

## **Online Data Supplement**

### **Accuracy of Nasal Nitric Oxide Measurement as a Diagnostic Test for Primary Ciliary Dyskinesia: A Systematic Review and Meta-Analysis**

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**Appendix 1:** Initial search strategy for PCD articles

**Appendix 2 :** PRISMA 2009 checklist

**Appendix 3:** Fagan normogram - Assuming a 35% pre-test probability (in blue, based upon prevalence data in several large PCD cohort studies (17, 18, 29, 31, 33)) and a 10% pre-test probability (in red) with the corresponding post-test probabilities for a pooled PLR of 24.3 and NLR of 0.03.

## Supplementary Material

### Appendix 1: Initial search strategy for PCD articles

The following databases were searched for relevant studies from inception until July 28, 2016: MEDLINE (Ovid), PubMed (NLM), Embase Classic + Embase (Ovid), AMED (Ovid), Africa-Wide Information (Ebsco), BIOSIS (Ovid), Cochrane (Wiley), Global Health (Ovid), Scopus (Elsevier), and Web of Science (ThomsonReuters).

The search strategy uses variations of Primary Ciliary Dyskinisia terms (*ranging from PCD, to ciliastatis, kartagener syndrome, etc.*) found within the Subject Headings as well as text words found within the titles, abstracts and keyword fields. The full MEDLINE strategy below was applied to all databases, with modifications to search terms as necessary.

### Medline Search strategy

#### Medline [Ovid] (July 28, 2016)

#### Ovid MEDLINE(R) In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

1	exp Ciliary Motility Disorders/	2039
2	((cili* or cilli*) adj3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*)).tw,kf.	2373
3	(ciliostasis* or ciliastasis* or ciliestasis or ((cili* or cilli*) adj stasis*)).tw,kf.	149
4	(PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil* ).tw,kf.	213069
5	Cilia/ or exp Nasal Mucosa/	32924
6	limit 5 to yr="1966 - 1983"	6923
7	4 and 6	260
8	1 or 2 or 3 or 7	3614
9	exp Kartagener Syndrome/	1204
10	((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) adj4 (disorder* or syndrom* or diseas* or triad*)).tw,kf.	829
11	((bronchiectas* or bronchoectas*) adj4 (polynesian* or dextrocardia* or dextro-cardia*)).tw,kf.	27
12	exp Bronchiectasis/ and exp Dextrocardia/	1217
13	Afzelius*.tw,kf.	40
14	exp Situs Inversus/ and exp Cilia/	380
15	((situs* adj3 (invers* or ambig* or total*)) and (cili* or cilli*)).tw,kf.	384
16	9 or 10 or 11 or 12 or 13 or 14 or 15	1553
17	8 or 16	3778
18	Animals/ not (Animals/ and Humans/)	4249555
19	17 not 18	3035
20	remove duplicates from 19	2951

Databases Searched

**Africa-Wide Information [EBSCO] (July 28, 2016)**

#	Query	Results
S10	S8 NOT S9	63
S9	<p>TI animals or animal or mice or mus or mouse or murine or woodmouse or rats or rat or murinae or muridae or cottonrat or cottonrats or hamster or hamsters or cricetinae or rodentia or rodent or rodents or pigs or pig or porcine or swine or swines or piglets or piglet or boar or boars or "sus scrofa" or ferrets or ferret or polecat or polecats or "mustela putorius" or "guinea pigs" or "guinea pig" or cavia or callithrix or marmoset or marmosets or cebuella or hapale or octodon or chinchilla or chinchillas or gerbillinae or gerbil or gerbils or jird or jirds or merione or meriones or rabbits or rabbit or hares or hare or diptera or flies or fly or dipteral or drosophila or drosophilidae or cats or cat or carus or felis or nematoda or nematode or nematoda or nematode or nematodes or sipunculida or dogs or dog or canine or canines or canis or sheep or sheeps or lamb or lambs or mouflon or mouflons or ovis or goats or goat or capra or capras or rupicapra or chamois or haplorhini or monkey or monkeys or macaque or macaques or primate or primates or anthropoidea or anthropoids or saguinus or tamarin or tamarins or leontopithecus or hominidae or ape or apes or pan or paniscus or "pan paniscus" or bonobo or bonobos or troglodytes or "pan troglodytes" or gibbon or gibbons or siamang or siamangs or nomascus or symphalangus or chimpanzee or chimpanzees or prosimians or "bush baby" or prosimian or "bush babies" or galagos or galago or pongidae or gorilla or gorillas or pongo or pygmaeus or "pongo pygmaeus" or orangutans or pygmaeus or lemur or lemurs or lemuridae or horse or horses or pongo or equus or cow or cows or calf or bull or chicken or chickens or gallus or quail or bird or birds or quails or poultry or poultries or fowl or fowls or reptile or reptilia or reptiles or snakes or snake or lizard or lizards or alligator or alligators or crocodile or crocodiles or turtle or turtles or amphibian or amphibians or amphibia or frog or frogs or bombina or salientia or toad or toads or "epidalea calamita" or salamander or salamanders or eel or eels or fish or fishes or pisces or catfish or catfishes or siluriformes or arius or heteropneustes or sheatfish or perch or perches or percidae or perca or trout or trouts or char or chars or salvelinus or "fathead minnow" or minnow or cyprinidae or carps or carp or zebrafish or zebrafishes or goldfish or goldfishes or guppy or guppies or chub or chubs or tinca or barbels or barbuis or pimphales or promelas or "poecilia reticulata" or mullet or mullets or seahorse or seahorses or "mugil curema" or "atlantic cod" or shark or sharks or catshark or anguilla or salmonid or salmonids or whitefish or whitefishes or salmon or salmons or sole or solea or "sea lamprey" or lamprey or lampreys or pumpkinseed or sunfish or sunfishes or tilapia or tilapias or turbot or turbots or flatfish or flatfishes or sciuridae or squirrel or squirrels or chipmunk or chipmunks or suslik or susliks or vole or voles or lemming or lemmings or muskrat or muskrats or lemmus or otter or otters or marten or martens or martes or weasel or badger or badgers or ermine or mink or minks or sable or sables or gulo or gulos or wolverine or wolverines or minks or mustela or llama or llamas or alpaca or alpacas or camelid or camelids or guanaco or guanacos or chiroptera or chiropteras or bat or bats or fox or foxes or iguana or iguanas or "xenopus laevis" or parakeet or parakeets or parrot or parrots or donkey or donkeys or mule or mules or zebra or zebras or shrew or shrews or bison or bisons or buffalo or buffaloes or deer or deers or bear or bears or panda or pandas or "wild hog" or "wild boar" or fitchew or fitch or beaver or beavers or jerboa or jerboas or capybara or capybaras or ovine or ewe* or chick* or slug or slugs or pigeon or pigeons or dalmation* or feline* or bovine</p>	467,979
S8	S1 OR S2 OR S3 OR S4 OR S5 OR S6 OR S7	86

S7	( TI ((situs* N3 (invers* or ambig* or total*)) and (cili* or cilli*)) ) OR ( AB ((situs* N3 (invers* or ambig* or total*)) and (cili* or cilli*)) ) OR ( KW ((situs* N3 (invers* or ambig* or total*)) and (cili* or cilli*)) ) )	8
S6	TI Afzelius* OR AB Afzelius* OR KW Afzelius*	16
S5	( TI ((bronchiectas* or bronchoectas*) N4 (polynesian* or dextrocardia* or dextro-cardia*)) ) OR ( AB ((bronchiectas* or bronchoectas*) N4 (polynesian* or dextrocardia* or dextro-cardia*)) ) OR ( KW ((bronchiectas* or bronchoectas*) N4 (polynesian* or dextrocardia* or dextro-cardia*)) ) )	2
S4	( TI ((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) N4 (disorder* or syndrom* or diseas* or triad*)) ) OR ( AB ((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) N4 (disorder* or syndrom* or diseas* or triad*)) ) OR ( KW ((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) N4 (disorder* or syndrom* or diseas* or triad*)) ) )	24
S3	( TI((cili* or cilli*) N0 stasis*)) ) OR ( AB((cili* or cilli*) N0 stasis*)) ) OR ( KW((cili* or cilli*) N0 stasis*)) )	1
S2	( TI( ciliostasis* or ciliastasis* or ciliestasis ) OR AB ( ciliostasis* or ciliastasis* or ciliestasis ) ) OR ( KW( ciliostasis* or ciliastasis* or ciliestasis ) )	3
S1	TI(((cili* or cilli*) N3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*))) OR AB(((cili* or cilli*) N3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*))) OR KW(((cili* or cilli*) N3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*)))	48

## AMED (Allied & Complementary Medicine) [Ovid] (July 28, 2016)

AMED (Allied and Complementary Medicine) 1985 to July 2016

1	((cili* or cilli*) adj3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*)).ti,ab,et.	2
2	(ciliostasis* or ciliastasis* or ciliestasis or ((cili* or cilli*) adj stasis*)).ti,ab,et.	0
3	((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) adj4 (disorder* or syndrom* or diseas* or triad*)).ti,ab,et.	1
4	((bronchiectas* or bronchoectas*) adj4 (polynesian* or dextrocardia* or dextro-cardia*)).ti,ab,et.	0
5	Afzelius*.ti,ab,et.	0
6	((situs* adj3 (invers* or ambig* or total*)) and (cili* or cilli*)).ti,ab,et.	0
7	1 or 2 or 3 or 4 or 5 or 6	3
8	Animals/ not (Animals/ and Humans/)	5801
9	7 not 8	3

## Biosis [Ovid] (July 28, 2016)

BIOSIS Previews 1969 to 2016 Week 35, BIOSIS Previews Archive 1926 to 1968

1	((cili* or cilli*) adj3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*)).ti,ab,mi.	2192
2	(ciliostasis* or ciliastasis* or ciliestasis or ((cili* or cilli*) adj stasis*)).ti,ab,mi.	153
3	((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) adj4 (disorder* or syndrom* or diseas* or triad*)).ti,ab,mi.	418
4	((bronchiectas* or bronchoectas*) adj4 (polynesian* or dextrocardia* or dextro-cardia*)).ti,ab,mi.	23

5	Afzelius*.ti,ab,mi.	67
6	((situs* adj3 (invers* or ambig* or total*)) and (cili* or cilli*)):ti,ab,mi.	275
7	(animal* not (animal* and hominidae)).st,tn.	8548116
8	1 or 2 or 3 or 4 or 5 or 6	2702
9	8 not 7	1676
10	remove duplicates from 9	1578

### Cochrane [Wiley] (July 28, 2016)

#1	((cili* or cilli*) near/3 (PCD or dyskin* or diskyn* or dys-kin* or dis-kin* or motil* or immotil* or immobil*)):ti,ab,kw	48
#2	(ciliostasis* or ciliastasis* or ciliestasis or ((cili* or cilli*) near/0 stasis*)):ti,ab,kw	0
#3	((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) near/4 (disorder* or syndrom* or diseas* or triad*)):ti,ab,kw	12
#4	((bronchiectas* or bronchoectas*) near/4 (polynesian* or dextrocardia* or dextrocardia*)):ti,ab,kw	0
#5	Afzelius*:ti,ab,kw	0
#6	((situs* near/3 (invers* or ambig* or total*)) and (cili* or cilli*)):ti,ab,kw	0
#7	#1 or #2 or #3 or #4 or #5 or #6	52
#8	(animals or animal or mice or mus or mouse or murine or woodmouse or rats or rat or murinae or muridae or cottonrat or cottonrats or hamster or hamsters or cricetinae or rodentia or rodent or rodents or pigs or pig or porcine or swine or swines or piglets or piglet or boar or boars or "sus scrofa" or ferrets or ferret or polecat or polecats or "mustela putorius" or "guinea pigs" or "guinea pig" or cavia or callithrix or marmoset or marmosets or cebuella or hapale or octodon or chinchilla or chinchillas or gerbillinae or gerbil or gerbils or jird or jirds or merione or meriones or rabbits or rabbit or hares or hare or diptera or flies or fly or dipteral or drosophila or drosophilidae or cats or cat or carus or felis or nematoda or nematode or nematoda or nematode or nematodes or sipunculida or dogs or dog or canine or canines or canis or sheep or sheeps or lamb or lambs or mouflon or mouflons or ovis or goats or goat or capra or capras or rupicapra or chamois or haplorhini or monkey or monkeys or macaque or macaques or primate or primates or anthropoidea or anthropoids or saguinus or tamarin or tamarins or leontopithecus or hominidae or ape or apes or pan or paniscus or "pan paniscus" or bonobo or bonobos or troglodytes or "pan troglodytes" or gibbon or gibbons or siamang or siamangs or nomascus or symphalangus or chimpanzee or chimpanzees or prosimians or "bush baby" or prosimian or "bush babies" or galagos or galago or pongidae or gorilla or gorillas or pongo or pygmaeus or "pongo pygmaeus" or orangutans or pygmaeus or lemur or lemurs or lemuridae or horse or horses or pongo or equus or equus or cow or cows or calf or bull or chicken or chickens or gallus or quail or bird or birds or quails or poultry or poultries or fowl or fowls or reptile or reptilia or reptiles or snakes or snake or lizard or lizards or alligator or alligators or crocodile or crocodiles or turtle or turtles or amphibian or amphibians or amphibia or frog or frogs or bombina or salientia or toad or toads or "epidalea calamita" or salamander or salamanders or eel or eels or fish or fishes or pisces or catfish or catfishes or siluriformes or arius or heteropneustes or sheatfish or perch or perches or percidae or perca or trout or trouts or char or chars or salvelinus or "fathead minnow" or minnow or cyprinidae or carps or carp or zebrafish or zebrafishes or goldfish or goldfishes or guppy or guppies or chub or chubs or tinca or barbels or barbus or pimephales or promelas or "poecilia reticulata" or mullet or mullets or seahorse or seahorses or "mugil curema" or "atlantic cod" or shark or sharks or catshark or anguilla or salmonid or salmonids or whitefish or whitefishes or salmon or salmons or	7146

	sole or solea or "sea lamprey" or lamprey or lampreys or pumpkinseed or sunfish or sunfishes or tilapia or tilapias or turbot or turbots or flatfish or flatfishes or sciuridae or squirrel or squirrels or chipmunk or chipmunks or suslik or susliks or vole or voles or lemming or lemmings or muskrat or muskrats or lemmus or otter or otters or marten or martens or martes or weasel or badger or badgers or ermine or mink or minks or sable or sables or gulo or gulos or wolverine or wolverines or minks or mustela or llama or llamas or alpaca or alpacas or camelid or camelids or guanaco or guanacos or chiroptera or chiropteras or bat or bats or fox or foxes or iguana or iguanas or "xenopus laevis" or parakeet or parakeets or parrot or parrots or donkey or donkeys or mule or mules or zebra or zebras or shrew or shrews or bison or bisons or buffalo or buffaloes or deer or deers or bear or bears or panda or pandas or "wild hog" or "wild boar" or fitchew or fitch or beaver or beavers or jerboa or jerboas or capybara or capybaras or ovine or ewe* or chick* or slug or slugs or pigeon or pigeons or dalmation* or feline* or bovine):ti	
#9	#7 not #8	51

### Embase [Ovid] (July 28, 2016)

Embase Classic+Embase 1947 to 2016 July 27

1	ciliary dyskinesia/	1903
2	((cili* or cilli*) adj3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*)).tw,kw.	3202
3	(ciliostasis* or ciliastasis* or ciliestasis or ((cili* or cilli*) adj stasis*)).tw,kw.	163
4	1 or 2 or 3	3842
5	Kartagener syndrome/	1383
6	((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) adj4 (disorder* or syndrom* or diseas* or triad*)).tw,kw.	1229
7	((bronchiectas* or bronchoectas*) adj4 (polynesian* or dextrocardia* or dextro-cardia*)).tw,kw.	43
8	bronchiectasis/ and dextrocardia/	120
9	Afzelius*.tw,kw.	54
10	situs inversus/ and eukaryotic flagellum/	110
11	((situs* adj3 (invers* or ambig* or total*)) and (cili* or cilli*)).tw,kw.	564
12	5 or 6 or 7 or 8 or 9 or 10 or 11	1970
13	4 or 12	5047
14	(exp animal/ or exp juvenile animal/ or adult animal/ or animal cell/ or animal experiment/ or animal model/ or animal tissue/ or nonhuman/) not human/	6491399
15	13 not 14	4082
16	remove duplicates from 15	3967

### Global Health [Ovid] (July 28, 2016)

Global Health 1973 to 2016 Week 28, Global Health Archive 1910 to 1972

1	((cili* or cilli*) adj3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*)).ti,ab,id.	94
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2	(ciliostasis* or ciliastasis* or ciliestasis or ((cili* or cilli*) adj stasis*)).ti,ab,id.	7
3	((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) adj4 (disorder* or syndrom* or diseas* or triad*)).ti,ab,id.	18
4	((bronchiectas* or bronchoectas*) adj4 (polynesian* or dextrocardia* or dextrocardia*)).ti,ab,id.	0
5	Afzelius*.ti,ab,id.	14
6	((situs* adj3 (invers* or ambig* or total*)) and (cili* or cilli*)).ti,ab,id.	14
7	Animals/ not (Animals/ and Humans/)	819009
8	1 or 2 or 3 or 4 or 5 or 6	123
9	8 not 7	93

### Medline [Ovid] (July 28, 2016)

Ovid MEDLINE(R) In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

1	exp Ciliary Motility Disorders/	2039
2	((cili* or cilli*) adj3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*)).tw,kf.	2373
3	(ciliostasis* or ciliastasis* or ciliestasis or ((cili* or cilli*) adj stasis*)).tw,kf.	149
4	(PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*).tw,kf.	213069
5	Cilia/ or exp Nasal Mucosa/	32924
6	limit 5 to yr="1966 - 1983"	6923
7	4 and 6	260
8	1 or 2 or 3 or 7	3614
9	exp Kartagener Syndrome/	1204
10	((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) adj4 (disorder* or syndrom* or diseas* or triad*)).tw,kf.	829
11	((bronchiectas* or bronchoectas*) adj4 (polynesian* or dextrocardia* or dextrocardia*)).tw,kf.	27
12	exp Bronchiectasis/ and exp Dextrocardia/	1217
13	Afzelius*.tw,kf.	40
14	exp Situs Inversus/ and exp Cilia/	380
15	((situs* adj3 (invers* or ambig* or total*)) and (cili* or cilli*)).tw,kf.	384
16	9 or 10 or 11 or 12 or 13 or 14 or 15	1553
17	8 or 16	3778
18	Animals/ not (Animals/ and Humans/)	4249555
19	17 not 18	3035
20	remove duplicates from 19	2951

PubMed [NLM] (July 28, 2016)

#24	#22 AND #23	52
#23	Publisher[sb] NOT pmcbook	493206
#22	#20 NOT #21	3552
#21	(animals[Title] OR animal[Title] OR mice[Title] OR mus[Title] OR mouse[Title] OR murine[Title] OR woodmouse[Title] OR rats[Title] OR rat[Title] OR murinae[Title] OR muridae[Title] OR cottonrat[Title] OR cottonrats[Title] OR hamster[Title] OR hamsters[Title] OR cricetinae[Title] OR rodentia[Title] OR rodent[Title] OR rodents[Title] OR pigs[Title] OR pig[Title] OR porcine[Title] OR swine[Title] OR swines[Title] OR piglets[Title] OR piglet[Title] OR boar[Title] OR boars[Title] OR "sus scrofa"[Title] OR ferrets[Title] OR ferret[Title] OR polecat[Title] OR polecats[Title] OR "mustela putorius"[Title] OR "guinea pigs"[Title] OR "guinea pig"[Title] OR cavia[Title] OR callithrix[Title] OR marmoset[Title] OR marmosets[Title] OR cebuella[Title] OR hapale[Title] OR octodon[Title] OR chinchilla[Title] OR chinchillas[Title] OR gerbillinae[Title] OR gerbil[Title] OR gerbils[Title] OR jird[Title] OR jirds[Title] OR merione[Title] OR meriones[Title] OR rabbits[Title] OR rabbit[Title] OR hares[Title] OR hare[Title] OR diptera[Title] OR flies[Title] OR fly[Title] OR dipteral[Title] OR drosophila[Title] OR drosophilidae[Title] OR cats[Title] OR cat[Title] OR carus[Title] OR felis[Title] OR nematoda[Title] OR nematode[Title] OR nematoda[Title] OR nematode[Title] OR nematodes[Title] OR sipunculida[Title] OR dogs[Title] OR dog[Title] OR canine[Title] OR canines[Title] OR canis[Title] OR sheep[Title] OR sheeps[Title] OR lamb[Title] OR lambs[Title] OR mouflon[Title] OR mouflons[Title] OR ovis[Title] OR goats[Title] OR goat[Title] OR capra[Title] OR capras[Title] OR rupicapra[Title] OR chamois[Title] OR haplorhini[Title] OR monkey[Title] OR monkeys[Title] OR macaque[Title] OR macaques[Title] OR primate[Title] OR primates[Title] OR anthropoidea[Title] OR anthropoids[Title] OR saguinus[Title] OR tamarin[Title] OR tamarins[Title] OR leontopithecus[Title] OR hominidae[Title] OR ape[Title] OR apes[Title] OR pan[Title] OR paniscus[Title] OR "pan paniscus"[Title] OR bonobo[Title] OR bonobos[Title] OR troglodytes[Title] OR "pan troglodytes"[Title] OR gibbon[Title] OR gibbons[Title] OR siamang[Title] OR siamangs[Title] OR nomascus[Title] OR symphalangus[Title] OR chimpanzee[Title] OR chimpanzees[Title] OR prosimians[Title] OR "bush baby"[Title] OR prosimian[Title] OR "bush babies"[Title] OR galago[Title] OR galago[Title] OR pongidae[Title] OR gorilla[Title] OR gorillas[Title] OR pongo[Title] OR pygmaeus[Title] OR "pongo pygmaeus"[Title] OR orangutans[Title] OR pygmaeus[Title] OR lemur[Title] OR lemurs[Title] OR lemuridae[Title] OR horse[Title] OR horses[Title] OR pongo[Title] OR equus[Title] OR cow[Title] OR cows[Title] OR calf[Title] OR bull[Title] OR chicken[Title] OR chickens[Title] OR gallus[Title] OR quail[Title] OR bird[Title] OR birds[Title] OR quails[Title] OR poultry[Title] OR poultries[Title] OR fowl[Title] OR fowls[Title] OR reptile[Title] OR reptilia[Title] OR reptiles[Title] OR snakes[Title] OR snake[Title] OR lizard[Title] OR lizards[Title] OR alligator[Title] OR alligators[Title] OR crocodile[Title] OR crocodiles[Title] OR turtle[Title] OR turtles[Title] OR amphibian[Title] OR amphibians[Title] OR amphibia[Title] OR frog[Title] OR frogs[Title] OR bombina[Title] OR salientia[Title] OR toad[Title] OR toads[Title] OR "epidalea calamita"[Title] OR salamander[Title] OR salamanders[Title] OR eel[Title] OR eels[Title] OR fish[Title] OR fishes[Title] OR pisces[Title] OR catfish[Title] OR catfishes[Title] OR siluriformes[Title] OR arius[Title] OR heteropneustes[Title] OR sheatfish[Title] OR perch[Title] OR perches[Title] OR percidae[Title] OR perca[Title] OR trout[Title] OR trouts[Title] OR char[Title] OR chars[Title] OR salvelinus[Title] OR "fathead minnow"[Title] OR minnow[Title] OR cyprinidae[Title] OR carps[Title] OR carp[Title] OR zebrafish[Title] OR zebrafishes[Title] OR goldfish[Title] OR goldfishes[Title] OR guppy[Title] OR guppies[Title] OR chub[Title] OR chubs[Title] OR tinca[Title] OR barbels[Title] OR barbus[Title] OR pimephales[Title] OR promelas[Title] OR "poecilia reticulata"[Title] OR mullet[Title] OR mullets[Title] OR seahorse[Title] OR seahorses[Title] OR "mugil	2482879



	curema[Title] OR "atlantic cod"[Title] OR shark[Title] OR sharks[Title] OR catshark[Title] OR anguilla[Title] OR salmonid[Title] OR salmonids[Title] OR whitefish[Title] OR whitefishes[Title] OR salmon[Title] OR salmon[salmon] OR sole[Title] OR solea[Title] OR "sea lamprey"[Title] OR lamprey[Title] OR lampreys[Title] OR pumpkinseed[Title] OR sunfish[Title] OR sunfishes[Title] OR tilapia[Title] OR tilapias[Title] OR turbot[Title] OR turbots[Title] OR flatfish[Title] OR flattfishes[Title] OR sciuridae[Title] OR squirrel[Title] OR squirrels[Title] OR chipmunk[Title] OR chipmunks[Title] OR suslik[Title] OR susliks[Title] OR vole[Title] OR voles[Title] OR lemming[Title] OR lemmings[Title] OR muskrat[Title] OR muskrats[Title] OR lemmus[Title] OR otter[Title] OR otters[Title] OR marten[Title] OR martens[Title] OR martes[Title] OR weasel[Title] OR badger[Title] OR badgers[Title] OR ermine[Title] OR mink[Title] OR minks[Title] OR sable[Title] OR sables[Title] OR gulo[Title] OR gulos[Title] OR wolverine[Title] OR wolverines[Title] OR minks[Title] OR mustela[Title] OR llama[Title] OR llamas[Title] OR alpaca[Title] OR alpacas[Title] OR camelid[Title] OR camelids[Title] OR guanaco[Title] OR guanacos[Title] OR chiroptera[Title] OR chiropteras[Title] OR bat[Title] OR bats[Title] OR fox[Title] OR foxes[Title] OR iguana[Title] OR iguanas[Title] OR "xenopus laevis"[Title] OR parakeet[Title] OR parakeets[Title] OR parrot[Title] OR parrots[Title] OR donkey[Title] OR donkeys[Title] OR mule[Title] OR mules[Title] OR zebra[Title] OR zebras[Title] OR shrew[Title] OR shrews[Title] OR bison[Title] OR bisons[Title] OR buffalo[Title] OR buffaloes[Title] OR deer[Title] OR deers[Title] OR bear[Title] OR bears[Title] OR panda[Title] OR pandas[Title] OR "wild hog"[Title] OR "wild boar"[Title] OR fitchew[Title] OR fitch[Title] OR beaver[Title] OR beavers[Title] OR jerboa[Title] OR jerboas[Title] OR capybara[Title] OR capybaras[Title] OR ovine[Title] OR ewe*[Title] OR chick*[Title] OR slug[Title] OR slugs[Title] OR pigeon[Title] OR pigeons[Title] OR dalmation*[Title] OR feline*[Title] OR bovine[Title])	
#20	#1 OR #2 OR #3 OR #4 OR #5 OR #6 OR #7 OR #8 OR #9 OR #10 OR #11 OR #12 OR #19	4013
#19	#15 and #18	388
#18	#16 OR #17	39562
#17	(cili*[Other Term] OR cilli*[Other Term])	2062
#16	(cili*[Title/Abstract] OR cilli*[Title/Abstract])	39562
#15	#13 OR #14	2651
#14	(situs*[Other Term]) AND (invers*[Other Term] OR ambig*[Other Term] OR total[Other Term])	389
#13	(situs*[Title/Abstract]) AND (invers*[Title/Abstract] OR ambig*[Title/Abstract] OR total[Title/Abstract])	2651
#12	Afzelius[Other Term]	0
#11	Afzelius[Title/Abstract]	40
#10	((bronchiectas*[Other Term] OR bronchoectas*[Other Term])) AND (polynesian*[Other Term] OR dextrocardia*[Other Term] OR dextro-cardia*[Other Term])	2
#9	((bronchiectas*[Title/Abstract] OR bronchoectas*[Title/Abstract])) AND (polynesian*[Title/Abstract] OR dextrocardia*[Title/Abstract] OR dextro-cardia*[Title/Abstract])	56
#8	((kartagener*[Other Term] OR cartagener*[Other Term] OR zivat*[Other Term] OR sivert*[Other Term] OR siewert*[Other Term] OR zivert*[Other Term])) AND (disorder*[Other Term] OR syndrom*[Other Term] OR diseas*[Other Term] OR triad*[Other Term])	166
#7	((kartagener*[Title/Abstract] OR cartagener*[Title/Abstract] OR zivat*[Title/Abstract] OR sivert*[Title/Abstract] OR siewert*[Title/Abstract] OR zivert*[Title/Abstract])) AND (disorder*[Title/Abstract] OR syndrom*[Title/Abstract] OR diseas*[Title/Abstract] OR triad*[Title/Abstract])	881
#6	(ciliary-stasis[Other Term]) OR ciliary-stasis[Other Term]	0

#5	(ciliary-stasis[Title/Abstract]) OR ciliary-stasis[Title/Abstract]	6
#4	(ciliostasis*[Other Term] OR ciliastasis*[Other Term] OR ciliestasis[Other Term])	1
#3	(ciliostasis*[Title/Abstract] OR ciliastasis*[Title/Abstract] OR ciliestasis[Title/Abstract])	143
#2	((cili*[Other Term] OR cilli*[Other Term])) AND (PCD[Other Term] OR dyskin*[Other Term] OR diskina*[Other Term] OR dys-kin*[Other Term] OR dis-kin*[Other Term] OR motil*[Other Term] OR immotil*[Other Term] OR immobil*[Other Term])	130
#1	((cili*[Title/Abstract] OR cilli*[Title/Abstract])) AND (PCD[Title/Abstract] OR dyskin*[Title/Abstract] OR diskina*[Title/Abstract] OR dys-kin*[Title/Abstract] OR dis-kin*[Title/Abstract] OR motil*[Title/Abstract] OR immotil*[Title/Abstract] OR immobil*[Title/Abstract])	3157

### Scopus [Elsevier] (July 28, 2016)

17	#15 NOT #16	814
16	PMID ( 0* OR 1* OR 2* OR 3* OR 4* OR 5* OR 6* OR 7* OR 8* OR 9* )	#####
15	#13 and not #14	3,252
14	TITLE(animals or animal or mice or mus or mouse or murine or woodmouse or rats or rat or murinae or muridae or cottonrat or cottonrats or hamster or hamsters or cricetinae or rodentia or rodent or rodents or pigs or pig or porcine or swine or swines or piglets or piglet or boar or boars or "sus scrofa" or ferrets or ferret or polecat or polecats or "mustela putorius" or "guinea pigs" or "guinea pig" or cavia or callithrix or marmoset or marmosets or cebuella or hapale or octodon or chinchilla or chinchillas or gerbillinae or gerbil or gerbils or jird or jirds or merione or meriones or rabbits or rabbit or hares or hare or diptera or flies or fly or dipteral or drosophila or drosophilidae or cats or cat or carus or felis or nematoda or nematode or nematoda or nematode or nematodes or sipunculida or dogs or dog or canine or canines or canis or sheep or sheeps or lamb or lambs or mouflon or mouflons or ovis or goats or goat or capra or capras or rupicapra or chamois or haplorhini or monkey or monkeys or macaque or macaques or primate or primates or anthropoidea or anthropoids or saguinus or tamarin or tamarins or leontopithecus or hominidae or ape or apes or pan or paniscus or "pan paniscus" or bonobo or bonobos or troglodytes or "pan troglodytes" or gibbon or gibbons or siamang or siamangs or nomascus or symphalangus or chimpanzee or chimpanzees or prosimians or "bush baby" or prosimian or "bush babies" or galagos or galago or pongidae or gorilla or gorillas or pongo or pygmaeus or "pongo pygmaeus" or orangutans or pygmaeus or lemur or lemurs or lemuridae or horse or horses or pongo or equus or cow or cows or calf or bull or chicken or chickens or gallus or quail or bird or birds or quails or poultry or poultries or fowl or fowls or reptile or reptilia or reptiles or snakes or snake or lizard or lizards or alligator or alligators or crocodile or crocodiles or turtle or turtles or amphibian or amphibians or amphibia or frog or frogs or bombina or salientia or toad or toads or "epidalea calamita" or salamander or salamanders or eel or eels or fish or fishes or pisces or catfish or catfishes or siluriformes or arius or heteropneustes or sheatfish or perch or perches or percidae or perca or trout or trouts or char or chars or salvelinus or "fathead minnow" or minnow or cyprinidae or carps or carp or zebrafish or zebrafishes or goldfish or goldfishes or guppy or guppies or chub or chubs or tinca or barbels or barbus or pimephales or promelas or "poecilia reticulata" or mullet or mullets or seahorse or seahorses or "mugil curema" or "atlantic cod" or shark or sharks or catshark or anguilla or salmonid or salmonids or whitefish or whitefishes or salmon or salmons or sole or solea or "sea lamprey" or lamprey or lampreys or pumpkinseed or sunfish or sunfishes or tilapia or tilapias or turbot or turbots or flatfish or flatfishes or sciuridae or squirrel or squirrels or chipmunk or chipmunks or suslik or susliks or vole or voles or lemming or	3,346,202

	lemmings or muskrat or muskrats or lemmus or otter or otters or marten or martens or martes or weasel or badger or badgers or ermine or mink or minks or sable or sables or gulo or gulos or wolverine or wolverines or minks or mustela or llama or llamas or alpaca or alpacas or camelid or camelids or guanaco or guanacos or chiroptera or chiropteras or bat or bats or fox or foxes or iguana or iguanas or "xenopus laevis" or parakeet or parakeets or parrot or parrots or donkey or donkeys or mule or mules or zebra or zebras or shrew or shrews or bison or bisons or buffalo or buffaloes or deer or deers or bear or bears or panda or pandas or "wild hog" or "wild boar" or fitchew or fitch or beaver or beavers or jerboa or jerboas or capybara or capybaras or ovine or ewe* or chick* or slug or slugs or pigeon or pigeons or dalmation* or feline* or bovine)	
13	#1 or #2 or #3 or #4 or #5 or #6 or #7 or #8 or #9 or #10 or #11 or #12	3,623
12	ABS((situs*) W/3 (invers* or ambig* or total*)) and ABS(cili* or cilli*)	436
11	TITLE((situs*) W/3 (invers* or ambig* or total*)) and TITLE(cili* or cilli*)	29
10	TITLE-ABS Afzelius*	0
9	ABS((bronchiectas* or bronchoectas*) W/4 (polynesian* or dextrocardia* or dextro-cardia*))	28
8	TITLE((bronchiectas* or bronchoectas*) W/4 (polynesian* or dextrocardia* or dextro-cardia*))	9
7	ABS((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) W/4 (disorder* or syndrom* or diseas* or triad*))	553
6	TITLE((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) W/4 (disorder* or syndrom* or diseas* or triad*))	692
5	ABS((cili* or cilli*) W/0 (stasis*))	10
4	TITLE((cili* or cilli*) W/0 (stasis*))	1
3	TITLE-ABS (ciliostasis* or ciliastasis* or ciliestasis)	166
2	ABS((cili* or cilli*) W/3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*))	2,439
1	TITLE((cili* or cilli*) W/3 (PCD or dyskin* or diskin* or dys-kin* or dis-kin* or motil* or immotil* or immobil*))	1,188

## Web of Science [Thomson Reuters] (July 28, 2016)

Indexes=SCI-EXPANDED, SSCI, A&HCI, CPCI-S, CPCI-SSH Timespan=All years

# 9	#7 not #8	3,039
# 8	Tl=(animals or animal or mice or mus or mouse or murine or woodmouse or rats or rat or murinae or muridae or cottonrat or cottonrats or hamster or hamsters or cricetinae or rodentia or rodent or rodents or pigs or pig or porcine or swine or swines or piglets or piglet or boar or boars or "sus scrofa" or ferrets or ferret or polecat or polecats or "mustela putorius" or "guinea pigs" or "guinea pig" or cavia or callithrix or marmoset or marmosets or cebuella or hapale or octodon or chinchilla or chinchillas or gerbillinae or gerbil or gerbils or jird or jirds or merione or meriones or rabbits or rabbit or hares or hare or diptera or flies or fly or dipteral or drosophila or drosophilidae or cats or cat or carus or felis or nematoda or nematode or nematoda or nematode or nematodes or sipunculida or dogs or dog or canine or canines or canis or sheep or sheeps or lamb or lambs or mouflon or mouflons or ovis or goats or goat or capra or capras or rupicapra or chamois or haplorhini or monkey or monkeys or macaque or macaques or primate or primates or anthropoidea or anthropoids or saguinus or tamarin or tamarins or leontopithecus or hominidae or ape or apes or pan or paniscus or "pan paniscus" or bonobo or bonobos or troglodytes or "pan troglodytes" or gibbon or gibbons or siamang or siamangs or nomascus or symphalangus or chimpanzee or chimpanzees or prosimians or "bush	3,820,570

	<p>baby" or prosimian or "bush babies" or galagos or galago or pongidae or gorilla or gorillas or pongo or pygmaeus or "pongo pygmaeus" or orangutans or pygmaeus or lemur or lemurs or lemuridae or horse or horses or pongo or equus or cow or cows or calf or bull or chicken or chickens or gallus or quail or bird or birds or quails or poultry or poultries or fowl or fowls or reptile or reptilia or reptiles or snakes or snake or lizard or lizards or alligator or alligators or crocodile or crocodiles or turtle or turtles or amphibian or amphibians or amphibia or frog or frogs or bombina or salientia or toad or toads or "epidalea calamita" or salamander or salamanders or eel or eels or fish or fishes or pisces or catfish or catfishes or siluriformes or arius or heteropneustes or sheatfish or perch or perches or percidae or perca or trout or trouts or char or chars or salvelinus or "fathead minnow" or minnow or cyprinidae or carps or carp or zebrafish or zebrafishes or goldfish or goldfishes or guppy or guppies or chub or chubs or tinca or barbels or barbuis or pimephales or promelas or "poecilia reticulata" or mullet or mullets or seahorse or seahorses or "mugil curema" or "atlantic cod" or shark or sharks or catshark or anguilla or salmonid or salmonids or whitefish or whitefishes or salmon or salmons or sole or solea or "sea lamprey" or lamprey or lampreys or pumpkinseed or sunfish or sunfishes or tilapia or tilapias or turbot or turbots or flatfish or flatfishes or sciuridae or squirrel or squirrels or chipmunk or chipmunks or suslik or susliks or vole or voles or lemming or lemmings or muskrat or muskrats or lemmus or otter or otters or marten or martens or martes or weasel or badger or badgers or ermine or mink or minks or sable or sables or gulo or gulos or wolverine or wolverines or minks or mustela or llama or llamas or alpaca or alpacas or camelid or camelids or guanaco or guanacos or chiroptera or chiropteras or bat or bats or fox or foxes or iguana or iguanas or "xenopus laevis" or parakeet or parakeets or parrot or parrots or donkey or donkeys or mule or mules or zebra or zebras or shrew or shrews or bison or bisons or buffalo or buffaloes or deer or deers or bear or bears or panda or pandas or "wild hog" or "wild boar" or fitchew or fitch or beaver or beavers or jerboa or jerboas or capybara or capybaras or ovine or ewe* or chick* or slug or slugs or pigeon or pigeons or dalmation* or feline* or bovine)</p>	
# 7	#1 or #2 or #3 or #4 or #5 or #6	3,440
# 6	TS=((situs* near/3 (invers* or ambig* or total*)) and (cili* or cilli*))	494
# 5	TS=Afzelius*	69
# 4	TS=((bronchiectas* or bronchoectas*) near/4 (polynesian* or dextrocardia* or dextrocardia*))	30
# 3	TS=((kartagener* or cartagener* or zivat* or sivert* or siewert* or zivert*) near/4 (disorder* or syndrom* or diseas* or triad*))	642
# 2	TS=(ciliostasis* or ciliastasis* or ciliestasis or ((cili* or cilli*) near/0 stasis*))	83
# 1	TS=((cili* or cilli*) near/3 (PCD or dyskin* or diskina* or dys-kin* or dis-kin* or motil* or immotil* or immobil*))	2,854

## Appendix 2 : PRISMA 2009 checklist

Section/topic	#	Checklist item	Reported on page #
<b>TITLE</b>			
Title	1	Identify the report as a systematic review, meta-analysis, or both.	Ok
<b>ABSTRACT</b>			
Structured summary	2	Provide a structured summary including, as applicable: background; objectives; data sources; study eligibility criteria, participants, and interventions; study appraisal and synthesis methods; results; limitations; conclusions and implications of key findings; systematic review registration number.	Ok
<b>INTRODUCTION</b>			
Rationale	3	Describe the rationale for the review in the context of what is already known.	Ok
Objectives	4	Provide an explicit statement of questions being addressed with reference to participants, interventions, comparisons, outcomes, and study design (PICOS).	Ok
<b>METHODS</b>			
Protocol and registration	5	Indicate if a review protocol exists, if and where it can be accessed (e.g., Web address), and, if available, provide registration information including registration number.	NA
Eligibility criteria	6	Specify study characteristics (e.g., PICOS, length of follow-up) and report characteristics (e.g., years considered, language, publication status) used as criteria for eligibility, giving rationale.	Ok
Information sources	7	Describe all information sources (e.g., databases with dates of coverage, contact with study authors to identify additional studies) in the search and date last searched.	Ok
Search	8	Present full electronic search strategy for at least one database, including any limits used, such that it could be repeated.	Ok
Study selection	9	State the process for selecting studies (i.e., screening, eligibility, included in systematic review, and, if applicable, included in the meta-analysis).	Ok
Data collection process	10	Describe method of data extraction from reports (e.g., piloted forms, independently, in duplicate) and any processes for obtaining and confirming data from investigators.	Ok
Data items	11	List and define all variables for which data were sought (e.g., PICOS, funding sources) and any assumptions and simplifications made.	Ok
Risk of bias in individual studies	12	Describe methods used for assessing risk of bias of individual studies (including specification of whether this was done at the study or outcome level), and how this information is to be used in any data synthesis.	Ok
Summary measures	13	State the principal summary measures (e.g., risk ratio, difference in means).	Ok

Synthesis of results	14	Describe the methods of handling data and combining results of studies, if done, including measures of consistency (e.g., $I^2$ ) for each meta-analysis.	Ok
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Section/topic	#	Checklist item	Reported on page #
Risk of bias across studies	15	Specify any assessment of risk of bias that may affect the cumulative evidence (e.g., publication bias, selective reporting within studies).	Ok
Additional analyses	16	Describe methods of additional analyses (e.g., sensitivity or subgroup analyses, meta-regression), if done, indicating which were pre-specified.	Ok
<b>RESULTS</b>			
Study selection	17	Give numbers of studies screened, assessed for eligibility, and included in the review, with reasons for exclusions at each stage, ideally with a flow diagram.	Ok
Study characteristics	18	For each study, present characteristics for which data were extracted (e.g., study size, PICOS, follow-up period) and provide the citations.	Ok
Risk of bias within studies	19	Present data on risk of bias of each study and, if available, any outcome level assessment (see item 12).	Ok
Results of individual studies	20	For all outcomes considered (benefits or harms), present, for each study: (a) simple summary data for each intervention group (b) effect estimates and confidence intervals, ideally with a forest plot.	Ok
Synthesis of results	21	Present results of each meta-analysis done, including confidence intervals and measures of consistency.	Ok
Risk of bias across studies	22	Present results of any assessment of risk of bias across studies (see Item 15).	Ok
Additional analysis	23	Give results of additional analyses, if done (e.g., sensitivity or subgroup analyses, meta-regression [see Item 16]).	Ok
<b>DISCUSSION</b>			
Summary of evidence	24	Summarize the main findings including the strength of evidence for each main outcome; consider their relevance to key groups (e.g., healthcare providers, users, and policy makers).	Ok
Limitations	25	Discuss limitations at study and outcome level (e.g., risk of bias), and at review-level (e.g., incomplete retrieval of identified research, reporting bias).	Ok
Conclusions	26	Provide a general interpretation of the results in the context of other evidence, and implications for future research.	Ok
<b>FUNDING</b>			
Funding	27	Describe sources of funding for the systematic review and other support (e.g., supply of data); role of funders for the systematic review.	Ok