An Investigation of Polycythemia Vera in the Tamaqua Area of Northeast Pennsylvania

#### **SUMMARY**

In 2004 four cases of a rare blood cancer called polycythemia vera (PV) were found in people who lived on one road near Tamaqua in Northeast Pennsylvania. This finding led the Pennsylvania Department of Health (PADOH) to review cases of cancer reported from the Tamaqua area to the state cancer registry. PADOH found that, although the overall rates of cancer in the Tamaqua area were the same as in other parts of the state, more PV cases than expected had occurred in the three counties around Tamaqua. In October 2006 PADOH asked the Agency for Toxic Substances and Disease Registry (ATSDR) to help study the patterns of PV in the three-county area. The goals of the ATSDR investigation were to find all residents of the three counties who were diagnosed with PV between 2001 and 2005, confirm the diagnosis of PV among these persons, and collect information from people found to have PV.

PV is a blood disease that occurs in about 1 out of every 100,000 people each year. Although PV is a cancer, it can be controlled with proper medical care. The cause of PV is not known. In 2004 a genetic marker called JAK2 was found in nearly all persons with PV. A test for JAK2 helps to confirm the diagnosis of PV and was used by ATSDR in its investigation.

The survey was conducted from December 12, 2006, to July 31, 2007. Of the 97 PV cases reported to the cancer registry, 38 patients agreed to participate. The remaining 59 individuals declined, were deceased, or could not be found. Another 24 people who were not in the registry were found in other ways. Thus, a total of 62 people agreed to be surveyed. Of these 62 people, 33 were confirmed to have PV, while 17 were found not to have this disease. The other 12 people did not have enough information in their medical records to determine whether or not they had PV. No differences that would suggest a cause for the disease were observed between the people found to have PV and those who did not.

Three areas in the region had more confirmed PV cases than would be expected. One area was near Pottsville, another was near Tamaqua, and the third was in eastern Carbon County. In two of these areas, the number of PV cases was very small. Only the area near Tamaqua had enough cases for the excess to be meaningful. Most of the people who were found not to have PV were from the Wilkes-Barre area. The survey did not find a link between the PV cases and reported chemical exposure at work or at home. ATSDR studied information on air, water, and soil to look for hazardous material sources located in all the highrate areas. No sources were common to all of the high-rate areas. Coal mining, waste-coal power plants, and Superfund sites were present in two of the high-rate areas. The investigation found no other connections besides location that link these sources to the PV cases.

The findings of this investigation are hard to interpret for several reasons:

PV rates from the investigation cannot be directly compared to standard PV incidence rates because different methods were used to count the cases.

- The PV diagnosis was confirmed using a new genetic test. This test was not used for most patients who were reported to the cancer registry or in other PV studies.
- A large number of persons reported to have PV in the state cancer registry did not take part in this study.
- Some areas may have had more people take part in the survey due to greater local interest in the study.
- The total number of PV cases in the three-county area is unknown.
- Zip code and census tract rates for rare diseases such as PV can be misleading when small numbers of cases are involved.

Although three areas were found with higher rates of PV than in the rest of the three-county area, the persons with PV did not have any jobs, leisure activities, or other factors in common that were different from persons without PV. The study was not designed to look for environmental exposures or other factors that could explain the high rates of PV. Cancers can take many years to develop; therefore, if an environmental exposure led to PV in the high-rate areas, it likely occurred well in the past. More studies are needed to identify reasons for the high numbers of PV cases and to ensure that any exposures that may be linked to the disease no longer pose a threat to the public.

# An Investigation of Polycythemia Vera in the Tamaqua Area of Northeast Pennsylvania

## Final Report

## **Background**

History. The Tamaqua area of northeastern Pennsylvania includes the towns of McAdoo, Hometown, Still Creek, and Tamaqua. Nearly 75,000 people live in the area. Tamaqua is near the juncture of Luzerne, Carbon, and Schuylkill counties. The 2000 US Census counted 528,388 residents in the three counties. The Tamaqua area, like much of Pennsylvania, has a long history of environmental concerns. These concerns include many past and current industrial and mining operations. Six U.S. Environmental Protection Agency (EPA) Superfund sites are located within 15 miles of the Tamaqua town center. These sites were active between 1960 and 1990 (Table 1). EPA has finished the cleanup at some, but not all, of these sites. Other industries in this area also produce hazardous waste materials. These operations include the Big Gorilla coal ash reclamation project and a number of waste-coal power plants (Table 2). A few reports of contaminated water (private wells, streams, and rivers) in areas near the Superfund sites were reported a number of years ago. Recent tests of municipal and private wells in the area have not found dangerous levels of contaminants. Acid mine drainage (AMD) into local creeks and streams continues to occur. AMD is a potential public health hazard if exposures occur.

In 2004 four cases of a rare blood cancer called polycythemia vera (PV) were found in people who lived on one road in the Tamaqua area. These cases occurred near a former Superfund site. A local health-care provider reported that other cases of PV were in the area and that rates of other cancers were also high. Some Tamaqua area residents believed that an environmental cause explained the problem. These concerns were expressed later that year at a town meeting with local and state officials. The residents thought that toxic waste from the nearby Superfund site entered their private wells and the Still Creek Reservoir. This reservoir is the drinking water source for more than 7,500 people.

The Pennsylvania Department of Health (PADOH) met with community leaders

in October 2004. PADOH agreed to review the cancer rates in Schuylkill, Carbon, and Luzerne counties from 1996–2002. PV rates could be studied for only the years after 2001 because the state cancer registry began collecting information on this diagnosis in that year. The average PV rate in Luzerne County was 3.1 cases per 100,000 persons per year. (PV rates in this report are per 100,000 persons per year.) The PV rate in Schuylkill County was 3.4. These rates were higher than the statewide rate of 1.5. No other cancers listed in the state cancer registry were higher than expected in the tri-county area. Later, PADOH looked at the 2003–2004 cancer registry data and found that the PV rates in Luzerne and Schuylkill counties remained high. Concerned residents claimed that there were cases of PV in the area that were not found in the cancer registry. As a result, PADOH asked ATSDR to help count the cases of PV in the three counties, confirm the diagnosis of PV in these persons, and collect information about these cases. Polycythemia Vera and the JAK2 Mutation. PV is a blood disorder in which the body makes too many red blood cells and sometimes too many white blood cells and/or platelets as well. The high number of red blood cells causes thick blood with symptoms such as headaches, high blood pressure, tiredness, leg cramps, and redness/itching of the skin. PV occurs slightly more often in men than in women and is more common in older persons. PV has no known cause. The general nature of PV symptoms can result in long delays in its diagnosis. PV is often a chance finding when a patient is being tested for other illnesses. If untreated, PV can lead to strokes or other life-threatening events due to stress on the heart and blood vessels. With proper medical care, which includes phlebotomy (removing blood) and/or drug therapy, PV patients are able to function normally and can live for more than 20 years after the diagnosis is made. Sometimes an excess number of red blood cells can be due to other health problems, including lung damage caused by heavy smoking or working in mines. The body's response to these lung problems is to produce more red blood cells to help carry oxygen, which is referred to as secondary polycythemia. PV is classified as a cancer because it involves mutated cells that do not respond to normal signals in the body to control the number of red blood cells. Unlike other cancers, PV does not spread to other parts of the body, and the red blood cells that are produced are normal.

In 2004 a single mutation in the bone marrow stem cells was found in nearly all PV patients. This mutation occurs on the JAK2 gene, which controls signals to start or stop producing red blood cells. Bone marrow cells with the JAK2 mutation do not respond to the "off" signal from the body and, thus, continue to make red blood cells. Experts believe that the JAK2 mutation happens early in the course of the PV disease. What causes this mutation or how it occurs is not known. A tendency to have the JAK2 mutation seems to run in families. The JAK2 mutation can be found through a blood sample and is now a routine part of PV diagnostic criteria. The JAK2 mutation can also be found in other disorders in the PV family (myeloproliferative disorders) but is not found in people with secondary polycythemia or other cancers and is otherwise very rare.

#### **Investigation Goals**

In October 2006 ATSDR and PADOH conducted a survey to locate the PV cases and to ask patients basic questions about place of residence, work history, and other lifestyle patterns. The main goals of the ATSDR investigation were to:

- 1) locate all PV cases diagnosed from 2001–2005 in persons in the tri-county area,
- 2) confirm the PV diagnosis, and
- 3) collect information about PV to identify factors in common among the cases.

## **Methods**

*Participants.* All persons in the Pennsylvania state cancer registry who were diagnosed with PV between 2001 and 2005 and who lived in the tri-county area at the time of diagnosis were asked to take part in the survey. Ads were run through local papers and on radio and television stations to help locate PV patients who had not been reported to the state cancer registry. Tri-county doctors who treat persons with blood disorders were asked to report all PV cases and to encourage their PV patients to take part in the survey. Those persons who responded to the initial request to participate were interviewed by the field investigation team. Any person with PV found after that time was interviewed by telephone. All persons who took part in the survey were asked to provide a blood sample for the JAK2 test, which was done at no charge. The field survey ended on July 31, 2007.

*Confirmation of PV Diagnosis.* A panel of medical experts on PV was formed to review the results of the JAK2 tests and the patients' medical records to identify whether each patient actually had PV.

*Data Analysis.* The survey responses were reviewed to identify differences between those persons with PV and those who did not have PV. PV rates for zip codes and census tracts were calculated for both the confirmed PV cases and all of the registry cases. These data were used to identify high-rate areas of PV.

*Environmental Analysis.* The Pennsylvania Department of Environmental Protection (DEP) and the U.S. EPA provided current environmental data for the identified high-rate areas. ATSDR reviewed the data to see if any exposures were common to the high-rate areas and might explain the findings. All potential exposure sources with available data were evaluated (Table 3), which included:

• air pollution sources;

- water pollution sources;
- solid waste sources;
- coal mining sources; and
- hazardous waste/storage sites.

#### **Results**

*Participants.* A total of 72 persons were interviewed, including 38 persons who were found through the cancer registry and 34 persons found through other means. The 38 persons from the cancer registry represented 39% of the 97 persons eligible to be surveyed. The remaining persons from the registry could not be found, did not want to be surveyed, or had died. The 34 persons found through other means included persons who self-reported their illness and those referred by their doctor. Ten of the 34 persons found through other means did not meet criteria for being in the survey, either because they did not live in the three-county area or were not diagnosed during 2001–2005. A survey group of 62 persons was left (Figure 1). Those 62 persons averaged  $65 \pm 13$  years in age. There were more males (60%) than females. All persons surveyed were white, and most were of European descent.

*PV Diagnosis Confirmation.* The PV diagnosis was confirmed in 33 (53%) of the 62 eligible persons who were surveyed. Of the remaining 29 persons, 17 had secondary polycythemia and 12 did not have enough information in their records to make a firm diagnosis (Figure 2). Five of the 10 patients who were not eligible to be surveyed also had PV. The fact that just more than half of those persons given a diagnosis of PV were confirmed to have the disease is similar to other studies that used the JAK2 test to verify an existing PV diagnosis. The participant and diagnosis confirmation data appear in Table 4.

*Survey.* The survey responses were analyzed by splitting them into two groups: answers from the 42 patients who had confirmed or probable PV and answers from the 20 persons who either did not have PV or had a negative JAK2 result. The 10 persons with unknown PV status were not included. Those persons found to have PV were more likely than those without the disease to report an enlarged spleen, which is a common finding with PV. The not-confirmed group (those who did not have PV) was more likely to report a history of smoking and shortness of breath, both of which are linked to secondary polycythemia. The two groups were similar with respect to exposure to chemicals on the job, other health problems, and leisure activities (Table 5).

*PV Case Locations*. The address at the time of diagnosis was mapped for each person in the survey according to disease status (PV, not PV, unknown) (Figure 3). Most of those in the survey who did not have PV were from the Wilkes-Barre region. When looking at only those persons with confirmed disease, researchers found several areas that had PV rates higher than expected. These high-rate areas were identified using zip code and census tract boundaries. A second analysis was done using all cases from the state cancer registry (whether the patients took part in the survey or not). Results of both methods were compared. Three potential high-rate areas were seen (Figure 4). In two of these areas (Areas 1 and 3), the number of PV cases was very small, making the likelihood of a chance occurrence high. The third area (Area 2) contained 12 of the confirmed cases and satisfied the standard requirements for statistical significance. As a result, these cases probably did not occur by chance.

*Environmental Analysis.* The high-rate area in eastern Carbon County (Area 3) contained few or no exposure sources. It is a densely forested, sparsely populated area. The high-

rate areas near Pottsville and Tamaqua (Areas 1 and 2), however, were found to have common potential exposure sources:

1) <u>Extensive coal mining operations.</u> Coal mining is common throughout the tri-county area, not just the high-rate zones. Higher rates of PV were not seen in other tri-county areas affected by mining.

<u>Waste-coal power plants</u>. Seven of these plants were in operation in or around the two high-rate areas since the early 1990s. Cancers generally take many years to develop, and no other factual data connect the waste-coal plants to increased PV rates in these areas.
 <u>Hazardous waste/Superfund sites</u>. Seven EPA Superfund sites and numerous other hazardous waste sites are within the two high-rate areas. These sites are inactive and are contained or are in the process of remediation by state and federal agencies. Sampling of local rivers and streams, public reservoirs, private wells, and soils does not suggest that people are currently exposed to hazardous chemicals. It is unknown if residents in the area were exposed to toxic substances from these or other sources in the past. Pre-1990 environmental exposure data are not available.

Figure 5 shows the proximity of Superfund sites and waste-coal power plant locations to the high-rate zones.

#### **Limitations**

Several factors limit the conclusions drawn from the investigation:

1. *There are no standard rates with which to compare the study's findings*. To look at disease rates in a population, researchers compare the disease rate in the target group or area of concern to a standard, or background, rate. For this comparison to be valid, both rates need to be based on the same collection and analysis methods. The current incidence rates for PV (0.9 nationally and 1.5 in Pennsylvania) are based solely on state and national cancer registry data. Cancer registries are based on cases diagnosed and reported by doctors. Investigations are not done on each reported case to find out if the diagnosis in the cancer registry is accurate. The ATSDR survey in the tri-county area included non-registry cases in addition to registry cases. Therefore, a direct comparison to standard cancer rates is invalid, and the absolute increase above background levels cannot be known.

2. *The survey used new methods to verify the PV diagnosis*. The PV diagnosis among those who took part in the survey was confirmed by a new genetic test that has not been available for most persons reported to the cancer registry or in other PV studies. The JAK2 mutation was only discovered in 2004 and has not been available to most doctors until recently. Up to 50% of persons diagnosed with PV before the JAK2 test became available may not in fact have this disease. These patients usually have secondary polycythemia or some other disease with similar symptoms. Because of this problem, comparing the rates found in the current survey to other published rates is not valid as the methods used to include cases are not the same.

3. *The PV status of many registry cases is unknown*. The disease status of 75 registry cases (66 patients who did not take part in the survey and 9 patients who lacked adequate medical records) is unknown. Some of these persons likely have PV, but determining how many is not possible. The number and location of these true cases could alter many conclusions of the survey, including the size, location, and significance of any case clusters.

#### 4. The effects of selection bias and unreported cases are not known.

a) Participation bias occurs when persons from certain areas are more likely to take part in a survey due to an increased awareness or interest, affecting the findings of a study. Efforts to find cases and to ask persons to take part in the survey were applied evenly throughout the tri-county area to reduce this form of bias. The impact of participation bias among non-registry cases is very hard to evaluate, because it is not known how many additional people with PV chose not to take part. However, this problem was probably relatively minor because 1) levels of participation among the registry cases were similar throughout the area; 2) there were no patterns among the callers to the PV hotline set up by ATSDR—in fact, few of the calls to the hotline were from tri-county residents; and 3) the patterns of registry versus non-registry cases reflected the way area doctors care for patients with PV. The cancer registry relies on reports from hospitals. In some parts of the tri-county area, doctors are less likely to admit PV patients to the hospital to diagnose their disease. In these areas, more of the cases were found outside the state cancer registry. b) Unreported cases likely exist in the tri-county area. Such cases could alter the survey findings, depending on their number and location. However, it is not likely there are many such cases. Nearly all patients with PV are seen and treated by a specialist, and the diagnosis is not normally made by a primary care doctor. All PV specialists in the tri-county area were contacted and asked to report their cases. No self-reported, confirmed PV cases were seen from the Pottsville or Wilkes-Barre areas, two of the three largest locations in the tri-county area. This is likely due to the high reporting frequency of the physicians in those areas.

5. *Spatial analysis of a rare disease is difficult.* One or two cases of PV in an area with few people will result in an elevated incidence rate, yet the rate itself has little meaning when based on such small numbers. To identify areas with elevated PV incidence below the county level, zip codes and census tracts were used. The zip codes in the tri-county area are of generally similar geographic size but have populations ranging from 10 to 59,418 persons. Census tracts in the tri-county area, although not uniform, are more consistent in population size (959–8,179 persons) but vary widely in geographic area (from less than 1 to nearly 400 square miles). These variations can result in misleading analyses, especially with rare diseases such as PV. The zip code/census tract analysis can have problems accurately identifying areas of environmental exposure or disease occurrence. However, it is a useful screening tool for identifying potential high-rate areas for further study. Only one of the high-rate areas that were found contained enough cases to satisfy the tests for statistical significance.

#### **Conclusion**

The investigation of PV in Carbon, Luzerne, and Schuylkill counties resulted in three main findings:

1) Thirty-three confirmed cases of PV diagnosed between 2001 and 2005 were identified. The patients with PV did not share traits, ancestry, or job experiences that were different from those patients who did not have PV. Three areas had higher PV rates than the rest of the tri-county area; however, only one of these areas contained enough cases to be considered statistically significant.

2) The reporting of PV to the cancer registry was incomplete because some PV patients are not hospitalized. Currently, PV is only reported to the registry through hospitals. Also, numerous misclassified cases were in the registry probably because the JAK2 test has only recently been available to community physicians.

3) There is no evidence to either confirm or refute that the increase in PV cases is related to environmental exposures. When potential environmental exposure sources were evaluated, Superfund sites and waste-coal power plants were common to some of the high-rate areas. Whether or not a relationship exists between any of these sites and the PV cases is not known. This investigation was not designed to study such relationships.

There is no known cause for PV nor was this investigation designed to identify a cause. Any environmental exposures, if they did contribute to the development of PV in the high-rate areas, would likely have occurred well in the past. Most cancers take many years to develop, and the diagnosis of PV is often delayed because of the mild and non-specific symptoms that are common in the early stages. Because the cause of PV is unknown, linking any environmental agent—or any other factor—to these cases is difficult. Further work is needed to study such questions.

#### **Recommendations**

This investigation resulted in three major recommendations:

1) Inform health-care providers about the new guidelines for the diagnosis of PV, which include the JAK2 test.

2) Improve the reporting of PV and other similar diseases to state registries.

3) Convene a roundtable of leading PV researchers to identify and prioritize studies regarding the high rates of PV found in this investigation.

ATSDR and PADOH convened an initial meeting of experts to address this issue. The August 25, 2008 meeting included leading medical researchers, environmental experts, and epidemiologists from local academic institutions. The group will be discussing and prioritizing the various studies that are necessary to better explain the observed cluster of PV cases. This work may include genetic analyses, environmental testing, ecological assessments, and more rigorous studies of the area population. In addition, local and national registry reporting of PV is currently being evaluated. PADOH will continue to monitor the PV incidence in the tri-county area. ATSDR and PADOH will provide technical support, when appropriate, to research partners.

Superfund Site Name	Country	Operation	EPA Cleanup	
Business Type	County	Period	Start	End
C & D Recycling telephone cable recycling	Luzerne	1960–1980	1987	1999
Eastern Diversified Metals wire recycling	Schuylkill	1966–1989	1987	-
McAdoo Associates hazardous waste recycling	Schuylkill	1975–1979	1988	1995
Metropolitan Mirror & Glass mirror manufacturing	Schuylkill	1959–1982	1997	1998
Tonolli Corp. battery recycling	Carbon	1974–1985	1989	-
Valmont (Chromatex) upholstery manufacturing	Luzerne	1978–1988*	1987	-

 Table 1. U.S. Environmental Protection Agency (EPA) Superfund sites located within 5

 miles of the polycythemia vera high-rate areas in Carbon, Luzerne, and Schuylkill counties

\*Company operated until 2001; trichloroethylene (TCE) use was discontinued in 1988.

Table 2. Waste-coal power plants within 5 miles of high-rate polycythemia vera areas in
Carbon, Luzerne, and Schuylkill counties

County	City	Plant Name	Capacity (MW)	Year Online
Carbon	Nesquehoning	Panther Creek Energy	83	1992
Schuylkill	Frackville	John B. Rich Memorial Power Station	80	1988
Schuylkill	McAdoo	Kline Township CoGen Facility	50	1989
Schuylkill	Shenandoah	St. Nicholas CoGen Project	88.6	1990
Schuylkill	Frackville	Wheelabrator Frackville Energy	43	1988
Schuylkill	Tremont	WPS Westwood Generation LLC	30	1987
Northampton	Northampton	Northampton Generating LP	108	1995

 Table 3. Environmental data reviewed to evaluate common potential exposure sources for

 the high-rate polycythemia vera zones in Carbon, Luzerne, and Schuylkill counties

Coal Mining Sources	Water Pollution Sources
Coal Mining Operation	Water Discharge Points
Discharge Point	Commercial
Mineral Prep. Plant	Electrical
Refuse Disposal Facility	Industrial
Underground Mine	Mineral
Surface Mine	Sewage Treatment
Refuse Reprocessing	
	Water Pollution Control Facility
Industrial Mineral Mining Operation	Discharge Point
Discharge Point	Groundwater Monitoring Site
Surface Mine	Production Service Unit
	Treatment Plant
Mine Drainage Treatment	
Land Reclamation Projects	Lake, River, and Creek Water Quality Test Results
Mine-Orphan Discharges	Private Well Test Results
Abandoned Mine Land	
Acid Mine Drainage Discharge Area	Solid Waste Sources
Underground Mine Fire	Residual Waste Operation
Untreated Discharge	Municipal Waste Operation
Treated Discharge	Landfill
	Abandoned Landfill
Hazardous Waste Sites/Storage	Transfer Station
Storage Tank Locations	
Manufacturing/Industrial	Land Recycling Cleanup Location
Petroleum	Air
Beneficial Land Use Areas	Groundwater
U.S. Environmental Protection Agency	
(EPA) Tri-Sites	Soil
Radiation Facilities	Surface Water
	Waste Media
Air Pollution Sources	
	Erosion and Sediment Control-
Air Emission Plant	Remediation/Restoration
Point of Air Emission	Sinkhole Locations
Fuel Material Location	Landslide Areas
Incinerator	
Combustion Unit	
Process	
Air Pollution Control Device	

	Registry	Non-Registry	Total
Participants			
Total	104 <sup>1</sup>	34	138
Eligible	97	24	128
Actual	38	24	62
Non-Participants			
Refused	16	-	16
Deceased	13	-	13
Not Found	30	-	30
Questionnaires			
Total	38	34	72
Eligible	38	24	62
PV Status <sup>2</sup>			
PV	18	15	33
Not PV	11	6	17
??? PV	9	3	12

# Table 4. Registry and non-registry participants, polycythemiavera (PV) status, and non-participants

<sup>1</sup> Includes seven cases added to the registry after the investigation was completed. <sup>2</sup>PV = polycythemia vera diagnosis confirmed by expert panel; not PV = secondary polycythemia or other non-PV diagnosis; ??? PV = medical record inadequate to Suggest a diagnosis.

-	Confirmed		Not-Confirmed	
Participants reporting:	Number	%	Number	%
exposure to toxic waste site	4	9.3	2	10.0
military service	16	37.2	7	35.0
blood transfusion	4	9.3	2	10.0
history of cigarette smoking <sup>*</sup>	28	65.1	19	95.0
eating local fish	20	46.5	8	40.0
eating local meat	32	74.4	12	60.0
eating local vegetables	40	93.0	18	90.0
regular consumption of alcohol	18	41.9	10	50.0
testing home for radon	11	25.6	3	15.0
high radon test result	1	2.3	1	5.0
working in a factory	31	72.1	13	65.0
exposure to solvents	20	46.5	11	55.0
exposed to chemicals at work	16	37.2	12	60.0
splenomegaly <sup>*</sup>	13	30.2	1	5.0
cataracts	15	34.9	4	20.0
deep vein thrombosis	5	11.6	2	10.0
thick blood	33	76.7	18	90.0
frequent headache	11	25.6	6	30.0
frequent dizziness	12	27.9	8	40.0
other visual symptoms	11	25.6	8	40.0
numbness/tingling	20	46.5	9	45.0
easily fatigued	24	55.8	10	50.0
abdominal discomfort-fullness	14	32.6	5	25.0
unintentional weight loss	12	27.9	2	10.0
night sweats	12	27.9	7	35.0
post-bath itching	27	62.8	12	60.0
facial fullness and redness	21	48.8	10	50.0
shortness of breath <sup>*</sup>	11	25.6	10	50.0
chest pain	4	9.3	4	20.0
frequent lower limb cramps	18	41.9	10	50.0
liver enlargement	5	11.6	2	10.0
blood relatives with PV	2	4.7	1	5.0
blood relatives with blood cancer	7	16.3	4	20.0

Table 5. Selected responses from polycythemia vera investigation questionnaire based on diagnosis status: confirmed vs. not-confirmed PV

Table 5 (continued)

Participants reporting having	Confirmed		Not-Confirmed	
ever lived within 1/2 mile of:	Number	%	Number	%
golf course	2	4.7	2	10.0
railroad	25	58.1	15	75.0
hazardous waste site	5	11.6	3	15.0
airport	4	9.3	1	5.0
gas station	22	51.2	15	75.0
nursery	2	4.7	4	20.0
high voltage tower	7	16.3	2	10.0
incinerator	4	9.3	1	5.0
factory	14	32.6	9	45.0
quarry/mine	22	51.2	10	50.0
coal-fired power plant	1	2.3	1	5.0
nuclear power plant	0	0.0	0	0.0
other sites	4	9.3	2	10.0
landfill/dump	5	11.6	5	25.0

\*statistically significant

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