

# Polycythemia Vera

## Polycythemia Vera Cancer Cluster Investigation in Northeast Pennsylvania

### *A Feasibility Assessment to Improve the Reporting of Hematopoietic Diseases*

#### Study Purpose

The purpose of this study was to find ways to increase reporting of PV and other blood cancers to cancer registries by physicians' offices and other outpatient facilities. Blood cancers are underreported because they are often diagnosed and treated in outpatient facilities. The reporting practices of physicians' offices and other outpatient facilities can affect the completeness and accuracy of cancer registry data.

#### Background

PV is part of a group of diseases called myeloproliferative neoplasms (MPN). MPNs are a group of blood cancers that cause the bone marrow to make too many blood cells. The MPN designation also includes essential thrombocytosis (ET) and primary myelofibrosis (PMF). In 2005, the Pennsylvania Department of Health (PADOH) used state cancer registry data to determine that there was a significantly higher incidence of PV in Luzerne and Schuylkill counties compared with the rest of Pennsylvania. In 2006, PADOH asked ATSDR to help study PV patterns among residents in an area of northeast Pennsylvania. ATSDR reviewed medical records from physicians' offices in the area, conducted genetic testing for the *JAK2V617F* mutation, and confirmed the presence of a cluster of PV at the center of three counties (Carbon, Luzerne, and Schuylkill) in northeast Pennsylvania. The study also found that nearly half (15/33) of the confirmed PV cases had not been reported to the Pennsylvania Cancer Registry.

#### What Was Studied

- In the current study, the state central cancer registries (CCRs) of Kansas, New York, and South Carolina were contracted by the Division of Cancer Prevention and Control at the Centers for Disease Control and Prevention to participate in this study. Each cancer registry recruited physician practices within their states that diagnosis and/or treat patients with reportable hematopoietic diseases, including PV and other myeloproliferative neoplasms (MPNs). Non-Hodgkin's lymphoma and Hodgkin's lymphoma were not included in the study. The registries then developed and delivered training to educate the physicians and their staff about cancer reporting requirements and processes. They also evaluated the way physicians' offices were reporting cases.

#### What Is a Cancer Registry?

*Cancer registries* collect information about cancer patients and the treatments they receive. This information is used to answer questions like:

- Are more or fewer people getting cancer this year compared with last year?
- What groups of people are most likely to get a specific cancer, such as PV?



## Features of this Study

Each registry had to identify and recruit doctors to participate in the study, train them to report cancer cases, and evaluate the data reported. All three registries recruited doctors with specialties in hematology and oncology. Additionally, Kansas' registry included a sample (11) of primary care clinics. Packages with information about the study and cancer reporting requirements were mailed to physicians' offices. Registries provided in person trainings and webinars. Mailings, telephone calls, emails, and Internet postings were also used. Using the required procedures for their state, doctors from each state reported cancer cases. In New York, doctors reported cases via the state's registry website. In Kansas, doctors reported cases via mail and computer software. In South Carolina, some doctors reported cases via a web-based application; other doctors provided the registry access to their medical records and registry staff abstracted case information.

## Gathering and Analyzing the Data

The three state cancer registries submitted data for 2010 on all reportable hematopoietic diseases included in the study. The data were reviewed to identify:

- Cases that were reported as a result of registries' routine follow up with doctors
- Cases that were reported as a result of the study
- Cases reported by more than one source (e.g., reported by targeted doctor and another source, such as hospital or laboratory)
- How the case was reported (e.g., reported by a medical practice or doctor in the study or by the registry)
- Demographic characteristics of cases
- Number of cases of PV (and other blood cancers) confirmed via *JAK2V617F* (JAK2) mutation testing

## Key Results

- In total, the three states collected data on 3,640 blood cancer cases diagnosed in 2010; 725 cases were reported as a result of this project. Of the 3,640 cases, 3,114 (86%) were in people aged 50 and older, and 22% of cases had a previous diagnosis of cancer.
- Of the 488 MPN cases, there were 166 PV cases (34%), 193 ET cases (40%), 31 PMF cases (6%), and 98 "other" cases (20%), including MPN, NOS, chronic neutrophilic leukemia, and chronic eosinophilic leukemia.
- Of the 379 cases coded as MPN, 51% (193) had the JAK2 mutation test ordered. Of the 193, 122 cases (63%) were JAK2 mutation-positive, 64 cases (33%) were negative for the mutation, and 7 cases (4%) did not have reported results available.

## Conclusions

- Although the approaches used by the three registries to target reporting of PV and other blood cancers varied by CCRs, they are broadly applicable to other CCRs.
- A substantial amount of effort and staff resources on the part of the registry is needed to enhance the reporting.

## Next Steps

Researchers from the Kansas Cancer Registry will be publishing their work in a scientific journal. When the results from all of the ongoing research projects related to this investigation are available, ATSDR will hold a public forum to share and discuss the results with interested stakeholders.

## For More Information

Visit ATSDR's Web page on PV: [http://www.atsdr.cdc.gov/sites/polycythemia\\_vera/index.html](http://www.atsdr.cdc.gov/sites/polycythemia_vera/index.html)

Call ATSDR's toll-free PV information line: 866-448-0242 or email Dr. Elizabeth Irvin-Barnwell ([jcx0@cdc.gov](mailto:jcx0@cdc.gov)), ATSDR Division of Toxicology and Human Health Sciences.

Contact Lora Siegmann Werner, ATSDR Region 3 (215-814-3141 or [lkw9@cdc.gov](mailto:lkw9@cdc.gov))