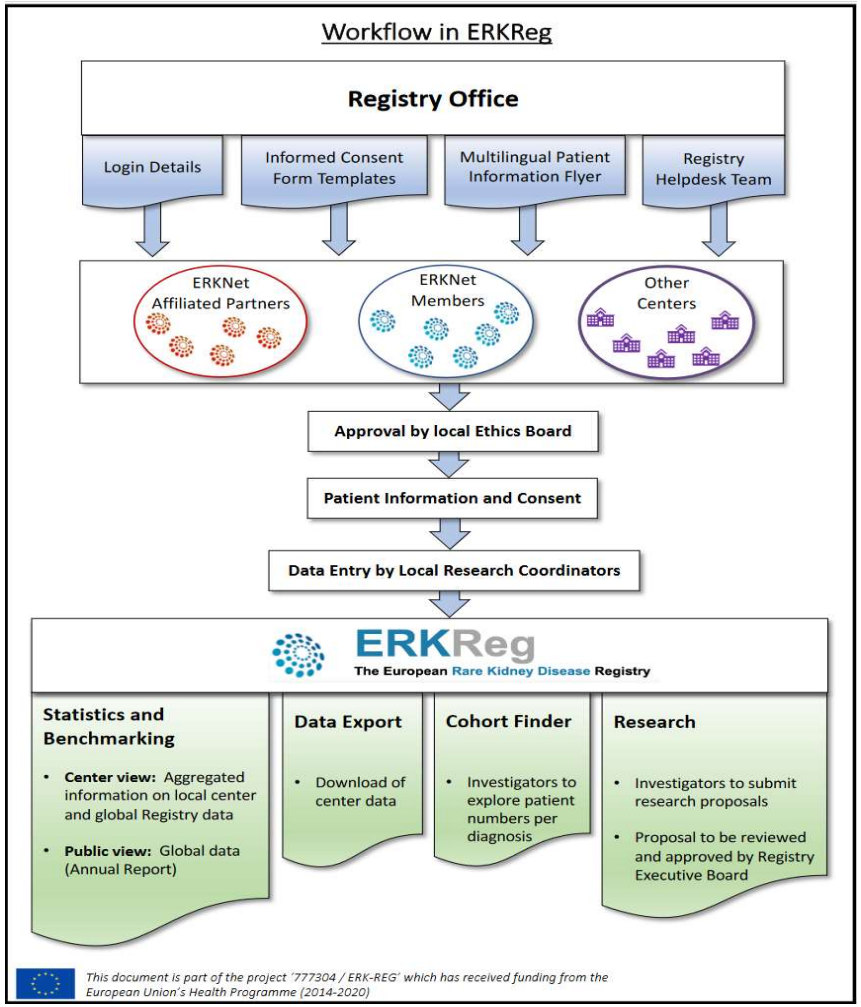
ORGANIZACJA REJESTRU



Co-financed by the Connecting Europe Facility of the European Union

ERKNet Helpdesk Team

ERKNet has established an operational **CPMS and Registry Helpdesk Team** to cover the different regions and languages of the Network. We are providing local assistance to all members, affiliated partners, and guest users in accessing and using both systems.

Check the table below and find your regional Helpdesk contact!

Name	HCP	Position	E-Mail	Regional contact for
Giulia Bassanese	Heidelberg University Hospital, Germany	MD, trainee in pediatrics	giulia.bassanese[at]med.uni-heidelberg.de	Italy
Tanja Wlodkowski	Heidelberg University Hospital, Germany	PhD, ERKNet Project Manager	tanja.wlodkowski[at]med.uni-heidelberg.de	Germany, UK, Affiliated Partners, Guests
Magdalena Drozynska-Duklas	Gdansk University Clinical Center, Poland	MD, PhD, pediatric nephrologist	magdalena.drozynska-duklas[at]gumed.edu.pl	Poland, Czech Republic, Sweden, Finland, Lithuania
Ilse Rood	Radbboud UMC Nijmegen, The Netherlands	MD, fellow nephrology	ilse.rood[at]radboudumc.nl	The Netherlands, Belgium
Victor Perez Beltran	Barcelona University Hospital Vall d`Hebron, Spain	MD, pediatric nephrologist	victorpbeltran[at]gmail.com	Spain, France

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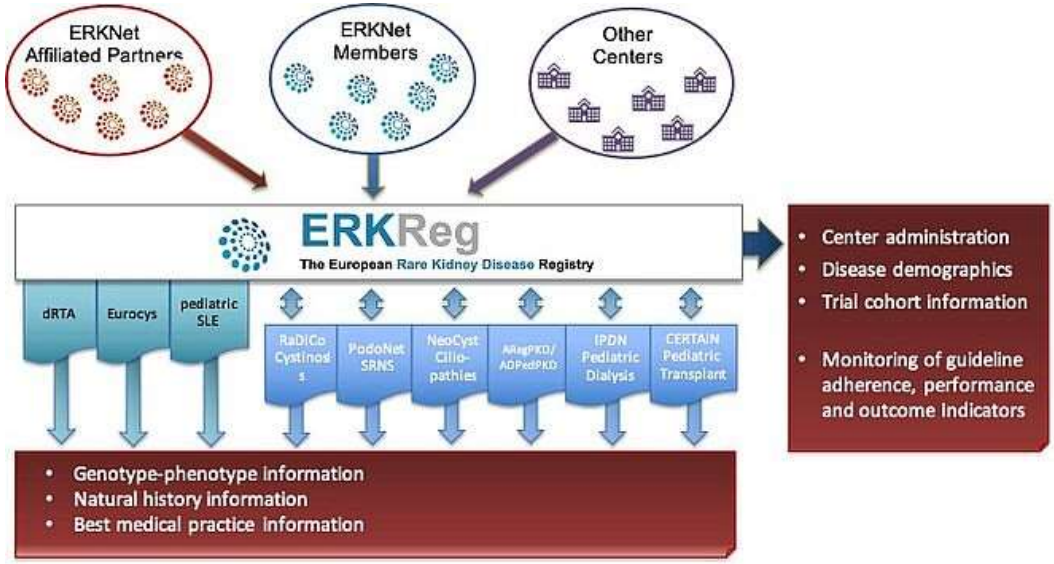
ORGANIZACJA REJESTRU EUROPEJSKIEGO

Current Number of Patients enrolled in the ERKNet-Registry

	All ERKNet centers					
	pediatric		adult		total	
	total	active	total	active	total	active
Glomerulopathies	2257	2071	2154	1392	4411	3463
Tubulopathies	791	690	307	205	1098	895
Metabolic nephropathies	305	279	113	86	418	365
Thrombotic microangiopathies	387	313	74	37	461	350
CAKUT	2591	2341	231	128	2822	2469
AD structural diseases	575	495	1432	967	2007	1462
Recessive cystic nephropathies	399	378	96	63	495	441
Rare causes of hypertension	96	93	6	4	102	97
TOTAL	**7401**	**6660**	**4413**	**2882**	**11814**	**9542**
Pediatric CKD and dialysis	6399	6062				
Pediatric transplantation	863	839				



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ERKNET REG : REJESTR DLA RZADKICH CHORÓB NEREK

W ciągu 24 miesięcy od powstania do Rejestru wpisano prawie 10 000 chorych z 45 nefrologicznych ośrodków pediatrycznych oraz 12 dla dorosłych z 21 krajów.

Ostateczne rozpoznanie miało 97.1% chorych w chwili wpisania.

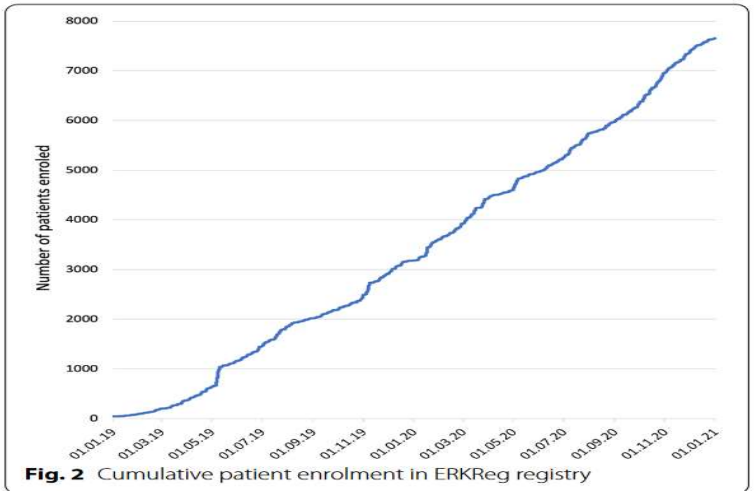

Fig. 2 Cumulative patient enrolment in ERKReg registry

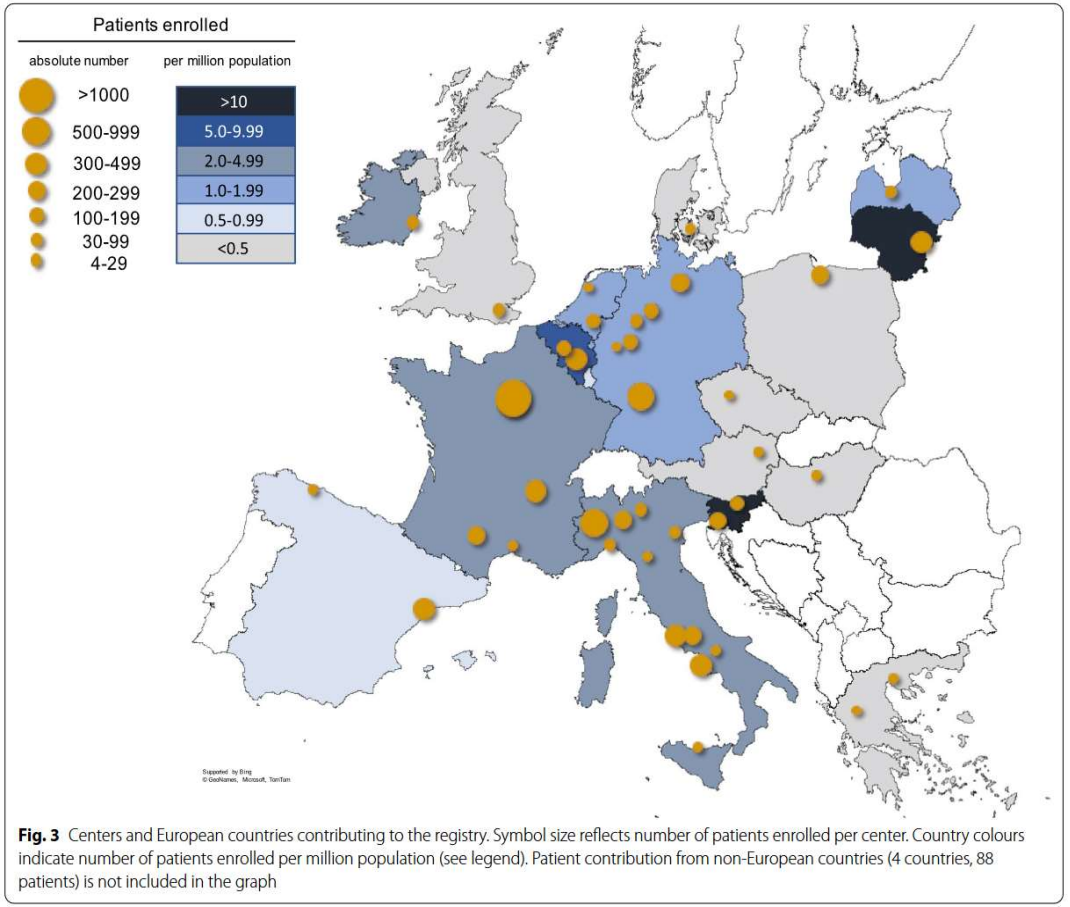

Fig. 3 Centers and European countries contributing to the registry. Symbol size reflects number of patients enrolled per center. Country colours indicate number of patients enrolled per million population (see legend). Patient contribution from non-European countries (4 countries, 88 patients) is not included in the graph

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DZIECI I DOROŚLI OPISYWANI W REJESTRZE

Bassanese et al. Orphanet J Rare Dis (2021) 16:251
https://doi.org/10.1186/s13023-021-01872-8

Orphanet Journal of Rare Diseases

RESEARCH Open Access

The European Rare Kidney Disease Registry (ERKReg): objectives, design and initial results

Giulia Bassanese[1†], Tanja Wlodkowski[1†], Aude Servais[2], Laurence Heidet[3], Dario Roccatello[4], Francesco Emma[5], Elena Levtchenko[6], Gema Ariceta[7], Justine Bacchetta[8], Giovambattista Capasso[9], Augustina Jankauskiene[10], Marius Miglinas[11], Pietro Manuel Ferraro[12], Giovanni Montini[13,14], Jun Oh[15], Stephane Decramer[16], Tanja Kersnik Levart[17], Jack Wetzels[18], Elisabeth Cornelissen[19], Olivier Devuyst[20,21], Aleksandra Zurowska[22], Lars Pape[23], Anja Buescher[23], Dieter Haffner[24], Natasa Marcun Varda[25], Gian Marco Ghiggeri[26], Giuseppe Remuzzi[27], Martin Konrad[28], Germana Longo[29], Detlef Bockenhauer[30], Atif Awan[31], Ilze Andersone[32], Jaap W. Groothoff[33] and Franz Schaefer[1*]

Abstract

Background: The European Rare Kidney Disease Reference Network (ERKNet) recently established ERKReg, a Web-based registry for all patients with rare kidney diseases. The main objectives of this core registry are to generate epidemiological information, identify current patient cohort for clinical research, explore diagnostic and therapeutic management practices, and monitor treatment performance and patient's outcomes. The registry has a modular design that allows to integrate comprehensive disease-specific registries as extensions to the core database. The diagnosis (Orphacode) and diagnostic information (clinical, imaging, histopathological, biochemical, immunological and genetic) are recorded. Anthropometric, kidney function, and disease-specific management and outcome items informing a set of 61 key performance indicators (KPIs) are obtained annually. Data quality is ensured by automated plausibility checks upon data entry and regular offline database checks prompting queries. Centre KPI statistics and benchmarking are calculated automatically.

Results: Within the first 24 months since its launch, 7607 patients were enrolled to the registry at 45 pediatric and 12 specialized adult nephrology units from 21 countries. A kidney disease diagnosis had been established in 97.1% of these patients at time of enrolment. While 199 individual disease entities were reported by Orphacode, 50% of the cohort could be classified with 11, 80% with 43 and 95% with 92 codes. Two kidney diagnoses were assigned in 6.5% of patients; 5.9% suffered from syndromic disease. Whereas glomerulopathies (54.8%) and ciliopathies including autosomal dominant polycystic kidney disease (ADPKD) (31.5%) were the predominant disease groups among adults, the pediatric disease spectrum encompassed congenital anomalies of the kidney and urinary tract (CAKUT) (33.7%), glomerulopathies (30.7%), ciliopathies (14.0%), tubulopathies (9.2%), thrombotic microangiopathies (5.6%), and metabolic nephropathies (4.1%). Genetically confirmed diagnoses were reported in 24% of all pediatric and 12% adult patients, whereas glomerulopathies had been confirmed by kidney biopsy in 80.4% adult versus 38.5% pediatric glomerulopathy cases.

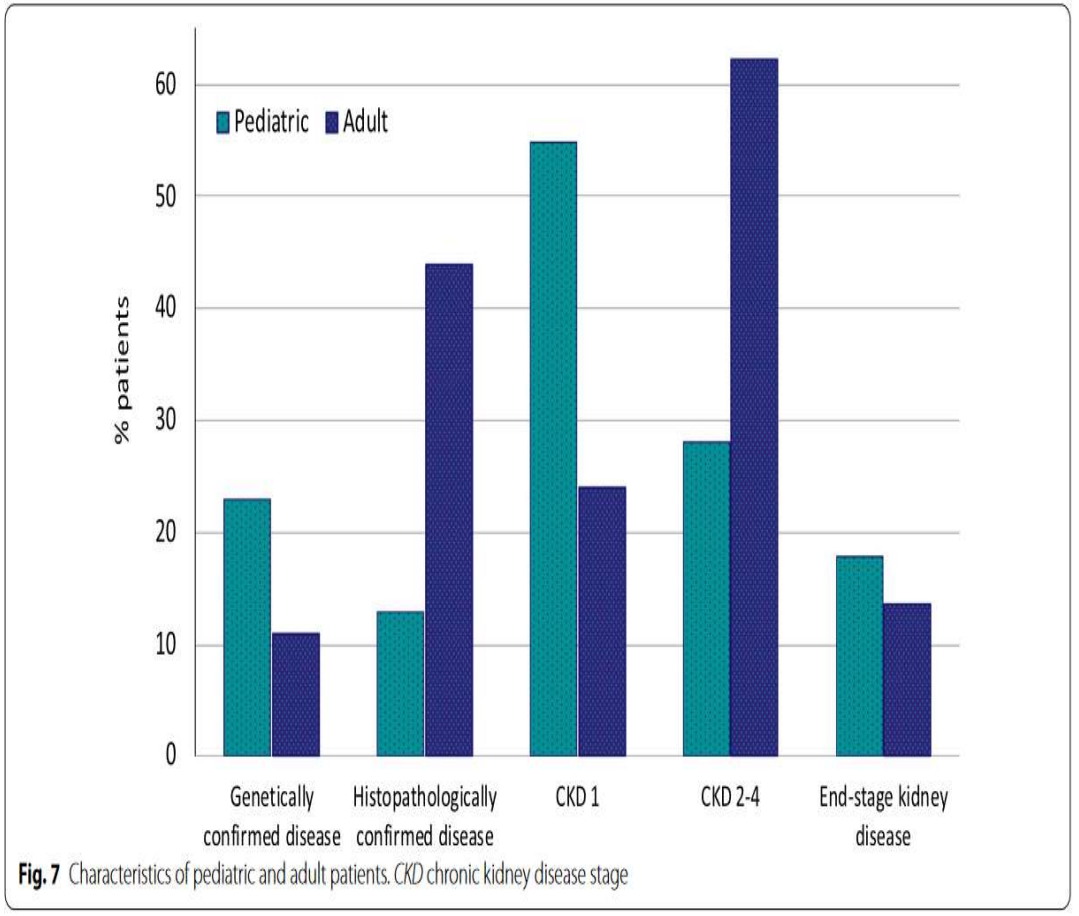

Fig. 7 Characteristics of pediatric and adult patients. *CKD* chronic kidney disease stage

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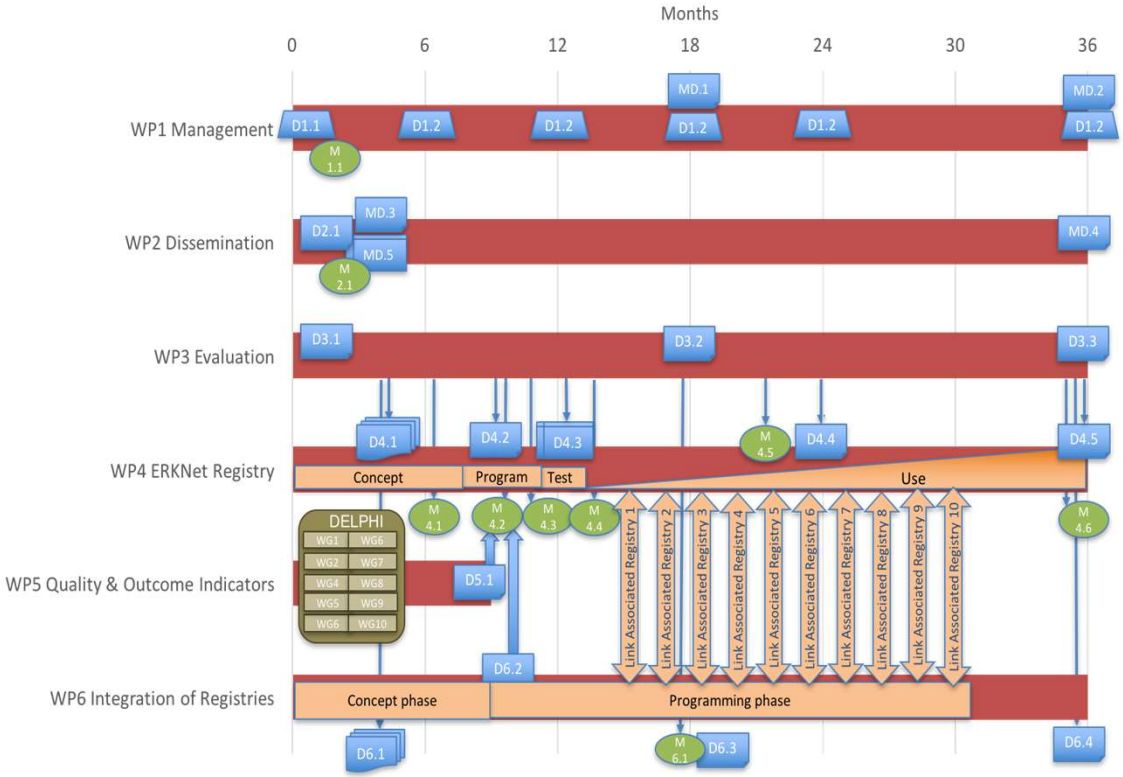
KPI MONITORING: SEMI-ANNUAL REPORTS

GRANT 5 OŚRODKÓW EUROPEJSKICH DLA PROWADZENIA REJESTRU
488 000 EU

Title of Proposal: The ERKNet Rare Kidney Disease Registry

List of applicants

Applicant No	Applicant organisation name	Country
1 (Coordinator)	University Medical Center Heidelberg (UKL-HD)	Germany
2	Medical University Gdansk, Poland (GUMed)	Poland
3	Amsterdam Medical Center, NL (AMC)	Netherlands
4	Hospital Universitario Vall d'Hebron, Barcelona, Spain (ICS-HUVH) and Fundació Hospital Universitari Vall d' Hebron - Institut de Recerca (VHIR)	Spain
5	INSERM - Hopital Necker, Paris, France (APHP)	France



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ROZLICZANIE GRANTU (PAN MATEUSZ KIRJAK GUMED)

WP No	Del Rel.	Del No	Title	Descrip							
WP2	D2.1	D3	Dissemination Plan	Detailed Project Diss							
WP2	D2.2	D16	Leaflet	A leaflet to promote the pro							
WP2	D2.4	D18	Website	Each project must have a de							
WP1	D1.1	D1	Minutes of the Kick-off M	Initial face-to-face meetin							
WP4	D4.2	D7	Informed Consent Form a	Informed Consent Form appr							
WP1	D1.2	D2	Minutes of project Meeti	Face-to-face meeting of pro							
WP6	D6.1	D12	Informed consent survey	Survey among all associatec							
WP3	D3.1	D4	Evaluation plan	Detailed Project Ev							
WP4	D4.3	D8	List of registry data item	Complete list of common an							
WP5	D5.1	D11	Lists of key quality and c	Validated consensus list of							
WP6	D6.4	D13	List of data sharing item	List of data available for sharing between asso...	AP-H	Report	Public	30 Sep 2018	15 Apr 2(	15 May 201	Approved
WP4	D4.4	D9	Web-based registry	web-based registry fully functional and online.	UKL-	Websit	Public	31 Dec 2018	30 Aug 2(	12 Sep 201	Approved
WP3	D3.2	D5	1st Evaluation Report	Detailed report on all evaluations performed in...	AMC	Report	Public	30 Jun 2019	27 Sep 2(	08 Oct 201	Approved
WP6	D6.2	D14	Report on informed cons	Report on informed consent harmonization activi...	AP-H	Report	Public	30 Jun 2019	27 Sep 2(	08 Oct 201	Approved
WP4	D4.1	D10	ERKNet Registry Annual F	Detailed report with demo-graphic figures and p...	UKL-	Report	Public	31 Dec 2019	26 Aug 2(	26 Aug 202	Approved
WP2	D2.3	D17	Layman report of the reg	This is a short (e.g. 10 pages) version of the ...	GUM	Report	Public	31 Dec 2020			Pending
WP3	D3.3	D6	2nd Evaluation Report	Detailed report on all evaluations performed in...	AMC	Report	Public	31 Dec 2020			Pending
WP6	D6.3	D15	Report on registry integr	Report on data exchange procedures established ...	AP-H	Report	Public	31 Dec 2020			Pending

Number	Name	Lead Beneficiary	Delivery Date (Annex I)	Achieved	Delivery Date (actual)
1	Project Office available	UKL-HD	28 Feb 2018	☑	01 Jan 2018
2	Dissemination channels work	GUMed	31 Mar 2018	☑	11 Mai 2018
3	Project approved by lead Eth	UKL-HD	31 Jul 2018	☑	07 Mär 2018
4	List data items	UKL-HD	30 Sep 2018	☑	30 Jun 2018
5	Test version	UKL-HD	30 Nov 2018	☑	30 Jun 2018
6	Testing completed	UKL-HD	31 Jan 2019	☑	28 Sep 2018
7	50% of ERKNet centers report	UKL-HD	30 Sep 2019	☑	16 Mai 2019
8	90% of ERKNet centers report	UKL-HD	31 Dec 2020	☐	
9	Delphi processes successfully	ICS-HUVH	30 Sep 2018	☑	31 Jul 2018
10	All Informed consents adapte	AP-HP	30 Jun 2019	☐	

Open Tasks	Work Package / Beneficiary
Layman report of registry acitvities	WP2 / GuMed
2nd evaluation report (months 18-36)	WP3 / AMC
Report on registry integration status	WP6 / AP-HP

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